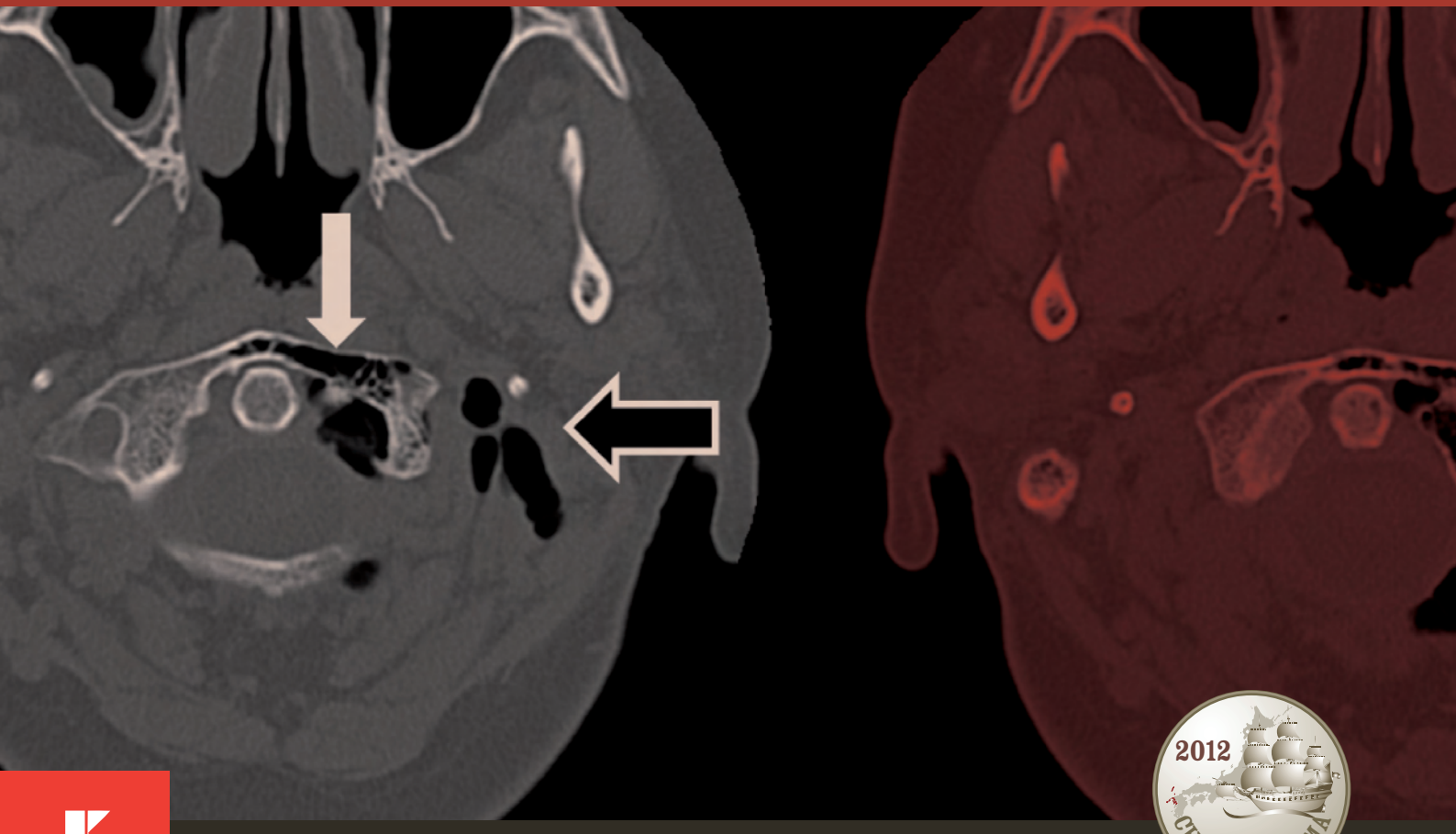
Cholesteatoma and Ear Surgery

an update

Edited by
Haruo Takahashi



CHOLESTEATOMA AND EAR SURGERY – AN UPDATE

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Proceedings of the 9th International Conference
on Cholesteatoma and Ear Surgery.
June 3-7, 2012, Nagasaki, Japan.

Kugler Publications/Amsterdam/The Netherlands

ISBN 13: 978-90-6299-237-9

Distributors:

For the USA and Canada:
Pathway Book Service
4 White Brook Road
Gilsum, NH 03448
U.S.A.
email: pbs@pathwaybook.com

For all other countries:
Kugler Publications
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PREFACE

It was indeed a great pleasure and honor for me to have hosted the 9th International Conference on Cholesteatoma and Ear Surgery in Nagasaki, Japan. There were 558 participants; four full-day programs provided them with rich scientific programs including six keynote lectures, seven symposia, 26 panel discussions, 12 mini lectures on recent topics, 23 instruction courses, two live cadaver temporal-bone dissections, three sponsored sessions, three other sessions, and three days of temporal-bone dissection courses. I would like to take this opportunity to express my greatest gratitude to all the faculties who contributed enormously to these scientific programs only to facilitate and improve the academic quality of all the participants. We also thank all the staff members who worked relentlessly for several years to help make this Conference a success, and the participants who took part in the Conference. I hope that the Conference was really fruitful to everybody.

It has been a tradition to publish paper proceedings after each International Conference on Cholesteatoma and this is the first time the proceedings were published as e-book on the website of the Conference. The advantages of this e-book are that it is of high quality, and any article can be downloaded by participants for free. It will be usable on multiple devices: computer/laptop, tablet PCs (iPad), iPhone, e-readers, etc. It is fully searchable and has easy navigation. If you want a copy (paper book) Kugler Publications can also provide POD (Publishing on Demand) copies for a reasonable price. The e-book will be kept open on the web at least until the next Cholesteatoma Conference.

I hope that you will enjoy this e-book as much as the traditional paper proceedings, and that it will stimulate scientific and academic accomplishments of all of you.

Finally, I am very much looking forward to seeing all of you at the 10th Conference in Edinburgh in 2016!

Haruo Takahashi, MD

KEYNOTE LECTURES

PREVENTIVE MEASURES AGAINST CHOLESTEATOMA RECURRENCE IN CANAL-WALL-UP TYMPANOPLASTY: STAGING THE OPERATION AND CHOICE OF MASTOID OBLITERATION

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Introduction

For the last 41 years, the senior author (NY) operated on 1053 new cases of middle-ear cholesteatoma at three institutions, where he followed up all these patients as long as possible. Since he saw the video of the planned staged tympanoplasy for cholesteatoma by Dr. Sheehy, Los Angeles, he has decided to adopt the staged canal-up operation and since 1970 he treated more than 500 fresh cases of advanced middle-ear cholesteatoma with this technique. Improvements of the surgical technique were presented to the international conference on cholesteatoma and mastoid surgery in Tel Aviv in 1981[1] and in Antalya in 2008.[2]

Based on the surgical experiences he has come to the conclusion that surgical management of cholesteatoma should satisfy the following four objectives: 1) To restore a normal external ear canal with self cleaning function; 2) To restore an aerated middle ear lined with normal mucosa; 3) To restore socially adequate hearing; and 4) To eliminate all possible causes of recurrence. As these four objectives are interdependent, only a canal-wall-up operation remains as a choice to achieve all four objectives. For this purpose, we have developed the staged canal-wall-up tympanoplasty with or without total mastoid obliteration.

Cholesteatoma recurrence

For a good understanding of the staged canal-wall-up tympanoplasty, we first describe why cholesteatoma recurs. Cholesteatoma recurrence is classified into two types: residual cholesteatoma that arises from a residue of the matrix, and recurrent cholesteatoma that originates from a post-operative retraction pocket.

Residual cholesteatoma

Complete elimination of residual cholesteatoma is not always easy because: 1) A tiny residual matrix is hidden in the area, difficult to observe directly; 2) A tiny matrix buried in the granulation tissue is difficult to identify; and 3) A tiny matrix of open-type cholesteatoma is difficult to differentiate from diseased thick mucosa. In our previous study, 40% of residual cholesteatoma was found in the tegmen of the attic and another 40% in the tympanic sinus and peristapedial area.[3] With the aid of oto-endoscopy we must carefully clean these risky areas so as not to overlook even a very tiny residue of the matrix.

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Recurrent cholesteatoma

Retraction pocket is a precursor of recurrent cholesteatoma. We believe that at least three factors, *i.e.*, chronic inflammation in the middle ear, hypoventilation of the tympanic cavity, and defect of the tympanic scute, are related to the development of a retraction pocket. Once self cleaning in the pocket is inhibited, cholesteatoma recurs. To prevent post-operative retraction pocket, therefore, chronic inflammation in the middle ear must be eliminated, an aerated tympanic cavity with ventilation function must be restored and a smooth and stable tympanic scute must be reconstructed.

Because background factors relating to cholesteatoma recurrence are many and complex in advanced cholesteatoma, we can not eradicate all of them with a one-stage canal-wall-up operation. For this reason, a carefully planned and individualized staged operation is needed to achieve the four operational objectives described above. The staged canal-wall-up tympanoplasty with or without mastoid obliteration as described below is a surgical method we have developed to treat the cases with high risk for recurrence due to destructive extension of cholesteatoma beyond the site of origin into the antrum and mastoid cavity involving the ossicular chain.[4,5]

Surgical method

The first-stage operation

Through a retro-auricular incision along the auricular attachment a transcanal attico-tympanotomy and a transcortical mastoidectomy with posterior hypotympanotomy are carried out. While drilling the mastoid cortex bone paté is collected using a paté collector. To control chronic inflammation in the middle-ear granulation tissue and edematous mucosa in the mastoid antrum are cleaned as much as possible. With the combined transcanal and a transmastoid approach cholesteatoma is eliminated totally, together with the malleus and incus, as shown in Figure 1. Oto-endoscopic inspection of the total hidden area is important to minimize the risk of recurrence due to residual cholesteatoma. A sufficiently large silastic sheet of 0.3 or 0.5 mm in thickness is cut in oval shape and folded to insert into the tympanic cavity from the antrum through the aditus ad antrum. As shown in the Figure 2, the lateral side of the folded silastic sheet attaches to the bony edge of the external ear canal and the tympanic membrane to prevent pocket formation. Then the scutum defect is closed by covering over the defect with a plate of bone paté mixed with fibrin glue (Fig. 3). Using a piece of the temporalis fascia, the tympano-meatal defect is closed with an underlay technique. The defect of the mastoid cortical bone is closed using a plate of bone paté hardened with fibrin glue (Fig. 4).[6] Then the retro-auricular wound is closed.

The procedures illustrated in Figures 2, 3 and 4 are important to facilitate aeration of the middle ear and to prevent post-operative retraction of the ear drum.

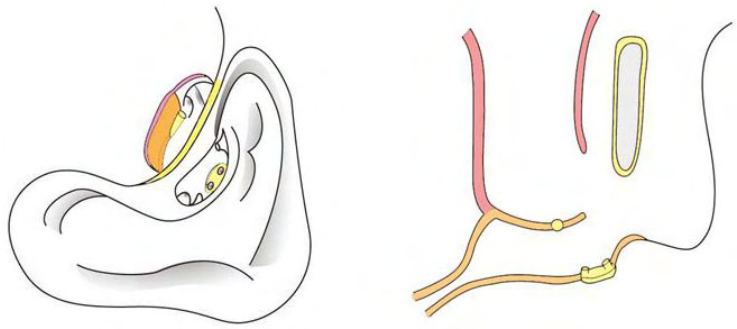

Fig. 1. With the combined transcanal and transmastoid approach, cholesteatoma and diseased tissue are removed, together with the malleus and incus. Through the wide posterior hypotympanotomy opening the attic and tympanic sinus must be carefully cleaned not to miss residual cholesteatoma. The superstructure of the stapes is missing in this case.

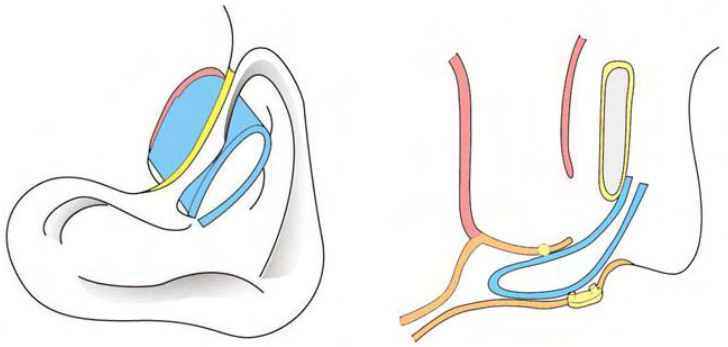

Fig. 2. A sufficiently large silastic sheet of 0.3 or 0.5 mm in thickness is cut in oval shape. The sheet is folded and inserted into the tympanic cavity from the antrum through the aditus ad antrum.

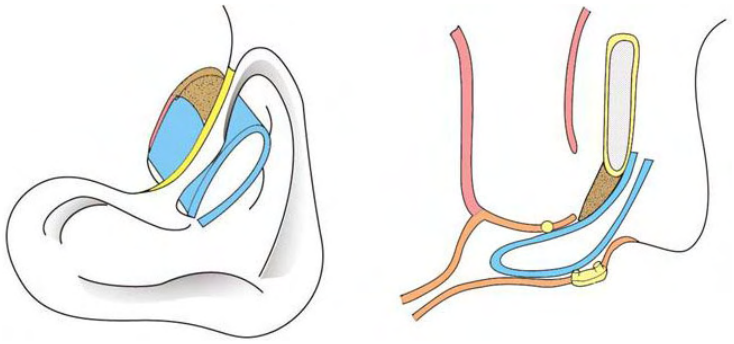

Fig. 3. View after the scutum plasty. Bone paté mixed with fibrin glue is pasted over the silastic sheet to close the scutum defect.

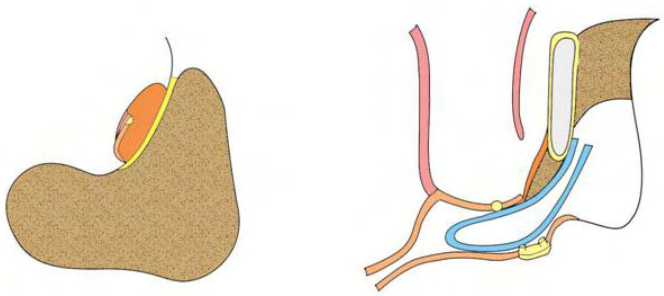

Fig. 4. View after the mastoid cortex plasty. Using a bone paté plate hardened with fibrin glue, the defect of the mastoid cortical bone is closed. The tympanomeatal defect is closed with a piece of temporal fascia by an underlay technique.

Timing of the second-stage operation and choice of mastoid obliteration

The second-stage operation is performed one year after the first operation. A year of interval is needed for the following reasons: 1) Residual cholesteatoma, if any, grows to an appropriate size to remove easily and completely; 2) Chronic inflammation in the middle ear cures; 3) The middle-ear cavity is lined by mucosa and aerated. To assess healing of the middle ear and the grade of aeration, computed tomography (CT) of the temporal bone is indispensable.

The middle-ear aeration is graded as follows: Grade 1, only the mesotympanum is aerated; Grade 2, the entire tympanic cavity, including the attic, is aerated; Grade 3, the tympanic and mastoid cavities are aerated.[7] Total mastoid obliteration is indicated for ears in which aeration is limited to the mesotympanum and attic (grade 1 and 2), whereas canal-wall-up tympanoplasty without mastoid obliteration is chosen for ears with grade-3 aeration.

The second-stage operation

Through the retro-auricular incision along the scar from the first operation, the reconstructed mastoid cortex is exposed and drilled out to open the mastoid cavity. Bone paté is collected again using a paté collector.

Canal-wall-up tympanoplasty without mastoid obliteration

In the well-aerated middle ear of grade-3 aeration, regenerated mucosa around the silastic sheet is thin and the mastoid antrum is aerated. The silastic sheet is pulled out of the tympanic cavity without difficulty and the aerated tympanic cavity is widely accessible through the hypotympanotomy opening. With the transcanal approach, the scutum reconstructed in the first operation is recruited with a piece of auricular cartilage. The ossicles are reconstructed using either a partial or a total ossicular replacement prosthesis (Fig. 5). The mastoid cortex plasty is carried out to keep good middle-ear aeration using bone paté.[6,7] Finally, the retro-auricular wound is sutured.

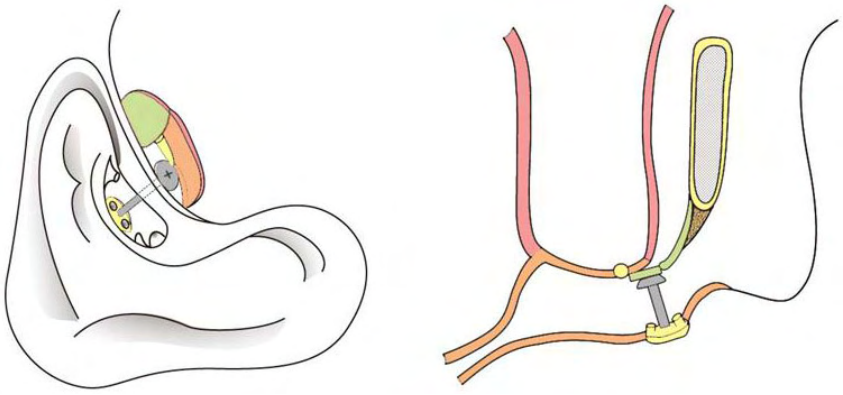

Fig. 5. With transcanal approach, the scutum reconstructed in the first operation is recruited with a piece of auricular cartilage. The sound conduction system is reconstructed, using either a partial or a total ossicular replacement prosthesis. In this case TORP is used and a piece of cartilage is inserted between the top and the tympanic membrane.

Canal-wall-up tympanoplasty with mastoid obliteration

In the poorly-aerated middle ear of grade-1 or grade-2 aeration the regenerated mucosa around the silastic sheet is thick and little air space is observable in the middle ear. The thick, edematous mucosa in the mastoid and antrum is removed together with the silastic sheet and the narrow air space in the tympanum is opened. The scutum and the ossicules are reconstructed as shown in Figure 5. Then the mastoid cavity is obliterated completely in the following way: 1) Using fibrin glue, two or three small pieces of auricular cartilage are glued to the posterior external ear canal from behind to block communication between the antrum and tympanic cavity (Fig. 6); 2) The antrum is obliterated with bone paté. When there is insufficient bone paté, hydroxyapatite granules can be added to obliterate the cavity (Fig. 7). Finally, the rest of the mastoid cavity is obliterated completely with bone paté to reconstruct the mastoid cortex[5] (Fig. 8) and the retro-auricular wound is closed.

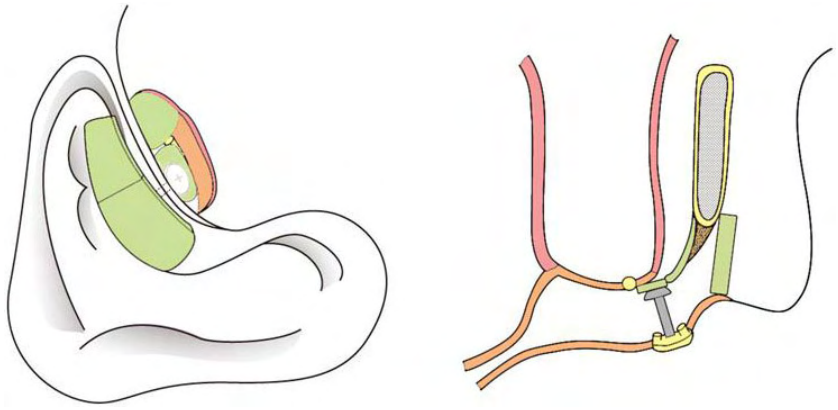

Fig. 6. Using fibrin glue, two or three small pieces of auricular cartilage are glued to the posterior external ear canal from behind to block communication between the antrum and tympanic cavity.

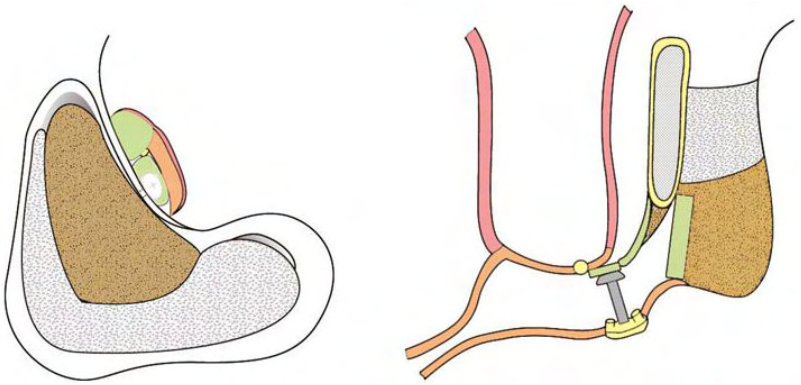

Fig. 7. The antrum is obliterated with bone paté. When there is insufficient bone paté, hydroxyapatite granules can be added to obliterate the cavity.

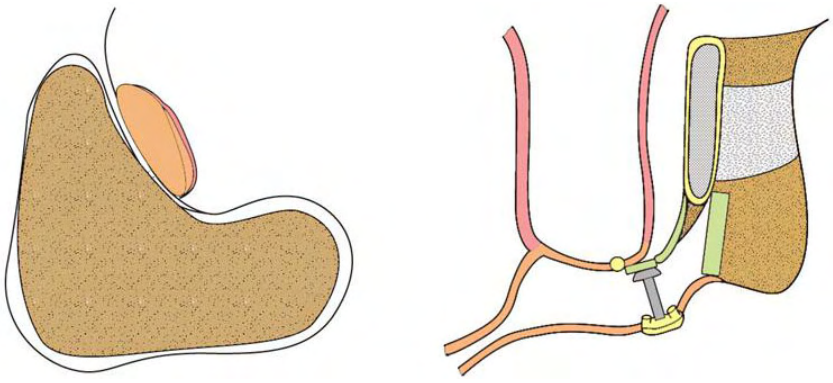

Fig. 8. The rest of the mastoid cavity is obliterated completely with the bone paté plate to reconstruct the mastoid cortex.

Results

Between December 1995 and November 2006, we have performed the staged canal-wall-up tympanoplasty on 80 new cases with advanced cholesteatoma. All cases were followed up as long as possible more than a year after the second-stage operation. No post-operative complications such as ear drum perforation, infection, otorrhea and extrusion of the ossicular prosthesis has been noted in any one of the 80 ears. In 20% of the ears a small residual cholesteatoma was found at the second-stage operation, but was removed easily as a pearly mass. Thanks to the staged procedure none of the 80 ears had residual cholesteatoma at the last follow-up examination.

Table 1. Demographic data of the patients.

	Obliteration	No Obliteration
	(n=27)	(n=21)
age (yrs)	46.3±15.0 (8-68)	36.6±21.8 (10-71)
gender	M:10, F: 17	M: 13, F: 8
numbers of age <15 years	1	8

Among the 80 cases we have completed follow-up examinations with otomicroscopy and otoendoscopy and a temporal bone CT in 48 cases (60%), 27 cases with mastoid obliteration and 21 without mastoid obliteration, for longer than five years after the second operation. Table 1 shows demographic data of the two groups of the patients. As to post-operative retraction pocket we classified eardrum finding into three categories as shown in Figure 9: without retraction pocket, with shallow retraction pocket and with deep retraction pocket. Table 2 summarizes the incidences of the pockets after the second-stage operation with and without obliteration. In total, 65% of the ears had no retraction pocket. They were totally free from fear of recurrence. Twenty-nine percent of the ears had a shallow but stable and self-cleaning pocket. They were also free from recurrence, but continuation of follow-up examinations once or twice a year was advised. Hence 94% of the cases restored trouble-free ears without fear of recurrence. While there was no ear with deep retraction and a result without retraction pocket was obtained more in the ears without obliteration, no statistically significant difference in the incidences of retraction pocket was verified between the groups with and without mastoid obliteration.

In our follow-up study, the incidences of retraction pocket three years and five years after the second-stage operation are the same. This result indicates that post-operative follow up for at least three years is mandatory to assess the surgical outcome.

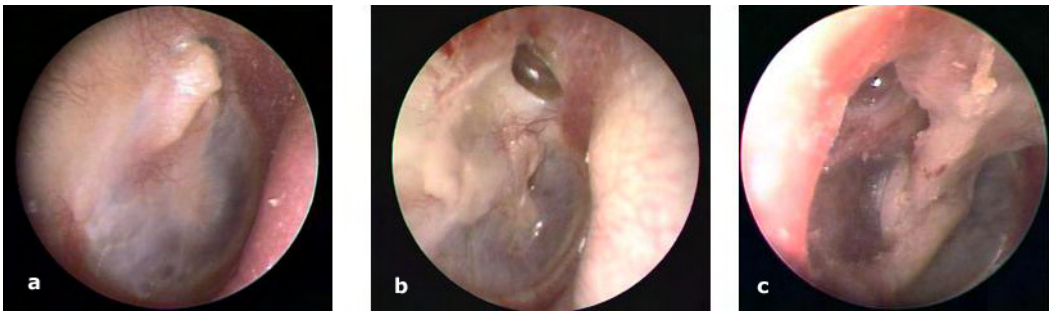

Fig. 9. Classification of ear drum retraction three years after the second stage operation: no retraction (a); sallow retraction, shallow, smooth, innocent, and did not require cleaning (b); and deep retraction with a crust requiring cleaning (c).

Table 2. Incidence of retraction pocket in the ears with or without mastoid obliteration.

	Witn obliteration	Without obliteration	Total
No retraction pocket	15 (56%)	16 (76%)	31 (65%)
Shallow retraction pocket	9 (33%)	5 (24%)	14 (29%)
Deep retraction pocket	3 (11%)	0 (0%)	3 (6%)
Total	27 (100%)	21 (100%)	28 (100%)

Comment

In this lecture we presented the staged canal-wall-up tympanoplasty that we have developed for the last 40 years aiming to eliminate all possible causes of recurrence of cholesteatoma and to restore a trouble-free ear, keeping the external ear canal intact. The updated surgical measures are described. We strictly limited the indication of the procedures to advanced middle-ear cholesteatoma extending from the attic or the tympanic cavity to the mastoid antrum involving the ossicular chain and destroying the tympanic scute. Careful otomicroscopic and otoendoscopic examinations and occasionally a CT of the temporal bone were carried out for five years or more after the second operation in 60% of the cases operated on.

In our investigations in 1992, incidence of recurrent cholesteatoma was 40% following a canal-wall-up one-stage operation.[8] As a result of improvements of the staged canal-wall-up procedures for the last 20 years,[9] we have achieved the purpose of the operation in 100% of the new cases when the middle ear restored good aeration after the first operation. Even in the ears with continued poor middle-ear ventilation we could reduce incidence of deep retraction pocket from 37% to 10% of the ears thanks to the improvement of the staged surgical maneuver and the mastoid obliteration method. The present result shows the effect of the total mastoid obliteration for tympanic aeration using sliced cartilage flap, bone paté and hydroxyapatite grain on one hand and importance of stable aeration of the tymapanic cavity to prevent post-operative retraction pocket on the other. Still, further study is needed to ameliorate middle-ear aeration and to stabilize it to prevent post-operative retraction pocket formation and eradicate the resultant cholesteatoma recurrence.

Post-operative hearing outcome is another important aspect of the surgery for cholesteatoma. Although it is out of the scope of the present lecture, we would like to mention that staging the operation and mastoid obliteration do not harm the hearing result. Post-operative hearing improvement also largely depends upon post-operative middle-ear aeration and mucosal condition at the second-stage operation.

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MANAGEMENT STRATEGY FOR CHRONIC OTITIS MEDIA WITH CHOLESTEATOMA IN 2012

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Introduction

Management of chronic otitis media with cholesteatoma has been the subject of numerous publications in the past 40 plus years. The open-cavity tympanomastoidectomy has been the gold standard for safety, but not necessarily the easiest for the patient. Intact-canal-wall strategies have emerged, but usually have a very high re-operation rate for residual or recurrent disease. We have elected to use a canal-wall reconstruction and mastoid obliteration strategy first described by Merke. Our long-term results with this technique are described.

Methods

A retrospective review was performed of all patients undergoing CWR tympanomastoidectomy with mastoid obliteration at a single institution from 1997 to 2011. The data included pre- and post-operative audiometry, residual cholesteatoma at second-look surgery with ossiculoplasty, post-operative complications, recurrence rate and location.

Results

Two hundred eighty-five ears in 273 patients underwent CWR tympanomastoidectomy with a mean age of 35 years (range 2 to 80 years). A second-look ossiculoplasty was performed in 245 (86%). Recurrent otorrhea occurred in 18 (7.3%) ears. Most otorrhea were secondary to attic retractions due to insufficient bone blocking the attic and were managed with atticotomy (n = 10). Only seven ears (2.5%) required a revision open-cavity mastoidectomy (n = 5) or subtotal petrousectomy (n = 2) for recurrent cholesteatoma. No ears developed recurrent cholesteatoma in the obliterated mastoid cavity. Audiometric follow up on 148 patients who underwent second-look ossiculoplasty demonstrated small improvement in pre-operative versus post-operative air-bone gap (ABG), 28 dB vs. 23 dB respectively. Post-operative infection occurred in 16 (5.6%) patients with one patient requiring conversion to open-cavity mastoidectomy.

Conclusions

A CWR tympanomastoidectomy provides excellent intra-operative exposure of the middle ear and mastoid without the long-term disadvantages of a canal-wall-down mastoidectomy. Long-term follow up demonstrates low rates of recurrent cholesteatoma with stable or improved ABG. Recurrent attic retractions have been eliminated by use of mastoid tip bone instead of bone chips.

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SYMPOSIA

EPIDEMIOLOGICAL STUDY ON CHOLESTEATOMA IN FUKUOKA CITY TO REVEAL THE PATHOGENESIS OF CHOLESTEATOMA

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It is true that the retraction theory mainly gets support as a pathogenesis of primary acquired cholesteatoma. It has been said that retraction of the ear drum is caused by Eustachian tube blockage as a result of otitis media. Since Eustachian tube blockage usually causes otitis media with effusion, the idea that cholesteatoma develops from OME is widely accepted. However, in our daily clinical practice, we do not usually observe the process from OME to cholesteatoma. Furthermore, several studies have reported that a patulous Eustachian tube may contribute to the pathogenesis of cholesteatoma. So it is still unclear what causes the retraction of the ear drum.

To reveal the pathogenesis of cholesteatoma, we carried out a epidemiological study. At first, we considered a cohort study, which has the advantage that it allows us to perform a prospective study which provides the direct measurement of absolute risk. However, a cohort study is time consuming and costly to carry out, and if the incidence is rare, it is impossible to carry out in a reasonable number of patients. Therefore, it is very difficult to select the right group for the study and very difficult to establish an appropriate system to follow up the patients.

Fortunately, we have a good example of a cohort study group near our University in Fukuoka (Japan), next to the town of Hisayama. Hisayama is the place where a prospective cohort study of lifestyle-related diseases has been carried out since 1961. Since the Hisayama study has established a great system for a cohort study, we planned to use the same population group in that study. However, if the incidence of cholesteatoma is too low, it would be impossible to perform a cohort study in Hisayama with its population of only 8,000. It is hard to determine how high the incidence of cholesteatoma is, because the epidemiological survey of cholesteatoma is seldom performed, and in particular not much study has been done including the non-operated cases. So, we first tried to predict the annual incidence of primary acquired cholesteatoma including the non-operated cases in a larger population, which is 1.4 million in the city of Fukuoka.

We collected the information of patients newly diagnosed with primary acquired cholesteatoma in Fukuoka City in 2008 during the period of six months. We sent questionnaires to all otolaryngologists in Fukuoka City and included those in surrounding towns, in case residents of Fukuoka city had consulted them. The surveyed area includes two university hospitals, 20 general hospitals and 87 ENT clinics. As a result, we collected 70 cholesteatoma cases during this six-month period: 37 males and 32 females. The mean age was 52 and biggest number of incidence was found amongst the age group of 60 years and older. Since the population of Fukuoka City is about 1.4 million, the annual incidence of acquired cholesteatoma was calculated to be 10.0 cases in a population of 100,000.

However, more detailed investigation revealed the difficulty of cholesteatoma diagnosis. From the clinical side, 20 cases were reported to be sent to hospitals. On the other hand, the hospital side reported 39 cases were sent to them by the clinics. Furthermore, only 13 cases out of 20 were found in these 39 cases reported from the hospitals, which means that seven cases were missing, and 26 other cases were not diagnosed as cholesteatoma in the clinics and sent to a hospital. In many cases, inflammation and otorrhea were severe. Therefore, it is possible that these cases were diagnosed as other otitis media cases in clinics without appropriate equipment, such as a CT scan. When we only use 48 cases from the hospital side, which seem to be

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Edited by Haruo Takahashi

highly reliable in diagnosis, the annual incidence of cholesteatoma was calculated as 6.8 cases in a population of 100,000. Among 48 cases, 27 were operated cases.

So, the annual incidence of acquired cholesteatoma was calculated as 3.9 from operated cases, 6.8 from hospital cases and 10.0 from the total number of cases in a population of 100,000.

When we compare our results with previous reports, this value is higher than those reported before from only operated cases, however, not considerably higher. When we apply this value to the population of 8,000 in Hisayama, it would take ten years to find only five to eight cholesteatoma cases! This number is too low to elucidate the pathogenesis of cholesteatoma by a prospective cohort study in the town of Hisayama.

Then we started a case-control study on cholesteatoma. A case-control study can be carried out only by retrospective study and it cannot provide direct measurement of absolute risk. However, even if the incidence is rare, it is possible to carry out.

To perform the case-control study, we selected the cholesteatoma patients at Kyushu University Hospital since 2009. As control cases, we chose newly-diagnosed patients with other diseases than cholesteatoma in the same hospital. We carried out a questionnaire survey. The items of the questionnaire were divided into three categories: environmental factors; past history of otitis media; and past history of other diseases related to Eustachian-tube functions. In a case-control study, selection bias of cases and controls can be a problem. So we tested the profiles of both groups and found no significant difference in age and sex between cholesteatoma and control cases.

First, we asked about environmental factors. A history of smoking and drinking are general factors. A history of breast feeding and group nursing are factors believed to be connected with infantile recurrent otitis media. A family history of ear surgery is a factor for potential genetic factors. We found no statistically significant differences in these environmental factors.

Next, we asked about past history of otitis media. Interestingly, 73% of cholesteatoma patients have suffered from otitis media before, while only 26% of the control group have. Our results suggest that a past history of otitis media may contribute to the pathogenesis of cholesteatoma. Among those who have a history of otitis media, we asked what kind of otitis media they have suffered from, and compared cholesteatoma cases with controls. We expected more cholesteatoma cases than controls have suffered from OME, however, we did not find statistical evidence that cholesteatoma cases develop through OME.

We also expected that frequent recurrence of otitis media may increase the risk of cholesteatoma development. However, our results showed no significant difference in frequency of otitis media between cholesteatoma cases and controls.

Then we inquired about past history of other diseases related to Eustachian tube functions. We found no significant differences in adenoid diseases, GERD and nasal diseases that may cause Eustachian tube blockage. On the other hand, we found significant differences in patulous Eustachian tube-related items between cholesteatoma cases and controls.

Twenty-three percent of cholesteatoma cases complain of ‘hearing own breathing sounds’, while only 9% of the control cases do. Forty-three percent of the cholesteatoma cases have ‘habitual sniffing’ while only 23% of control cases do. ‘Hearing own breathing sounds’ directly indicates patulous Eustachian tube. But ‘habitual sniffing’ can also be caused by nasal symptoms. So we asked the patients with habitual sniffing why they sniff. ‘Ear fullness’ and ‘autophonia’ that could indicate patulous Eustachian tube were significantly high in the cholesteatoma cases. On the other hand, we did not see a difference in nasal symptoms. Our results suggest that patulous Eustachian tube is highly suspected as a cause of habitual sniffing in the cholesteatoma cases.

Based on our case-control study, the otitis media may contribute to the pathogenesis of cholesteatoma although the process is still unclear. Our results also suggest that patulous Eustachian tube may contribute to it.

In summary, our epidemiological study showed the annual incidence of cholesteatoma including non-operated cases was calculated as 6.8 to 10.0 in a population of 100,000 in Fukuoka City, Japan. And the results of our case-control study suggest that a past history of otitis media and patulous Eustachian tube may contribute to the pathogenesis of cholesteatoma.

THE ROLE OF THE MASTOID IN MIDDLE EAR PRESSURE REGULATION

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Introduction

The mastoid consists of a series of aerated cells lying mainly behind the middle ear (ME) cavity and the ear canal. These air cells are formed after birth by successive expansion from the antrum; the air cell system is fully developed by the age of 14 to 16 years.[1] Since the air cells are all interconnected as well as connected to the ME cavity by the antrum, these structures are all connected both in terms of structure and function referred to as the ME cleft.[2] The pressure of the ME cavity is identical to the mastoid, and thus, the mastoid is likely to participate in the overall regulation of ME pressure.[2]

The role of the mastoid in middle ear physiology is relatively unknown, which may partly be explained by its inaccessibility. However, during recent years more studies have reported more detailed descriptions of its structure, whereas functional studies are still few.

Mastoid structure

Studies on the mastoid structure based on 3D reconstruction of clinical CT scans have given valid estimates of its properties. A majority of these studies are limited by only determining the mastoid volume, while relatively few have undertaken the more complicated task of determining its surface area. Whereas the mastoid volume may be relevant for some of its functions, its surface area is important for the capacity of gas exchange across its mucosa.[2,3] While the ME cavity exhibits a surface area and a volume of 7.7 cm^2 and 0.7 cm^3, respectively, the mastoid has corresponding values of 85 cm^2 and 6 cm^3 in normal ears.[4] Thus, the mastoid surface area is much larger, and its area-volume-ratio is also larger than for the ME cavity (17 vs. 12 cm^{-1}).[4] However, the resolution of CT scans applied in a clinical context is limited to around 0.6 mm, so that smaller air cells may not be detected.

Micro-CT scanning has been applied for description of the human ME structures such as the ossicles,[5] but no data have been reported yet for mastoid surface area and volume. Most recently, however, micro-CT scans of temporal bones have revealed a distinct set of numerous micro-channels connecting the exterior surface of the mastoid directly with its underlying air cells (Fig. 1).[6] The dimension of these channels shows an average diameter of 150 μm, which coincides with a content of a venole and an arteriole. Based on the development of the mastoid from birth, its vascular supply would reasonably expand together with the air cells from the antrum;[1] thus, this set of micro-channels seems to suggest an additional and separate vascular supply for the mastoid, but its role remains unknown.

The histological structure of the mastoid is different from the ME cavity itself, so that the surface epithelium is lower with flat or cuboidal cells, and the underlying connective tissue is more loose with an ample superficial vascular network.[7] This results in a short diffusion distance as well as enabling a high perfusion; these properties, including its larger surface area, seem to indicate that the mastoid is adapted to gas exchange.[3,4,7]

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Edited by Haruo Takahashi

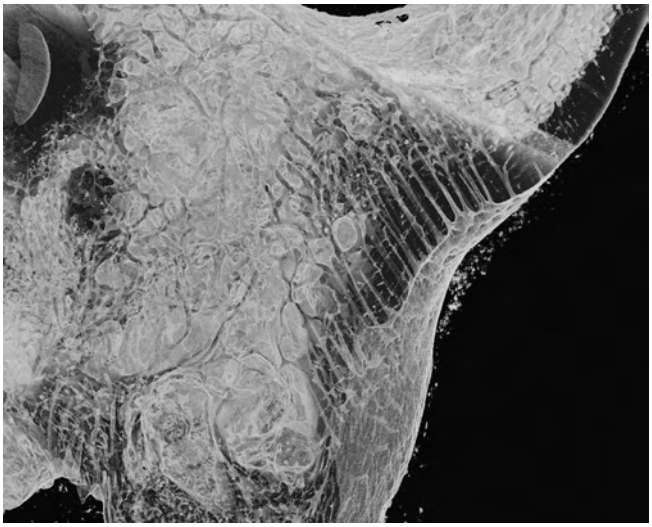

Fig. 1. Micro CT-scanning of a human temporal bone (tangential view of lateral surface; the mastoid tip is seen at the bottom). Volume rendering enhances the transitional voxels, *i.e.*, the surfaces between air and bone. Multiple micro-channels are seen connecting the surface of the bone directly with the underlying air cells; these micro-channels transverse the superficial compact bone (black area between surface and air cells).

Mastoid function – passive aspects

The larger part of the literature considers the functions of the mastoid to be merely passively related to its air volume. It has been suggested to act as a buffer for pressure changes or a gas reservoir for pressure regulation.[8,9] In addition, its cellular structure may result in damping of acoustic pressures and act as an acoustic filter decreasing noise and increasing hearing sensitivity.[10]

The pressure regulation of the ME cleft has traditionally focused on the function of the Eustachian tube and gas exchange between its air and the mucosal blood of the ME cleft.[11] In this context, its gas exchange has been considered passive, only driven by the differences in partial pressures of the gases.

Mastoid function – active aspects

Basically, the gas exchange depends on both the diffusion properties of the gases and the barrier as well as the perfusion of the mucosa.[11] Whereas the diffusion itself is a passive process, there is also evidence that gas exchange may be dependent on the mucosal perfusion.[12] Thus, changes in perfusion may be altered by vasomotor actions, which may alter the exchange of gases; this may ultimately alter the ME pressure. Altogether, this implies that mucosal perfusion may contribute in an active regulation of the pressure.

Recent clinical experiments on direct measurements on ME pressure have revealed two distinct patterns of systematic counter regulation of experimental pressure changes.[13] These pressure changes consist of *step-wise pressure changes* towards ambient pressures explained by Eustachian tube openings,[14] and *gradual pressure changes* explained by the mastoid found in both positive and negative directions.[13] These latter changes cannot be explained by gas exchange only, since the experiments were performed in awake subjects, where the gas exchange normally would result in gas absorption (negative direction), therefore, other mechanisms seem to be involved.

An alternate hypothesis has been proposed for ME cleft pressure regulation, where changes in the congestion of the mastoid mucosa result in changes in the pressure.[15,16] Diving mammals such as the hooded seal and the sea lion exposing their ME's to high pressures, when diving to high depths of water, seem to employ such a mechanism; their ME mucosa is rich in sinusoids or cavernous venoles, which are thought to fill with blood during diving.[16,17] Based on measurements of human mastoid surface areas and volumes,[3] it has been calculated that a small change in mucosal thickness of only 6 μm can alter the ME pressure by 1 kPa.[15] Therefore this mechanism seems very effective, and may contribute to pressure regulation also in humans. Since vasomotor

action may control the congestion, this process may also be active, but in essence it may also be passively driven by the pressure gradient.[16]

We have recently found support for the hypothesis of mucosal congestion playing a role in pressure regulation by histological examination of the mucosa from the lateral part of the mastoid in a normal human ear (Fig. 2). The specimen has been immuno-stained with CD-31 displaying the endothelial cells, and a collection of broad venous structures are seen, which appear similar to sinusoid. Moreover, the connective tissue is loose, which may allow for expansion of the mucosa. Finally, the surface epithelium is low with flat cells, as well as the distance to the underlying vascular structures is short.[7] Previous studies on the histological properties are few, and seem to be focused on the medial parts of the mastoid,[7] whereas the lateral parts may exhibit specialized vascular structures represented by sinusoids (Fig. 2). This has not been reported earlier.

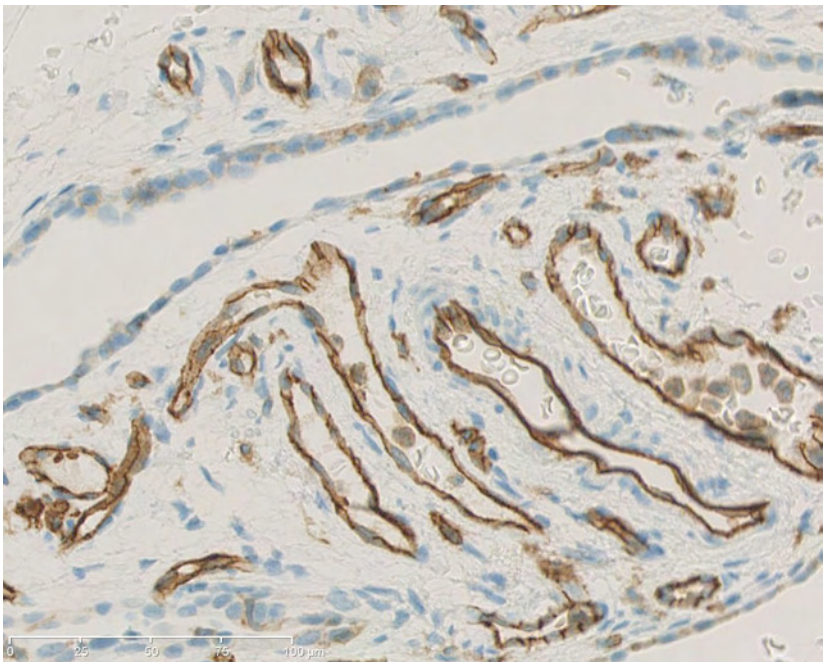

Fig. 2. Histological image of the mucosa at the lateral part of a normal mastoid (×40). The specimen has been immuno-stained with CD-31 coloring the endothelial cells with brown. The surface epithelial lining is thin with flat cells, while the subepithelial tissue contains a loose connective tissue with numerous broad venous structures.

Conclusions

The mastoid air cells are likely to participate in the overall regulation of ME pressure; passive buffering of the pressure may be obtained merely by its volume, but also gas exchange is facilitated by its structure with a large surface area together with the properties of its mucosa. In addition, congestion of the mucosa may play a role, and the distinct set of micro-channels at the lateral part of the mastoid implying an abundant separate blood supply together with a specialized vascular structures in the same area may represent an entity which seem important in ME pressure regulation. The systematic existence of such structures will have important functional implications for the role of the mastoid in normal and diseased ears.

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MIDDLE EAR PRESSURE NEURAL FEEDBACK CONTROL

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Introduction

The normal function of the middle ear (ME) depends on a pressure close to ambient pressure; however, impaired regulation of the ME pressure often leads to underpressure in diseased ears. Thus, underpressure is found in relation to clinical conditions such as retraction pocket of the tympanic membrane (TM), atelectasis, and formation of cholesteatomas.[1] Basically, this underpressure is explained by gas absorption from the ME-mastoid air to the mucosal blood which is insufficiently replenished due to a decreased air supply related to an impaired function of the Eustachian tube (ET).[2]

The overall regulation of the ME pressure is basically unknown. Whereas it may depend solely on local factors such as the gas exchange of the ME and the ET function, it has also been suggested that an overall neural feedback control exists similar to, for instance, respiratory control (Fig 1).[3] According to this hypothesis, mechano-receptors in the TM, the ME and/or the mastoid convey afferent neural information on the pressure status of the ME by the tympanic nerve (NIX). This information is transmitted to areas related to the respiratory centers of the brain stem nucleus of the solitary tract. These centers are also close and connected to the nucleus ambiguus and NV motor nuclei which control the opening of the ET. Thus, these brain stem centers obtain afferent information on the ME pressure from the NIX and exert their efferent activity via NV motor neurons activating the mm. levator palati and the mm. veli palatini. Therefore, opening of the ET can be elicited resulting in an equilibration of any differences between ME and ambient pressure.

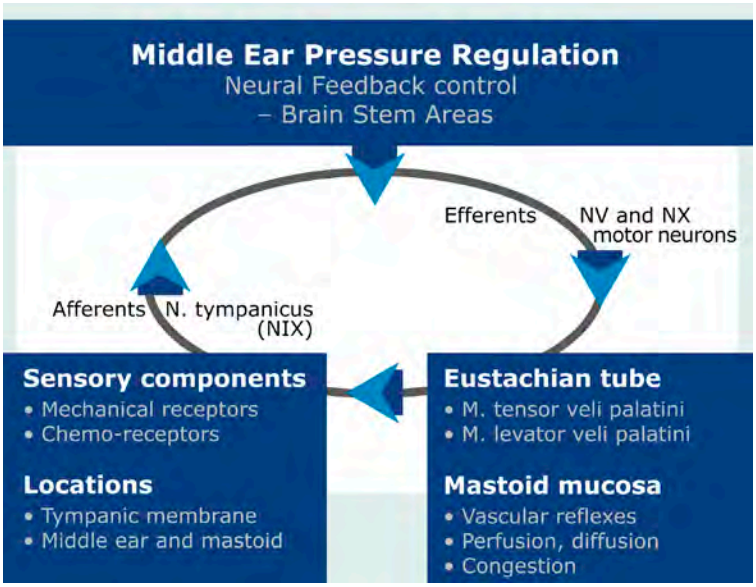

Fig. 1. Description of the overall mechanisms involved in neural feedback control of ME pressure.

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Edited by Haruo Takahashi

This hypothesis has initially been based on animal studies, and further documentation is still demanded. However, over time a series of both basic and clinical experiments have supported this idea. These studies contain both anatomical components as well as physiological evidence for an overall neural control of ME pressure.

Anatomical studies

The early evidence on neural feedback control of ME pressure came from histological studies in rabbits using horseradish peroxidase as a neural tracer. The tracer was applied in two places: 1) at the promontory of the ME around the tympanic plexus; and 2) at the ET muscles in the palate.[3] The subsequent findings of these tracers were: 1) in the brain stem at the nucleus of the solitary tract in areas of respiratory control; and 2) in the brain stem at the nucleus ambiguus and the NV motor nucleus.[3] Since the same brain stem areas are all connected and involved in respiratory control, it has been suggested that similar mechanisms function in ME pressure control.[3] Later, the same findings have been reported also for primates.[4]

The receptors for monitoring the pressure have been suggested to be baro- or stretch-receptors, but also glomus chemo-receptors; however, the latter are inconsistently found in human temporal bones.[5] Modified Vater-Pacinian corpuscles have been described in the pars tensa of the TM,[6] and similar corpuscles have been reported in the mucosa of the ME itself as well as in the mastoid in temporal bones.[7] The pars flaccida of the TM has been suggested as a logical site for pressure monitoring because of its higher elasticity,[8] and the region is rich in both myelinated and un-myelinated nerve fibers; however, no specialized nerve endings have been found here.[7]

In summary, the exact mechano-receptors playing a role in monitoring the pressure status of the ME have not been identified yet. However, the tympanic nerve innervates the mucosa of the entire areas of the ME including the tympanic plexus, the mastoid, and the ET, so that it is likely to convey any afferent information to the brain stem centers.[9]

Physiological studies

Important physiological studies have further supported the idea of a central neural feedback control for ME pressure regulation. Thus, electrical stimulation of the tympanic plexus has been demonstrated to elicit EMG recorded activity of the ET muscles with latencies of nine to 28 ms in primates.[10] The connections between the ME and the ET muscles have also been suggested after clinical experiments anesthetizing the TM by iontophoresis which resulted in a decreased ability of the ET to equilibrate deviating ME pressures.[11] Similar findings have been reported, where the anesthetics were applied directly at the promontory in subjects with a TM perforation or injected into the ME through a puncture of the TM.[12]

Clinical experiments with anesthesia of the TM lead to increasing thresholds of pressure sensation, when experimental pressures were applied to the ear canal; especially patients with pathological TM's showed increasing thresholds, so that it has been suggested that depletion of neural receptors in the TM may result in an impaired pressure regulation.[13]

Animal experiments with sectioning of the tympanic nerve have been followed by formation of retraction pockets in a series of rabbits; moreover, ME effusion was formed in more of these animals.[14] It was suggested that inactivation of the tympanic nerve resulted in a decreased aeration of the ME as well an impaired clearance function of the ET.[14]

Clinical neuro-physiological studies of evoked potentials with 3D recordings and source analysis from up to 128 electrodes have demonstrated distinct activation patterns of the brain stem related to static pressure stimulation of the TM in normal subjects.[15] These static activations of the brain stem were clearly different from similar evoked potentials related to acoustic pressure stimulation, so that separate neural pathways to the brain stem have been presented for static pressures.[15] In addition, activation of cerebellar centers has been demonstrated suggesting a coordination of related muscular activities.[15] Further, these experiments also included wavelet analysis, which reflects the frequency contents of neural activities; these frequency contents are specific for different neural systems. Static pressures mainly result in θ-band activity (0-4 Hz), whereas acoustic pressures result in α-band activity (7-10 Hz).[16] In conclusion, there are separate activation patterns related to static and acoustic pressure stimulations of the TM.

Finally, fMRI studies have demonstrated cortical activation in normal subjects in response to static pressure stimulation of the TM. This activation includes the post-central gyrus of the Brodmann area 43 which is related to control of the pharyngeal muscles. It has been suggested that these results represent a connection between the ME and activation of the ET muscles.[17]

Conclusions

More anatomical and physiological studies have produced evidence supporting the idea of an overall neural feedback control of ME pressure in humans. Considering the many similarities between these findings and respiratory control, it seems reasonable to conclude that ME pressure in humans is subjected to such a regulatory mechanism. In addition to the efferent activation of the ET, the mastoid may also take part in pressure regulation by vasomotor control of the perfusion and the congestion of its mucosa (Fig. 1).[18] Further research is needed to increase our basic knowledge, and especially studies which can link the efferent and afferent parts of such a feedback regulation.

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EVALUATION OF ENDOLYMPHATIC HYDROPS ON MAGNETIC RESONANCE IMAGING IN PATIENTS WITH OTOSCLEROSIS

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Introduction

In otosclerosis surgery, post-operative vertigo occasionally occurs though it is usually transient. Profound sensorineural hearing loss after otosclerosis surgery has been reported in about two percent of the patients.[1] The mechanism of the inner ear disturbance is unknown. Short distance between the stapes footplate and endolymphatic space may be associated with the post-operative vertigo or sensorineural hearing loss. We attempted to evaluate the distance between the stapes footplate and the endolymphatic space with the endolymphatic space size.

Methods

In four patients with otosclerosis, magnetic resonance imaging (MRI) was taken four hours after intravenous gadolinium (Gd) injection using a 3-tesla MR unit. We applied heavily T2-weighted 3D fluid attenuated inversion recovery (3D-FLAIR),[2,3] 3D-inversion recovery sequence with real reconstruction (3D-real IR),[4] positive endolymphatic imaging (PEI)[5] and HYbriD of Reversed image Of Positive endolymph signal and native image of positive perilymph Signal (HYDROPS).[6]

Heavily T2-weighted 3D-FLAIR after intravenous standard-dose Gd administration was reported to achieve the visualization of endolymphatic hydrops.[2,3] On 3D-FLAIR, perilymph with Gd distribution shows a high signal and endolymph without Gd distribution shows a low signal similar to that of surrounding bone. The sensitivity of this technique to low Gd concentration is better than that of the 3D-real IR.[7] However, the 3D-real IR at 3 tesla makes it possible to separately visualize endolymph, perilymph and bone using a single-pulse sequence.[4] In PEI, optimal shortening of the inversion time in 3D-FLAIR suppressed the signal of the Gd-containing perilymph and instead gave a high signal for the endolymph that contains no Gd.[5] HYDROPS images allowed a 3D-real IR-like image presentation even after intravenous standard-dose Gd administration.[6]

Results

Endolymphatic hydrops was frequently observed in both the cochlea and vestibule. The distance between the stapes footplate and the endolymphatic space in the vestibule could be evaluated in all patients.

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Edited by Haruo Takahashi

An example of MRI is shown in Figure 1 A-C. In this patient, endolymphatic hydrops was observed in both the cochlea and vestibule on the right side.

The endolymphatic space size in the right ear was clearly larger than that in the left ear. The hearing level was not different between the right and left ears. Stapedotomy was performed on the left side in which the distance between the stapes footplate and the endolymphatic space was larger than that in the other side.

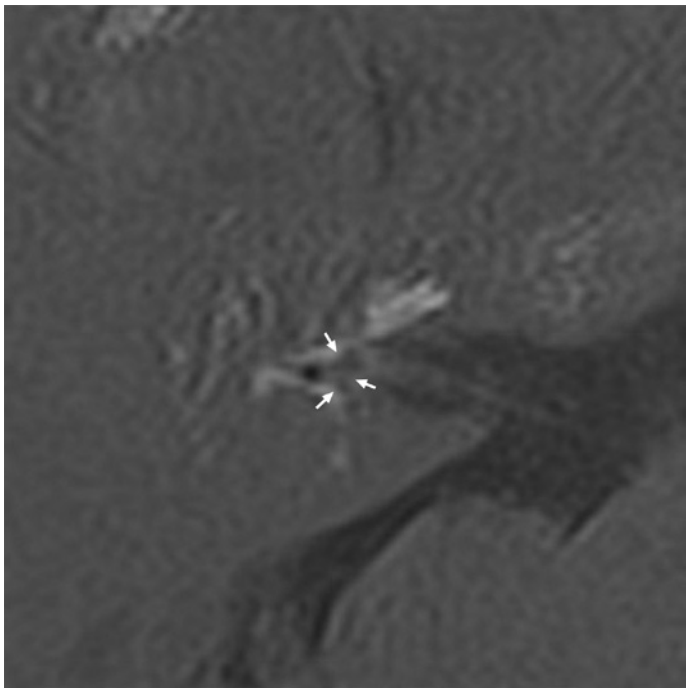

Fig. 1A. Endolymphatic hydrops in the right vestibule (arrows) in a 48-year-old female with otosclerosis (3D real IR).

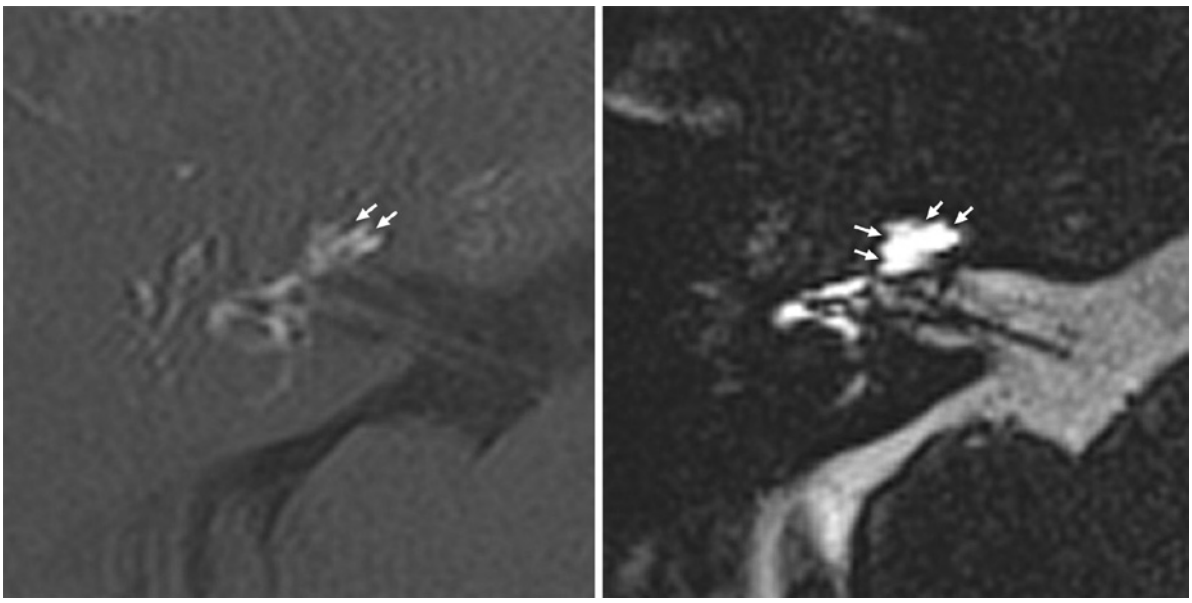

Fig. 1B. Endolymphatic hydrops in the right cochlea (arrows) in a 48-year-old female with otosclerosis (left side image; 3D real IR, right side image; heavily T2-weighted 3D FLAIR).

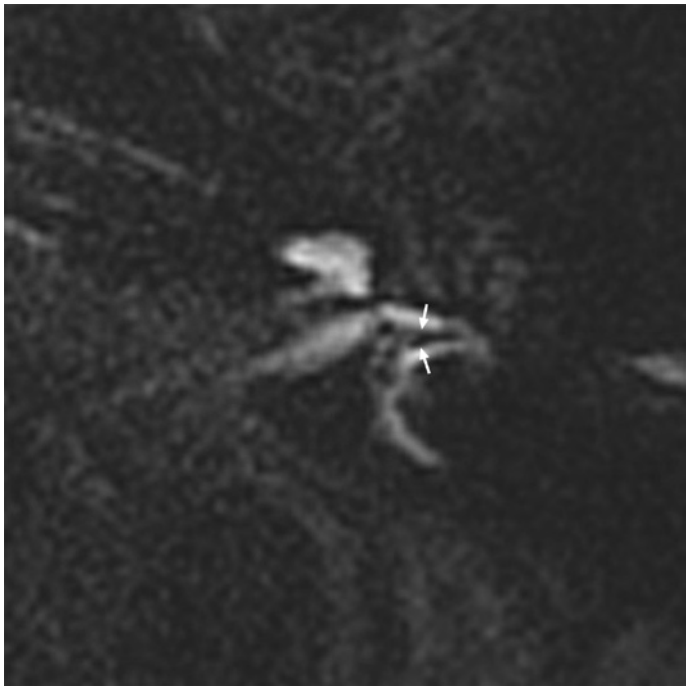

Fig. 1C. Endolymphatic space in the left vestibule (arrows) in a 48-year-old female. The space size is smaller than that in the right vestibule shown in Fig. 1A.

Discussion

Endolymphatic hydrops can be visualized by MRI after intra-tympanic or intravenous Gd injection. In general, the intra-tympanic method has higher enhancement of the perilymph and better ability to predict intratympanic drug transition to the inner ear. The intravenous method is less invasive and enables stable bilateral observation of the condition of the cochlea.

We reported many cases with asymptomatic endolymphatic hydrops using advanced techniques of MRI.[8] Although the mechanism is not clear, endolymphatic hydrops was often recognized in temporal bone specimens in patients with otosclerosis.[9,10] Accordingly, it was felt that evaluation of endolymphatic space size with use of MRI is important even if the patients had no sensorineural hearing loss.

It has been postulated that otosclerosis may produce vertigo by several mechanisms. One mechanism is by causing endolymphatic hydrops.[9,10] To investigate the anatomic relationship between utricle, saccule, and stapes footplate, and adapt the stapes prosthesis tip to reduce post-operative vertigo, temporal bones were serially sectioned and stained, and the distance from the inner lining of stapes footplate to saccule and utricle, respectively, were measured.[11] Based on these measurements, the Fisch prosthesis was modified with a slope of 45 degrees at its tip to adapt to the anatomic configuration of the vestibule. Post-operative vertigo was significantly reduced when the modified prosthesis was inserted.[11]

Endolymphatic hydrops may be associated with pre-operative and post-operative vertigo in patients with otosclerosis. There is a possibility that the distance between the stapes footplate and the vestibular endolymphatic space is also related to the occurrence of post-operative sensorineural hearing loss. Because the endolymphatic space imaging is now possible after intravenous standard-dose Gd administration, the imaging is expected to contribute to elucidation of the mechanism of pre- and post-operative sensorineural hearing loss and vertigo.

Conclusions

It is now possible to evaluate endolymphatic space size in patients with otosclerosis after intravenous standard-dose Gd injection using 3-tesla MRI. The MRI may contribute to prevention and treatment of the inner ear disorders in patients with otosclerosis.

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CHOLESTEATOMA GROWTH AND PROLIFERATION: EXPRESSION OF HGF (HEPATOCYTE GROWTH FACTOR) AND ITS HIGH-AFFINITY RECEPTOR C-MET

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Introduction

Cholesteatoma is epidermally-derived temporal bone lesion that is locally destructive and frequently recurrent. There are no medical therapies for cholesteatoma and currently the only available treatment is surgical resection. Despite many studies, the molecular events governing cholesteatoma formation are not well established.[1] It is clear that when squamous epithelium lining the external ear canal (EAC) or the tympanic membrane (TM) becomes trapped into the middle ear (ME)(retraction pockets), a process of hyper-proliferation ensues. Cholesteatoma is actually a hyper-proliferative disease in which keratinocytes (KC) at all levels of the epithelium show both increased proliferation rates and decreased rates of programmed cell death (apoptosis).[2,3]

The application of molecular biology to clinical investigation of cholesteatoma should bring us to a new understanding of this disease with implied benefits of new therapies.

Materials and methods

Total RNA (tRNA) was extracted from cholesteatoma and control EAC skin for each patient (n = 10). Then cDNAs derived from tRNA were hybridized with GeneChip Human Genome U133 Plus 2.0 Array for both tissues. The data were analyzed with GeneSpring GX, and both up-regulated (three times higher than the control) and down-regulated (1/3 lower than the control) genes expressed in cholesteatoma were determined.

Results

DNA microarray analyses indicated that both HGF and C-MET were up-regulated by five to ten times in all cholesteatomas (n = 10). Real-time PCR analysis confirmed that mRNAs of both HGF and C-MET were up-regulated by five to15 times in all cholesteatomas.

Immunohistochemical studies indicated that HGF was predominantly expressed in infiltrated monocytes and macrophages just beneath the cholesteatoma epithelium, while C-MET was expressed in the entire cholesteatoma epithelium (Fig. 1A,B).

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Edited by Haruo Takahashi

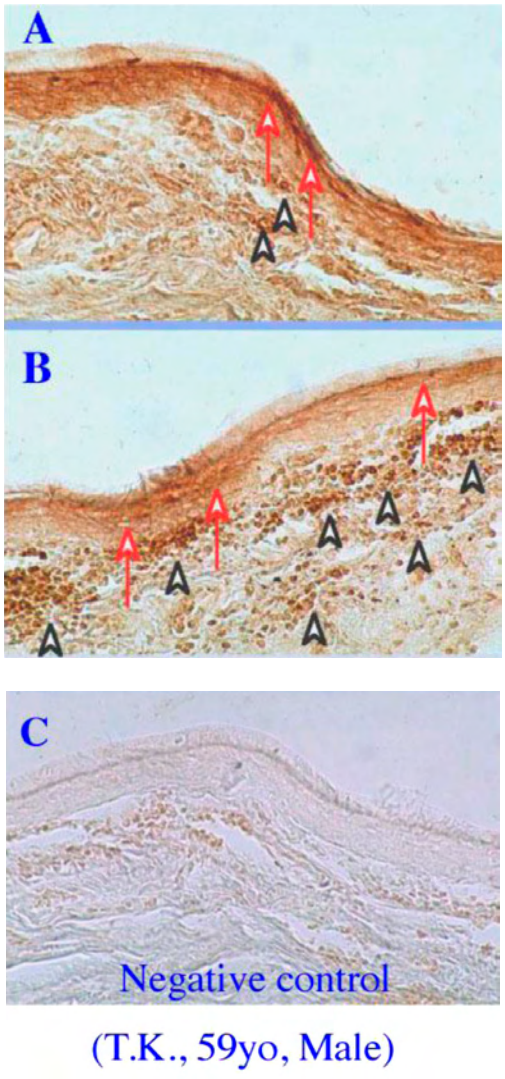

Fig. 1. Expressions of HGF & C-MET in cholesteatoma tissue. A: C-MET expression in the epithelium (red arrows) and sub-epithelial layers (black arrow heads); B: HGF expressions in the cholesteatoma epithelium (red arrows) and sub-epithelial layers (black arrow heads); C: negative control.

Discussion

HGF was first identified as a potent mitogen for fully differentiated hapatocyte, and it is currently known to have multiple biological activities on a wide variety of cells via its binding to C-MET receptor.[4] HGF has been shown to have critical roles in both KC proliferation and apoptosis. HGF may stimulate phosphatidylinositol 3-kinase (PI3K), leading activation of Akt. HGF may protect KC against apoptosis by maintaining of levels of Bcl-2 and Bcl-XL and by activating Akt. HGF could promote KCs proliferation and prevent KCs apoptosis, resulting in abnormal KCs growth.[2] The present study suggests that HGF might induce KC proliferation in cholesteatoma epithelium through up-regulations of both HGF and C-MET in autocrine as well as paracrine manner (Fig. 2A).

The discovery of NK4 as an HGF-antagonist has promoted research in cancer biology, pathology and therapy. Growing evidence indicates that NK4 inhibits tumor growth, invasion and metastasis in animal models of various types of malignant tumors. Several approaches to inhibit HGF-C-MET signaling are now being developed as an anti-cancer agent. In future, the relatively easy access of the ME to intra-tympanic drug delivery makes HGF/C-MET-related RNA/DNA-based topical inhibitors of cholesteatoma an attractive mode of primary or adjunctive medical therapy (Fig. 2B).

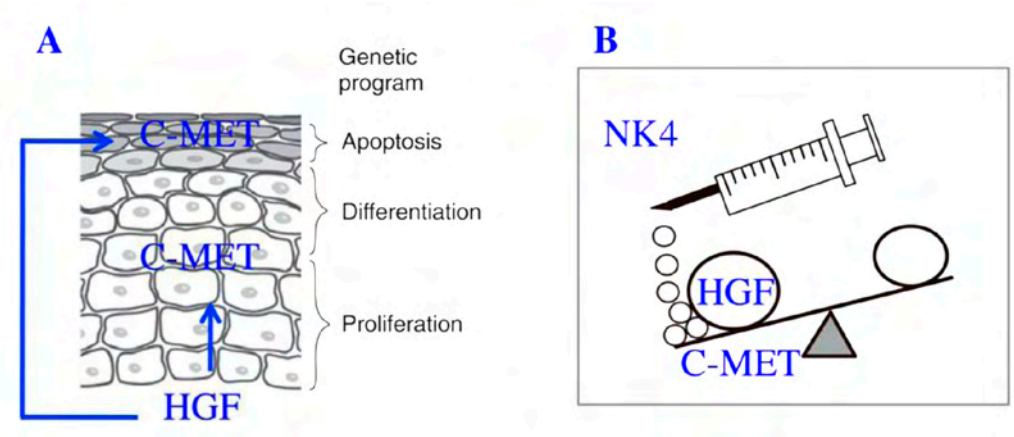

Fig. 2. Keratinocytes hyperproliferation and HGF. A: Autocrine and paracrine actions of HGF; B: Development of a HGF/C-MET-related new medical therapy.

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REGULATING OSTEOCLASTS FOR THE MAINTENANCE OF AUDITORY OSSICULAR MORPHOLOGY, THE MIDDLE EAR AND HEARING

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Abstract

Currently little is known about whether and how osteoclasts play a role in the vibration of auditory ossicles. This review focuses on morphological change of auditory ossicles and hearing function when osteoclasts increase or decrease.

Prevention of bone resorption may be implicated with clinical application of several middle-ear diseases.

Introduction

Bone remodeling may be important for auditory ossicular structure. Osteoclasts are specialized multinuclear macrophages that secrete HCL and hydrolases causing bone resorption. Osteoclasts are upregulated after inflammation, and ossicles are eroded by repeated otitis media. Therefore, regulating of the number of osteoclasts is critical for maintaining the shape and size of auditory ossicles. For bone remodeling to occur, receptor activator of nuclear factor-kB (NF-kB) ligand (RANKL) must bind with its receptor (RANK), located on osteoclasts. The potentially continuous bone loss is mitigated by the decoy receptor osteoprotegerin (OPG) which competitively binds RANKL and blocks the interaction of RANKL-RANK. The osteoclastic bone resorption in adults is balanced by osteoblastic bone formation through 'coupling' mechanisms, which maintain bone integrity (bone remodeling)(Fig.1). [1]

We reviewed the role of osteoclasts in the maintenance of the ossicular structure and how hearing function is impaired in knock-out mice with increased or decreased osteoclast numbers.

Auditory ossicles in osteopetrotic mice

Conductive and/or sensorineural hearing loss is frequently seen in humans with ostepetrosis.[2-5] We analyzed hearing function and the morphology of the auditory ossicles in osteopetrotic mice, which lack osteoclasts due to either a c-Fos or RANKL deficiency. The auditory brainstem response showed that mice of both genotypes had hearing loss, and that the mobility of the malleus was dramatically reduced as revealed by laser Doppler vibrometry.[6] Although involvement of the nervous system cannot be excluded, impaired vibration of the malleus is the most plausible explanation for the hearing loss of osteopetrotic mice.[6] The impaired vibration was apparently due to contact of the malleus with the promontory, the reduced volume of the tympanic cavity and the increased volume of auditory ossicles in comparion to control animals.[6]

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Cholesteatoma and Ear Surgery – An Update, pp. 33-35
Edited by Haruo Takahashi

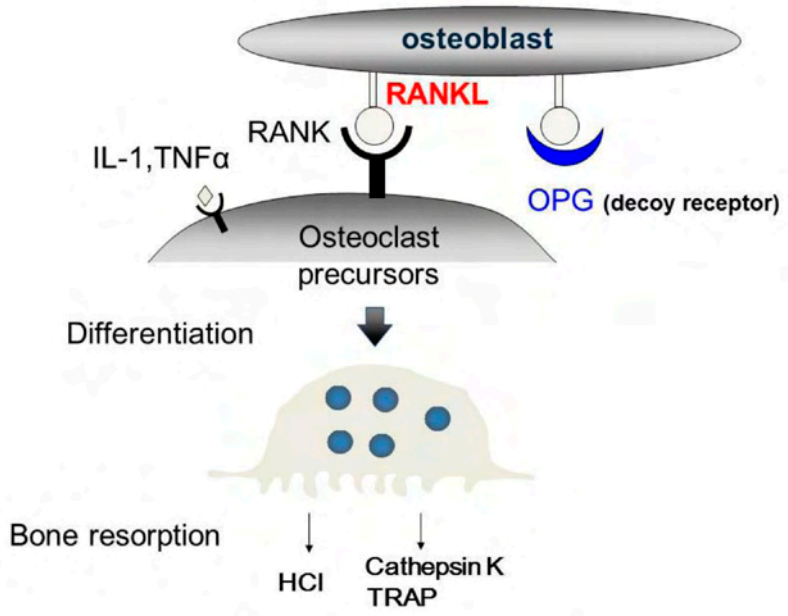

Fig. 1. Summary of osteoclast differentiation and bone resorption – RANK-RANKL lignad Osteoprotegerin osteoclast differentiation. RANKL: receptor activator of nuclear factor-kappa B ligand. Osteoclasts differentiate from precursors of the monocyte macrophage lineage in the presence of the cytokine RANKL.

Excessive numbers of osteoclasts in ossicles

However, too much of a 'good thing' also leads to hearing loss. The shape of ossicles and hearing were also degraded when the numbers of osteoclasts were increased. Mice lacking OPG (*Opg*-/-mice), also known as a model for juvenile Paget's disease, exhibited excessive numbers of osteoclasts resulting in abnormal bone remodeling of the otic capsule.[7,8] Auditory ossicles in *Opg*-/-mice are massively resorbed by the abundant osteoclasts which may also result in impaired hearing function.[7,8] In *Opg*-/-mice, the ligament at the junction of the stapes and the otic capsule is lost by bony ankylosis (fusion).[7,8]

Osteoclasts in cholesteatoma

We counted the number of Cathepsin K positive cells which are markers of osteoclasts. There are significant differences of the number of cathepsin K cells between normal people and patients with cholesteatoma.

Bisphosphonate therapy in otosclerosis

Opg -/-mice were intra-peritonally injected with risedronate, one of the widely-used bisphosphonates, for five days/week over nine weeks. The treatment significantly inhibited bone loss in auditory ossicles as well as in long bones of *Opg*-/-mice compared to untreated control mice.[9] Thinning of malleus handle and bony fusion of the junction between the stapes and the otic capsule were reduced by the treatment. In addition, hearing loss in *Opg*-/-mice was significantly reduced by risedronate treatment.[9]

The clinical controlled trial is ongoing. We investigate whether bisphophonate can prevent progressive hearing loss in Pts with otosclerosis or not.

Conclusion

We have shown that both osteopetrosis and osteoporosis impacts the structure of the middle-ear ossicles and impairs hearing function. The modification of the ossicles provides an explanation for the impaired vibration

of auditory ossicles seen in both osteoporotic and osteopetrotic mouse models. Anti-bone-resorptive therapy can prevent osteoclastic bone resorption of ossicles and progressive hearing loss. The number of Cathepsin K positive cells and/or osteoclasts are significantly increased in the ossicles in cases of cholesteatoma. The anti-bone-resorptive medication may prevent bone resorption of cholesteatoma.

Acknowledgements

This work was supported by Grants-in-Aid for Young Scientists B (17791198, 21791643 to S.K.), for Scientific Research C (23592495 to S.K.), and Grants-in-Aid for Scientific Research B (17390420 and 21390425 to K.M.) from JSPS; and a Keio University Special Grant-in-Aid for Innovative Collaborative Research Projects.

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SURGICAL TREATMENTS FOR PATULOUS EUSTACHIAN TUBE: AUTOLOGOUS FAT GRAFTING AND ARTIFICIAL EUSTACHIAN TUBE

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Introduction

The origins of patulous Eustachian tube (PET) have not been fully elucidated. However, more than half of the patients with PET have experienced weight loss, and their CT imaging studies have shown a depletion of the soft tissue (*i.e.*, fat tissue), surrounding the medial two-thirds of the cartilaginous portion in the Eustachian tube (ET). We had developed autologous fat grafting (AFG) techniques for the refractory PET and have been using them since 1999. In this study, we presented the methods of using our self-developed specialized needle instrument for the injection of fat into the sub-mucosal tissue surrounding the ET from the nasopharyngeal orifice. Moreover, we have developed an artificial Eustachian tube (AET) which is able to alleviate various types of tubal dysfunctions such as tubal stenosis and PET. The AET can restore the ventilatory and drainage functions of the ET. The AET can be inserted into the cartilaginous ET via the bony ET, through the tympanic membrane, or directly into the ET in the case of a tympanoplasty with a guide wire.

In the present study, efficacies of surgical treatment using AFG were evaluated, and the AET operation was added in some patients who had a low response to AFG.

Method

Fourteen cases (20 ears) of refractory PET, who complainwed of ear symptoms like ear fullness and autophony, were evaluated in this study. Diagnosis of PET was made with the ET functioning tests, which includes sonotubometry, and the tubo-tympanum-aerodynamogram (TTAG) with a Valsalva maneuver developed by Kumazawa *et al*. The effects of both treatments with AFG and AET on PET were also evaluated by using these ET functioning tests. Figure 1 shows the intra-tubal insertion point of fat on the orifice of the right ET. As shown in the right photo, injection from the intra-ET and into the infra-mucous membrane is in an upper-lateral direction.

We developed a specialized two-prong needle for autologous fat grafting techniques in order to improve the methods of injection over the currently used single needle. The top of the specialized needle is divided into two branches, one, a needle for fat injection, the other, a dichotomic stopper. A needle with a dichotomic stopper is an instrument for the injection of fat into the sub-mucosal tissue surrounding the ET at approximately a ten-mm distance from the nasopharyngeal orifice. This new type of needle with a dichotomic stopper is especially adapted for the injection of fatty tissue, from the intra-ET into the infra-mucous membrane of the ET cavity. The stopper helps to allow for a precise placement of the needle at the ideal injection point. The needle for the operation of AFG was inserted into the infra-mucous membrane from the intra-tubal cavity.

The AET was added to the AFG operation in some of those patients when the operation did not effectuate a ‘more than slightly effective’ result for the patients with refractory PET. The AET consists of a tubular body,

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Edited by Haruo Takahashi

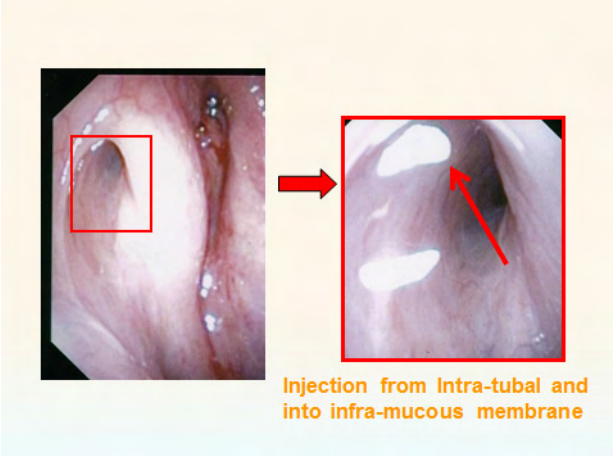

Fig. 1. Intra-tubal insertion point of fat on the orifice of the right ET.

and is made from polyurethane. The proximal end can be placed protruding out of the tympanic membrane, or just beneath, while the distal end is inserted into the cartilaginous ET passing through the isthmus.

An AET was inserted into the ET through the incised tympanic membrane, as shown in Figure 2. The AET, equipped with an intra-tubal guide wire, could be inserted firmly along the curve of the bony ET. The distal end of the AET has micro-holes in the side near its tip facing the inside of the cartilaginous ET for ventilation.

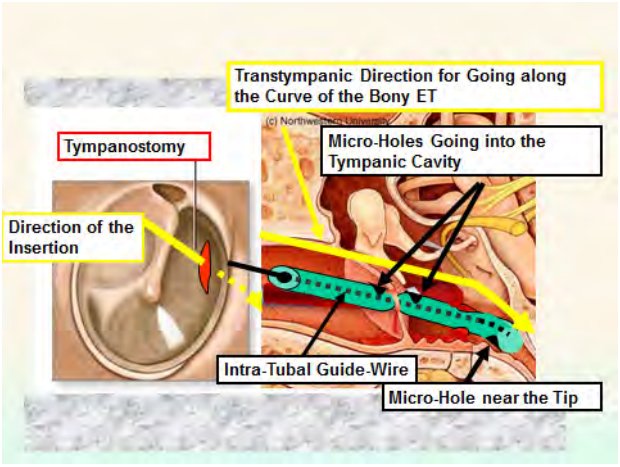

Fig. 2. Schematic illustration of AET: insertion of AET with a guide wire into the ET through the incised tympanic membrane.

Results

The effects of the AFG using the specialized needle were evaluated more than one year after the operation in 14 cases (19 ears). Effects classed as moderate or exceptional were observed in 74% of the ears including those receiving the operation more than once. Five ears (71%) improved after additional grafts, usually one operation, but up to three in certain cases, if the AFG effects were not evident after the initial graft.

The AET operation was added to the AFG operation in three cases (three ears) when the AFG operation did not effectuate a ‘more than slightly effective’ result for these patients with refractory PET. The AET operation in all three ears that did not get more than moderately effective by the AFG operation, was moderately or more than moderately effective. The combined results of the AFG and AFT operation had quite a high rate of effectiveness on PET patients. Total effects were 90%, which were moderately and remarkably effective as shown in Figure 3.

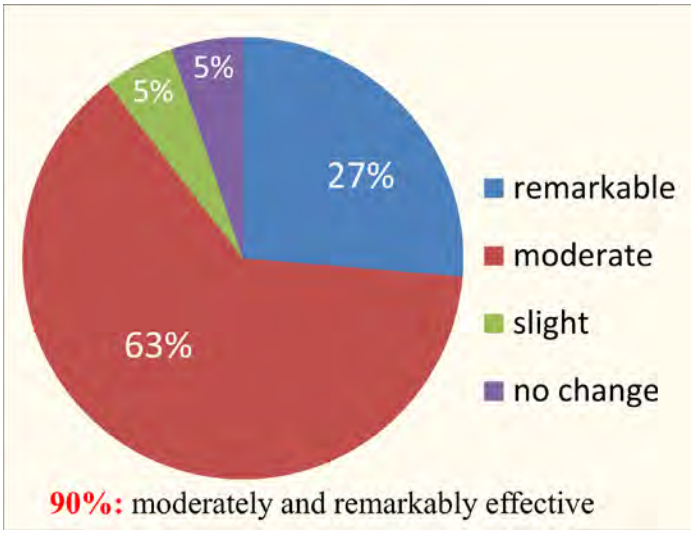

Fig. 3. Combined results of AFG and additional AET operation.

Possible complications in this operation, like an impairment to opening the mouth, was not found at all in any of the 19 ears using our specialized needle, though some impairment, like temporal sensory weakness of the submandibular portion, was found in five out of 36 ears operated on using the current method of a single needle, in our past study. There was no complication, like diplopia, concerned with the central nerve associated with any of the ears treated with our specialized needle.

Discussion

Patients with severe PET, who have had ear symptoms more than half a day, usually tend to be very difficult to treat with any types of medication. We presented minimally invasive methods for the treatment of refractory PET that involves AFG into the ET lumen from the nasopharyngeal orifice, in combination with or without an AET from the middle ear.

Topical medical therapy, such as boric acid and salicylic acid powders, for treatment of PET is estimated to increase congestion of the nasopharyngeal ET orifice.[1,2] However, most of these topical medications did not achieve remarkable relief in patients with severe PET, and tended to lose their effectiveness within several days, and needed to be administered repeatedly even in patients with mild PET. Therefore, we have developed a surgical procedure for the treatment of PET, and the effects of AFG and AET operations on refractory PET have been evaluated. Autologous fat grafting for the refractory PET had been reported by Doherty *et al*.[3] They did not only treat with fat grafting, but also with other treatments of fat plugging, or cauterization the entire circumference of the ET orifice.

In other studies on surgical treatment, trans-tympanic insertion of a new silicone plug, developed by Kobayashi, seems to be useful for controlling a chronic PET.[4]

Poe reported that Endoluminal patulous ET reconstruction was performed by using the techniques with autologous cartilage graft.[5]

The effects of an AFG operation has only been reported of a few cases, probably because its injection point is very difficult to identify in an AFG operation, and for the placement and fixing at the adequate injection point, specialized skills are needed with the current method of a single needle.

We have found two factors that affect the outcome of the AFG operation. The first factor is age. In patients over 50 the outcomes were not as positive as those in patient under 50. Only three out of the13 ears (23%) of patients of 50 years and older showed either remarkable or moderate outcomes.

The second factor is BMI. Severe emaciation makes extraction of periumbilical adipose tissue very difficult, especially in males.

We conclude that we need to get much more clinical research for the expansion of these two types of treatment, AFG and AET.

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LONG-TERM EUSTACHIAN TUBE DYSFUNCTION IN POST-RADIOTHERAPY NASOPHARYNGEAL CARCINOMA PATIENTS

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Introduction

Chronic middle ear effusion (MEE) is a common sequela of nasopharyngeal carcinoma (NPC) patients who have been treated with radical radiotherapy (RT). Although knowledge of how MEE develops in these patients is important in determining appropriate treatment options, its pathogenesis is still not well understood. This study aimed to find out if there was a correlation between obliteration of the Eustachian tube (ET) and the development of MEE in long-term post-irradiated NPC patients.

Methods

NPC patients who have been successfully treated by RT for at least five years were prospectively studied. Flexible nasal endoscopy focusing on visualization of the opening of the Eustachian tubal orifice was carried out, to see if the tubal lumen was obliterated. Microscopic examination of the tympanic membrane was also performed in each patient. Findings of MEE were confirmed by pure-tone audiometry and tympanometry.

Results

Of the 23 patients studied, 45 ears were available for evaluation. One of the patients had undergone surgery with blind-sac closure in one ear for cerebrospinal leak post-RT; hence only 45 ears were assessed for this study. Thirty-four were noted to have obliterated Eustachian tubes, and there was a tendency for effusion to develop in ears with obliteration of the Eustachian tube (Table 1)($p < 0.0001$, chi-squared test).

Table 1. The relationship between obliteration of the Eustachian tube and the presence of ipsilateral middle ear effusion.*

	Effusion present	*Effusion absent*	*Total*
Obliterated ET	33	1	34
Patent ET	4	7	11
Total	37	8	45

*ET- Eustachian tube; Two-tailed P value < 0.0001.

Discussion

MEE is a common clinical condition and its development has been attributed to various mechanisms. Although physical obstruction of the Eustachian tubal lumen has been the most popular, alternative mechanisms have

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Edited by Haruo Takahashi

been proposed. Bluestone suggested altered cartilage compliance could impact tubal function and lead to effusion.[1] The same mechanism applies in Down's syndrome, which has defective cartilage formation.[2] In cleft palate, involvement of the muscles relating to the Eustachian tube leads to tubal dysfunction and effusion.[2]

We studied the existence of MEE in NPC patients who have received radiotherapy for a minimum period of five years. Of the ears studied, 34 out of 45 ears (73.3%) were found to have obliteration of the tubal lumen. Not uncommonly, there was associated distortion or even absence of Eustachian cushion (Fig 1). These observations could be attributed to tumor destruction of tubal cartilage and/or post-irradiation changes. MEE was noted to be present in almost all the ears with associated obliterated Eustachian tubes, suggesting that physical obstruction of the Eustachian tube could be a key mechanism in the development of MEE in long-term post-radiotherapy NPC patients.

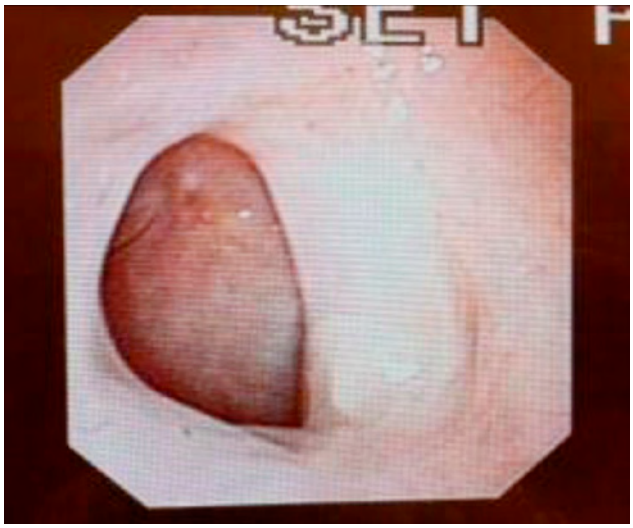

Fig. 1. Endoscopic image of an obliterated left Eustachian tube with destruction of the cartilaginous cushion.

However, it has been argued that mere anatomical obstruction of the Eustachian tube does not always culminate in effusion.[3] In fact, the 'hydrops-ex-vacuo' theory of attributing MEE to the highly negative middle-ear pressure formed as a result of continuous gaseous absorption in a closed biological space has largely been discredited.[4] In the development of MEE in long-term post-irradiated NPC patients therefore, besides obliteration of the Eustachian tube, other factors may play a role as well. These include the detrimental effects of radiation on mucosal function as radiation could cause irreversible ciliary loss, intra and inter cellular vacuolation and ciliary dysmorphism.[5]

As long-term post-radiotherapy NPC patients are likely to have chronic MEE because of irreversible Eustachian tube dysfunction from multiple etiologies, one should exercise caution in recommending ventilation tube insertion as this may lead to chronic or recurrent troublesome otorrhea.[6] These patients should be appropriately counseled, as hearing amplification devices may be preferred alternative treatment options.[7]

Conclusion

Eustachian tubal cartilage destruction and obliteration is an important factor in the development of MEE in long-term post-radiotherapy NPC patients. Together with other factors such as radiation-induced impairment of the mucosal function, these patients develop irreversible Eustachian tube dysfunction, which has implications in management of the resulting middle-ear effusion.

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PANEL DISCUSSIONS

TYMPANOPLASTY WITH SOFT POSTERIOR MEATAL WALL RECONSTRUCTION: CHANGING THE WAY OF THINKING FOR PREVENTION OF RETRACTION POCKET RECURRENCE

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Introduction

There are a few reports of soft posterior wall reconstruction, but no report of tympanoplasty with soft wall reconstruction to prevent post-surgical retraction pocket formation before our short report.[1] Smith *et al*.[2] reported reconstruction using a Palva flap and a large piece of fascia, totally obliterating the middle ear and mastoid cavity with Gelfoam. Their purpose was not to prevent post-operative retraction pocket formation, but to simplify reconstruction. We changed our way of thinking and tried to make a soft posterior meatal wall without the aim of fortification but with the aim of preventing post-operative retraction pocket formation.[1,3]

With our technique, instead of a retraction pocket formation, a balloon-like retraction may occur on the soft posterior meatal wall when there is aeration disturbance of the middle ear. The purpose of this study was to assess the effectiveness of our surgical method for the prevention of retraction pocket formation and to study long-term post-operative results pertaining to reconstructed soft posterior meatal wall.

Surgical procedures

After eradication of the lesion by means of canal wall down technique, every effort is made to leave the posterior meatal skin as intact as possible in order to preserve the original cylindrical shape of the external auditory canal. Myringoplasty is performed using one end of the fascia sheet, and the posterior wall is reconstructed by gluing the other end of the fascia sheet to the reverse side of the peeled-off posterior meatal wall skin with fibrin adhesive[1,3] (Fig. 1).

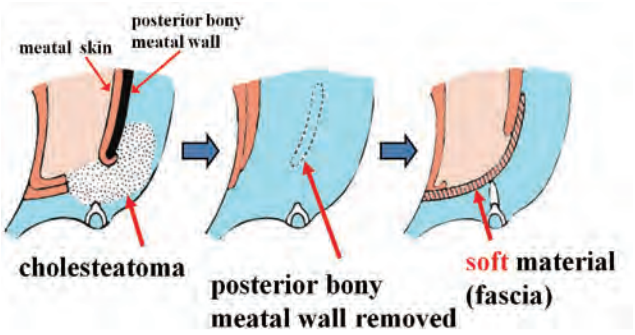

Fig. 1. Surgical procedures. Following the elevation of the meatal skin from the posterior bony wall, the bony wall is removed. This is followed by eradication of the lesion in the middle ear. The posterior meatal skin should be as intact as possible in order to keep original cylindrical shape of the canal. Myringoplasty is performed using one end of a fascia sheet, and the posterior meatal wall is reconstructed by gluing the other end of this fascia sheet to the reverse side of the peeled-off posterior meatal skin using fibrin adhesive.

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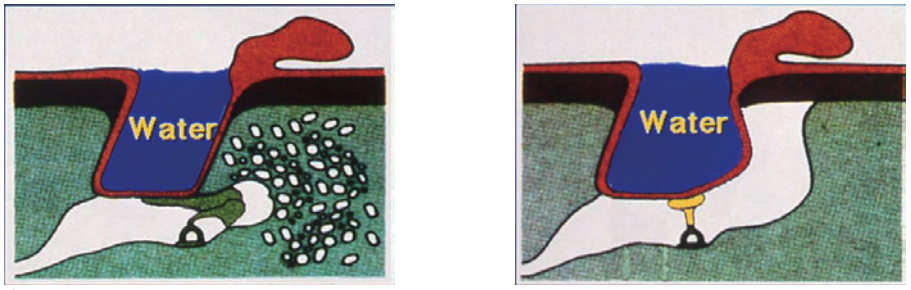
Subjects and methods

Subjects of this study were 322 patients with unilateral cholesteatoma who were treated using this method of tympanoplasty between October 1989 and February 2011 and followed up more than fifteen months.

Observation was to determine whether or not the post-operative retractions in narrow-neck pockets occurred and what kind of changes occurred on the reconstructed soft posterior meatal wall. Since we observed that a balloon-like retraction occurred in some patients, the degree of retraction on the soft posterior meatal wall in each case was investigated by calculating the ratio of the volume of the post-operative external auditory canal and the canal on the normal side. The volume was measured by pouring water into the external auditory canal (Fig. 2A,B).



A B

Fig. 2. Method of calculating the degree of retraction. Scheme for explaining the method of calculating the ratio of the volume of the post-operative external auditory canal and canal of the normal side. Volume is measured by pouring water into the external auditory canal. A: normal side; B: post-operative balloon-like retraction.

Results

None of the patients manifested retraction pocket formation, and whenever changes occurred on the soft posterior meatal wall, a balloon-like retraction was observed (Fig. 3A,B).

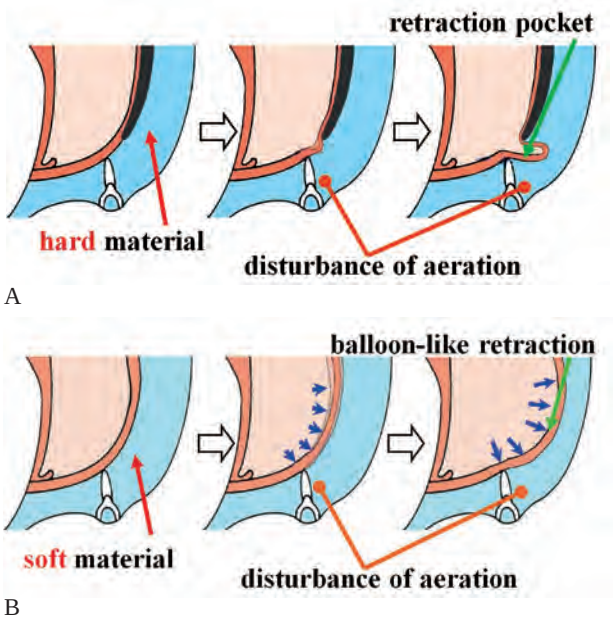


Fig. 3. Postoperative retraction due to postoperative middle ear aeration disturbance. A Hard wall: When the hard material is used for the posterior meatal wall reconstruction, negative pressure acts to form the retraction pocket which would lead cholesetatoma to recur. B Soft wall: When the wall is reconstructed with a soft material, negative pressure acts on the entire posterior meatal wall and pushes back the whole wall. Retraction cholesteatoma may not recur in such a balloon-like retraction wall.

Judging from the change in the ratio of the volume over time, retraction of the posterior meatal wall appears to occur mainly within one year of surgery.

Figure 4 shows the relationship between the degree of retraction of the posterior meatal wall at the final observation of each patient and the number of ears affected. Regarding the X axis, the larger the ratio of volume, the larger the degree of retraction. 'The ratio of the volume is 1.0' means no retraction. The soft posterior meatal wall was maintained in a normal position in 27 ears in which the ratio of the volume was 1.0. The remaining ears had various degrees of balloon-like retraction. However, in the majority of these cases, the degree of retraction was within the ratio of 2.0.

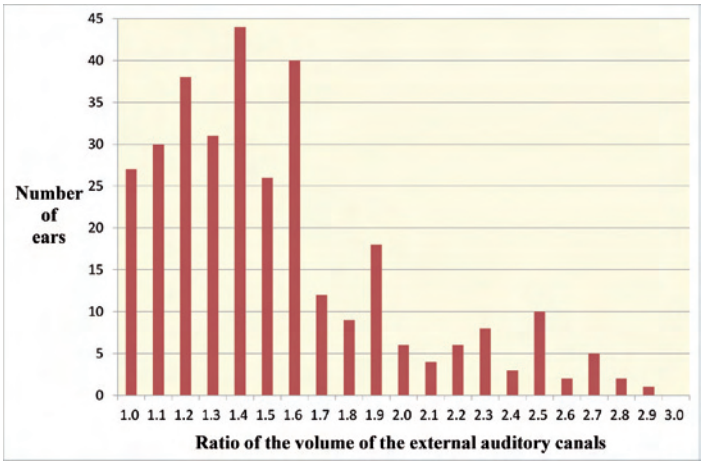


Fig. 4. Number of ears and degree of retraction of the posterior meatal wall at the final observation (322 cases).

Discussion

All other methods of tympanoplasty with canal wall up or canal reconstruction are performed based on the idea of fortification of the posterior meatal wall for prevention of retraction pocket recurrence. Changing this way of thinking, I could avoid the retraction pocket formation. What I would like to underline here is not the material of posterior meatal wall reconstruction but the way of thinking for prevention of cholesteatoma.

With this technique, instead of retraction pocket formation, a balloon-like retraction occurs on the soft posterior meatal wall when there is aeration disturbance of the middle ear, which prevents the recurrence of cholesteatoma.

In comparison to the mastoid obliteration technique, this method also has advantages. Mastoid obliteration is difficult to perform in patients with intracranial complications and metabolic disorders, such as diabetes mellitus. In addition to a wider range of applications, our method allows for easier exposure of residual cholesteatoma, and dry ears can be obtained in a shorter post-operative period.

After our previous papers,[1,3,4] some papers concerning soft wall reconstruction were publishued.[5,6] The number of ear surgeons who adopt this method is now increasing in Japan. I hope this way of thinking becomes widely used in the world.

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METHODS FOR PREVENTION OF RECURRENT CHOLESTEATOMA

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In cholesteatoma surgery, the primary goal is the complete eradication of the pathology with no residual disease, while secondarily the prevention of recurrences and the preservation or improvement of hearing are searched. After cholesteatoma surgery, relapsing of the disease is due either to residual or recurrent cholesteatoma and this may occur anywhere from the ear canal, to the eardrum, the middle ear, the mastoid, to the petrous apex. Residual cholesteatoma results from leaving squamous epithelium in the mastoid and middle-ear cleft. Therefore, in order to reduce the risk of new cholesteatoma formation, the radical removal of all the matrix is mandatory.[1,2]

The incidence of recurrent cholesteatoma is reported to vary from 5 to 71%, depending on the type of cholesteatoma, the surgical technique, the follow-up rate, the length of the post-operative observation period and the statistical method applied.[3-6]

The surgical techniques used in cholesteatoma surgery can be divided in those where the posterior canal wall is removed to achieve a better exposure of the surgical field (open technique or canal-wall down, CWD), and those where the posterior canal wall is kept intact (closed technique or canal-wall up, CWU). The choice between them has largely been debated over the years, without reaching any general agreement, thus leaving each otologist free to decide.[7]

CWD tympanoplasties, as radical cavities, rarely require a revision surgery and offer hypothetically better chances for reducing the recurrence rate of cholesteatoma. On the other hand they offer less chances for the reconstruction of a functional sound-conducting system and for fitting of a hearing aid. Moreover, the creation of a large cavity not infrequently results in annoying symptoms such as ear discharge, water intolerance, dizziness or headache, with frequent need to have the mastoid bowl periodically cleaned.[8-10]

In CWU tympanoplasties, preservation of the normal anatomy should allow better functional results, avoiding those disadvantages associated with large cavities, although it is susceptible to a higher rate of recurrences, often requiring a second look surgery.[5,11]

Some authors, in order to achieve the best exposure of the disease and to prevent recurrences advice to apply 'à la demande' techniques that encompass a partial drilling the posterior canal wall, its temporary removal,[10] or its total removal.

The use of a large variety of materials and autologous tissues, like flaps, silastic sheeting, cement, hydroxyapatite, cartilage, bone paté, demineralized bone matrix,[12] silicon block, titanium (or a combination) has been described in order to cover defects of the posterior canal wall, reaching good functional results and preventing recurrences of the disease.[13,14,15-18]

Some authors have stated that recurrent cholesteatoma after CWU procedures occurs in four sites: the attic, with erosions of the canal, at the posterosuperior pars tensa, and as borderline patterns, between the attic and the pars tensa (occurring after purely attic disease).[19] It usually happens when the newly reconstructed tympanic membrane undergoes a progressive retraction toward the attic, the antrum and the mastoid cavity.[20] This condition has been associated with a malfunctioning Eustachian tube, the blockade of communication between the Eustachian tube and the attic-antrum region, a bony defect of the tympanic scutum and the

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Edited by Haruo Takahashi

chronic inflammation in the attic.[1,11,21] In order to prevent these lesions it is mandatory to close all canal defects, to stiffen a collapsed tympanic membrane and carefully repair attic and epitympanic defects.[4,22,23] Many techniques that include the use of bone paté,[24] autologus cartilage,[25-27] hydroxyapatite plates, silastic and bone chips,[28] have been described in literature, especially for the repair of the attic defects, with a high reduction in the recurrence rate.[5,22,29]

Personal experience

In our department, type-II epitympanic retraction pockets, that are at risk for developing a secondary acquired cholesteatoma, have been preventively treated by using the lateral attic reconstruction (LAR) technique in order to prevent the recurrence of a retraction pocket.[30] This technique has not only proved to enable to prevent the relapse of these pre-cholesteatomatous conditions, but it has also proved not to impair the hearing function. After primary surgery, which in our experience has privileged the CWU technique, an extensive use of cartilage for reconstructing the surgical defects can play a crucial role for the prevention of cholesteatoma recurrence, also considering that the follow up is nowadays carried out by diffusion non-epi (HASTE) MRI that, in negative cases, allows to avoid a second-look surgery.

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CWU, CWD – RECONSTRUCTION OR OBLITERATION?

Canal-wall-down mastoidectomy for cholesteatoma

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Introduction

The incidence of cholesteatoma in the Indian sub-continent is higher than that reported by western literature. In the past, most cholesteatoma patients reported late, which resulted in a late diagnosis and many patients with complications due to extensive cholesteatoma. There is a recent trend in the early diagnosis of cholesteatoma due to improved diagnostic procedures and improved patient literacy. Nowadays, more cases of cholesteatoma are being diagnosed and operated in Indian clinics. In this author's clinical practice, the same trend has been observed recently times and cholesteatoma seems to have an incidence rate of 15-20% among all chronic otitis media cases.

The intricate anatomy around the sinus tympani and the posterior mesotympanum poses surgical challenges to the otologist, who often relies upon his surgical experience to fully eradicate the disease while preserving the vital structures in its vicinity.[1] There is an inherent risk of spread of mesotympanic cholesteatoma in a variety of ways, a.o. the risk of ossicular involvement including stapes supra-structure, facial nerve and labyrinthine windows. In many cases, the foot plate is found to be spared and this provides the possibility of using the mobile footplate for hearing reconstruction as a single-stage procedure.[2]

A one-stage dissection of all cholesteatoma matrix in the mesotympanum and hearing reconstruction as above would be most ideal in the Indian context, due to problems of patient follow up. It is prudent to do a complete canal-wall-down (CWD) mastoidectomy with adequate lowering of the facial ridge and clearance of all disease covering the sinus tympani and vertical segment of the facial nerve course.[2] The surgeon needs to emphasize the need for diligent follow-up with oto-microscopy in these patients to identify any recurrence or recidivism at the earliest.

Principles of cholesteatoma surgery

The basic goal of cholesteatoma surgery is the complete removal of squamous epithelium to minimize the risk of recurrence while maximizing hearing gain / preservation. The best method would be a CWD mastoidectomy, in which the posterior wall of the external auditory canal is removed and the mastoid and external auditory canal become a common communicating cavity exposed to the outside. This approach in management of cholesteatoma has been well emphasized in literature published since the last century.[3] Bezold and Hartmann stated this in their manuscript on 'Office management of discharging ears' in the 1880s. Schwartz, Kuster and Zaufal (1889 emphasized the need for 'exteriorization and fistulization' in ear surgery and Hartmann, Jansen and Stacke (1890-91) proposed the true beginnings of CWD surgery, which still remains the 'Gold-standard' operation for unsafe ears, since then till date.[3]

The basic indications for CWD mastoidectomy include: extensive cholesteatoma involving mastoid air cells, sclerotic, contracted mastoid with limited access to epitympanum, cholesteatoma in the only hearing ear,

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Edited by Haruo Takahashi

recurrence of cholesteatoma after closed-cavity procedure, non-reconstructable posterior canal wall, otologic or CNS complications, labyrinthine fistula, poor ET function, patients with unreliable follow up, cases of extensive pediatric cholesteatoma.[3] The fact that CWD mastoidectomy is still an important tool in managing cholesteatoma is testament to certain clear advantages.

The salient investigations would include otomicroscopy, radiological and audiological tests. Cholesteatoma surgery requires the nuances of an experienced otologist in order to completely eradicate the disease from the entire middle ear cleft, while taking utmost care for preserving and reconstructing the hearing mechanism of the disease-free ear either in the same sitting or as a staged procedure. These decisions lie with the operating surgeon and intra-operative decisions must be made judiciously with concern towards the patient factors like age, pre-op hearing thresholds, presence of co-morbidities, and financial status.

The major advantages of CWD mastoidectomy include: better access to meso- and epitympanum, facial recess gets exteriorized well, there is decreased incidence of residual/recurrent disease, CWD facilitates better post-operative inspection and second-look surgery is usually not required. CWD mastoidectomy may be performed by retro-auricular or endaural approach with trans-cortical or trans-meatal route. There are a few demerits to CWD mastoidectomy, *e.g.*, the open cavity needs frequent ear canal cleaning and shallow middle ear space makes ossiculoplasty difficult with a possibility of increasing loss of hearing over time.[4] In the Indian sub-continent, residual disease and recurrent disease occur in an estimated 11-27% and 5-13% of patients undergoing the closed procedure, whereas residual or recurrent disease occurs in 2-10% of patients undergoing the open procedure.

Management philosophy in cholesteatoma surgery

In this author's extensive experience with cholesteatoma surgery over the past thirty years, small cholesteatomas limited to the post-mesotympanum are best treated with canal-wall-up mastoidectomy with reconstruction of scutum with cartilage, while large cholesteatomas in a small sclerotic mastoid should be treated with the retrograde approach or inside-out CWD mastoidectomy. Extensive cholesteatoma in a well-pneumatized mastoid should be treated with a thorough CWD mastoidectomy. In this author's experience, there is a bias towards CWD mastoidectomy, due to the following results: MERF experience: January 1999-December 2006 (eight years); total number of cholesteatoma surgeries: 480 – CWD = 390, intact CW = 90.

Table 1. MERF Experience with Cholesteatoma Surgery (Jan 1999 - Dec 2006)

Type of Surgery	*Recurrence of Disease*	*Residual Disease*	*Total %*
CWU	9 (10%)	5 (6%)	14 (15%)
CWD	12 (3%)	6 (1.5%)	18 (4.5%)

Operating pearls for Cholesteatoma Surgery

In this survey, salient steps while performing cholesteatoma surgery have been highlighted and the identification of important anatomical landmarks – like the second genu of the facial nerve which is located inferomedial to the prominence of the lateral semicircular canal, and the cochleariform process which is a reliable landmark for tympanic segment of facial nerve – have been emphasized. The author also pointed out that the micro-drill burr should be moved parallel to important structures, *e.g.*, facial nerve, sigmoid sinus, LSCC; and drilling over the facial nerve must be with large burrs and with continuous irrigation to cool the drilled area. Special care needs to be taken while the anterior buttress is removed, when the facial ridge is lowered to the level of the nerve, and when the perilabyrinthine cells in the attic are drilled out.[5,6]

The importance of saucerization of the entire cavity and removal of overhanging edges which allows soft tissue to fill in the bony defect which makes the cavity smaller, and the fact that an adequate meatoplasty is an essential prerequisite to CWD procedures are also points that have been described. Performing such a meticulous dissection would essentially create the ideal CWD cavity which will be small, rounded in shape, self-cleansing, smooth, without niches, with low facial ridge, having a wide meatoplasty which is dry and well epithelialized. This cavity should essentially be disease-free for a life time.[6]

Conclusion

Incomplete exteriorization of cellular tracts, inadequate removal of the bone over the middle fossa dura, sigmoid sinus, facial nerve, or mastoid tip and inadequate meatoplasty are the major causes for cavity problems in most of the CWD procedures. Since diagnostic modalities to identify early cholesteatomas accurately have advanced in the present day, operative techniques should also be further refined by otologists, in order to cure the patients of all disease by the best possible approach and provide them with the best audiological outcomes. A correctly performed CWD technique together with a sufficiently large meatoplasty, has proven to be successful in the management of extensive cholesteatomas and has paved the way for achieving the above-mentioned desired goals.

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EVALUATION OF MIDDLE EAR PNEUMATIZATION AFTER PLANNED TWO-STAGED TYMPANOPLASTY FOR CHOLESTEATOMA: ITS CORRELATIONS WITH HEARING RESULTS AND RECURRENCE RATES

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Introduction

The middle ear (ME) is lined by a mucosa that embeds a network of blood vessels. The ME should function to passively exchange gases with the local blood across their respective mucosa in the normal condition – therefore, decrease in ME volume and poor ME mucosal function might predispose to certain pathological conditions, including cholesteatoma.[1,2]

After a planned two-staged tympanoplasty for cholesteatoma cases, the reconstructed and pneumatized ME could be lined again by a mucosa and the restored ME function might lead to a good hearing result as well as a low recurrence rate of cholesteatoma.

Materials and methods

A planned two-staged tympanoplasty was performed in most cases with advanced cholesteatoma. At the first stage of surgery (operation 1), cholesteatoma was completely eradicated and the anterior attic bony plate was widely opened (Fig. 1A,B), then two silicon sheets were placed into the ME (Fig. 1C,D), and finally the tympanic membrane was formed with a fascia of temporal muscle. The external ear canal was reconstructed using the soft-tissue wall technique. At the second stage (operation 2), after taking the silicon sheets out and searching for residual cholesteatoma, both the ossicular chain and the external ear canal were reconstructed with cartilage plates from auricle or tragus.

The ME pneumatization was determined by several CT scans according to the 0-5 rating system (Fig. 2) (score 0: none; 1: pneumatized ET orifice; 2: mesotympanum; 3: attic; 4: antrum; 5: mastoid). Correlations of ME pneumatization with hearing results as well as recurrence rates of cholesteatoma were evaluated.

Results

In 76 cases (82 ears) with acquired cholesteatoma among the patients who received the surgery at Osaka University Hospital between 2000 and 2009, CT scans were examined at least twice at any time of before operation 1, before operation 2, and after operation 2. There were 35 males (37 ears) and 43 females (45 ears), and the median age was 42 years (range 4-75 years). In one case (50-year-old male), the scores of the

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Edited by Haruo Takahashi

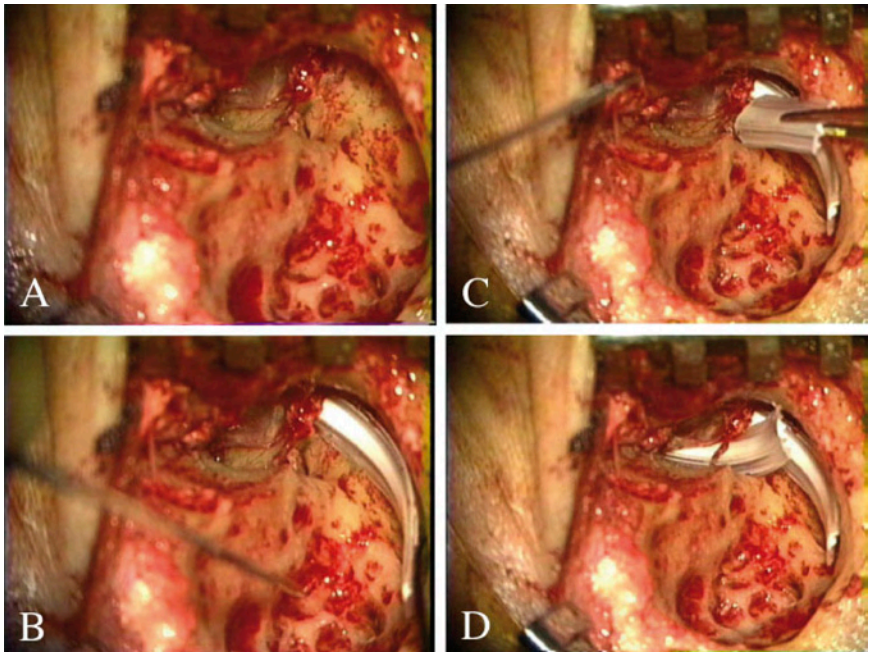

Fig. 1. Insertion of two silicon sheets into the ME at the first-stage surgery.

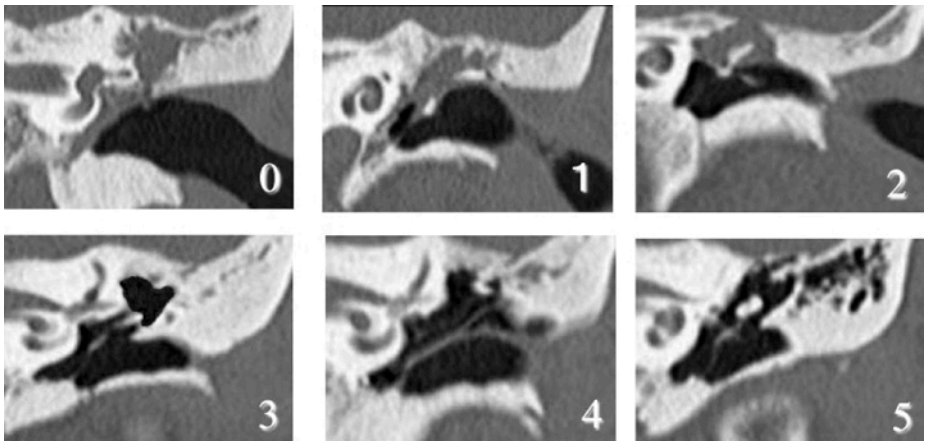

Fig. 2. The 0-5 rating system of ME pneumatization.

ME pneumatization were rated at 0 before operation 1 and at 5 before operation 2, indicating its significant improvement within just one year (Fig. 3).

The average ME pneumatization was 2.65 before operation 1 and 3.45 before operation 2 (Fig. 4A, N = 35), and it was 3.14 before operation 2 and 4.29 one year after operation 2 (Fig. 4B, N = 21). These improvements of the ME pneumatization were statistically significant ($P < 0.05$) for both evaluations. The ME pneumatization tended to continuously improve for a certain period of time even after operation 2 (N = 34). The mean follow up was 727 days after operation 2.

Hearing results (80% success) in patients with well-pneumatized ME (rated at 3-5; N = 27) after operation 2 was not significantly better ($P > 0.05$) than those (50% success) in patients with poorly pneumatized ME (rated at 0-2; N = 29). The mean follow up was 941 days after operation 2.

The ME pneumatization after operation 2 (2.8) in patients with recurrences/retraction pockets (N = 7) was significantly ($P < 0.01$) worse than that (4.55) in patients with no recurrence of cholesteatoma (N = 59). The recurrence rate of cholesteatoma was 5.6% (mean follow up: 1497 days after operation 2).

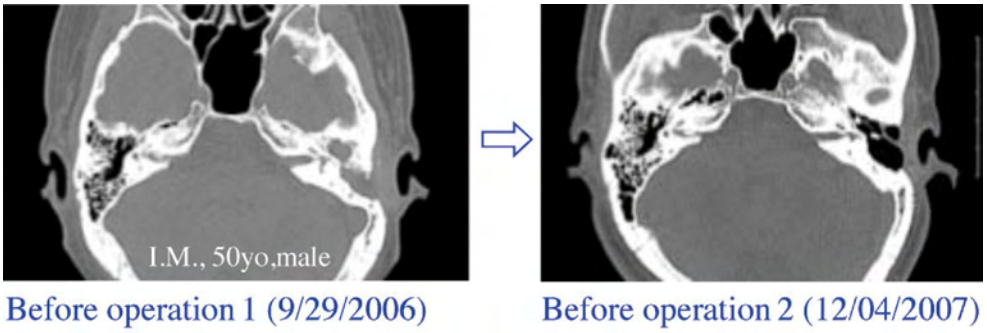

Fig. 3. Improvement of the ME pneumatization after operation 1.

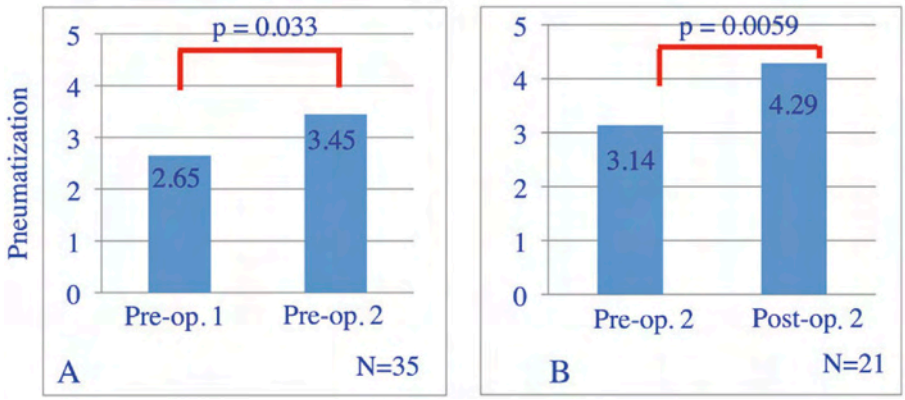

Fig. 4. Improvement of the ME pneumatization after operation 1 and operation 2.

Discussion

With a planned two-staged tympanoplasty, the ME pneumatization improved significantly one year after operation 1, and it tended to improve continuously for a certain period of time even after operation 2. Good vibrations of TM, stable placements of columella resulting from a reconstruction of the well pneumatized ME should contribute to a good hearing result and a low recurrent rate of cholesteatoma. In some cases, the ME pneumatization was poor before and after operation 2, presumably due to a dysfunction of the Eustachian tube. A ventilation tube should be inserted at the second stage, if the ME pneumatization is found to be poor before operation 2.

Reconstruction of the well-pneumatized ME and restoration of the ME function might be crucial to good hearing results and low recurrence rates in tympanoplasty for cases with cholesteatoma.

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HOW TO DEAL WITH CHOLESTEATOMA IN A DEVELOPING COUNTRY

Will the canal-wall-up (CWU) technique provide an opportunity to control the disease?

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Introduction

Early diagnosis, accurate CT scan data, good training and surgical skills, and good communication with patients (or parents in case of children) are key elements that influence the successful treatment of cholesteatoma. Although the procedures for diagnosing and treating cholesteatoma in the developing countries are the same as in developed countries, the possibilities for earlier diagnosis and treatment are not the same.[1,2]

In developing countries, like Albania, canal-wall-down (CWD) mastoidectomy is still the preferred technique for control of the disease. However, in the last decade, with the improvement of medical care and hospital infrastructure, we more often perform the closed technique.

The goal of this study is to evaluate the likelihood of success for controlling disease after performing the closed technique in our clinic.

Material and method

The study considered 23 cases of patients with cholesteatoma, including a cross section of children and adults that underwent surgery between 2007 and 2010. The main focus of the analysis was to study the management of cholesteatoma and not the functional outcomes. Cases of simple retraction pockets were excluded. All patients received a CT scan prior to the surgery. Follow-up CT scans and/or MRI were performed 1-1.5 years after the surgery. CT scans were considered conclusive in cases where the middle ear and mastoid cavity were well aerated without presence of soft tissue (Fig. 1), or in cases with a round, well-delineated lesion which was highly suggestive of residual cholesteatoma (Fig. 2). When the CT scan was not considered conclusive, an MRI procedure was performed to identify the possibility of residual cholesteatoma. The MRI sequences obtained to evaluate the soft tissue in the middle ear and mastoid cavity are 'delayed gadolinium enhanced T1 weighted' and 'non-echo-plannar (non-EPI) diffusion weighted (DW)' imaging (Fig. 3).

Closed mastoidectomy was the operating technique in all cases. The removal of cholesteatoma was done through the transcanal-transmastoid approach. Cartilage-shield tympanoplasty was the preferred technique for the reconstruction of the tympanic membrane.

Statistical analysis was performed using SPSS statistical software (version 15; SPSS, Chicago, IL, USA).

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Edited by Haruo Takahashi

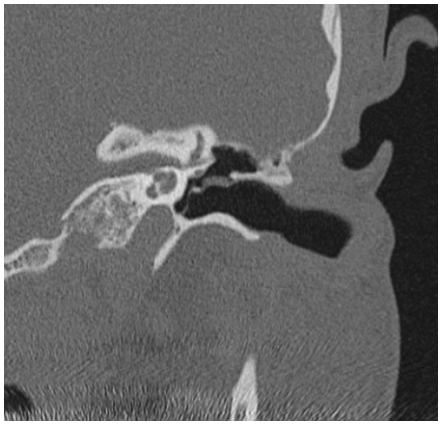

Fig. 1. The CT scan shows a well-aerated middle ear cavity and attic without presence of soft tissue.

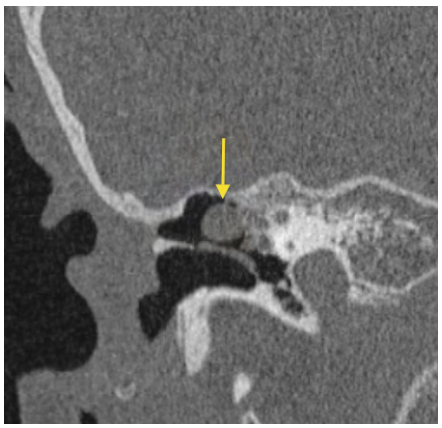

Fig. 2. This round lesion is highly suggestive for residual cholesteatoma (arrow).

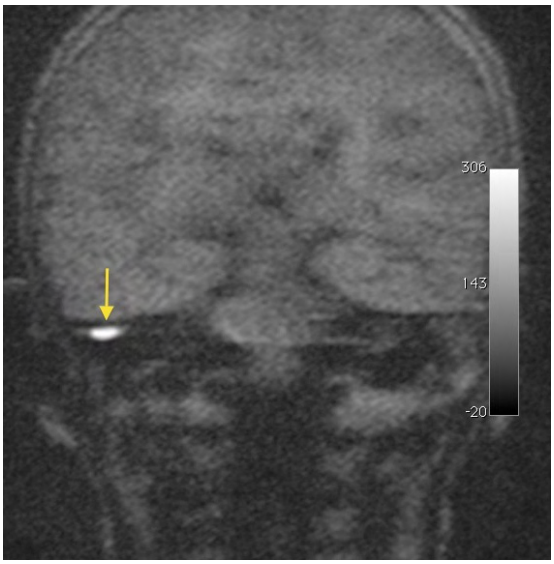

Fig. 3. Non-echo-plannar (non-EPI) diffusion weighted (DW) imaging shows cholesteatoma as a hyper-intense lesion (arrow).

Results

The average age of 23 operated patients was 26 years old (range 6-59). Of the patients, 17.4% was child and 82,6% adults. Sex ratio was 1:1.5 (9 males, 14 females). Three patients (13 %) never returned for the follow up.

Regarding the surgical findings: in 17 patients (73.9%) diffuse cholesteatoma was observed; in the remaining cases (26.1%) cholesteatoma was well encapsulated.

Cholesteatomas were epitympanic (attic) in nine patients (39.1%); mesotympanic (middle ear) in eight patients (34.8%) and were arising from both areas in six cases (26.1%).

Although the patients were treated pre-operatively for three to six months, inflammation was present at the time of surgery in six of them (26.1%).

Considering the availability of the devices, a CT scan was the method of choice for the follow up. A CT scan was performed in 19 patients (95%) at an interval of 1-1,5 years following the first operation. One patient presented with retro-auricular swelling 2,5 years after the first surgery and was operated on immediately. In seven cases (36.8%), the data of CT scan were considered conclusive to rule out the residual cholesteatoma (middle ear and mastoid cavity well aerated) or for the positive diagnosis of residual cholesteatoma (round, well-delineated lesion).

MRI exams were done in 12 cases (63.2%). In three cases (25%), the MRI images were suggestive for residual cholesteatoma.

Seven patients (35%) underwent second-look surgery. In five cases (71.4%, or 25% of all patients) residual cholestatoma was present; radical mastoidectomy was the treatment of choice for three cases (42.9%) (in two cases cholesteatoma was diffuse and the mucosa inflammatory; in one case the Eustachian tube was invaded by epidermis); in two cases (28.6%) the residual cholesteatoma was presented as round lesions that were easily removed with the preservation of canal wall. Residual cholestatoma was not found in two patients (28.6%). The ossiculoplasty was done in four patients (57.14%).

Thirteen patients (65%) refused second-look surgery with the purpose to perform the ossiculopasty.

Discussion

During their practice and surgical treatment of cholesteatoma, physicians in developing countries like Albania may face a lot of difficulties, like lack of infrastructure and inadequate training in the field of otosurgery or imaging, patients with huge cholesteatomas that are often diagnosed at an advanced stage with life-threatening complications,[1,2] or patients that never return for different reasons.

Considering all this, CWD, as the ‘gold standard’ for control of cholesteatoma[3,4] is still the preferred treatment of the disease in Albania. But with the economic development and sociocultural changes of the last two decades, patients are becoming more aware of the benefits of early diagnosis and more often are seeking treatment solutions that can offer them better quality of life. CWU mastoidectomy, as it has been shown in several studies,[3,4] is the best technique that offers a better quality of life and the possibility of hearing improvement. The results of our study (residual cholesteatoma in 25% of cases), compared to bigger series,[5,6] gave us an optimistic view regarding the eradication of the cholesteatoma with the closed technique, although we still had a significant loss of follow-up patients (13%) and a majority of patients that refused the second-look surgery (65%).

Despite initial difficulties, due to lack of experience in the field of imaging of temporal bone, the close collaboration with radiologists permitted us to obtain appropriate pre-operative CT images and, most importantly, suitable radiological approach for follow-up patients. MRI, with its high sensitivity and specificity,[7-9] has become the best exam for the detection of residual cholesteatoma. Considering the lower cost and higher accessibility, the CT scan remains the test of choice in Albania. We think that a CT scan might be conclusive in cases with perfect pneumatization of the middle ear and mastoid cavity as well as in cases with round, well-delineated lesion. In other cases, MRI and/or second look are mandatory to rule out the possibility of residual cholesteatoma (Fig. 3).

Conclusion

The results of the study indicate that the closed technique can be considered as a good option for treatment of cholesteatoma in developing countries. The role of the patient is essential in succeeding.

The likelihood of eradication of cholesteatoma using this approach is directly related to anatomical patterns (pneumatization of the mastoid, position of the sigmoid sinus, position of the dura, exposed dura or dehiscent facial nerve), extension of the cholesteatoma (on supratubal recess, retrotympanum, superstructure or footplate of stapes), and concomitant pathology (inflammation, polyps, fistula of LSCC).

The CT scan, and especially MRI with its high sensitivity, have obviously changed our attitude toward the management of cholesteatoma. However, the necessity of the second look still depends on the opportunities to control the disease.

FACIAL-NERVE TUMOR COMBINED WITH CHOLESTEATOMA

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Case 1

A 37-year-old male with facial weakness and hearing loss on the right side. In August, 2009, he felt abnormal blinking of his right eye and complained of drooling while eating. He consulted a nearby ENT doctor, who performed a CT scan and suspected a facial nerve neuroma. The patient was then referred to our hospital for further treatment.

Clinical examinations

At the initial examination, a very tiny retraction pocket at pars flaccida of his right tympanic membrane was noted along with a mixed hearing loss of 38.8 dB (a+2b+c/4) hearing level. His left ear was normal. His facial nerve palsy was evident as shown in Figure 1, which was evaluated as Class V of the House-Brackman Grading System. The value of electro-neuronography (ENoG) was 5%. A CT scan revealed a small space-taking mass in the epitympanic space, with bony erosion at the base of the middle cranial fossa (Fig. 2a). Also a labyrinthine portion of the facial nerve became somewhat wider, and a small fistula was found at the basal turn of the cochlear (Fig. 2b). MRI showed that the main location of the space-taking lesion with peripheral enhancement (Fig. 3a) was at the horizontal portion of the facial nerve, extending to surrounding bony tissue, including the semicircular canal (Fig. 3b). Based upon these radiological findings, we thought that the most likely diagnosis could be intra-tympanic facial-nerve neuroma.

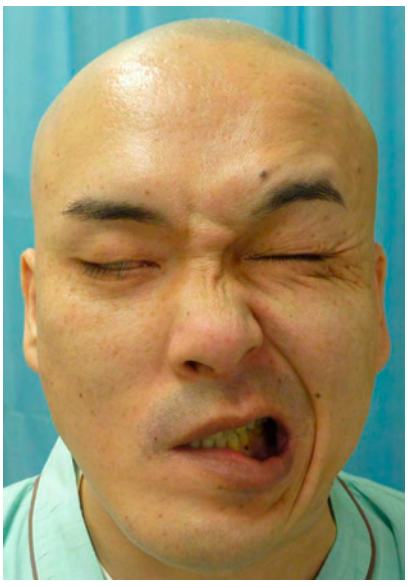

Fig. 1. Evident right facial nerve palsy.

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Edited by Haruo Takahashi

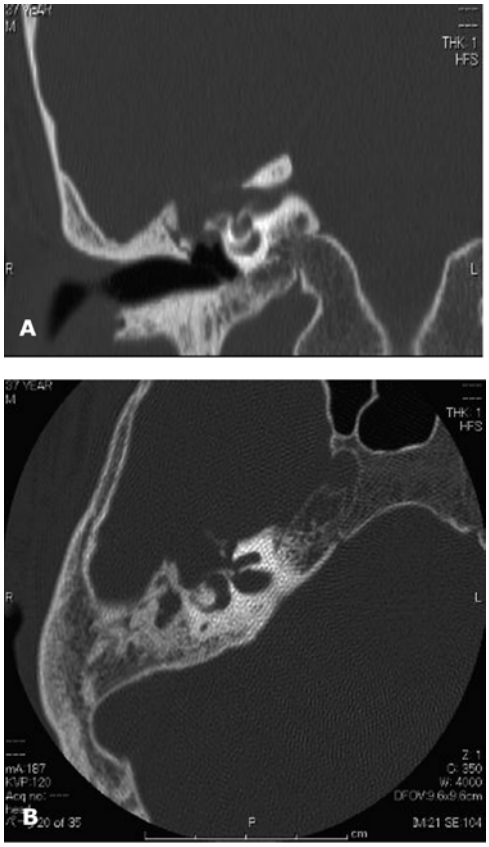

Fig. 2. CT findings. a: epi-tympanic mass lesion with bony erosion; b: small cochlear fistula.

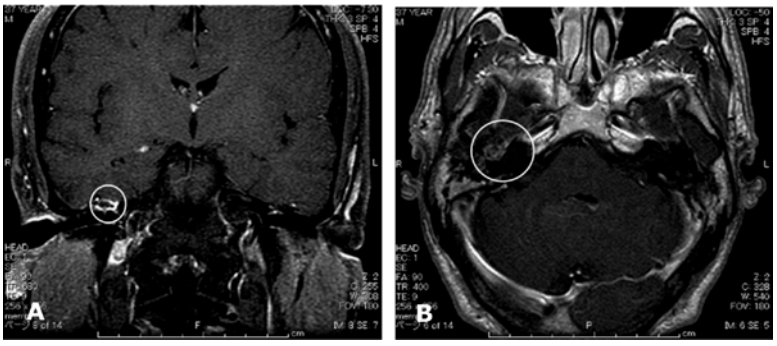

Fig. 3. MRI findings with contrast enhancement. a: coronal view showing the lesion (open circle); b: axial view showing the lesion (open circle).

Operation

An inverted S-shaped incision from the temporal portion to the retro-auricular region was made to give access to the middle cranial fossa and mastoid. The lower part of the incision was extended to just below the mandibular angle for facial-nerve reconstruction. When opening the epi-tympanic space from the middle cranial base, the facial-nerve tumor embedded in the cholesteatoma was found and removed. The facial nerve was severed from the labyrinthine portion to the horizontal portion. During this manipulation, especially while elevating the labyrinthine portion of the facial nerve, a cochlear fistula was confirmed and sealed by fascia. By frozen section during the surgery, the incised edge of the facial nerve was found to be free from tumor cells. Then by trans-mastoid approach, the vertical portion of the facial nerve was exposed and elevated, followed by performing a facial nerve-hypoglossal nerve end-to-side anastomosis. A large defect of the MCF base was covered by free cortical bone.

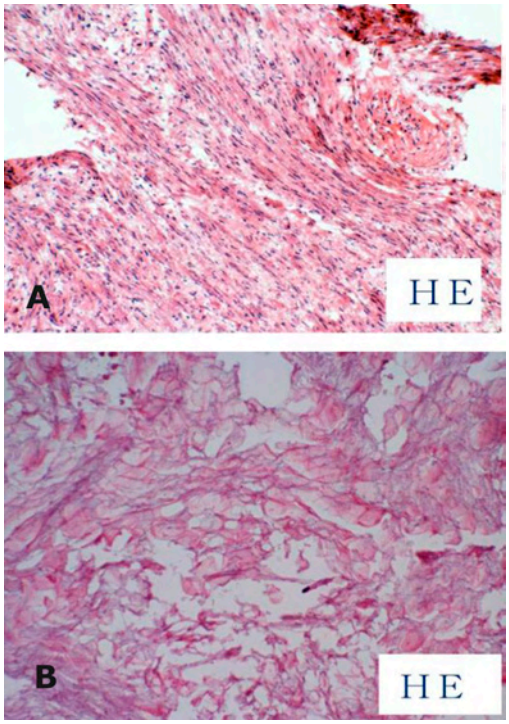

Fig. 4. Pathological findings (HE: Hematoxylin Eosin Staining). a: sample taken from the tumor: schwannoma; b: one sample of cholesteatoma.

Post-operative histology of the removed specimen demonstrated that the tumor was a combination of facial-nerve neuroma and cholesteatoma (Fig. 4A,B).

Discussion

It is quite rare that a facial-nerve tumor and cholesteatoma coexist in the tympanic cavity. The pathology in this case caused facial-nerve palsy and mixed hearing loss, although it might be difficult to make a correct diagnosis based upon the clinical findings obtained above before surgery.[1] However, Sellari-Franceschini reported that the presence or absence of ossicular-bone erosion could be an important finding to differentiate cholesteatoma from other tumors.[2] When considering the findings of ossicular erosion and a tiny retraction pocket, it could have been possible to suspect cholesteatoma along with a facial-nerve tumor. In this case, careful image study could have led to the correct diagnosis. As for the surgical approach, it could have been performed through trans-mastoid approach, but we selected a combined approach of middle cranial fossa and trans-mastoid. In this case, this combined approach should be safe, albeit invasive to some extent, considering unexpected laceration of the dura mater during dissecting tumor and cholesteatoma. Pre-operatively, we have noticed that the tumor eroded the bony wall of the basal turn of the cochlear to some degree, and thought that the tumor could be elevated from this part. It appeared that the patient lost his hearing post-operatively, which shows that it is quite difficult to preserve hearing acuity in cases of cochlear fistula.[3] When considering the nature of this tumor, operative manipulation of the cochlear fistula should not have been performed and the tumor removal from the geniculate ganglion to the pyramidal portion might be sufficient to preserve hearing. Finally, regarding the facial-nerve reconstruction after the facial-nerve tumor removal, the vertical portion of the facial nerve was elevated and then the hypoglossal nerve was exposed. The stump of the facial nerve was sutured with epineurium of the hypoglossal nerve (end-to-side anastomosis) with 9-0 nylon. We have already reported that this method, if properly done, might show fairly well recovery with minimum associated movement.[4]

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A PROPOSAL ON THE CLASSIFICATION SYSTEM OF AURAL CHOLESTEATOMA IN KOREA

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Introduction

The incidence of cholesteatoma is difficult to estimate accurately due to wide spectrum and long duration of the disease. Sadé[1] presumed five million people were affected with cholesteatoma world-wide in 1982, and Ruben[2] estimated its incidence was approximately 4.2 of 100,000 per year in the USA. Tos[3] reported incidence corresponding to three children and 13 adults of 100,000 per year in Denmark, two decades ago. On the other hand, the Korean otologic society[4] reported that 10,000 cases of chronic otitis media (COM) surgery were performed in 2005 in Korea. As about one fourth of those were cholesteatoma cases, and Korean census reported fifty million of population in Korea, an incidence of cholesteatoma would be approximatetly five of 100,000 in a year.

Various classifications or staging of cholesteatoma have been described, although none became complete, so far. As for congenital cholesteatoma of the middle ear, Postic[5] proposed the staging system, in 2002, in which stage I was designated as involvement of a single quadrant of the eardrum and no ossicular involvement or mastoid extension; stage II as multiple quadrant, but no ossicular involvement or mastoid extension; stage III as ossicular involvement without mastoid extension; and stage IV as mastoid extension. As for acquired cholesteatoma, one category consisted of primary acquired cholesteatoma, and another of acquired cholesteatoma. In 1989, Tos[6] proposed a new classification: attic-, sinus-, and tensa-type cholesteatoma, which became popular world-wide.

In this study, the author established the modified classification systems for congenital and acquired cholesteatoma, respectively, by analyzing the clinical database of cholesteatoma surgery, and by assessing whether they could provide reliable guides for surgical interventions, and postoperative outcomes.

Materials and methods

For this study, 1069 surgically-treated cases of cholesteatoma were retrieved from the database of 3880 COM clinical records, the database program[7] of which was released by the Korean Otologic Society in 2005, and all 1069 cases of surgery in the database were done by one surgeon. Ninety-eight of those cases were congenital cholesteatoma cases (83 primary and 15 recurred; 2.5%), and 971 were acquired cholesteatoma (826 primary and 145 recurred; 25.0%). Using the database of 1069 records, surgical approach, and postoperative outcomes including recurrence and hearing success were analyzed for the congenital and acquired cholesteatoma group, respectively.

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Edited by Haruo Takahashi

Congenital cholesteatoma

As Postic's staging system was somewhat overlapping and confusing, a combined posterior group was set up instead of Postic's stage II and III, and a group representing recurrent or residual disease after surgery was included. The new staging system consisted of an anterior quadrant group without ossicular involvement or mastoid extension (A); a posterior quadrant group with/without ossicular involvement but no mastoid extension (P); a mastoid extension group (M); and a recurrent group (R) (Fig. 1). In total there were 98 cases of

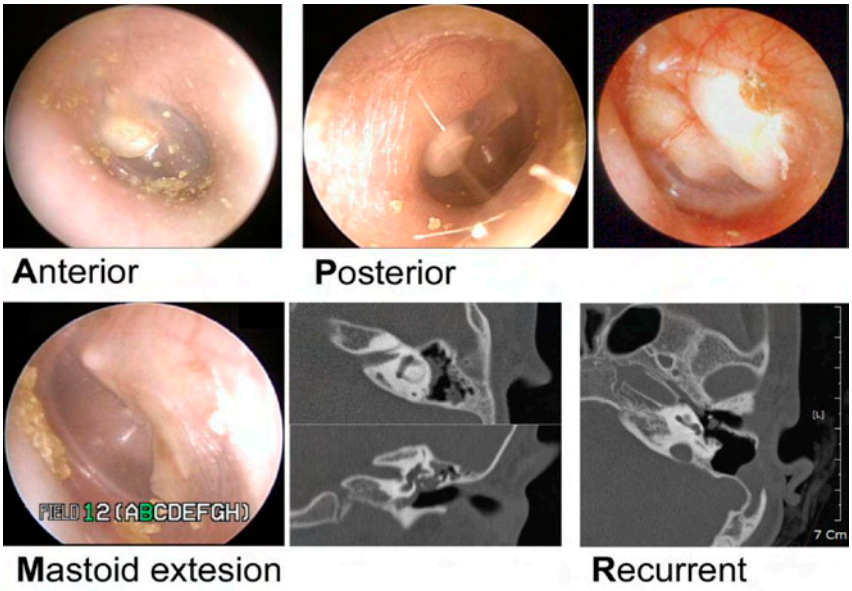

Fig. 1. A proposed classification system of congenital cholesteatoma of the middle ear: **A**nterior group; **P**osterior group; **M**astoid extension group; **R**ecurrent or residual group.

congenital cholesteatoma: primary congenital cholesteatoma in 29 cases of the anterior location group; 34 cases of posterior location without mastoid extension; 20 cases of mastoid extension; and 15 cases of recurrent cholesteatoma.

Acquired cholesteatoma

Modifying a recently proposed classification by Semaan and Megerian,[8] the author established a classification system of acquired cholesteatoma. The new system consisted of retraction pocket with aerated middle ear (RPa); retraction pocket with unaerated middle ear (RPua); epithelial migration cholesteatoma (EM); combined type; recurrent or residual type; and unclassified cholesteatoma (Fig. 2). In total there were 971 clinical cases of acquired cholesteatoma: acquired middle-ear cholesteatoma including 227 cases of the RPa type, 415 cases of the RPua type, 163 cases of the EM type, and 21 unclassifiable primary cases of the acquired group; recurred or residual cholesteatoma of the acquired type included 145 cases.

Results

Congenital cholesteatoma

Of the 29 congenital anterior cases, exploratory tympanotomy was performed in 22, and mastoidectomy in seven cases. Of the 34 posterior cases, exploratory tympanotomy was performed in ten, and mastoidectomy in 24 cases. Of the 20 mastoid extension cases, canal-wall-up (CWU) mastoidectomy was performed in 19 cases, and canal-wall-down (CWD) mastoidectomy in one case. Seven cases of residual cholesteatoma of

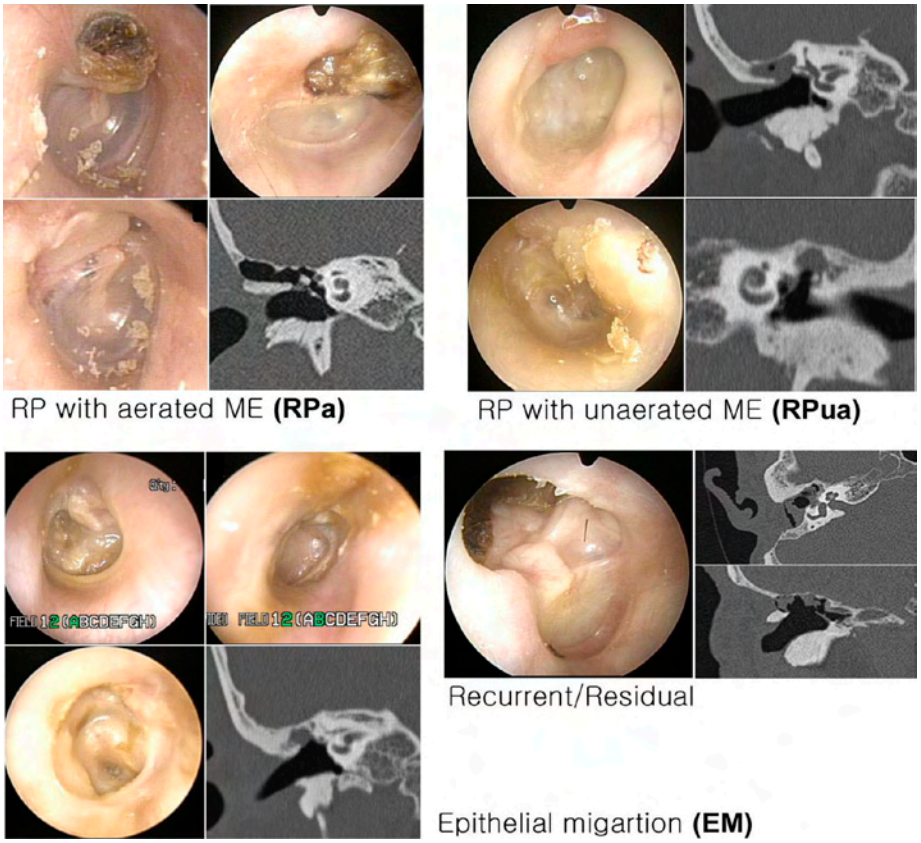

Fig. 2. A proposed classification system of acquired cholesteatoma of the middle ear. RP: retraction pocket; ME: middle ear.

congenital origin were operated with exploratory tympanotomy, and eight with CWU mastoidectomy. More exploratory tympanotomy surgeries were performed in the anterior group than in the posterior and mastoid groups; in the mastoid extension group and the recurrent group more mastoidectomy surgeries were performed (Fig. 3). In the anterior group either no ossiculoplasty or T1 tympanoplasty was performed; in the posterior and mastoid extension groups more ossiculoplasty (Fig. 4). Overall results of recurrence or perforation, or adhesion after surgery were five to 20%. The mastoid extension and recurrent groups showed worse results than the anterior and posterior groups (Fig. 5).

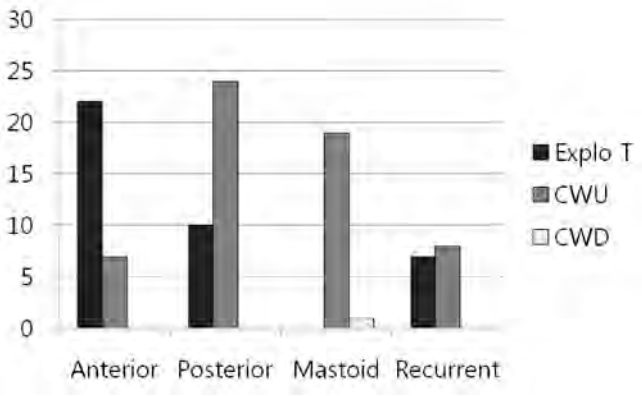

Fig. 3. Surgical approach according to the four groups of congenital cholesteatoma. Explo T: exploratory tympanotomy; CWU: canal-wall-up mastoidectomy; CWD: canal-wall-down mastoidectomy.

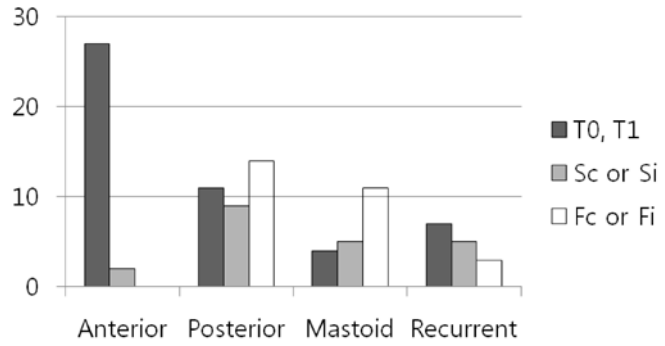

Fig. 4. Surgical approach according to the four groups of congenital cholesteatoma. T: tympanoplasty; Sc: stapes columella; Si: stapes interposition; Fc: footplate columella; Fi: footplate interposition.

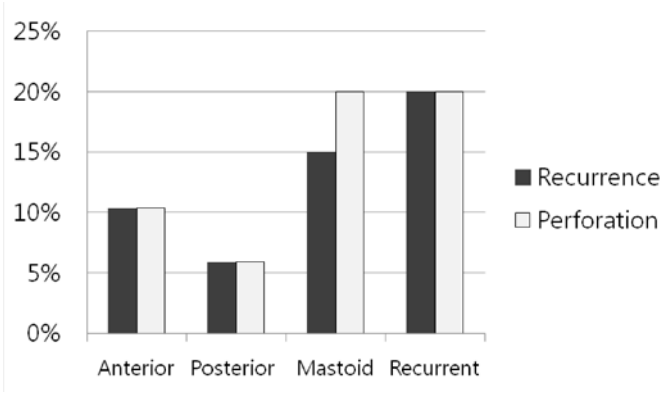

Fig. 5. Post-operative results according to the four groups of congenital cholesteatoma.

Acquired cholesteatoma

Of the 227 cases of the RPa type, CWU mastoidectomy was performed in 128 cases and CWD mastoidectomy in 99 cases. Of the 415 cases of the RPua type, CWU mastoidectomy was performed in 224 cases and CWD mastoidectomy in 191 cases. Of the 163 cases of the EM type, CWU mastoidectomy was performed in 58 cases, and

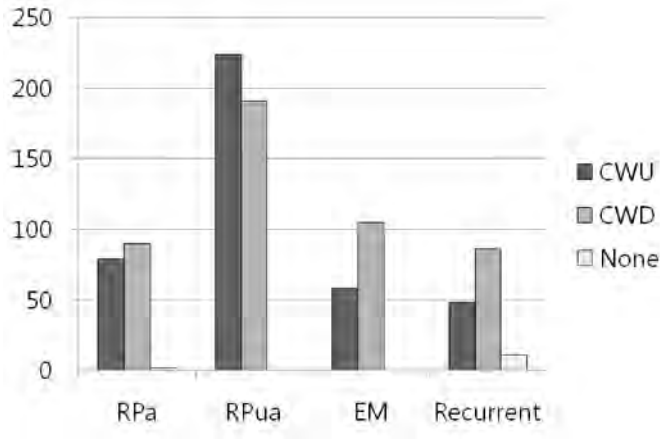

Fig. 6. Mastoidectomy approach according to the group of acquired cholesteatoma. Rpa: retraction pocket with aerated middle ear; Rpua: retraction pocket with not aerated middle ear; EM: epithelial migration; Recurrent: recurrent or residual type; CW:; canal-wall-up mastoidectomy; CWD: canal-wall-down mastoidectomy; None: no mastoidectomy.

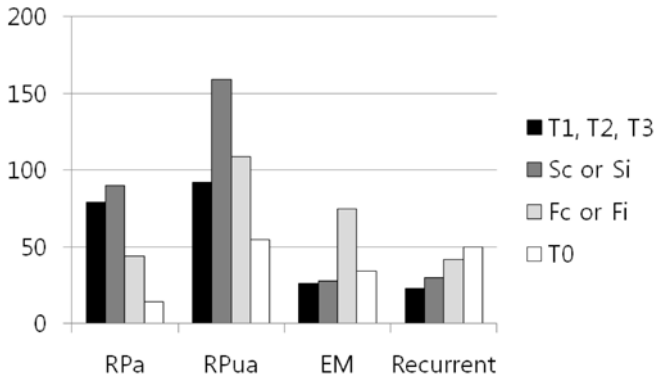

Fig. 7. Ossiculoplasty approach according to the group of acquired cholesteatoma. Rpa: retraction pocket with aerated middle ear; Rpua: retraction pocket with not aerated middle ear; EM: epithelial migration; Recurrent; recurrent or residual type; T: tympanoplasty; Sc: stapes columella; Si: stapes interposition; Fc: footplate columella; Fi: footplate interposition; T0: no tympanoplasty.

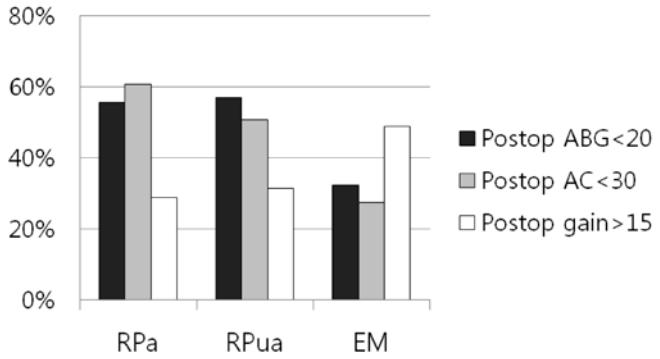

Fig. 8. Post-operative hearing results according to the group of acquired cholesteatoma. Rpa: retraction pocket with aerated middle ear; Rpua: retraction pocket with not aerated middle ear; EM: epithelial migration; Recurrent: recurrent or residual type; ABG: air bone gap; AC: air conduction threshold.

CWD mastoidectomy in 105 cases. Residual or recurrent cholesteatoma from acquired origin were operated via exploratory tympanotomy in 25, CWU mastoidectomy in 46, and CWD mastoidectomy in 74 cases. As for the type of surgical approach, more CWU mastoidectomy was performed in the RPa or RPua types than in the EM and recurrent types (Fig. 6). More T1 tympanoplasty or Sc or Si was performed in the RPa and RPua types, and more footplate ossiculoplasty was performed in EM and recurrent type (Fig. 6). Overall post-operative hearing success rates were 30-60% using the 2005 guideline of the Korean Otological society. The EM type showed worse results than the RPa type (Fig. 7). Using post-operative air bone gap grading, the EM type also showed worse results than the RPa or RPua types (Fig. 8).

Conclusion

These modified classifications for congenital and acquired cholesteatoma could play a useful role in planning the surgical procedures, and estimating outcomes after surgery.

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ADVOCATING THE USE OF THE TERM EOSINOPHILIC OTITIS MEDIA (EOM)

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In 1993, we reported intractable otitis media in three cases with bronchial asthma.[1] In 1997, we reported on seven adults, including three previously reported patients, who suffered from a combination of bronchial asthma and intractable otitis media.[2] These conditions were initially characterized by retention of serous effusion, such effusion soon being replaced by a gelatin-like secretion. A heavy infiltration of eosinophils was observed in the otorrhea, middle ear effusion, and inflammatory granulation tissue. Tympanoplasty without attention to mucosal abnormality was ineffective. Frequent severe hearing loss included sudden onset of deafness. This type of otitis media usually lasted for a long period and was progressive. Progression of otitis media in these patients was controllable to a certain degree with systemic steroid therapy. However, long-term administration of systemic steroids required considerations of side effects. Therefore, we considered it necessary to detect this type of otitis media.

The condition suffered by our patients seems to be quite similar to what Shambaugh[4] and Derlacki[3] termed middle-ear allergy or allergic otitis media. However, these terms were used before the detection of IgE in 1967 Tomonaga *et al.*[5] reported that an immune response in the middle-ear cavity was generated by antigen via the ear drum, but not by antigen via the Eustachian tube due to the barrier of the tube.

In 2006, Iino reported that the tubal opening duration was significantly longer in eosinophilic otitis media (EOM) patients than in the control group.[6] She pointed out that a patulous Eustachian tube in EOM patients easily allows the entry of antigenic materials into the middle ear. However, IgE-mediated hypersensitivity by antigen via the Eustachian tube had not yet been confirmed. Also, extremely viscous gelatin-like effusion was another intractable factor. Such a gelatin-like effusion was difficult to remove from the middle-ear cavity, and such remaining viscous effusion included harmful granules from eosinophiles which caused mucosal damage. Another problem is the complication due to bacterial infection. Methicillin-resistant *Staphylococcus aureus* (MRSA) infection is associated with a high incidence of EOM. It is difficult to treat EOM in the face of such repeated bacterial infections. Hence, simple IgE-mediated hypersensitivity for the pathogenesis of this type of otitis media still seems debatable. So, I concluded that the term allergic otitis media is inadequate. The most characteristic features of these cases were the presence of significant infiltration of eosinophiles in the inflammatory granulation tissue and middle-ear effusion. In the etiopathology of this otitis media, eosinophiles are thought to play an important role. Therefore, we advocated the term of eosinophilic otitis media (EOM) in 1997.[2]

Early-stage Churg-Strauss syndrome limited to the middle ear is difficult to differentiate from EOM. Generally, over the years, various vasculitic symptoms have become associated with middle-ear lesions in Churg-Strauss syndrome. However, no patients suffering from vasculitic symptoms were included in our over 25-year series of EOM. Thus, we considered EOM to be another middle ear disease entity. Churg-Strauss syndrome should be necessary to exclude after development of vasculitis.

Lino[7] declared EOM to be the new middle ear disease entity in 2008. Her analysis was based on retrospective clinical data of patients in a study by an EOM study group, *i.e.*, a Japanese multi-centre study involving five referral centers. The EOM study group is composed of Yukiko Iino (Department of Otolaryngology,

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Jichi Medical University), Atsushi Matsubara (Department of Otolaryngology, Hirosaki University), Takashi Nakagawa (Department of Otolaryngology, Fukuoka University), Manabu Nonaka (Department of Otolaryngology, Nippon Medical School) and me (Department of Otolaryngology, Sendai Red Cross Hospital). Diagnostic criteria of EOM by this study group are the following.

Major: Otitis media with effusion or chronic otitis media containing eosinophils in the effusion.

Minor: Association with bronchial asthma; extremely gelatinous middle ear effusion; resistance to conventional treatment of otitis media with effusion or chronic otitis media; association with eosinophil dominant nasal polyposis.

Definitive case: positive for major + two minor criteria.

Exclusion criteria: Churg-Strauss syndrome; Hypereosinophilic syndrome.

Lino *et al.*[8] reported the results of the analysis of 138 patients with EOM and 134 age-matched patients with the common type of otitis media with effusion or chronic otitis media as control. High odds ratios were obtained from an association with bronchial asthma (584.5), resistance to conventional treatment for otitis media (232.2), viscous middle ear effusion (201.6), association with nasal polyposis (42.17), association with chronic rhinosinusitis (26.49), bilaterality (12.93), and granulation tissue formation (12.62). The percentage of patients with EOM who were positive for two or more of the highest four items was 98.55%. A patient who shows otitis media with effusion or chronic otitis media containing eosinophils in the effusion and two or more minor criteria can be diagnosed as having EOM.

An efficacious to achieve stability over a long period of time has not been established. However, the proper detection of EOM is the first step towards adequate treatment, resulting in prevention of severe hearing loss. It is hoped that better management of EOM can be established in the near future.

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EOSINOPHILIC RHINOSINUSITIS AND OTITIS MEDIA IN PATIENTS WITH ASTHMA – FOCUS ON EOSINOPHILIC NASAL POLYPOSIS

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Epidemiology and clinical features of eosinophilic otitis media (EOM) with asthma and eosinophilic nasal polyposis (ENP) in Japan

Eosinophilic nasal polyposis (ENP), in which there are prominent infiltration of eosinophils and submucosal edema, is linked to comorbidities such as non-atopic asthma, adult-onset asthma, aspirin intolerance, or may represent a part of a systemic disease such as Churg-Strauss syndrome.[1,2,3] Eosinophilic otitis media (EOM) is an intractable otitis media characterized by the presence of a highly viscous yellow effusion containing eosinophils.[4] It mainly occurs in patients with adult asthma.[4,5] Table 1 shows the frequencies of severe asthma,

Table 1. Frequency of asthma, ENP and EOM in Japanese patients with adult asthma.

	Severe asthma	ENP	EOM
Atopic asthma (n = 200)	9%	2%	3%
Non-atopic asthma without AIA (n = 100)	31%	21%	6%
AIA (n =100)	56%	96%	66%

ENP, and EOM in Japanese adult asthmatics obtained from our hospital database of 400 patients. The frequencies of severe asthma, ENP, and EOM are low in atopic asthmatics. In non-atopic asthmatics, the frequency of severe asthma is 31%, and that of ENP is 21%. The frequency of EOM is also low in non-atopic asthmatics. In aspirin-induced asthma (AIA) patients, the frequencies of severe asthma, ENP, and EOM are very high. Figure 1 shows the frequencies of ENP and EOM in patients with severe and non-severe aspirin-tolerant asthma (ATA) and AIA. Almost all patients with severe and non-severe AIA show ENP. The frequencies of ENP and EOM in non-severe ATA patients are very low, while the frequency of EOM in severe AIA patients is very high. These data indicate that EOM is very common in AIA patients, particularly in severe AIA patients, and is rare in non-severe ATA patients. Figure 2 shows the frequency of EOM in AIA patients with or without ENP. EOM is very rare in AIA patients without ENP, but very common in AIA patients with a history of ENP surgery. These data suggest that EOM is common particularly in AIA patients with severe ENP. Many AIA patients firstly manifest only ENP symptoms at a mean age of 36. A few years later, asthma symptoms appear,[6,7] and five to ten years after the onset of asthma, EOM symptoms appear (Fig. 3).[8]

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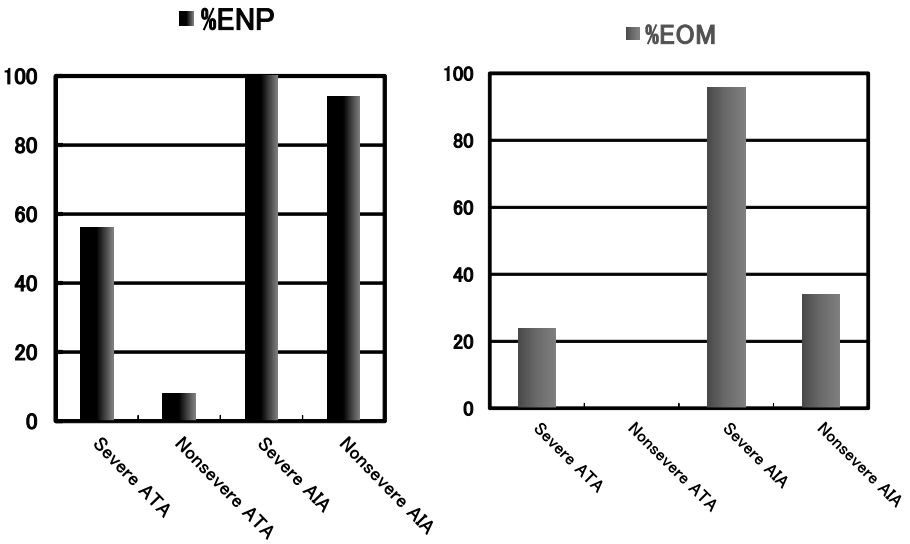

Fig. 1. Frequency of ENP and EOM in patients with (severe and non-severe) ATA and AIA.

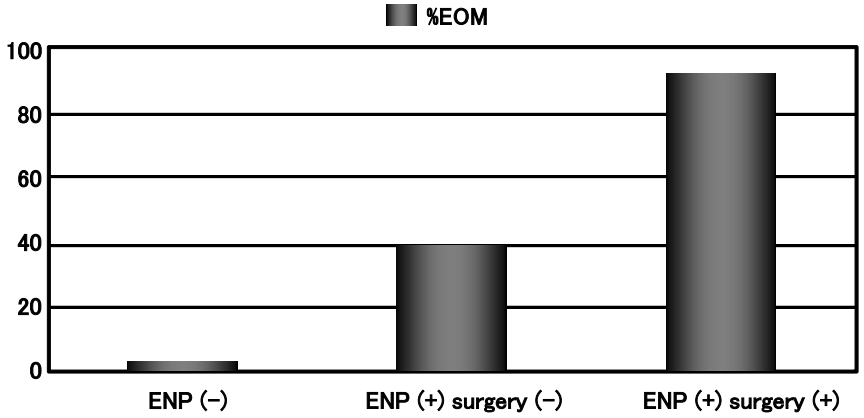

Fig. 2. Frequency of EOM in AIA patients with and without history of ENP surgery.

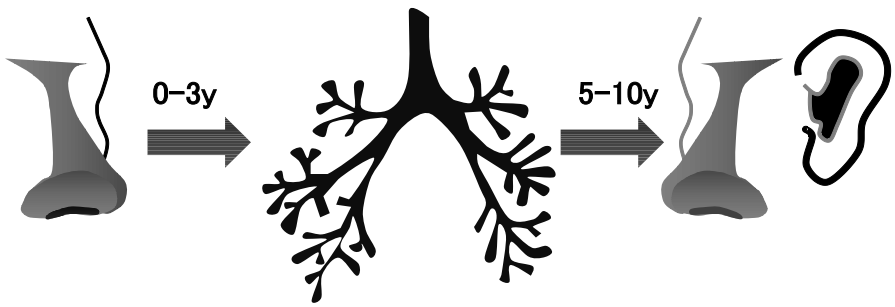

Fig. 3. The typical courses of onset of ENP, asthma, and EOM in Japanese AIA patients: Many AIA patients firstly manifest only ENP symptoms at a mean age of 36. A few years later, asthma symptoms appear, and lastly, five to ten years after the onset of asthma, EOM symptoms appear.

Figure 4 shows the annual changes in the number of hospitalizations due to asthma exacerbation and the frequency of EOM in our AIA patients. The number of hospitalizations due to asthma exacerbation has decreased to one-fourth over the last twelve years. On the other hand, the percentage of EOM patients with AIA has increased fourfold over the last twelve years. These findings suggest that stabilization of asthma may lead to EOM manifestation in Japanese asthmatics. We do not know the precise mechanism underlying this

phenomenon, but many asthma specialists in Japan believe that the increased EOM frequency is due to the stabilization of bronchial inflammation by recent powerful inhaled corticosteroids (ICS). In Japan, old-type ICS such as beclomethasone were commonly used in the 1990s. However, the new-type powerful ICS, such as fluticasone (FP) and budesonide (BUD) have been widely used from 2000 to the present (Fig. 4).

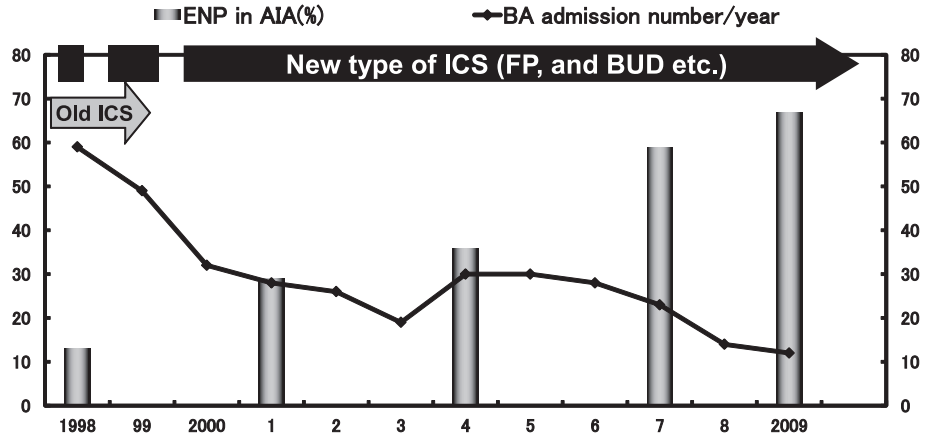

Fig. 4. The annual changes in the number of hospitalizations due to asthma exacerbation and the frequency of EOM in our AIA patients.

To summarize the epidemiology of EOM and ENP in Japanese asthmatics:

1. EOM is closely related to adult-onset asthma, ENP, and AIA.
2. EOM appears in almost all patients with both ENP and asthma, mainly at middle age.
3. EOM frequently develops in severe ENP with asthma, particularly in AIA with severe ENP.
4. AIA patients firstly manifest only ENP symptoms; a few years later, asthma symptoms appear, and lastly, EOM symptoms appear.
5. Over the last twelve years, the frequency of EOM has markedly increased in Japan. The reason for this increase is perhaps asthma stabilization by new-type ICS treatment.

Difference between chronic rhinosinusitis (CRS) with and without ENP

Inflammatory mechanisms of ENP[1,2]

Nasal polyps are edematous semi-translucent masses in the nasal and paranasal cavities, mostly originating from the mucosal linings of the sinuses and prolapsing into the nasal cavities. The results of immunohistochemical analysis in previous studies demonstrate that activated eosinophils and B cells increase in number in polyp-tissue samples from patients with CRS with ENP. Activated eosinophils, B cells, and plasma cells increase in number in polyp tissue in ENP, particularly in ENP with AIA. Increased levels of cytokines and chemokines, such as IL-5 and eotaxin, were also confirmed in ENP tissue. B cell-activating factor (BAFF) level also increases in ENP tissue. On the other hand, IL-1beta and IL-10 levels decrease in ENP tissue. Polyclonal production of local immunoglobulins, not only IgE but also IgG and IgA, increases in ENP, particularly in ENP with AIA. Iino *et al*. reported that EOM fluid show also increased IgE and ECP concentration.[9]

From these findings, we may ask, which factor plays the central role in these imbalances in ENP and EOM tissue? We hypothesized that these imbalances may be due to a decreased cyclo-oxygenase (COX) activity, particularly COX-2 activity.

Findings that support our hypothesis are:

1. Marked eosinophil infiltration and increased leukotriene production are observed in the lungs of COX-KO asthmatic mice[10] or COX-2-antagonist-treated mice.[11]
2. Increased local immunoglobulin (IgE, IgG, and IgA) production is also observed in COX-KO asthmatic mice [10,12]) or COX-2-antagonist-treated mice.[11]
3. Decreased COX-2 activity decreases production of strong anti-inflammatory mediators, such as PGE2 and lipoxin in both animal models and humans.[13]

4. A significant decrease in COX-2 activity is confirmed in ENP tissue, particularly in ENP tissue from AIA patients.[14,15]
5. There are significant positive correlations of COX activity with PGE2 and lipoxin concentrations in nasal polyp tissues. Moreover, there are significant negative correlations of COX activity with Il-5, IgE, and CysLT concentrations in ENP tissue.[16,17]

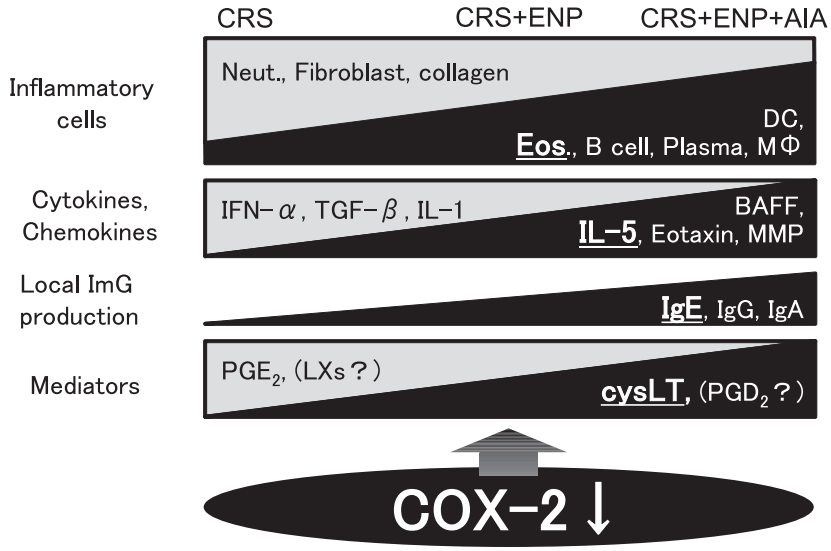

Fig. 5. Differences among CRS, CRS+ENP, and CRS+ENP+AIA.

However, unfortunately, the mechanism underlying the decrease in COX activity in the airway is as yet unclarified.

Perez-Novo *et al*. reported that the IL-5 concentrations and the eosinophil cationic protein (ECP) concentrations in four groups, which are the normal control, CRS without nasal polyps,[17] CRS with nasal polyps, and CRS with aspirin-sensitive nasal polyps. They reported that both IL-5 and ECP concentrations significantly increase in CRS with polyps in comparison with those in CRS without polyps. Moreover, aspirin-sensitive nasal polyp tissue shows a marked and significant increase in IL-5 concentration.[17]

Eicosanoid imbalance in nasal polyp tissue

Cysteinyl leukotrienes (CysLT: leukotrienes C4, D4, and E4) have long been implicated in the pathogenesis of asthma and several allergic diseases.[18] CysLT are potent bronchoconstrictors that have the additional effects of edema, mucous secretion, and eosinophilic accumulation, and airway remodeling.[18] Steinke *et al*. reported that the concentrations of CysLT significantly increase in ENP tissue compared with those in non-ENP tissue.[19] Perez-Novo *et al*. reported that the CysLT concentrations and prostaglandin E2 (PGE2) concentrations in four groups, which are the normal control, CRS without nasal polyps, CRS with nasal polyps, and CRS with aspirin-sensitive nasal polyps.[17] CysLT concentrations significantly increase in CRS with polyps in comparison with those in CRS without polyps. Moreover, aspirin-sensitive nasal polyp tissue shows marked and significant increases in CysLT concentrations. On the other hand, PGE2 concentration significantly decreases in CRS with aspirin-sensitive nasal polyps.[17]

Figure 6 shows a summary of the imbalance of eicosanoids in the nasal polyp tissue. Previous studies demonstrated that the concentration of the anti-inflammatory prostanoid PGE2 decreases in the nasal polyp tissue. The low concentration of PGE2 may be due to the decreased COX2 activity.[14,15] On the other hand, the high concentrations of strong inflammatory mediators, namely, CysLT, are probably due to the upregulation of LTC4 synthase.[19]

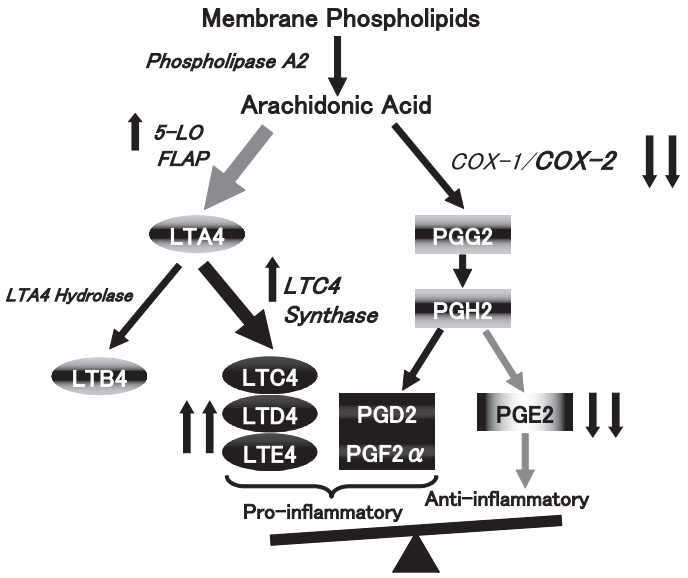

Fig. 6. The imbalance of eicosanoids in nasal polyp tissue.

Systemic leukotriene production in patients with nasal polyposis

LTE4 has been identified as a major metabolite of LTC4 and urinary LTE4 (U-LTE4) has been considered as the most reliable analytic parameter for monitoring the endogenous synthesis of CysLT.[20] We have demonstrated that nasal polyposis is one of the most important factors indicating hyperleukotrienuria.[21] Figure 7 shows U-LTE4 concentrations in adult asthmatics with and without ENP. U-LTE4 concentration is expressed on a log scale. The ATA patients with ENP showed higher U-LTE4 concentrations than the ATA patients without ENP.[22] In contrast, because there were only eight patients without ENP in the AIA group, no significant correlations between AIA with ENP and AIA without ENP were found.

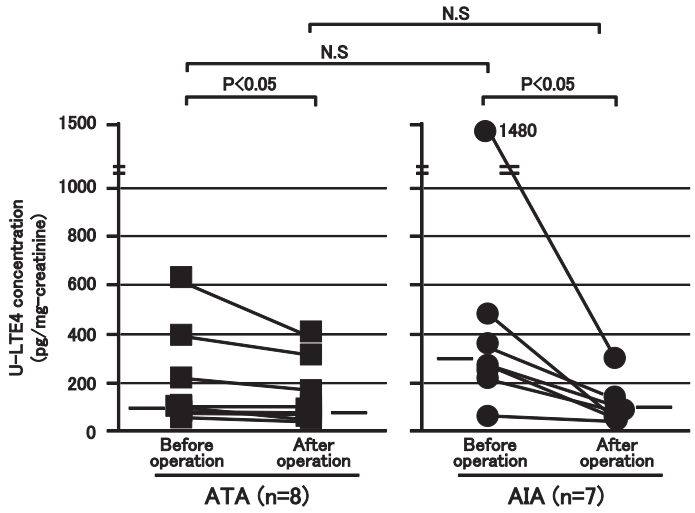

Fig. 7. Urinary LTE4 concentrations in adult asthmatics with and without ENP.

From these observations, we hypothesized that the ENP tissue is an important source of CysLT. To confirm this hypothesis, we compared the concentrations of U-LTE4 before and after the surgical treatment of nasal polyps. Figure 8 shows a significant decrease in the concentrations of U-LTE4 after endoscopic surgery of the sinuses in both the ATA and AIA groups.[22] Surprisingly, in a retrospective study, more than one-half of the patients with sinusitis showed normal U-LTE4 concentrations after endoscopic sinus surgery. In our prospective study carried out over the last six years, we have confirmed a significant decrease in U-LTE4 concentration after sinus surgery in AIA patients.

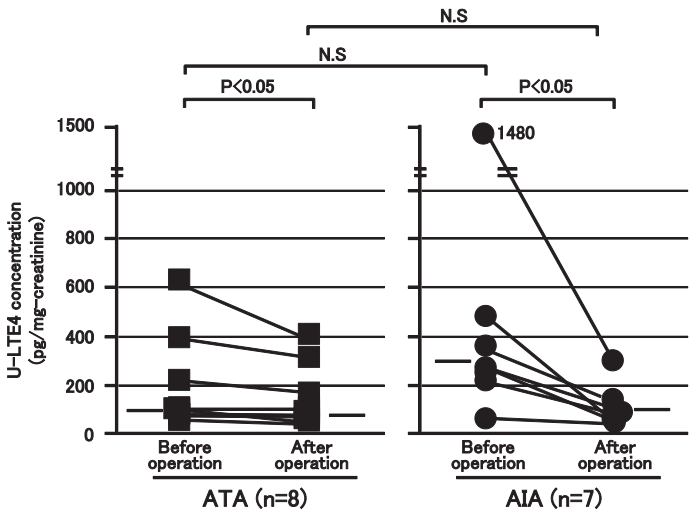

Fig. 8. Significant decrease in the concentrations of U-LTE4 after endoscopic surgery of the sinuses in both the ATA and AIA groups.

If our hypothesis, that is, the nasal polyp tissue is a major source of CysLT, is correct, there may be a positive correlation between sinus CysLT concentration and urinary LTE4 concentration. We confirmed that there is a strong and significant correlation between sinus tissue and urinary concentrations of leukotrienes. From these findings, CRS tissue with nasal polyps is a major source of CysLT in adult asthmatics with both AIA and ATA.

Summary of EOM and ENP in Japanese asthmatics

1. EOM is closely related to adult-onset asthma, ENP, and AIA.
2. The frequency of EOM has markedly increased in Japan. The reason for this increase is perhaps asthma stabilization by new-type ICS treatment.
3. Marked eosinophil infiltration and increased IgE, CysLT, IL-5 concentrations in ENP tissue may be due to a decreased COX-2 activity.
4. ENP is a major cause of CysLT overproduction in stable asthmatics both with AIA and ATA patients.
5. ENP may play an important role in not only cysLT production, but also aspirin sensitivity in AIA patients.

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FROM RETRACTION POCKETS INTO EARLY-STAGE CHOLESTEATOMA: PATHOGENESIS AND MANAGEMENT

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Introduction

Most of the early-stage cases of cholesteatoma have their genesis from retraction pockets; attic cholesteatoma from epitympanic retractions, sinus cholesteatoma from posterosuperior tensa retractions, and tensa cholesteatoma from tensa retractions. These retractions may vary from a simple retraction, to an atelectasis, a retraction pocket, an invagination or an adhesive otitis. Some of these retraction pockets are unsafe, if they are marginal, or with keratine debris or wet, and may lead to early cholesteatoma.

Pathogenesis of cholesteatoma

The epithelium covering the surface of the tympanic membrane has a property of clearence mechanism, which is called 'centrifugal migration' or 'keratin dispersion'. Once there is a formation of a atelectasis or retraction pocket, this clearance mechanism is disturbed and the surface dead cells that contain keratin are trapped and cannot clear themselves out. This is the essence of a cholesteatoma.[1]

When the drum has been retracted for a long time, the most important histological change is the disappearence of the fibrous layer of pars tensa. The fibers of the middle layer become thinner and longer under the effect of the negative pressure, and disappear in time, which means that the tympanic membrane will later be formed of two layers: outer epithelium and inner mucosa. This dimeric tympanic membrane is not anymore resistant enough to negative pressure and retraction pockets will be formed.[2] Furthermore, the epithelium sends papillas inside the mucosa (papillary ingrowth) which means that the skin touches the middle ear space.[3]

Should there be any difference in our clinical attitude between a cholesteatoma and debris accumulation? Is it important that debris may be cleaned or not by suction? There may be a lot of different comments on this subject but in our opinion, a retraction pocket which may not be cleaned by suction, accompanied by proliferation, or accompanied by bone resorption may develop a cholesteatoma.

The pathogenesis of cholesteatoma varies according to middle ear conditions, but also to immunological status; to mention some: cytokines and epidermal growth factors.

A short evaluation of retraction pockets

An evaluation of a retraction pocket and an early cholesteatoma should begin by stating its position. It is known that the posterosuperior quadrant is the most likely quadrant to be prone to pressure effects, to easy loose its lamina propria and to adhere to ossicules to cause ossicular problems.[2]

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Edited by Haruo Takahashi

The criteria should include the following points:

- Is the bottom of the pocket visible? – Stage I
- What is the depth of the pocket? (contact?) – Stage II
- Is the pocket fixed? (evaluated by Valsalva manoevre, by pneumatic otoscopy or by suction); is it a self-cleaning retraction? – Stage III
- Does the interior of the pocket indicate any wetness, irregularity, debris? – Stage IV

A radiological examination will be of great value to evaluate a retraction in case of an erosion at the bony annulus; a deep and invaginated retraction; a suspicion or presence of a cholesteatoma or symptoms of fistula; or when an operation is planned on the only hearing ear. This should cover a *multi-sliced, high-resolution CT in both coronal and axial plans.*

Management of retraction pockets

In a case of a retraction pocket we have three options: to observe, to ventilate, or to operate.

When to observe?

Observing and watchful waiting is almost always a rule in stage-I and -II retraction pockets. That is also true for deep retraction pockets if there is no infection, no hearing loss and no CT finding; and even for total adhesive otitis cases if there is no cholesteatoma, no hearing loss and under 12 years old.[4] This observation period should include a watchful waiting which will cover regular microscopic examinations, regular audiologic follow up, debridement of crusts, and so on.

During this observation period a medical treatment also may take part in case of allergy, or sinonasal disease; or an adenoidectomy should be done when indicated.

When to ventilate?

A ventilation tube should be considered in retractions type II or III, progressive, but also without adhesion, without keratin debris, without bony erosion, and without significant conductive hearing loss. But, the possible complications of ventilation tubes must be remembered.[5]

When to operate?

Surgery is absolutely indicated to prevent early-stage cholesteatoma in cases of retraction pockets with debris, with Herodion formation, or draining.

Indications for surgery cover retraction pockets stage III or IV, with unavoidable accumulation of keratin, repetitive inflammation, in evidence of progression or cholesteatoma, and/or in cases with significant conductive hearing loss.

Surgery of early-stage cholesteatoma (stage IV retraction pocket) may cover, in our hands, transcanal attico-antrotomies with scutum removal and reconstruction, or endaural approaches for attic cholesteatomas; or endaural or retro-auricular approach for sinus cholesteatomas; or retro-auricular approach for tensa cholesteatomas.

In cases with posterosuperior quadrant self-cleaning retractions of stage II, excision of the retraction may be sufficient for a self reparation or it may be reinforced by a cartilage tympanoplasty or a palisade technique, as in stage III-IV retractions.[6,7]

In cases of a retraction of the pars flaccida stage III or over, an atticotomy and cartilage reconstruction is convenient.

Cases with complete atelectasis and adhesion but without hearing loss may be followed up; or if operated, as needed for cases with hearing loss, a mastoid management should also be considered.[8]

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ON-DEMAND SURGICAL TECHNIQUE FOR CHOLESTEATOMA: ATTIC EXPOSITION ANTRUM EXCLUSION

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Introduction

The way to treat cholesteatoma is by surgical removal. Several techniques have been described with variable recurrence rates. Traditionally, they can be classified as open and closed techniques. Open techniques provide adequate control after removal of the cholesteatoma; however, the incidence of infections is higher, they require periodic follow up and the entrance of water is not advisable. A closed technique has a lower rate of infections and allows the entrance of water, but the rate of residual and recurrent cholesteatoma is higher and it also requires periodic follow up.

Several variables must be taken into account when choosing the surgical technique: anatomy of the temporal bone – particularly the aeration of the mastoid and the status of the mucosa – the kind of cholesteatoma, the presence of co-morbidities, environmental features, the age and so on.

Attic exposition and antrum exclusion (AE-AE) is an on-demand surgical technique for the treatment of cholesteatoma. Olaizola described this open technique as the surgical approach of choice for cholesteatoma.[1] It has several advantages compared to classical open techniques such as canal-wall-down mastoidectomy. The AE-AE totally exposes the attic by drilling the superior wall of the external auditory canal (classical atticotomy through the canal) and excludes the antrum and the mastoid cells by closing the additus with a cartilage graft (Fig. 1). This last maneuver makes the AE-AE similar to an obliterative technique of the mastoid.

The aim of this study is to describe the indications for the AE-AE technique and analyse its long-term outcomes after the surgical removal of the cholesteatoma.

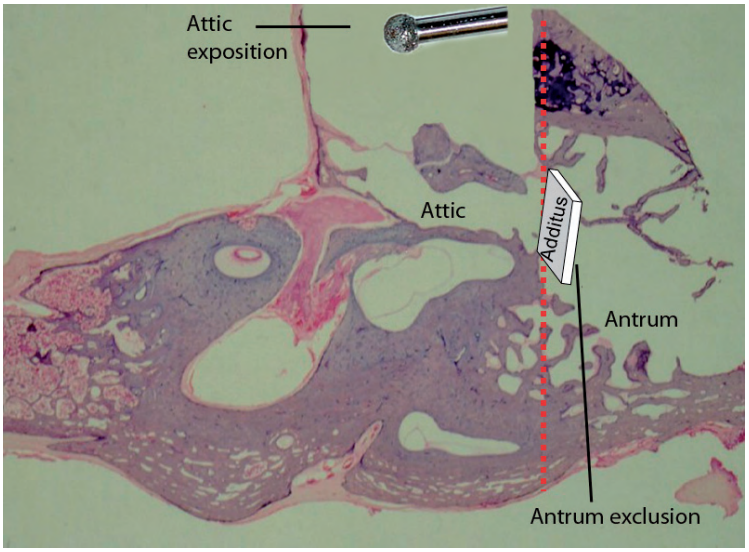

Fig. 1. Histological representation of the AE-AE technique. The red dotted line represents the division between attic and antrum, blocked by a cartilage.

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Edited by Haruo Takahashi

Material and methods

Demographics

A retrospective study was carried out in a tertiary center, including patients who have undergone an AE-AE surgery to remove a primary acquired cholesteatoma from November 2003 to January 2010. We analysed 42 patients whose ages range from six to 68 years old (average: 48 years), of which 25 were men and 17 woman; with an average follow-up period of 2.58 years (range: six months-seven years).

Indications, surgery and follow up

Depending on the features and location of the cholesteatoma, two types of AE-AE can be carried out. Either technique is applied based on the following criteria:

- AE-AE: 25 patients with cholesteatoma located in the attic, either medial or lateral to the malleus or incus, not invading the additus, with or without erosion of the atical wall (Fig. 2A)
- Extended AE-AE: 17 patients diagnosed of cholesteatoma entering into the additus until de mastoid antrum, where a well-defined cholesteatoma that does not damage the labyrinth (Fig. 2B)

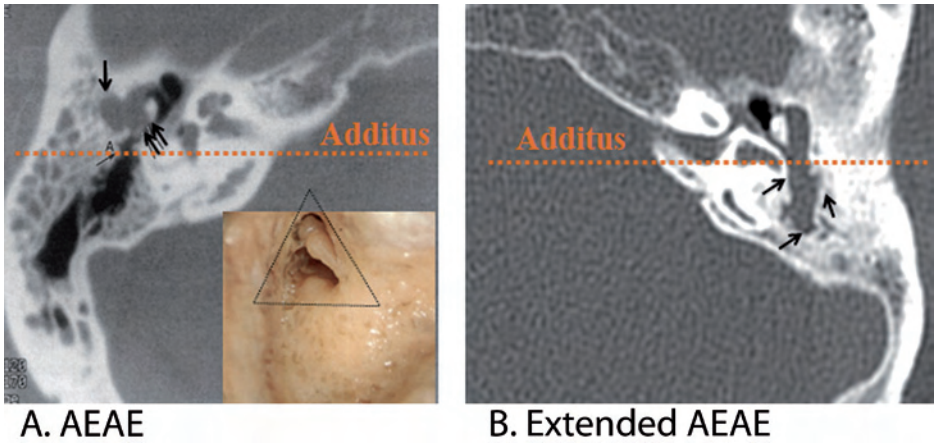

Fig. 2. A. AE-AE indication represented in a CT scan. The cholesteatoma is located in the attic and does not invade the additus. *B.* Modified AE-AE represented on a CT scan. The cholesteatoma is located on the mastoid antrum.

In case the cholesteatoma did not fulfil these criteria, other surgical techniques were carried out to remove it.

Olaizola,[1] Ramirez-Camacho[2] and López Villarejo[3] report the surgical technique in detail (Fig. 3).

Patients are followed up ten days after surgery, once a month during six months and annually afterwards. Physical examination (Fig. 4) and radiology (CT scan and non-EPI MRI since 2009) are undertaken, the latter after two years of follow-up. Nowadays, non-EPI MRI[4,5] is a non-invasive reliable test to diagnose recurrent or residual cholesteatoma.

Statistical analysis

SPSS v15.0 is used to determine the rate of recurrence, pathological findings during surgery and otomicroscopic examination, auditory performance and the reliability of imaging tests such as CT and non-EPI MRI.

Results

Surgical details, such as extension and location of the cholesteatoma, lesions of the ossicular chain and the presence or absence of the membranous labyrinth, are summarized in Table 1.

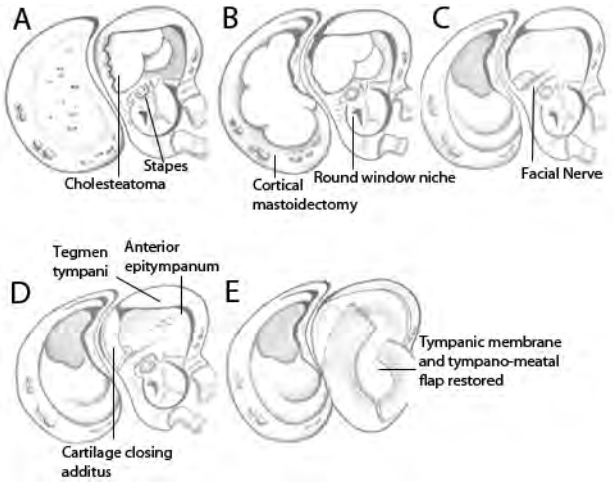

Fig. 3. Surgical procedure for performing the AE-AE. (Figure adapted with permission from Lopez Villarejo P et al.[3])

Pre-operative mean auditory thresholds are 41.04 for the air conductive and 26.3 dB for the bone conductive. The pre-operative air-bone gap is 29.69 ± 21.39 dB (range, 5-100 dB).

Post-operative auditory thresholds are 51,50 for the air conductive and 27.68 for the bone conductive. The post-operative air-bone gap is 25.17 ± 17.74 dB (range, 1,25-100 dB). The average difference pre-post-operative is 4.5 ± 16.9 dB (range [-22.5]- 56.6 dB).

From the forty-two patients included in the study, two patients have been diagnosed of residual cholesteatoma located in the posterior region of the mesotympanum (4.8%); both detected four years after surgery. They have been removed under local anesthesia through a trans-canal approach. No recurrences have been observed to date.

Imaging tests have been used to diagnose residual cholesteatoma, especially in those areas not visible through the otoscopy such as the mastoid. Before the non-EPI MRI was available, a CT was requested if a recurrence was suspected during the physical examination. It was carried out in six cases (14.3%). In one case (2.4%) out of six there was no evidence of disease in the mastoid cavity; in the rest of them (11.9%) there was evidence of mastoid occupation. A surgical second look was carried out in two cases, finding inflammatory tissue in the antrum. In another two cases a non-EPI MRI showed no evidence of disease and in the last one, a wait-and-see option was chosen. From January 2009, MRI's are conducted after two years of follow up. To date, it has been executed in 12/42 cases (28.6%). No signs of cholesteatoma have been described.

No pathological findings were encountered during otoscopy in 35/42 cases (83.3%). In the rest, the following lesions were observed: atelectasis in the mesotympanum in two cases (4.8%), a polyp in the posterior

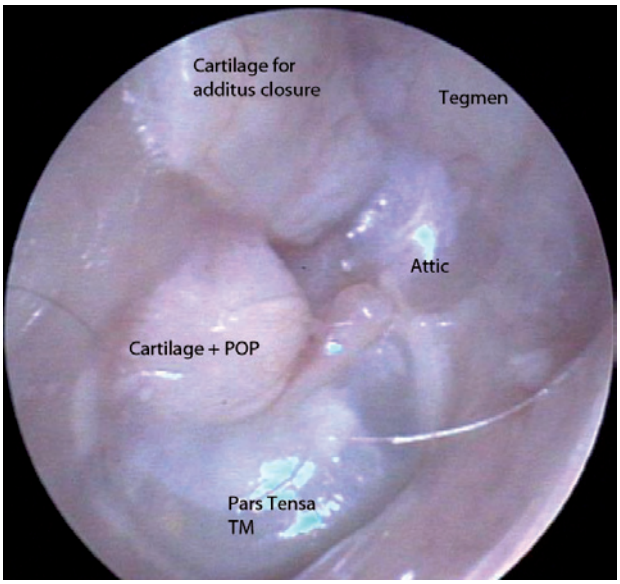

Fig. 4. Otoscopy of a three-month post-op AE-AE.

Table 1: Cholesteatoma characteristics between the two groups.

	AEAE	Extended AEAE	Total
Extension and localization			
Mesotympanum	8,3%	–	4,9%
Attic	62,5%	17,6%	43,9%
Attic and mesotympanum	25%	29,4%	26,8%
Antrum and attic	4,2%	41,2%	19,5%
Antrum, attic and mesotympanum	–	11,8%	4,9%
Ossicular chain integrity			
No damage	41,7%	17,6%	31%
Lesion in malleus and incus	50%	35,3%	42,9%
Lesion in malleus, incus and stapes	8,3%	29,4%	19%
Lesion in incus and stapes	–	17,6%	7,1%
Fistulae			
None	95,8%	82,4%	90,2%
Horizontal Semicircular Canal	–	11,8%	4,9%
Tegmen	4,2%	5,9%	4,9%

wall of the external auditory canal (2.4%), two cases of attic membranous partition (4.8%) and two cases of serous tympanic cavity occupation (4.8%).

Discussion

The principal objective of the cholesteatoma surgery is its removal. The surgical technique chosen for the procedure must provide a full visualization of the cholesteatoma. AE-AE, when properly indicated, fulfils this requirement. The atticotomy through the EAC gives excellent exposure of the attic. Also, the mastoidectomy in the context of an extended AE-AE allows a good control of a cholesteatoma that invades this region.

The ratio of residual cholesteatoma varies from 5% to 57%,[6-9] depending on the authors and the selected technique. In our series we had no residual cholesteatoma located in the attic or mastoid area. The two cases of residual cholesteatoma were located on the sinus tympani and were diagnosed four years after the AE-AE. We estimate these recurrences were not directly associated with the surgical technique, but with the poor visualization of the posterior area of the mesotympanum.[10] These results are similar to those of other authors who have used the AE-AE to treat cholesteatoma.[3,11]

The main advantage of this technique to avoid recurrence is the attic exposition towards the external auditory canal. This maneuver, aside from controlling the attic adequately to reduce the risk of residual cholesteatoma, also prevents the formation of retraction pockets, thereby reducing the risk of recurrent cholesteatoma. Also, the surgery creates a micro-tympanic cavity formed by the mesotympanic and hypotympanic space. This means less air volume for the Eustachian tube to ventilate, reducing the possibility of pars tensa atelectasis.

Antrum exclusion 'obliterates' the mastoid cavity. The normal epithelium is plain, with no cilium and no secretory glands.[12] Therefore, its obliteration should not generate any effusion unless inflammatory disease is left on the mastoid cavity. In our series, some cases of effusion have been detected either by CT scan or MRI. In any case, no pathological findings or clinical symptoms have been found. In the same way obliterative techniques,[13] avoiding an extensive removal of the EAC walls, have a faster recovery in terms of cicatrization, allowing the entrance of water, and shortening the follow-up period. Only two cases of othorrea have been recorded in our group.

Since 2009 we are using a specific diffusion-MRI protocol to detect cholesteatoma, as described by De Foer.[4,5] This technique is especially effective when the lesion is over two mm, and gives the surgeon the chance to avoid a second-look surgery. For this reason, we recommend this exploration two years after the initial cholesteatoma surgery.

It is very important to highlight that the AE-AE does not change the surgical attitude towards ossicular reconstruction, as it depends only on the condition of the remaining ossicules. The global analysis shows that the hearing levels during the pre- and post-surgical procedure are virtually the same.

Conclusion

AE-AE is a variant of an open technique, in which the attic is exposed through the EAC while the antrum is excluded from the tympanic cavity. With this surgical procedure, the disease control is excellent with low recurrence rates of cholesteatoma, allowing the entrance of water and offering a good quality of life.

Diffusion MRI is a reliable technique for cholesteatoma follow up and it is helpful to surgeons in avoiding unnecessary surgical procedures.

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USE OF CARTILAGE IN TYMPANOPLASTY

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Introduction

The use of cartilage in tympanoplasty was presented as used in author's otologic practice. It includes treatment and prevention of attic retraction, reconstruction of tympanic membrane (TM) perforation, especially revision cases and atelectatic TM reconstructions, placement over the ossicular prosthesis to prevent extrusion, and reduction of mastoid bowl size in canal wall down mastoidectomy. Illustrative cases were presented for the use of cartilage for these purposes.

Treatment and prevention of attic retraction

When there is attic retraction pocket without cholesteatoma, the tympanomeatal flap is developed to beyond the 12-o'clock position to 1 or 2 o'clock. The flap is extended to elevate the TM over the manubrium to the umbo (Fig. 1.1). The perichondrium/cartilage island flap is made either from tragal or conchal cartilage (Fig. 1.2) and grafted to the retraction defect. This procedure can prevent development of cholesteatoma.

Pitfalls of this procedure include: continuation of the Eustachian tube dysfunction ending up with atelectatic TM or recurrence of retraction pocket, or, even worse, covering up deeper cholesteatoma.

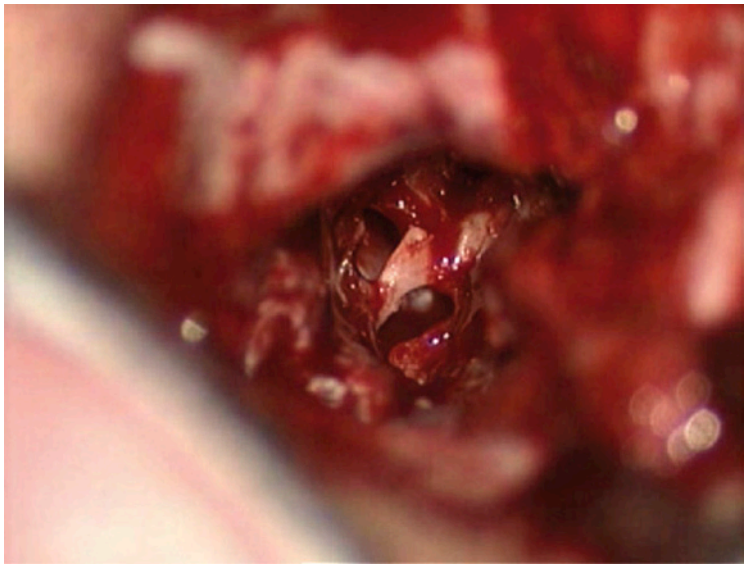

Fig. 1.1. The retraction pocket is elevated with an extended tympanomeatal flap down to the manubrium.

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Edited by Haruo Takahashi

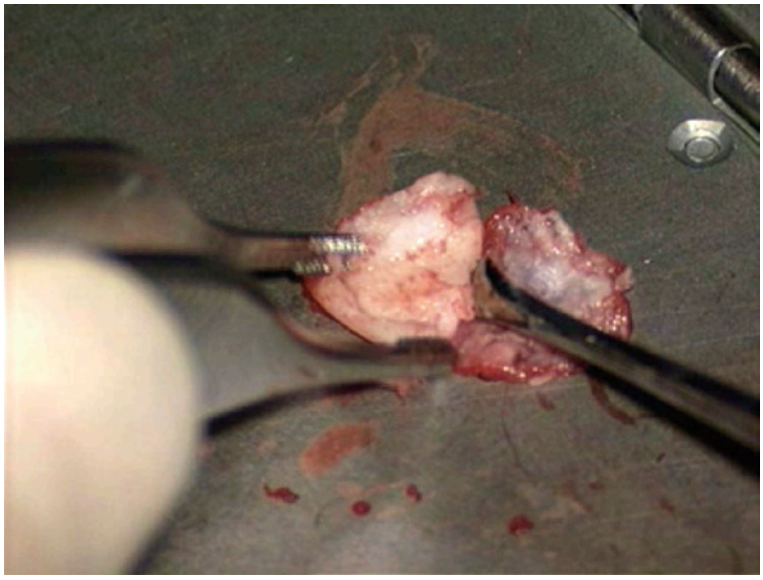

Fig. 1.2. Perichondrium/cartilage island flap from tragus to be grafted to the retraction pocket.

Reconstruction of TM perforation

Recently there have been many reports on the superior outcome of the use of cartilage for reconstruction of TM perforation. Cartilage is used as perichondrium/cartilage island flap[1,2] or palisade.[3] These studies revealed that cartilage with or without perichondrium is either superior or equal to a fascia graft.[1,2]

For posterior or central perforation, the edges of perforation are denuded of epithelial tissue (Fig. 2.1.1), a perichondrium/cartilage island flap is made (Fig. 2.1.2) and grafted as medial (underlay) graft (Figs. 2.1.3 and 2.1.4) with Gelfoam support under the graft. For anterior or subtotal TM perforation, a perichondrium/cartilage island graft can be placed as a medio-lateral technique.[4]

The use of cartilage is the more important for revision tympanoplasty and reconstruction of atelectatic TM.

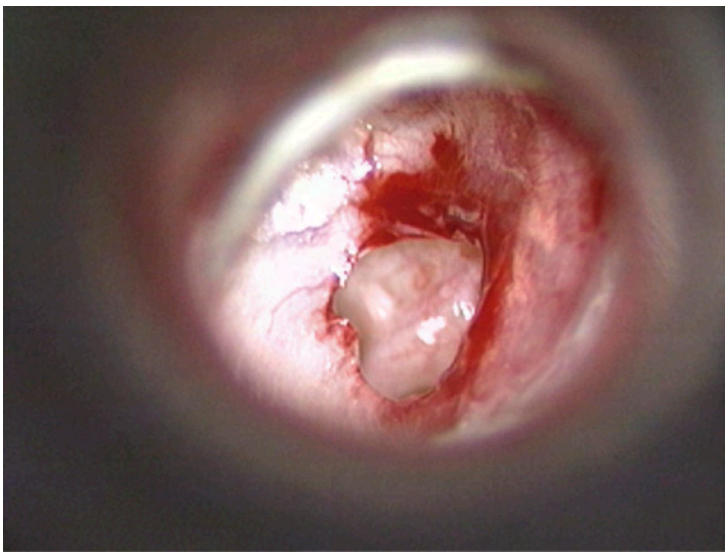

Fig. 2.1.1. Surgical steps to repair posterior or central TM perforation with medial graft tympanoplasty. First, the edges of perforation are denuded.

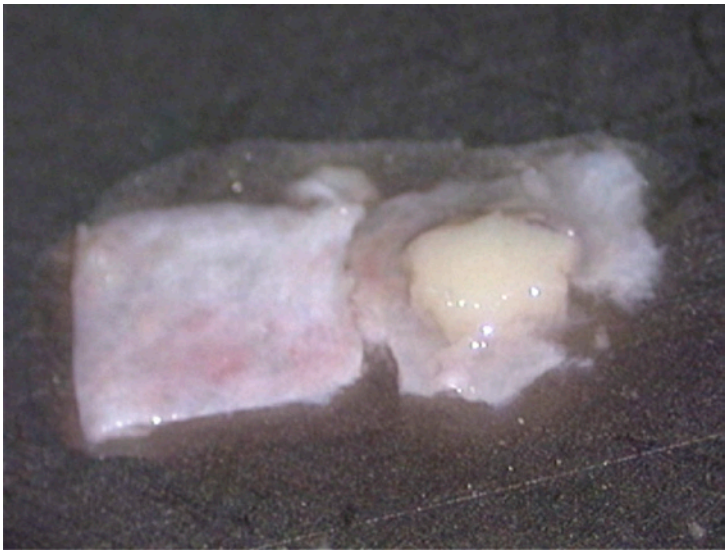

Fig. 2.1.2. The perichondrium/cartilage island graft is prepared.

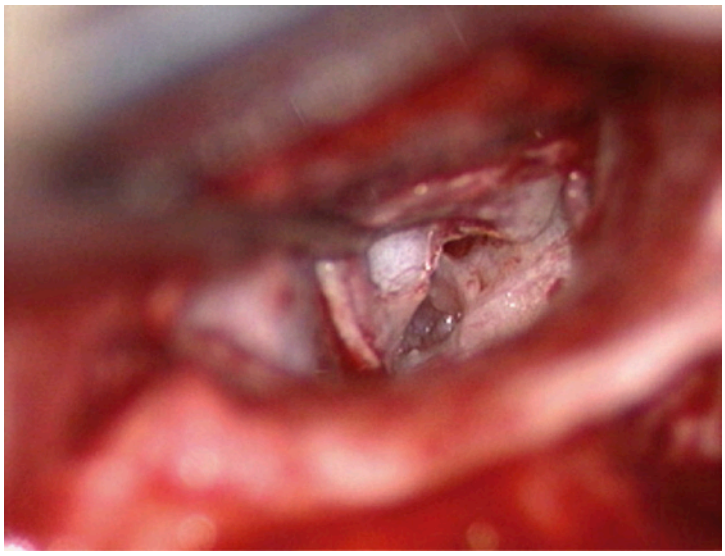

Fig. 2.1.3. The tympanomeatal flap is elevated.

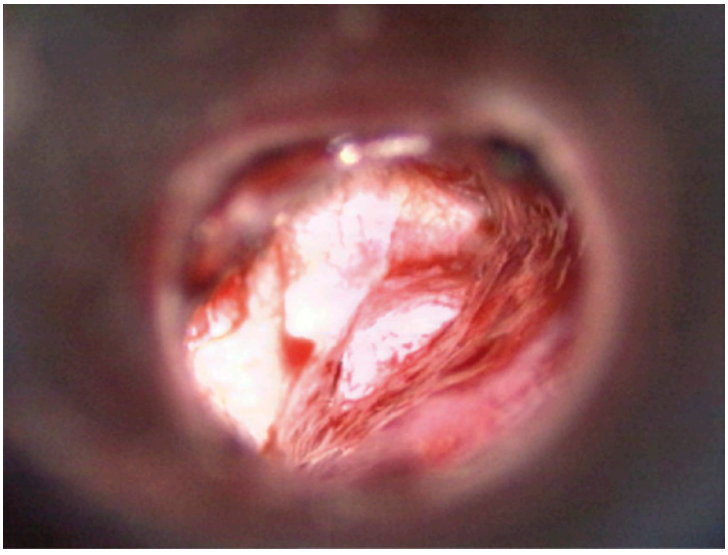

Fig. 2.1.4. The graft is placed medial to the TM perforation.

Placement over the ossicular prosthesis to prevent extrusion

When ossiculoplasty is performed using a titanium or hydroxyapatite partial ossicular replacement prosthesis (PORP) or a total ossicular replacement prosthesis (TORP), the cartilage graft is essential to prevent extrusion (Figs. 3.1 and 3.2).

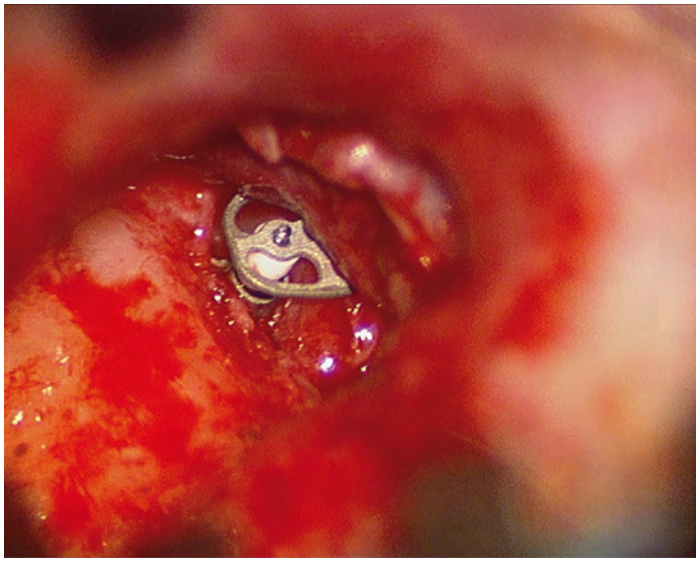

Fig. 3.1. The titanium total ossicular replacement prosthesis is in place.

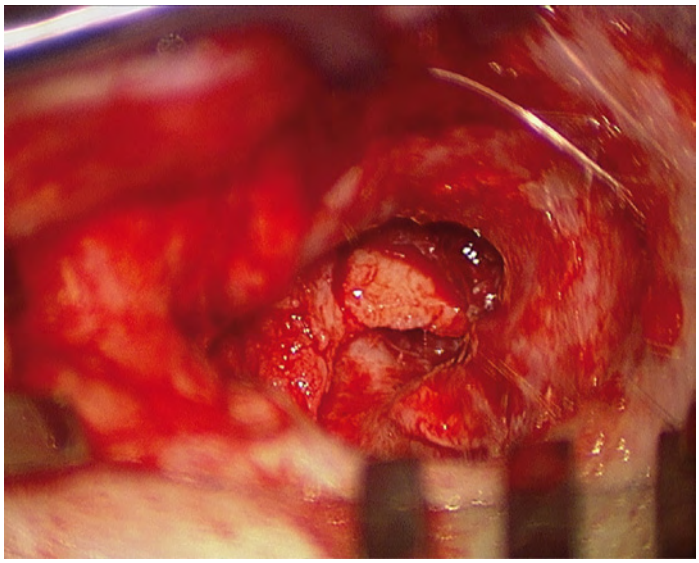

Fig. 3.2. The cartilage is grafted over the prosthesis to prevent extrusion.

Reduction of mastoid bowl size in canal wall down mastoidectomy

At the time of canal-wall-down mastoidectomy, cartilage removed from the concha to perform wide meatoplasty is cut and packed into the mastoid bowel reducing the size of the bowel. This is in effect partial reconstruction of the posterior canal wall.[5]

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TREATMENT OF CHOLESTEATOMA-INDUCED LABYRINTHINE FISTULA

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Introduction

Labyrinthine fistula (LF) is the most common complication of cholesteatoma, with an estimated incidence of 0.2-12.9%.[1-4] In 90% of the cases, LF occurs in the lateral semicircular canal (the remaining 10% involving the basal turn of the cochlea).

Symptoms such as vertigo, sensorineural hearing loss and even acute meningitis may ensue if a fistula is left untreated for a long period of time. Complications of cholesteatoma-induced LF can be divided into pathological and functional. The most common pathological consequence is a higher residual rate due to incomplete matrix removal; total deafness is the most threatening functional complication.

Adequate surgical management of LF is still debated, the majority of authors adopting routinely the open tympanoplasty technique;[5-8] this attitude is nowadays slowly changing, some surgeons preferring the closed technique.[9-11] A further option consists of applying the middle ear exclusion technique.[12]

The size of the fistula and the hearing threshold, as pure tone average (PTA), are the main factors to take in account in the choice of the surgical approach.

When the surgeon has to deal with a type I fistula (according to Dornhoffer and Milewski's fistula grading system[13]) (Fig. 1), being smaller than two mm with a mild to moderate conductive or mixed hearing loss, in our opinion the preferable surgical approach is intact-canal-wall tympanoplasty with ossicular reconstruction by mean of passive middle ear implant, associated to a fistulaplasty.

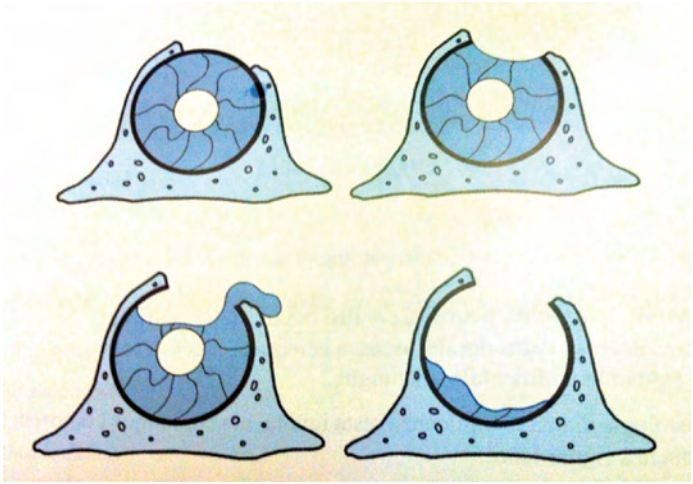

Fig. 1. Classification of the cholesteatoma-associated labyrinthine fistula according to Dornhoffer and Milewski. Type 1: bone erosion without penetration of the endosteal layer of the canal; Type 2a: bone erosion with penetration of the endosteal layer of the canal; Type 2b: as type 2a, with spontaneous perilymphatic leakage; Type 3: complete invasion of the canal.

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Edited by Haruo Takahashi

When considering a type IIb or III fistula, with a concomitant profound hearing loss or anacusis, a middle ear exclusion technique with fistulaplasty may be proposed, simultaneously associated to a bone conduction hearing prosthesis such as B.A.H.A. or similar, provided that the pre-operative audiological tests have considered it as a practicable option for rehabilitating as single-sided deafness.

Another point to take into consideration is to leave or not the cholesteatoma matrix over the exposed labyrinthine spaces. Traditionally, the majority of otologists advised to leave the cholesteatoma matrix over the bone defect, in order not to jeopardize the delicate structures of the membranous labyrinth, therefore avoiding a jatrogenic sensorineural hearing loss.[11,14]

On the other hand, leaving residual cholesteatoma tissue may increase the risk for the infective/inflammatory process to grow or spread from the cholesteatoma matrix to the perilymph and the membranous labyrinth, with a possible consequent late-occurring sensorineural hearing loss.[1,13,15,16]

In our personal experience, out of 153 patients who underwent surgical treatment for cholesteatoma at Sant'Andrea Hospital Sapienza University of Rome between 2005 and 2011, 22 LF (14.4%) were found. The location of the LF was in 85% the lateral semicircular canal (dome 70%, superior aspect 25%, anterior aspect 5%), in 10% the superior semicircular canal and in 5% of the cases the posterior semicircular canal.

The surgical technique was canal-wall-up in 80% of the patients, the modified radical cavity in 15% of them and the subtotal petrosectomy with middle ear exclusion by blind-sac closure of the external auditory canal in 5%.

Fistula repair was performed as one of the last surgical steps, after removal of the major cholesteatoma mass and before performing tympanic or ossicular reconstruction. Our three-layer sealing procedure consisted of:

- Gentle dissection of the matrix over the bony defect, helped by fine suction tip and dissector;
- Immediate covering with temporalis fascia;
- Placement of bone paté collected from the cortical bone and successive fixation with fibrin glue (Fig. 2).

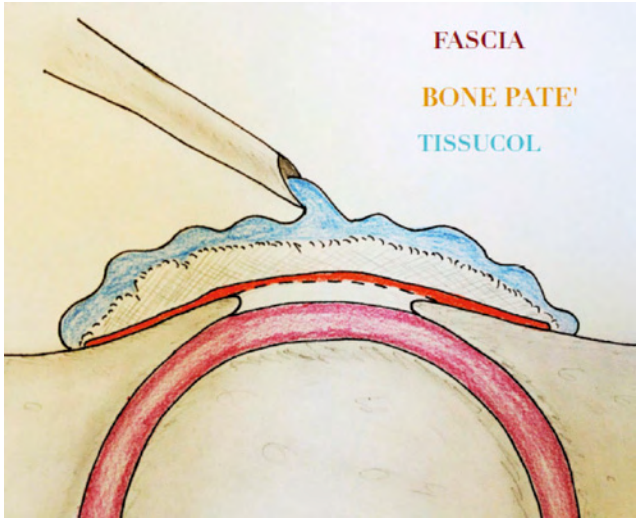

Fig.2. Three-layer fistulaplasty technique.

From a functional point of view, among these 22 patients, two were pre-operatively deaf, 12 (54.6%) showed a mixed hearing loss, seven (31.9%) a conductive hearing loss and one (4.5 %) had a sensorineural hearing loss. Post-operatively, one patient developed anacusis.

In most of the cases presenting with a conductive and mixed hearing loss, a serviceable post-operative hearing was preserved. Only one patient with a pre-operative conductive hearing loss developed anacusis after the surgery. Also, the only patient in our series affected with a sensorineural hearing loss developed anacusis.

Conclusion

Our experience seems to suggest that hearing function may be preserved in the majority of the cases, especially in early stage (grade I), associated with a conductive and mixed hearing loss.

Therefore, when dealing with small fistulas (< 2 mm), it is our attitude to apply always a canal-up technique with fistulaplasty.

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EFFICACY OF 3D FLAIR MRI FINDINGS IN EVALUATING CHOLESTEATOMA WITH LABYRINTHINE FISTULAE

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Introduction

Surgical management of cholesteatomas with labyrinthine fistulae has been reported and several treatment techniques have been advocated. These issues may have arisen as a result of selection bias in the reported techniques: there is neither a widely-used staging system for fistulae[1] nor any uniform surgical technique that can be adopted for all cases.[2] Computed tomography (CT) examinations are often conducted for preoperative evaluation for middle ear surgery. Additionally, diffusion-weighted magnetic resonance imaging (MRI) has been shown to be useful for the detection of cholesteatomas.[3] Three-dimensional (3D)- Fluid-attenuated inversion recovery (FLAIR) MRI can aid in identification of labyrinthitis[4] or with cholesteatoma and labyrinthine fistula.[5]

We have evaluated pre-operative 3D FLAIR MRI findings in relation to clinical features in cases of middle-ear cholesteatoma with a labyrinthine fistula, and to evaluate the suitability of MRI findings regarding fistula status and surgical management.

Materials and methods

Twenty-eight patients who underwent surgery for middle-ar cholesteatoma with one or more labyrinthine fistulae confirmed by CT were studied. Pre-operative imaging analysis was performed using 3D-FLAIR MRI before and after intravenous administration of a single dose of gadolinium. Fistula size measured by CT and the signal intensity (SI) in the affected lesion in the inner ear after contrast enhancement were evaluated with respect to the clinical features and surgical findings. SI was measured in each affected ear and in the cerebellum in the most artifact-free area, and The SI ratio (SIR) between the affected lesion and the cerebellum was then estimated.

Clinical features included hearing threshold of pre-operative bone conduction, and existence of fistula symptoms or active infection. Fistula status was classified into one of the following three stages: (I) no involvement of the endosteal membrane; (II) cholesteatoma matrix invading the endosteal membrane, so that removal of matrix would open the perilymphatic space; or (III) direct attachment or invasion of the cholesteatoma matrix or granulation tissue within the membranous labyrinth.

Results

Example image of CT and 3D-FLAIR MRI in a case with a labyrinthine fistula in the lateral semicircular canal are shown in Figure 1. There was no correlation between fistula size and the SIR. The hearing threshold determined by pre-operative bone conduction correlated with the SIR, especially in patients with acute sensorineural hearing loss, but it did not correlate with fistula size. Patients with fistula symptoms had a significantly higher SIR than those without symptoms (Table 1), and similar findings were observed in patients

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Edited by Haruo Takahashi

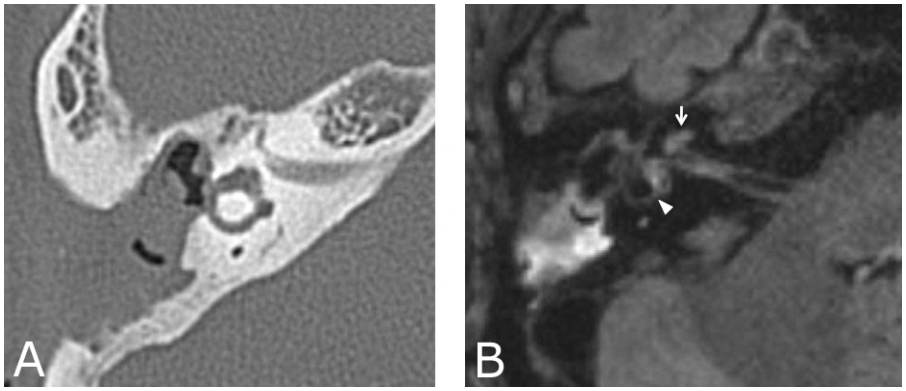

Fig. 1. Example images of CT (A) and 3D-FLAIR MRI (B) in a case with a labyrinthine fistula in the lateral semicircular canal. High signals in the lateral semicircular canal (arrowhead in B) and in the cochlea (arrow in B) of the affected ear.

Table 1. Fistula sizes and SIRs in the fistula lesion of patients with and without fistula symptoms or active infection

		Fistula size on CT mm (average)	SIR on MRI (average)	
Fistula symptoms	positive	0.9 ~ 5.1 (2.7)	0.71 ~ 1.38 (0.95)	*
	negative	0.8 ~ 7.1 (2.7)	0.16 ~ 1.35 (0.47)	
Active infection	positive	1.1 ~ 5.1 (2.9)	0.71 ~ 1.38 (0.94)	**
	negative	0.8 ~ 7.1 (2.6)	0.16 ~ 1.56 (0.52)	

SIR signal: intensity ratio
* *p<0.05*, ** *p<0.01*

Table 2. Relationship between fistula stages, fistula sizes and SIRs in the fistula lesion

Stage	Fistula size on CT mm (average)		SIR on MRI (average)	
I	0.9 ~ 3.9 (1.7)	*	0.2 ~ 0.77 (0.92)	**
II	0.8 ~ 5.3 (2.7)		0.16 ~ 1.35 (0.59)	
III	1.1 ~ 7.1 (3.8)		0.27 ~ 1.56 (0.33)	

SIR signal: intensity ratio
* *p<0.05*, ** *p<0.01*

with an active infection. Patients with a larger fistula or higher SIR tended to have a deeper fistula and a more adhesive fistula matrix at operation (Table 2).

Discussion

Removal of the matrix at initial surgery has been proposed for patients with small fistulae.[6] In the present study, sizes of fistulae detected by CT images did not related to SIRs, and the stronger signals were observed for the smaller fistulae. Thus, severe labyrinthitis can occur even in patients with a small fistula, and 3D-FLAIR images can help evaluate the severity of intra-labyrinth disturbances. Removal of the cholesteatoma might be more successful when the ear is not actively infected.[6,7] In other words, removal of the cholesteatoma matrix should be considered carefully when there is active inflammation, as this can accelerate the erosion of labyrinthine bone, and such inflammation within the inner ear could be observed on 3D-FLAIR images in the present study.

Higher elevations of hearing threshold were observed in patients with stronger SIRs, including those with preoperative acute SNHL. The prevalence of fistula symptoms was higher in those with stronger SIRs. This finding suggests that 3D-FLAIR signal represents the degree of labyrinthitis that can cause dysequilibrium.

We removed the cholesteatoma matrix at initial surgery in patients with relatively little adhesion of the matrix to the membranous labyrinth, which related to SIR. Removal of the matrix led to opening of the peri-lymphatic space in some cases with relatively weaker SIR, however, postoperative clinical symptoms including vertigo were observed less in these patients. New bone formation in the fistula site was frequently observed post-operatively in patients with relatively strong SIR, which indicated more severe inflammation in the lesions. Our impression is that 3D-FLAIR evaluation was beneficial for surgical management, especially in patients with large fistulae or symptoms related to inner ear disturbances.

Conclusion

SIR was more strongly correlated than CT findings to the clinical status of patients with labyrinthine fistulae caused by cholesteatoma. Adhesion of the cholesteatoma matrix to the membranous labyrinth correlated with the SIR. Information provided by 3D-FLAIR image is valuable in pre-operative evaluation of the status of labyrinthine fistulae caused by cholesteatoma and a useful indicator of fistulae stage and surgical management.

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STAPES SURGERY AND COCHLEAR IMPLANT SURGERY FOR SEVERE ORTOSCLEROSIS

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Introduction

Profound deafness has received increasing attention, because of the availability of cochlear implants (CI). Consequently, it is especially important to remember that a 'blank' audiogram does not necessarily mean absence of hearing. Severe otosclerosis (far-advanced otosclerosis; FAO) generally involves air conduction (AC) levels worse than 85 dB, and bone conduction (BC) levels beyond the limits of the audiometer.[1-4] If AC levels exceed 85 dB but BC levels are measurable at some frequencies but worse than 30 dB, the condition is called advanced otosclerosis (AO). Failure to recognize FAO or AO may result in unnecessary CI surgery.

Materials and methods

A retrospective analysis was conducted of the clinical charts of all patients who received stapes surgery (n = 306) and CI surgery (n = 536) at Osaka and Kinki University Hospitals from 1992 to 2012. Stapes surgery involved 210 ears in females and 96 ears in males. Otosclerosis accounted for 80% of the the stapes surgery. Objective improvement was noted in pure-tone audiogram (PTA), and subjective patients' satisfaction with amplification was the real measure of success because the stapes surgery was performed to restore a serviceable hearing with Hearing aid (HA) for these FAO and AO patients.

Results

Among 306 stapes surgery cases, one patient (NS, 45 years old, male) with FAO received stapedotomy on the right ear, and another patient (MS, 56 years old, male) with AO received bilateral stapedotomy. Both patients had a positive family history of progressive hearing loss. MS's daughter (KM, 28 years old) received partial stapedectomy on the left ear, and the result was excellent. AC levels were worse than 85 dB bilaterally in both patient, and BC levels were not measurable at most (not all) frequencies. The past audiograms and the family history help us to diagnose FAO and AO. Pre-operatively, both patients (NS and MA) were not successful hearing aid (HA) users, although both continued to use a HA anyway. Post-operatively, MS does not need HA any longer, while NS is still wearing HA unsuccessfully and considering CI surgery the left ear.

Among 536 CI surgery cases, just one patient (UH, 52 years old, male) had been found to have the history of otosclerosis preoperatively, and has been a good CI user postoperatively (Fig. 1A). HRCT demonstrated a massive sclerotic lesion bilaterally, indicating the presence of cochlear otosclerosis (Fig. 2A). Past audiograms

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Cholesteatoma and Ear Surgery – An Update, pp. 111-113
Edited by Haruo Takahashi

clearly suggest the presence of an air-bone gap and a progressive nature of HL (Fig. 2B). After cochleostomy onto the promontory, the scala tympani was found to be filled with soft connective tissues. A full insertion of CI24RCS electrodes into the scala vestibuli was successfully completed. Among 2558 CI surgery cases, bilateral otosclerosis accounted for just 1% of the causes of deafness in Japan (Fig. 1B), according to a survey by the Cochlear Corporation in 2006.

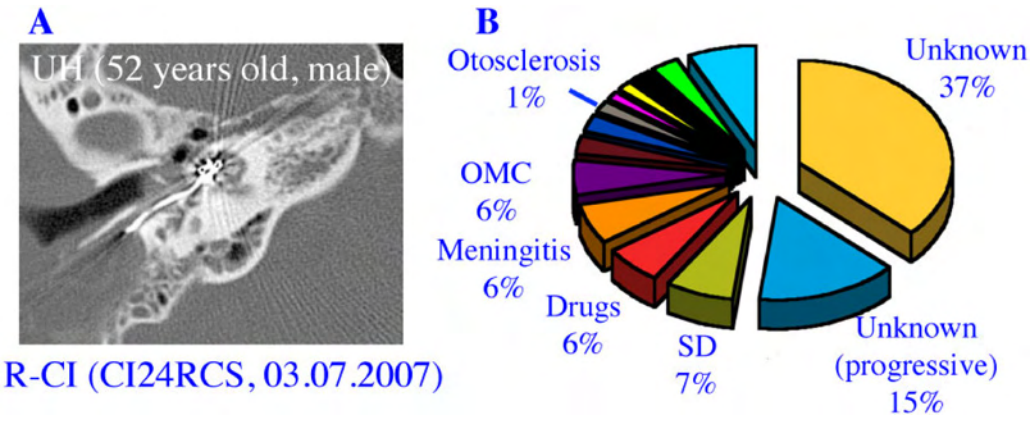

Fig. 1. Profound hearing loss caused by otosclerosis. A: a case of the CI surgery with FAO; B: the causes of deafness in Japanese CI cases (2006).

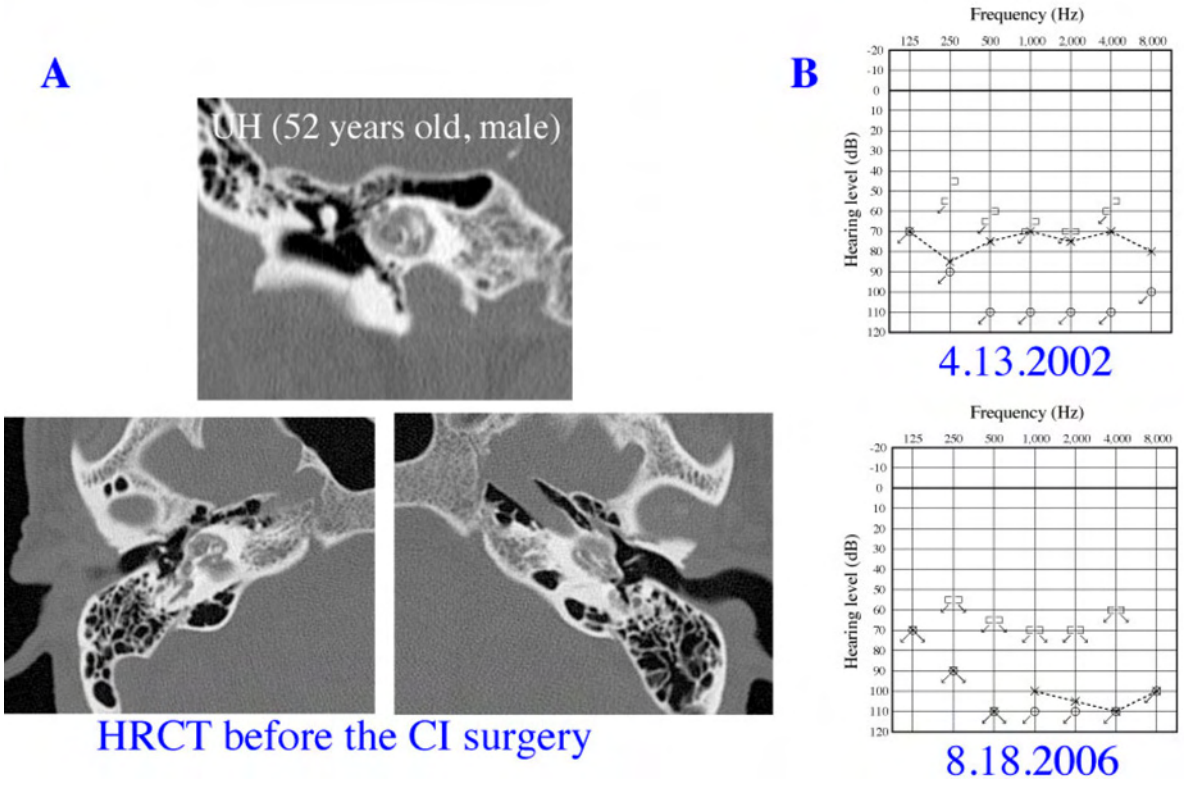

Fig. 2. HRCT and PTA of a patient with FAO who received CI surgery. A: massive sclerotic lesions within bilateral cochlea; B: past audiograms suggesting a progressive hearing loss.

Discussion

A convincing histological explanation for increased bone-conduction threshold in FAO remains an issue for continued investigation. There are two types of otosclerosis described: a conductive disturbance limited to specific areas of the oval and round windows, and a more aggressive form called ‘cochlear otosclerosis’ with

multiple foci developed relatively early in life. No correlation between BC thresholds and size of the lesion, activity of the lesion, involvement of endosteum or presence of a round window lesion in otosclerosis was found, while moderate diffuse loss of hair cells and cochlear neurons in the basal turn, and strial atrophy near the foci of otosclerosis were reported in FAO patients.

Sheehy[2] published specific diagnostic clues for FAO: 1) positive family history for otosclerosis; 2) progressive hearing loss beginning in early adult life; 3) paracusis during the early stage of the disease; 4) past use of bone-conduction hearing aid; 5) previous audiograms showing an air-bone gap. In addition, the following criteria can be obtained from the physical examination: 1) normal voice; 2) positive Schwarze's sign; 3) evidence of otosclerosis on HRCT; 4) a Weber test lateralizing to the poor ear or a negative Rinne test by a 512-Hz tuning fork; 5) no other apparent cause for hearing impairment. The diagnosis is just presumptive and can be confirmed only at surgery. All of our cases showed a positive family history of hearing loss, a progressive hearing loss on the past audiograms, and sclerotic findings of cochlea on HRCT.

Patients with FAO may appear to be suffering from profound sensorineural hearing loss and are frequently directed to CI programs. Specific clues shown above can lead the clinician to suspect FAO, and some FAO patients who had been unable to use a hearing aid (HA) preoperatively obtained serviceable hearing with a HA after the surgery.[1-4]

The most gratifying aspect of the stapes surgery for severe otosclerosis (FAO and AO) should be converting the patients' hearing from non-serviceable to serviceable with HA. The patients must be aware not only of the risks of the procedure, but also of the relatively limited goals. On the basis of the conventional criteria for stapedectomy surgery, objective results would be sometimes disappointing in FAO. However, some FAO patients clearly do benefit from the surgery and show marked improvement in HA performance. The success rate was reported to range within 70-100%.[1-4] If a successful outcome is not achieved, the patient might be suitable for the CI surgery.

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TREATMENT OF SEVERE OTOSCLEROSIS: COCHLEAR IMPLANTATION, STAPEDOTOMY, AND OTHER OPTIONS

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Introduction

Severe otosclerosis is relatively rare in Japan compared with Western countries. In European hospitals, otosclerosis was listed as the cause of deafness in about 7% of all patients who received cochlear implantation.[1] In Japan, otosclerosis was the only cause of deafness in only 1% of such patients according to Cochlear Company data. Although we only have a small number of cases with severe otosclerosis, we discuss the decision-making process with regard to severe otosclerosis based upon the findings in our cases and the literature.

Report of cases

Case 1

Case 1 is a 62-year-old female. Her hearing threshold was 120 dB in the right ear and 97.5 dB in the left ear (Fig. 1). In her past hearing records, we recognized bone conduction in the low frequency area. CT imaging of the petrous bone showed solely fenestral involvement (Fig. 2). It was classified as a so-called type 1 according to Rotteveel *et al*.[1] Stapedotomy was performed on both ears in 2004 and 2005.

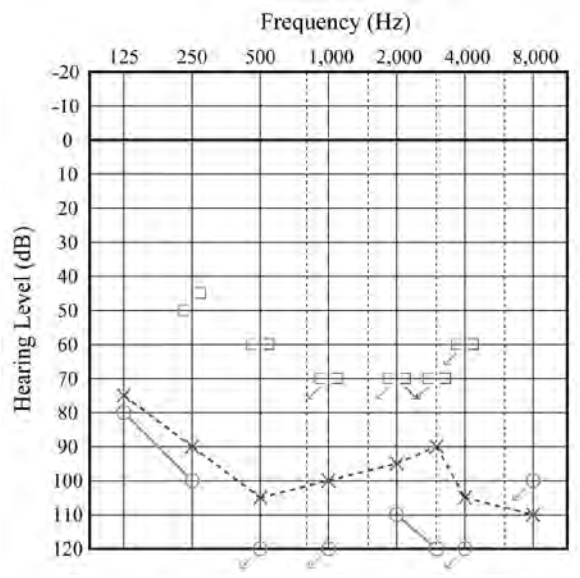

Fig. 1. Pre-operative audiogram of case 1.

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Edited by Haruo Takahashi

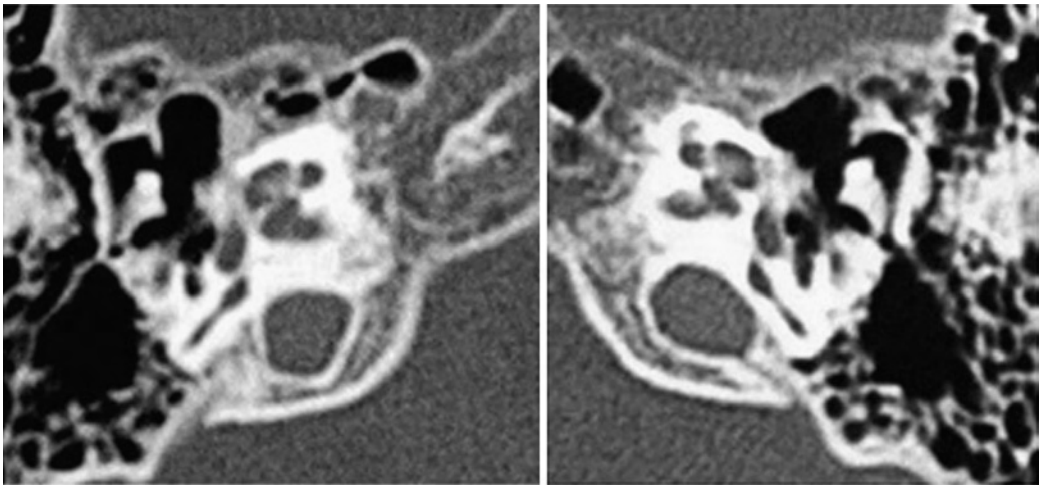

Fig. 2. Pre-operative CT findings (both side ears) of case 1.

Eight years after the first operation, the hearing threshold was 90dB in the right ear and 86.3 dB in the left ear. With hearing aids on both ears, maximum speech discrimination score (SD) was 75%. CT imaging of the petrous bone showed solely fenestral involvement which was the same as before the operation. She was satisfied with her hearing now.

Case 2

Case 2 is a 51-year-old male. His hearing threshold was 107.5 dB in the right ear and scale out in the left ear (Fig. 3). In his past hearing records, we did not observe an air-bone gap. CT imaging of the petrous bone showed diffuse confluent retrofenestral involvement (Fig. 4). It was classified as a so-called type 3 according to Rotteveel *et al*.[1] Cochlear implantation (CI) was performed on the right ear in 2007.

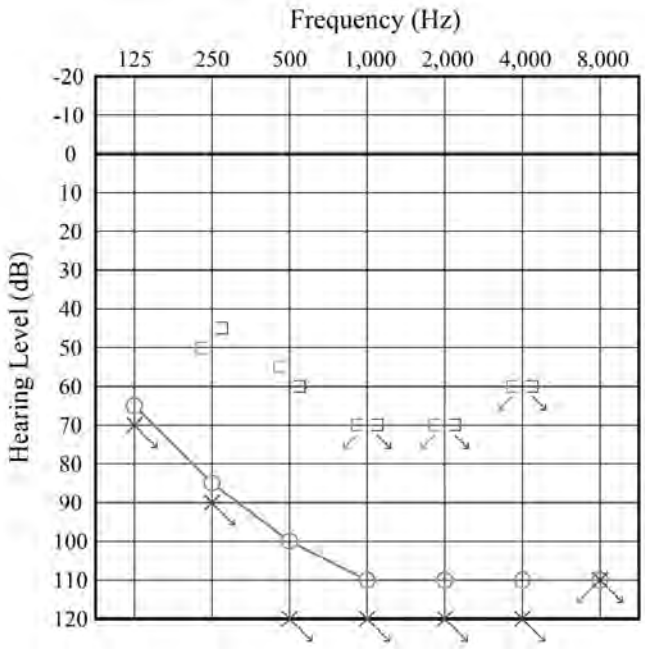

Fig. 3. Pre-operative audiogram of case 2.

Although his surgery was rather difficult because of bleeding and narrow scala tympani, the Cochlear Company device was inserted perfectly. Post-operatively, all electrodes could be used and the rehabilitation

course was good. His maximum SD was 90%. He was called ‘a star patient’ by the president of the Cochlear Company.

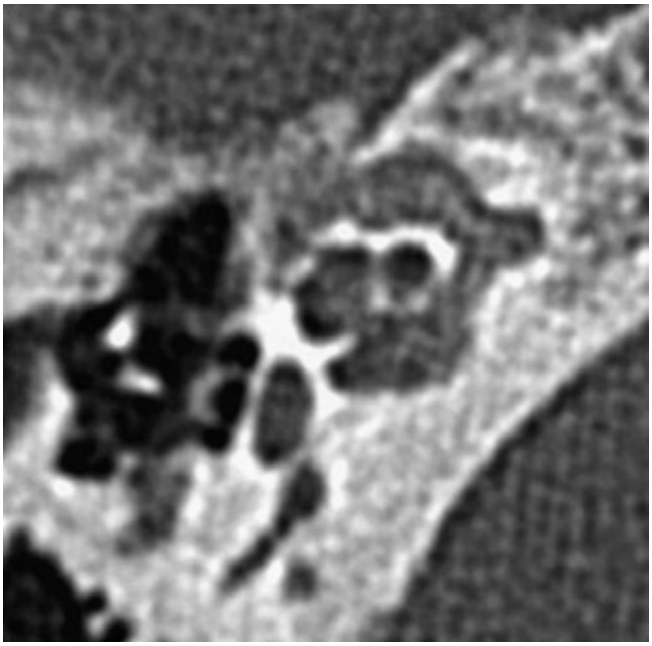

Fig. 4. Pre-operative CT finding (right ear) of case 2.

Case 3

Case 3 is a 59-year-old male. His hearing threshold was 105 dB in the right ear and 100 dB in the left ear. CT imaging of the petrous bone showed a double-ring effect (so-called type 2A according to Rotteveel *et al*.[1]). With a hearing aid on the left ear, his maximum SD was only 15%. Now we are planning a CI for him.

Discussion

In Japan, there are three treatment options for patients with severe otosclerosis.

- Continuing hearing aids;
- Stapedotomy and hearing aid use;
- CI.

Stapedotomy is a relatively simple and inexpensive procedure. However, the results are unpredictable because measurement of bone conduction is often difficult and because an air-bone gap can occur without stapes fixation. When surgery and rehabilitation have been successful, CI has provided excellent results. However, it is an expensive and complex procedure. Surgery is often difficult in patients with progressive cochlear otosclerosis because of the difficulty in achieving full and exact insertion into the perilymphatic space. Furthermore, rehabilitation of such patients is challenging because progressive otosclerotic changes in the cochlea can affect performance of the implant in aspects such as facial nerve stimulation.

If the lesions of the patients with otosclerosis are not progressive and adequate hearing gain is likely to be acquired, then stapedotomy will be recommended. If adequate hearing gain is not likely, then CI will be necessary. CT imaging is expected to be an indicator of the progression of otosclerosis, because the hearing ability is likely to deteriorate when extensive lesions are present in CT imaging. In patients with severe otosclerosis, measurement of bone conduction is sometimes difficult. In such cases, SD with a hearing aid may be helpful. If maximum SD with a hearing aid is poor, then stapedotomy will not be effective. In our hospital, the average maximum SD after CI in all adult patients was 54%; a result that should be considered when planning for CI. From these ideas, we think that CT imaging, air-bone gap and SD may become important indicators for decision making in cases of severe otosclerosis.

Recently, Mercus *et al*.[2] proposed the following algorithm guidelines based on SD, CT imaging and air-bone gap from their experience and the literature review. If maximum SD is under 30 dB, then CI should be considered. If maximum SD is between 30 dB and 50 dB and CT imaging shows type 2C or 3, then CI should be considered. If CT imaging does not show the findings, and air-bone gap is over 30 dB, then stapedotomy should be considered. If not, CT should be considered. If maximum SD is between 50 dB and 70 dB, and CT imaging shows type 2C or 3, then CI should be considered. If CT imaging does not show the findings, and air-bone gap is over 30 dB, then stapedotomy should be considered. If not, only hearing aid use will be considered. However, measurement of air-bone gap is sometimes difficult in patients with far-advanced otosclerosis because there are many scale-out cases in bone conduction. In such cases, past pure-tone average records will be helpful in addition to SD.

Conclusion

Therapy for severe otosclerosis should be considered with the use of CT imaging and air-bone gap values if the measurement of bone conduction is possible. Past pure-tone average records are also helpful, as is maximum speech discrimination with hearing aids before operation.

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BILATERAL CONGENITAL AURAL ATRESIA: SURGICAL RECONSTRUCTION VS. BAHA IMPLANTATION

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Introduction

Congenital aural atresia defines a clinical entity involving atresia of the ear canal and abnormalities in the middle ear usually in conjunction with microtia. These ear malformations may occur in isolation or in association with other craniofacial dysplasias. The reported incidence of aural atresia is around 1 in 10.000 and one third of cases are bilateral with a variety of complexity on each side. Surgical reconstruction has been the traditional approach to resolve the conductive hearing loss and the resulting handicap in bilateral cases, being the objective to restore sufficient hearing to avoid the necessity for amplification. Atresia surgery is usually performed after the age of 5 years old.[1-5] In some cases based on CT scan findings, patients are considered no candidates for surgical repair. It is this group of patients for whom the osseointegrated bone-conducting hearing aid is intended.[6-8] Currently the most popular osseointegrated hearing aid is the Bone-Anchored Hearing Aid (BAHA, manufactured by Cochlear Corporation, Molnlycke, Sweden, formerly Entific Corp.).

However, it is generally perceived that this operation is challenging, even for experienced otologic surgeons. Many of them suggest that it is a risky procedure, specially for the facial nerve and the cochlear function with non stable results over the time, even that many authorities have report large series of patients , some included long-term follow-up, suggesting that the BAHA implantation should be considered as the first option for this patients.

The present case revision intents to compare the experience of the author during 10 years performing both types of treatments for bilateral Congenital Aural Atresia.

Methods

Patients

From 1998 through 2008, 65 primary surgeries for congenital ear malformation were performed by the author, but just 34 cases were available for the long term analysis. During 2003 through 2008, the author performed 19 BAHA implantation surgeries for congenital aural atresia patients, 14 of which were available for the analysis. Revision surgeries and unilateral atresias were not included in this series. Cholesteatoma was found in three patients which was an additional reason to perform the surgery. Of the 48 patients comprising in this study 28 were males and 20 females. Mean age at surgery were 11 +/- 3 for the reconstruction group and 12+/-2 for the BAHA group. The side was chosen based on anatomic and radiologic considerations and the better ear was selected for the initial surgical procedure. In the BAHA group the side selection was based on anatomic issues, patients and parents preferences and patient dominance. The reconstruction was done in 18 patients in the left side and in 16 patients in the right side.

Temporal bone CT scans and adequate hearing documentation (complete audiogram and bone conduction auditory brainstem response testing) were performed on all patients for preoperative assessment. Patient selection criteria for surgery included normal sensorineural function bilaterally and a rating of 7 or above

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Edited by Haruo Takahashi

in the CT scan according to the Jahrsdoerfer grading system.[9] Patient selection criteria included normal sensorineural function bilaterally and at least 4 mm of cortical temporal bone thickness demonstrated in the radiologic evaluation.

The selection of the procedure was based on CT scan findings and patient and parents decision after a very careful consultation and explanation over the pros and cons of each procedure. Diagnoses included bilateral isolated and syndromes atresias. In this context the mean Jahrsdoerfer rate was 7.8+/-1.2 for the reconstruction group and a little lower 7.3+/-1.2 for the BAHA group.

Surgical Procedure

If an auricular reconstruction was done before, a post-auricular incision with preservation of the implanted rib cartilage framework is performed. If not, the skin and soft tissue incisions are decided based in a previous discussion with the plastic surgeon in order to preserve blood supply and minimize scars.

The temporalis fascia and the mastoid bone are exposed with a careful but generous elevation of the soft tissue. An external auditory canal is created in the atretic bone by drilling posterior to the glenoid fossa and inferior to the middle cranial fossa dura according to the anterior surgical approach described previously.[1,2,10] However it is very common to combine this approach with the transmastoid approach due to anatomical reasons.[11] All surrounding bone is removed from the ossicular chain so that it is centered in the new canal. The mobility of the ossicles is assured trying to preserve the malformed but commonly present ossicular chain. A thin fascia graft is placed over the mobilized ossicular chain. The goal is to maintain contact between the graft and the ossicles and prevent the lateralization. Several techniques can be used to prevent this complication: a notch incision in the graft to accommodate the malleus, a sulcus in the anterior canal wall can be drilled or sometimes cement or fibrin glue can be used to fix the graft to the malleus. A meatoplasty is performed with debulking of the subcutaneous tissues and the auricle is returned to its normal position leaving an anterior skin flap reflected over the anterior canal. A split-thickness skin graft is harvested from the upper arm and used to line the bony canal and placed over the fascial graft, then a Rosebud packing of the new external auditory canal is done.[12] The pack is removed 3 weeks later as an office procedure in older patients and under general anesthesia in patients younger than 12 at this time a second look of the canal is performed.

The proper technique of osseointegrated prosthesis placement has been described in detail elsewhere.[7,13] Briefly, only a single implant in a single stage operation is required to hold a BAHA securely in place. The marking of the implant site is crucial to preserve enough space to the auricular reconstruction or to avoid contact with the rib cartilage framework or the auricular prostheses (usually the post-auricular flap is elevated a little more posterior than the common technique in non malformed ears). Insertion of the implant must be performed with a high torque drill at slow speeds under maximal irrigation. Previously, an anteriorly based skin flap and careful removal of soft tissue is done. This allows the dermal layer to heal directly on the mastoid periosteum, creating a solid, immobile foundation for the implant. After placement of the implant, the flap is sutured and a hole is punched in the skin directly over the implant. The healing cap is placed and a dressing is positioned and secured. Three months later in order to allow for adequate healing and osseointegration, the bone conductor is placed.

Data Collection

For each patient, preoperative air-bone conduction PTA thresholds at 500, 1000 and 2000 Hz was recorded. The preoperative air-bone gap for each patient was measured by subtracting the best preoperative bone-conducting PTA from the best air-conduction PTA. Postoperative air-bone conduction PTA thresholds and postoperative air-bone gap were also measured for each patient in the reconstruction series. The postoperative BAHA aided PTA thresholds were recorded. Hearing improvement was calculated by subtracting the postoperative air thresholds from the preoperative air conduction PTA.[14]

The postoperative data were taken from the most recent audiogram beyond the first postoperative year and called long-term hearing results (mean follow-up in reconstruction series, 4.4 y, range, 1.0-10 y; mean follow-up in BAHA series, 3.5y, range, 1.0-6 y).

Major complications encountered in all cases were recorded, including facial nerve iatrogenic injury, sensorineural hearing loss or acquired cholesteatoma in the reconstruction group; and meningitis, failure of osseointegration or severe local skin complications in the BAHA group. The number of surgeries per patient

was established according to the number of revision surgeries in each case. It was necessary to review 2 BAHA patients due to skin problems; in one of these cases it was impossible to use the implant. The mean number of surgeries per patient in the reconstruction group was 1.9, but it is necessary to stipulate that a second operation under general anesthesia it was implemented for patients under 12 years old three weeks after the primary surgery, even in absence of complication.

Patient satisfaction was measured trough the question to the patient and parents if they would be able to do the same procedure in the contra lateral ear or in another family member? The answer was positive in 29 (85%) of the reconstruction patients and in 13 (93%) of the BAHA patients. Surgeon satisfaction was also taken into account, and this was defined as a long term postoperative air-bone gap less than 30 dB restoring sufficient hearing so amplification is no longer needed. These was accomplished in 24 (71%) of the reconstruction patients and in 13 (93%) of the BAHA cases.

Results

Operative Findings

In all cases, a fused and malformed malleus/incus complex attached to the bony atresia plate was found. There was one case in which the stapes was absent and a rudimentary oval window was identified. Ossicular replacement prosthesis was not used in any case in the primary surgery. In two cases it was impossible to complete the surgery due to the anatomic position of the facial nerve, both cases were syndromes. Cholesteatoma was found in three patients, two of them had widely excavated the bony external canal. In one cholesteatoma case a fistula was present and a subcutaneous abscess was identified in the post auricular region.

Two of the fourteen patients who underwent osseointegrated implantation had adverse tissue reaction. One patient required removal of the implant because of an infection and a skin graft was necessary.

Hearing Results

Preoperative an postoperative long-term (mean follow-up, 4.4 y) hearing results for 34 patients who underwent primary surgery only or primary surgery followed by revision surgery are summarized in Table 1. 71% of the patients in this series achieved and ABG (air-bone gap) of 30 dB or less in the long term (> 1 y). Furthermore, an average long-term improvement in the ear conduction PTA was 24.2 dB.

Preoperative and postoperative long-term (mean follow-up, 3.5 y) hearing results for 14 patients who underwent BAHA implantation are summarized in Table 2. 93% of the patients achieved and ABG of 30 dB or less in the long term. The average long-term improvement in the ear conduction PTA was 30.6 dB.

Complications

In the reconstruction group, no major complications were documented. Neither of these patients had facial nerve problems, significant sensorioneural loss of hearing or acquired cholesteatoma. Among the causes of initial failure were stenosis of the external auditory canal or meatus, fixation of the ossicular chain and lateralization of tympanic membrane; In general, the failure long-term rate was 28%. For the BAHA group, two patients experienced adverse tissue reactions and one patient required removal of the implant because of an infection. The failure rate with BAHA in this series was 7%.

Discussion

In infants with bilateral atresia, early amplification is essential and a bone conduction hearing aid must be fitted as soon as possible. Surgical decision is usually taken after the age of 5 or 6 years. Although some surgeons are reluctant to recommend surgical intervention in unilateral cases, there is no controversy about the early surgical reconstruction or the implantation of a bone-anchored hearing aid in bilateral cases.[15] Many otologic surgeons maintain that hearing recovery in atresia surgery is seldom achieved and in good cases the likelihood of maintaining the initial hearing result is poor. In addition, the perceived risks, especially to the

Table 1. Atresia surgery patients and hearing results

		SEX		SIDE				PTA(dB)		GAP(dB)				SATISFACTION			
	No	F	M	R	L	AGE(y)	ARSDOERF	PRE	POST	PRE	POST	ROVEMENT (	NUMBER OF SURGERIES	Y	N	CHOLESTEATOM/	FOLLOW-UP(y)
	1		X	X		8	8	65	25	50	10	40	2	X			10
	2	X		X		11	7	60	30	50	20	30	2	X			10
	3		X		X	6	7	55	25	45	15	30	2	X		X	3
	4	X		X		17	8	62	25	52	18	37	1	X			7
	5		X	X		16	7	65	65	60	60	0	2				8
	6	X			X	9	8	65	20	50	15	45	2	X			6
	7		X	X		14	8	62	40	52	25	22	3	X			2
	8	X			X	5	9	65	30	55	18	35	2	X		X	3
	9		X	X		7	7	60	58	50	50	2	4		X		5
	10	X			X	17	7	62	26	52	20	36	1	X			4
	11		X		X	13	8	60	20	50	15	40	1	X			2
	12	X		X		16	7	62	60	57	55	2	1		X		3
	13		X		X	15	7	54	48	44	38	6	1	X			9
	14		X	X		4	8	62	28	52	20	34	2	X	X	X	4
	15	X		X		7	7	60	18	45	10	42	2	X			6
	16	X		X		9	8	62	47	52	42	15	3		X		8
	17		X		X	8	7	58	42	48	34	16	2	X			2
	18		X		X	15	8	62	38	52	33	24	1	X			3
	19	X		X		17	7	60	60	55	55	0	1		X		6
	20		X		X	9	9	58	38	53	30	20	2	X			3
	21		X		X	10	8	60	28	50	23	32	1	X			2
	22	X		X		7	9	57	37	50	27	20	1	X			4
	23		X	X		16	7	63	38	53	28	25	1	X			2
	24		X		X	8	8	52	37	47	30	15	2	X			9
	25	X		X		7	9	63	40	53	30	23	2	X			2
	26	X			X	8	9	63	47	53	40	16	2	X			3
	27		X	X		10	8	60	32	50	22	28	2	X			1
	28		X	X		12	8	58	38	53	33	20	2	X			5
	29	X			X	16	9	62	32	57	27	30	1	X			3
	30		X		X	9	8	65	35	55	27	30	2	X			2
	31		X		X	14	7	62	38	52	28	24	1	X			6
	32		X		X	10	8	60	28	53	23	32	3	X			4
	33		X		X	7	8	63	37	57	27	26	2	X			3
	34	X			X	11	8	60	32	57	25	28	2	X			2
TOTAL		14	20	16	18									29	5		
MEAN		44,118	55,882	47,059	52,941	10,824	7,8235	60,794	36,529	51,882	28,618	24,264706	1,7941176	85,294			4,47058824

Table 2. BAHA patients and hearing results

	SEX		SIDE				PTA(dB)		GAP(dB)				SATISFACTION		
No	F	M	R	L	AGE	JHARSDOERFER	PRE	POST	PRE	POST	ROVEMENT	ER OF SURG	Y	N	OLLOW'UP(y
1	X			X	7	9	63	25	53	15	38	1	X		6
2		X		X	15	6	60	28	50	20	32	1	X		5
3		X	X		15	6	60	23	50	13	37	1	X		2
4		X		X	9	7	57	20	52	10	37	2	X		4
5	X			X	17	5	60	60	50	50	0	3		X	4
6		X	X		12	9	58	20	53	15	38	1	X		2
7	X			X	8	8	60	25	50	15	35	1	X		5
8		X	X		10	8	55	55	45	15	0	1	X		2
9	X		X		14	8	60	20	50	10	40	1	X		1
10		X	X		12	6	65	25	55	15	40	1	X		1
11	X		X		11	6	60	23	50	13	37	1	X		5
12		X	X		7	7	62	27	50	13	35	1	X		5
13		X	X		16	9	55	30	45	20	25	1	X		6
14	X		X		14	8	60	25	50	15	35	1	X		2
TOTAL	6	8	9	5									13	1	
MEAN	42,8571	57,1429	64,2857	35,7143	11,9286	7,285714286	59,6429	29	50,2143	17,0714	30,64286	1,214286	92,8571		3,5714286

facial nerve, are also cited as a deterrent to surgery. Apparently, the advances in imaging methods, microscopes, facial nerve monitoring and surgical technique have no changed this perception.[16] Consequently, some authors suggest the titanium anchored bone-conducting hearing aids as the option with more predictable and stable results with less likelihood of complications and a high rate of patient satisfaction.[17] The measure of the long-term outcomes with both types or treatments have very important implications for counseling patients about the benefits they would expect, thus enabling the patient and surgeon to make a more informed choice. Outcome measures are also important in patient selection because the BAHA is a relatively expensive device.

The results of these series show good long-term hearing results in both groups of patients with a very low incidence of major complications. The likelihood of achieve a long-term post-operative air-bone gap of 30 or less after, interpreted for the author as a good result, is 71% in the atresia surgery series and 93% in the BAHA group. The atresia surgery group had an average hearing improvement of 24.2 dB. By comparison, patients with BAHA had on average a 30.6 dB hearing improvement.

Although the number of surgeries per patient is clearly higher in the atresia surgery, the advantage of avoid the permanent use of a hearing aid and its implications in terms of cost, batteries replacement, exposition to wet environments and the interference of a metallic implant in subsequent radiologic studies make a very comparable rate of patient satisfaction in the long-term.[18]

In summary, surgical correction of congenital aural atresia has the potential to significantly improve hearing with low risk to the patient. Osseointegrated bone-anchored hearing aid offer to the otolaryngologist a viable alternative for hearing rehabilitation in patients who are not candidates for surgery or have failed to obtain hearing improvement with the surgical reconstruction.

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FACIAL PARALYSIS IN CHRONIC OTITIS MEDIA

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Introduction

Chronic suppurative otitis media is a destructive process that can involve the facial nerve and may result in an impairment of its function. In the pre-antibiotic era, facial paralysis occurred in 2.3% of patients with chronic otitis media. The frequency of otogenic facial palsy has decreased since the introduction of antibiotics, but it still occurs in about 1% of the patients with chronic middle-ear infection.

Mechanisms of the effects of chronic otitis media on the facial nerve are complex but can be summarized as follows: direct extension of the inflammatory process to the facial nerve, pressure from the pathological process (cholesteatoma, granulation), and toxic action upon the nerve itself. Although numerous studies have dealt with otogenic facial paralysis, this still is a controversial issue.[1-3]

Subjects and methods

The study was performed on 136 patients with otogenic facial paralysis who were surgically treated at our clinic. Ages of the patients ranged between 15 to 82 years. The medical records of the patients were analyzed in order to determine the factors of importance for facial paralysis. Attention was paid to the types of chronic infection, location of bone destruction of the facial canal, pre-operative and post-operative nerve function and follow up. Computed tomography was performed in most of the cases and magnetic resonance imaging was obtained in some patients. Evaluation of degree of recovery of facial function depended on the methods used in the follow-up period.

Results

Surgical intervention was performed as early as possible in all cases of chronic otitis media with a pre-operative facial paralysis. The period of time from onset of paralysis to surgery ranged from five days to two years. Surgery (canal-wall-up or canal-wall-down) and facial-nerve decompression was performed in the majority of cases. Cholesteatoma was found in 52 (80%) of the patients while in the remaining 12 (20%) chronic inflammation manifested as granulation or fibrocystic tissue. Decompression was done from geniculate ganglion to the stylomastoid foramen with or without epineural incision in 75% cases. In some patients, aggressive cholesteatoma and granulation fibrous tissue were involved which interrupted the facial nerve. Because of that, the patients underwent direct end to end neural anastomosis or neural graft with auricular nerve, after the resection of the involved segment of the facial nerve.

Otogenic facial paralysis was more common as an isolated complication than associated with other complications (68.8% : 31.2%). A fistula of the lateral semicircular canal was the most common complication (18.7%). Other complications, such as destruction on the promontory and intracranial complications, occurred less frequently (6.3% : 6.2%).

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Edited by Haruo Takahashi

The site of bone destruction of the facial canal was confirmed at surgery in 48 (75%) of the cases, while such a site was not discovered in 16 (25%) cases. The tympanic segment of he facial canal was the most common site of involvement (77.2%). In the majority of these the bone destruction was adjacent to the oval window. In some cases, the facial nerve was protruding in the tympanic cavity, and in some cases the nerve partly covered the oval widow itself. Destruction of the mastoid part of the facial canal was found in six (12.7%) of the cases. All these cases showed an opening in the lateral bone canal with exposed nerve in the facial recess. In five cases (10.1%), the facial canal was extensively destroyed and the nerve almost totally exposed from its proximal part to the stylomastoid foramen.

There may be a number of reasons why in 25% of our patients evidence of bone destruction of the facial canal was not present. In some cases, a pathological process can destroy the facial canal at a place which is not possible to see at surgery, such as the bony recess adjacent to the facial canal and the medial wall of the facial canal. Also, the presence of communications between the blood vessels of the facial canal and surrounded structures allowed spread of the inflammatory process into the facial nerve. In some cases, an osteitic process can involve the facial canal as localized focus of infection. The bony facial nerve canal was open and the nerve decompressed depending on the place of pathological involvement.

Follow up and evaluation of the degree of recovery of the facial function depends upon the method used and possibility.

In our study we recognized three conditions: 1) Complete recovery is characterized by a full restoration of the facial function without synkinesis (70%); 2) Partial recovery is characterized by slight asymmetry at rest and weakness of voluntary movements (24%); 3) Failure to recover was found in 6% of the cases. It manifested with asymmetry at rest, slight return of muscle function, contractures, hemi-facial spasms and synkinesis.

Discussion

Chronic otitis media causing facial paralysis is frequently associated with cholesteatoma. But, acute exacerbation of the inflammatory process plays an important role in the pathogenesis of facial palsy. Our previous human-temporal-bone studies have described that chronic otitis media is associated with degenerative changes in the facial nerve without clinical alteration of its function. Nonetheless, these changes may predispose to the development of facial paralysis in chronic otitis. The findings suggest that facial paralysis occurs in chronic supportive obits when the inflammatory process specifically involves the facial-nerve trunk.[4,5]

Infection may involve the facial canal at any point but most often occurred in the tympanic segment. The main reason for this occurrence is that this part of the facial canal is most frequently dehiscent and the wall of the facial canal at this site is extremely thin. A pathological process such as cholesteatoma or granulation tissue is very often localized around the tympanic part of the facial canal.

Concerning the management and prognosis of facial paralysis due to chronic middle-ear infection, some questions are still open. Firstly, the type of the pathological process, cholesteatoma or other (granulation, fibrosis). In cases with facial palsy, cholesteatoma is most commonly present. Also, at surgery, it is important to remove its matrix, not only for better recovery of facial-nerve function, but also to prevent potential recurrence and additional ear surgery. Secondly, the occurrence of facial paralysis does not depend on the extent of destruction of the facial canal. In some cases, destruction of the canal was very small, but in some cases it was very massive, indicating that the facial nerve may have been uncovered and exposed to the pathological process for a long period of time prior to the facial paralysis. Thirdly, it is important to make the incision on the epineurium and always leave the perineurim intact to form a protective barrier to infection. We believe that in some cases, after removal of pathological process, it is useful to open the epineurium in a limited area, which has a relieving effect in cases with severe edema and inflammation.The results of surgical intervention were better in cases with shorter duration of paralysis prior to the surgery. We believe that early surgical intervention is crucial for recovery of the facial function. However, eradication of cholesteatoma and possible occurrence of its residual/recurrence types, can be a concern for the treatment of chronic otitis and its complications.

Conclusion

Otogenic facial paralysis most commonly occurs due to chronic otitis media with cholesteatoma. We believe that early surgical intervention is crucial for better recovery of the facial-nerve function. Also, it is important to perform a successful surgical procedure with the aim to remove the pathological process from the middle-ear spaces and to prevent recurrence or serious complications.

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FACTORS INFLUENCING HEARING AFTER TYPE-III TYMPANOPLASTY USING COLUMELLA

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Introduction

The purpose of tympanoplasty for chronic ear diseases is to cure the disease, preserving/improving hearing. To obtain post-operative hearing, a variety of materials are used for a columella in type-III tympanoplasty, including autografts and prostheses. This surgery is usually performed with canal-wall-up or canal-wall-down mastoidectomy, which influences the post-operative middle ear conditions. Among middle ear conditions, aeration of the tympanic cavity is critical to the success of any type of tympanoplasty.[1] Therefore, both choice of the material of the columella and the middle ear aeration are important for successful tympanoplasty. The overall goal of this study is to better understand the keys resulting in good hearing outcomes with type-III tympanoplasty.

Patients and methods

From April 2004 to May 2011, 110 ears of 107 patients underwent type-III tympanoplasty using a columella at Kobe Medical Center General Hospital. All procedures were performed by the senior author (YN). Excluding nine patients of ossicular malformations, 101 ears of 99 patients were enrolled in this study (58 male and 41 female patients; age, 3-78 yr; mean age, 38 yr). Ossiculoplasty's with the columella on stapes head and on stapes footplate were performed in 74 and 27 ears, respectively. Fifty-one ears (50%) were pars flaccida type cholesteatoma, 25 ears (25%) were pars tensa type, combined, or secondary cholesteatoma, 13 ears (13%) were congenital cholesteatoma, and 13 ears (13%) were adhesive otitis media, tympanosclerosis and others. Seventy-five ears (74%) underwent canal wall down mastoidectomy with soft wall reconstruction.[2] Audiometric data obtained at least one year after the surgery were used by calculating the pure-tone average of 0.5, 1, 2, 4 kHz. Post-operative air-bone (A-B) gaps were calculated in each ear and divided into four bins: 0-10, 11-20, 21-30, and greater than 30 dB. We reviewed the relationship between the materials of columella and post-operative A-B gap. In addition, we reviewed the clinical records and the computed tomography (CT) findings of the ears of which postoperative A-B gaps were greater than 21 dB, and determine the cause of poor postoperative hearing.

Results

Hearing outcomes in the 101 ears according to the post-operative A-B gap were as follows (Table 1): 0-10 dB, 21 ears; 11-20 dB, 39 ears; 21-30 dB, nine ears; and greater than 30 dB, five ears in 74 ears undergoing type-III tympanoplasty using columella on stapes head, and 0-10 dB, six ears; 11-20 dB, 11 ears; 21-30 dB, seven ears; and greater than 30 dB, three ears in 27 ears undergoing type-III tympanoplasty using columella on stapes footplate. The relationship between the A-B gap and the materials for columella showed Table 2.

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Edited by Haruo Takahashi

Table 1. Postoperative hearing. Number of the ears divided according to postoperative air-bone gap (average: 500, 1000, 2000, 4000Hz)

	Columella on stapes head	*Collumella on stapes footplate*
0-10dB	21 ears	6 ears
11-20dB	39 ears	11 ears
21-30dB	9 ears	7 ears
31dB	5 ears	3 ears
Total ears	74 ears	27 ears
A-B gap average	13.8dB [0-43.8]	18.5dB [0-37.5]

Table 2. Postoperative air-bone gap (average: 500, 1000, 2000, 4000Hz)

	Columella on stapes head	*Collumella on stapes footplate*
Auto ossicle (incus or malleus)	14.0 dB [n=41]	18.5 dB [n=14]
Cortical bone	14.4 dB [n=24]	18.4 dB [n=7]
Hydroxyapptite	14.5 dB [n=4]	26.3 dB [n=3]
Cartilage	23.1 dB [n=4]	11.3 dB [n=3]
Silastic plate	30.0 dB [n=1]	
Total	15.4 dB [n=74]	18.5 dB [n=27]

There was no significant difference among materials for columella in hearing outcomes. Among the 24 ears (24%) of which post-operative A-B gaps were greater than 21 dB, 18 ears exhibited poor middle ear aeration caused by eardrum retraction, adhesion, inflammation or fibrosis (11 ears), tympanosclerosis and diseased stapes mobility (five ears), Eustachian tube dysfunction due to sniffing habit or cleft palate (two ears). Six ears had problems of columella: displacement (two ears), detachment from the eardrum or stapes head (two ears), ankylosis due to contacting with the bony wall of the tympanic cavity (two ears).

Discussion

In the present study, good hearing results were obtained by mainly using autologous tissues including ossicles, cortical bone, and cartilage, which were used for columella. These autologous tissues are biocompatible and can achieve long-term success with hearing results.[3] But, on the other hand, poor aeration of the middle ear space was associated with poor post-operative hearing more strongly than the problems of columella. Poor aeration of the middle ear space was due to Eustachian tube dysfunction, chronic infection, and mucosal adhesion, which result in severe attical and/or posterior wall retraction and atelectasis of the tympanic membrane. These problems can occur regardless of careful procedure for tympanoplasty. In such cases, it may be effective to use cartilage as a grafting material on account of its increased stability and resistance to negative middle ear pressure.[4] Further study is needed to determine long-term results for each surgical procedure.

Conclusion

Post-operative hearing may be influenced more by middle ear conditions than columella itself. Control of inflammation and good aeration of the tympanic cavity are important to obtain favorable post-operative hearing.

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OUTCOME OF RADICAL SURGERY AND POST-OPERATIVE RADIOTHERAPY FOR SQUAMOUS CARCINOMA OF THE TEMPORAL BONE

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Objective

To analyze the clinical data and outcome of all the patients treated surgically for squamous carcinoma of the temporal bone in a tertiary referral skull base department over 25 years.

Methods

Fifty-four patients with squamous carcinoma of the temporal bone were analyzed. The patients were staged according to the Pittsburgh system. The surgical technique, reconstruction of the surgical defect, post-operative radio-therapeutic treatment, follow-up regimen and results are all described in detail.

Results

Disease-free survival of T2N0M0 tumors or stage II was 100%. The survival of stage-III disease T3N0M0 was also 100%. Overall T3 tumors resulted in 50% survival since 50% had nodal involvement were stage-IV disease and none of those survived. There was 100% mortality where nodes were involved. There was a 54% survival for T4N0M0 tumors and 45% for T4 tumors overall. The stage-IV tumor survival was 42%.

Conclusion

The overall disease free survival in the whole series was 52%. Node positive disease, poorly differentiated squamous cell histology, brain involvement and salvage surgery were associated with a poorer outcome. The improved survival (66%) of patients treated de novo in this series compared with those treated as salvage (34%) suggests that early referral and aggressive primary surgical treatment with post-operative radiotherapy offers the greatest chance of cure.

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Edited by Haruo Takahashi

SURGICAL MANAGEMENT OF T1 AND T2 LESIONS WITH OUTCOMES

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Introduction

T1 or T2 lesion of squamous cell carcinoma (SCC) of the temporal bone is defined to be limited to the inside of the external auditory canal by the Pittsburg T-staging system.[1] It is an early stage and good outcomes in survival ratio were reported by several authors.[2,3] However, there is no consensus in operative procedure. The necessity of neck dissection remains to be demonstrated. Factors which affect outcomes have not been discussed yet. We have checked these points from outcomes in our facilities.

Method

We treated 56 patients with squamous cell carcinoma of external auditory canal (EAC) and middle ear. Settings were Kyushu University Hospital from January 1998 to March 2006 and Fukuoka University Hospital from April 2006 to March 2011. Mean age was 65 years (ranged between 41 and 93 years). There were 31 male and 26 female patients. Cases of a follow-up period of more than 12 months were included in the study. Patients who died of the disease were not included. Mean follow-up period was 37 months. Numbers of cases: T1N0 = one; T1N1 = none; T2N0 = 12; T2N1 = three; T3N0 = seven; T3N1 = two; T4N0 = 20; T4N1 = 11. Twenty-nine percent (n = 16) had lymph-node metastasis. There was only one case of T2N1M1.

Treatment strategy was determined by the extent of the tumor, evaluated by both CT and MRI before treatment. Fig. 1 shows the treatment strategy of our facilities at which the first author worked as a final decision maker of strategy for each patient.

Using the method of lateral temporal bone resection, the cartilaginous region including the cavum conchae of the auricle and the EAC, the bony part of the EAC and the tympanic membrane with the bony annulus were resected at the lateral side of the facial nerve. Upper neck dissection was in most cases added to the surgical procedure.

Using subtotal temporal bone resection, the facial nerve, inner ear, and internal auditory canal were sacrificed. The posterior and middle cranial fossa dura, sigmoid sinus, internal jugular vein, bulb and internal carotid artery were preserved. Modified neck dissection up to the skull base with total parotidectomy was commonly performed. As a consequence, the lesion inside the middle ear and the soft tissue containing lymph-flow draining from primary lesion down to the lower neck were totally removed.

Results

In 2006, we reported that disease-specific survival rate of the T1, T2, T3 and T4 lesions were 100%, 100%, 83% and 39%, respectively.[3] Including the previous six years, the disease-specific survival rates for 13 years were summarized (Fig. 2). The five-year survival rate of T3 and T4 was 89% and 52%, respectively. However, the five-year survival rate of T1 and T2 decreased to 68% and was lower than that of T3. Although treatment

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Edited by Haruo Takahashi

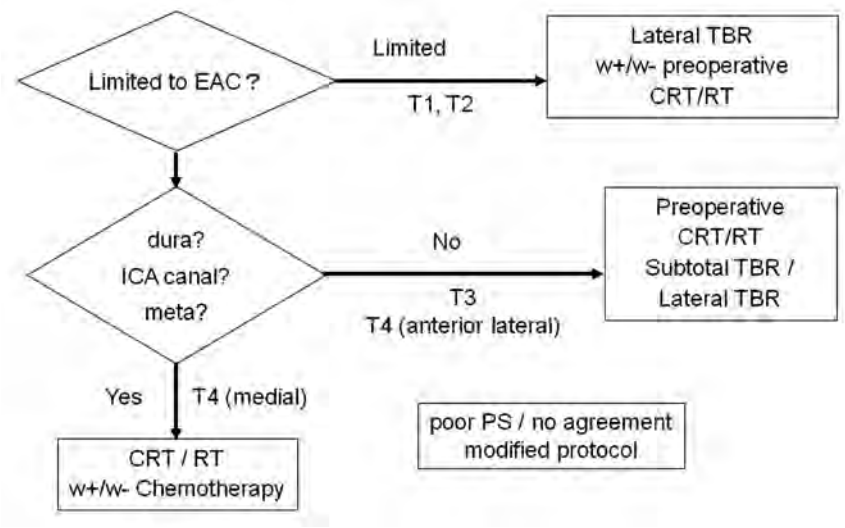

Fig. 1. Treatment strategy of temporal bone squamous cell carcinoma. Extent of tumor was evaluated by both CT and MRI before any treatment.

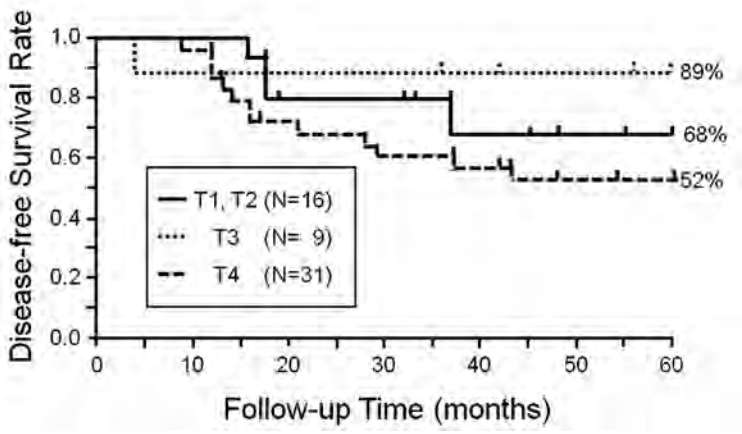

Fig. 2. Disease-free survival rate for T stage.

strategy was maintained during 12 years, the number of cases of T1 and T2 was roughly three times that of the previous report.

Fig. 3 shows details of T1 and T2 cases. Four patients died of the disease in the follow-up period of 16, 18, 18 and 37 months. Markers of epithelial-mesenchymal transition (EMT) in the SCC tumor cells were examined in seven out of 16 cases. Three out of four patients who died of the disease were included in the study. Vimentin was positive and laminin was diffusely stained in all cases.

Discussion

We reported the outcome of our treatment strategy for temporal-bone SCC from 1998 to 2010. Characteristics of our strategy are pre-operative chemo-radiotherapy to control minor lesion of surgical margin and as surgical procedure en-bloc dissection with neck dissection. The strategy works for operative cases in an advanced stage. Five-year survival rate of T3 and T4 elevated to 89% and 52%. However, the survival rate of early-stage cases decreased to that of a T3 stage case.

As shown in Fig. 1, when a patient did not agree with our strategy, we modified the protocol and usually selected a less invasive treatment. Patients' freedom of choice may modify the results. The disease-free survival rate was re-calculated by picking up on 46 cases which followed our treatment strategy. Early-stage, T3 and T4 cases were 83%, 100% and 55%, respectively. Although the survival rate of early-stage cases elevated over 80%, inversion still existed.

#	follow-up period (month)	age	gender	rT?	T	N	M	stage	dead or alive	Under Protocol	Laminin type of staining	Laminin <25%: 1+ 25-50%: 2+ 50-75%: 3+ >75%: 4+	vimentin
1	12	70	F	No	2	2b	0	4	alive	1			
2	16	58	M	No	2	0	0	2	DOD	1	diffuse	3	positive
3	18	74	M	No	2	0	0	2	alive	1	marginal	2	positive
4	18	70	F	No	2	0	0	2	DOD	1	diffuse	3	positive
5	18	62	F	Yes	2	0	0	2	DOD	1			
6	19	73	M	No	2	0	0	2	alive	1			
7	32	78	M	No	2	0	0	2	alive	1			
8	33	77	F	No	2	0	0	2	alive	1	marginal	1	negative
9	37	61	F	No	2	0	0	2	alive	1			
10	37	57	F	No	2	1	0	3	DOD	1	diffuse	3	positive
11	48	64	M	No	2	0	0	2	alive	1			
12	48	80	F	No	2	0	0	2	alive	1			
13	55	77	F	No	2	0	0	2	alive	1			
14	60	61	M	No	1	0	0	1	alive	1	marginal	2	positive
15	60	71	F	No	2	0	0	2	alive	1			
16	60	60	F	Yes	2	1	0	3	alive	1	marginal	1	negative

Fig. 3. Details of T1 and T2 cases DOD labeled ‘died of disease’.

One of the reasons for inversion could be the different extent of neck dissection beside temporal bone resection. All regional lymph nodes with soft tissue including the parapharyngeal space were salvaged with T3 lesion by sacrificing the facial nerve. This procedure was not performed to the T2 lesion because of avoiding facial nerve palsy. Like in other head-and-neck cancers, lymph-node metastasis can be a poor prognostic factor for early stage. Cases 1, 10 and 16 had node metastasis as shown in Fig. 3. Several lymph-node metastases were found at parotid and upper neck in case 1. The patient agreed to a total parotidectomy by sacrificing the facial nerve even though facial nerve palsy was not pre-operatively found. The patient survived for a year. In case 16, a single lymph-node metastasis was found in the upper neck. Treatment of choice was neck dissection without parotidectomy and the patient survived for five years without recurrence. However, case 4 refused parotidectomy in spite of lymph-node metastasis at the deep lobe of the parotid gland and chose chemotherapy combined with immunotherapy. The patient died 18 months after the onset of the disease.

These experiences also suggested a validity of the high survival ratio of T3 cases treated by our strategy. Modified neck dissection up to the skull base with total parotidectomy consequently salvages all soft tissue containing lymph-flow draining from primary lesion down to the lower neck. It indicated that neck dissection was the treatment of choice lymph-node metastasis was found. Although it should depend on each patient’s own decision, we should not be reluctant to select total parotidectomy with sacrificing facial nerve for a case with lymph node metastasis in the deep lobe of the parotid gland.

There is also the possibility that pre-operative chemo-radiotherapy might not be effective to microlesion of lymph-node metastasis. An insufficient neck dissection for early-stage cases lowered the survival ratio. Sugimoto and colleagues suggested that EMT in tumor cells can be an indicator of poor prognosis in temporal bone SCC.[4] Vimentin is one of the intermediate filament proteins expressed in mesenchymal cells related to EMT. Laminin is one of the proteins in the basal lamina. The translational activation of laminin during EMT was one of the prognostic factors of cancer. Our examination of immunostaining showed that vimentin was positive and laminin was diffusely stained in all early-stage DOD cases. It may suggest that an aggressive treatment strategy should be proposed when the biopsy specimen shows diffusion by laminin staining and vimentin-positivety. It could also be possible that a less invasive therapy might be selected when the immuno-pathological examination shows no lymph-node metastasis. Further study is necessary to investigate these options.

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CONGENITAL CHOLESTEATOMA: RADIOLOGIC EVALUATION AND PERSONAL EXPERIENCE

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The congenital origin of cholesteatoma has been hypothesized in 1800. Congenital cholesteatoma of the petrous portion of temporal bone was reported in 1938. Congenital cholesteatoma (CC) may locate in petrous portion of the temporal bone or in the middle ear. CC in the middle ear can be defined as a whitish mass in the middle ear with the intact tympanic membrane that arises from embryonal inclusions or rests of epithelial cells. Previous otologic procedure, otorrhea and perforation and abnormalities on the middle ear are exclusion criteria to diagnose middle ear cholesteatoma. It is the most frequent type and the symptom of conductive hearing loss appears in the young age, however, it can be seen in every age. Awareness of the problem is the main reason for an early diagnosis. Moreover, having the better diagnostic tools such as endoscopes, better otoscopes and otomicroscopes as well as better radiologic evaluation make the diagnosis easier. High-resolution computed tomography (CT) and magnetic resonance imaging (MRI) are the main radiological evaluation methods of congenital cholesteatoma. A CT scan confirms the location as well as the size of middle ear cholesteatoma. It gives us a lot of details for surgery, however, it is not enough to define any mass in the middle ear or cranium as cholesteatoma. All the findings are indirect. Moreover, the CT scan produces a high dose of radiation. This is especially harmful for young children, which is why the cone-beam CT scan has recently become the recommended modality. MRI shows a homogenous lesion that is hypo-intense in T1-weighted images and hyper-intense in T2-weighted images. A delayed contrast enhanced MRI sequence is another type of evaluation. However, it is time consuming and requires repeated scanning 30-120 minutes after contrast administration. Diffusion-weighted imaging (DWI) detects the molecular diffusion of water within the scanned tissue. Cholesteatoma exhibits a high-signal intensity on a DWI. The sensitivity and specificity of DWI has been found 69.4% and 92.8%, respectively in a recent study.[1] Between 2006 and 2012, six patients with congenital cholesteatoma were seen. Details of them can be found in Table 1. High-resolution CT scan and MRI were used for diagnosis. The MRI machine in the hospital has some major limitations such as low resolution and the production of relatively thick image sections. They have distorted shapes due to artifacts. Since it does not give us excellent details, we asked for a baseline MR scan after the first operation for follow up. Except for one patient, all cases presented with extensive cholesteatoma in the temporal bone. Fullness was the main symptom in the case with CC in the middle ear and trans-canal surgical management has been performed. The mass was removed without touching the ossicular chain. One patient has been diagnosed incidentally. Another one presented with recurrent facial palsy, while other patients complained about hearing loss or dizziness. Trans-mastoid surgery was performed. Congenital cholesteatoma may originate in different sites within the temporal bone. There are various symptoms. According to the location, different surgical techniques can be performed. Intensive and long-term follow up is mandatory for this group of patients.

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Edited by Haruo Takahashi

Table 1. Details of patients

	Age(yrs)	*Gender*	*Otoscopy*	*Complaints*
1	8	Male	Normal	Fullness
2	5	Female	Intact TM	Fall down
3	15	Female	Slightly retracted TM	CHL
4	24	Male	Slightly retracted TM	Dizziness when he sneeze
5	35	Female	Normal TM	Recurrent FP
6	28	Female	Normal	Incidental

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SURGICAL MANAGEMENT OF PETROUS APEX CHOLESTEATOMA: OUR EXPERIENCE OF 15 CASES

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Introduction

Petrous apex cholesteatoma (PAC) may involve the petrous bone as a congenital primary lesion or secondary to an acquired middle ear cholesteatoma.[1-5] PAC surgery is challenging because complete removal of PAC matrix from the dura, sigmoid sinus, jugular bulb, internal carotid artery (IAC) and facial nerve is often difficult. The choice of surgical approach should be based on the location and extent of the lesion, and pre-operative hearing and facial nerve functions.

Materials and methods

A retrospective analysis was conducted of the clinical charts of all patients with PAC (n = 15) at Osaka and Kinki University Hospitals from 1992 to 2012. During the same period, 1212 cases of cholesteatoma were surgically treated by the author. The prevalence of PAC, therefore, should be 15/1212 (1.2%). The mean age was 40 years and the age ranged from 19 to 80 years old. Female/male ratio was 4:11. Probable etiology was classified into congenital (N = 9), acquired (N = 4), and unknown (N = 1). Acquired cases had a history of chronic otitis media or had undergone previous middle-ear surgery elsewhere.

Results

The preoperative common symptoms were hearing loss (HL) (Fig. 1A; N = 15) and facial nerve palsy and weakness (N = 7). As for HL, mixed HL was present in three cases, conductive HL in four cases, profound sensorineural hearing loss (SNHL) in two cases, and total deafness in six cases (Fig. 1B). According to Sanna's classification,[2] the lesions confirmed at the surgery were classified into supra-labyrinthine (N = 9), massive labyrinthine (N = 5), and infra-labyrinthine (N = 1) groups (Fig. 2A). Middle cranial fossa (MFC) approach combined with lateral transtemporal approach was conducted in 12 cases, and transmastoid with/without trans-labyrinthine approach was selected in three cases (Fig. 2B).

None of the eight patients with a pre-operative normal facial nerve function developed a facial palsy post-operatively. In three out of seven patients with a facial nerve palsy pre-operatively, facial-nerve function recovered somewhat post-operatively. The removal of PAC and infected bone with appropriate decompression of the nerve, might improve the facial-nerve function. None of the four patients with a pre-operative conductive HL developed deterioration in bone conduction post-operatively (hearing results were successful in three cases, moderate in one). One out of three patients with a pre-operative mixed HL developed a profound SNHL post-operatively.

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Cholesteatoma and Ear Surgery – An Update, pp. 139-141
Edited by Haruo Takahashi

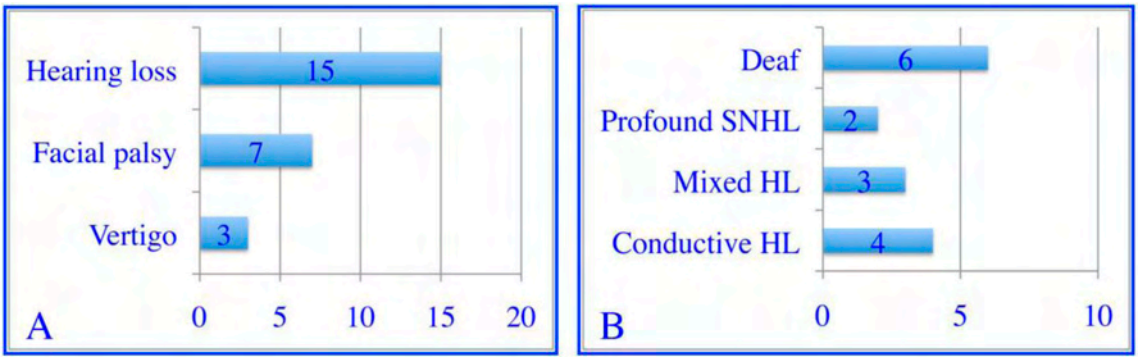

Fig.1. The preoperative common symptoms and types of hearing loss.

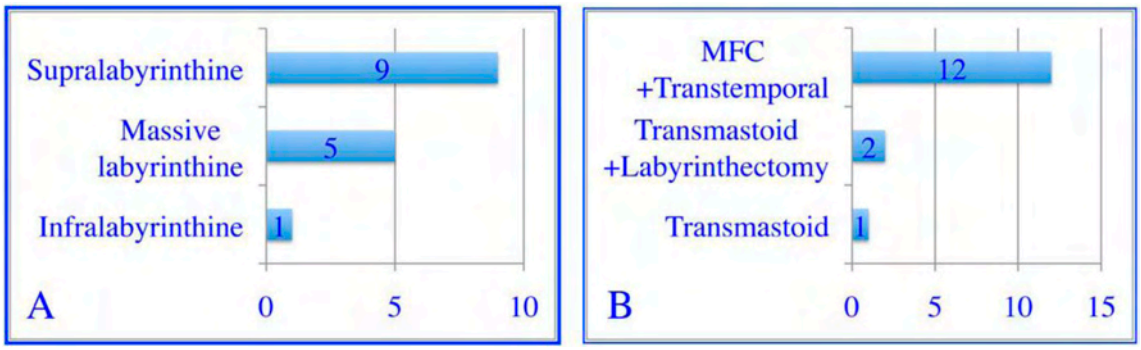

Fig.2. The classification of the lesions and the surgical approaches.

Total PAC removal was completed in 13 cases. Because thin PAC matrix was found to be adhered extensively and tightly to the dura of middle and posterior cranial fossa, the wall of sigmoid sinus, jugular bulb, bipolar coagulation of suspected portions had been advocated to denature of the matrix in two cases. The mastoid and middle ear were explored at the second surgery in eight cases one year after the first stage surgery. Three residual PACs were confirmed and removed. The mean follow up was 49 months (range three to 192 months) and annual MRI scans showed no apparent suggestion of PAC recurrence in all of the 15 cases.

Discussion

The MFC approach can be used successfully to remove supra-labyrinthine PAC with or without an apical extension. A MFC approach offers the possibility of hearing preservation, but the exposure is often inadequate for removal of the more extensive lesions. The lateral transtemporal approach has two main advantages: it is the most direct route to the pathology, bone removal avoids dural retraction. For complete PAC removal, the most suitable approaches might be the lateral transtemporal (trans-labyrinthine-trans-cochlear) approach combined with the MFC approach. Following total PAC removal, the defect is usually closed in either of two ways: obliteration of the cavity and blind sac closure techniques, and the more traditional open cavity. The advantages of reconstruction and establishment of a pneumatized middle ear have been suggested: one is an easy detection of recurrent cholesteatoma on CT and MRI during follow up; the other is easy access to the petrous apex recurrent lesion because of an absence of barriers in the approach.

The main factors affecting the choice of surgical approach are: inaccessible nature of the petrous apex, the extent of disease, the degree of facial nerve and hearing functions, the need for the prevention of CSF leaks as well as the recurrence of the lesion. We believe that the adoption of a lateral transtemporal approach combined with a MFC approach and subsequent reconstruction of a well-aerated middle ear allows the greatest opportunity for total removal of the lesion, preservation of good neurological functions, and prevention of PAC recurrence.

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CONGENITAL CHOLESTEATOMA OF THE MIDDLE EAR: A REPORT OF 54 CASES

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Introduction

Congenital cholesteatoma (CC) of the middle ear, first reported by House (1953), is a rare clinical entity that classically presents as a white mass situated in the anterior-superior quadrant of the middle ear behind an intact tympanic membrane (TM) associated with an ipsilateral conductive and/or sensorineural hearing loss.[1] Derlacki and Clemis established the diagnostic criteria for CC:[2] 1) A pearly white mass medial to an intact TM; 2) Normal pars tensa and pars flaccida; 3) No history of otorrhea, perforation or previous otologic procedures. Later Levenson *et al*. insisted the past history of acute otitis media should not be precluded from the diagnostic criteria.[3] CC is seen far more frequently in children, but House and Sheehy remarked adult patients with cholesteatoma behind an intact TM fit the criteria.[4]

Materials and methods

A retrospective analysis was conducted of the clinical charts of all patients with CC in both children (n = 50) and adults (n = 4) at Osaka and Kinki University Hospitals from 1992 to 2012. The male/female ratio was 36:18. CCs of the petrous apex (n = 9) were excluded. The age ranged from 2 to 40 years, with an average of ten years and a median of seven years. During 1992-2012, 1212 cases of acquired and congenital cholesteatomas were treated by the senior author. The prevalence of CC, therefore, should be 4.5% (54/1212).

Results

Histopathological studies demonstrate two distinct pathological types of CC. A ‘closed’ cyst in the anterior mesotympanum, which is easily removed. An ‘open’ infiltrative type in which the matrix is in direct continuity with the middle-ear mucosa. Surgical removal is difficult for this type and it is more likely to be associated with residual disease. We found 22 closed-type CCs and 34 open-type CCs (both pathologies were co-existent in two cases). Potsic *et al*.[5] proposed a staging system for CC: Stage I: single quadrant, no ossicular involvement or mastoid extension; Stage II: multiple quadrants, no ossicular involvement or mastoid extension; Stage III: ossicular involvement, no mastoid extension; Stage IV: mastoid extension. Based upon this system, 54 patients were classified in stages I-IV according to the surgical findings: eight cases in stage I, six in stage II, 22 in stage III, and 18 in stage IV (Fig. 1A). According to the growth pattern confirmed at the surgery, it was suggested that these CCs could be derived from the epidermoid formation (EF)[6] in 47 cases.

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Edited by Haruo Takahashi

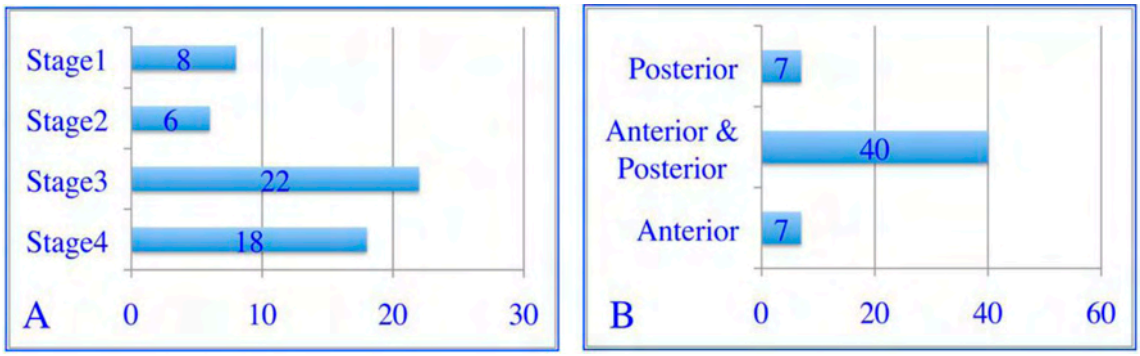

Fig. 1. Staging and location of CC.

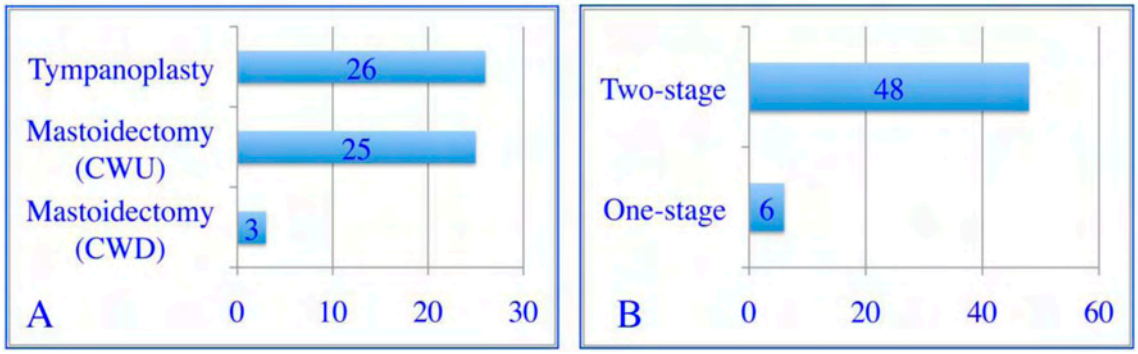

Fig. 2. Techniques used in the treatment of CC.

Regarding the classification of CC according to the location at the surgery, the CC lesion was divided into the anterior (A; n = 7), posterior (P; n = 7), and anterior-posterior (A-P; n = 40), using the malleus handle as the reference point (Fig. 1B). The favored position of the CC in the anterior-superior region of the middle ear in early cases be explained by its origin from the only cell rests (EF).

All patients were scheduled for tympanoplasty with or without mastoidectomy and all surgeries were performed with the post-auricular approach and lateral graft technique (Fig. 2A). A planned two-staged surgery was conducted in 48 cases (89%), while one-stage surgery was adopted in six cases (11%) including five cases of stage I (Fig. 2B). Three cases received multiple surgeries due to the residual and recurrent CCs several years after.

The residual cholesteatoma at the time of second-stage surgery was detected in 18 out of 43 cases (42%). The most common residual sites were at the oval window (n = 7), followed by attic (n = 6) and promontory (n = 2). The recurrence was found in two cases. Hearing assessment was done in 48 cases: success in 39 cases (81%), moderate in eight cases, and failure in one. The success rate was 100% both in type-1 and type-3 tympanoplasty, but went down to 70% in type-4 tympanoplasty.

Discussion

One thousand two hundred-twelve cases of acquired and congenital cholesteatomas were treated by the senior author during 1992-2012. The prevalence of CCs, therefore, should be 54/1212 (4.5%). About half of the cases were referred on the basis of abnormal otoscopic findings by outside physicians, others were referred for hearing loss. A planned two-staged surgery was conducted in most of our cases (89%), while one-stage surgery was adopted in six cases (11%). The rate of residual CC at the time of second surgery is very diverse (8-81%) in the literatures. In our cases, the residual cholesteatoma at the time of second stage surgery was detected in 18 out of 43 cases (42%).

Derlacki *et al*. advocate the use of a transcanal extended anterior tympano-meatal flap for limited anterior quadrant CCs and transcanal atticotomy for CCs with superior extension.[2] Levenson *et al*. recommend a transcanal anterior-superior tympanomeatal flap for CCs located antero-superiorly in the middle ear,[3] while

House and Sheehy prefer the post-auricular incision and lateral graft tympanoplasty.[4] They recommend two-stage tympanoplasty, in which replacement of the TM with a fascia graft, and removal of any attic or mastoid extension via mastoidectomy could be done. Only very limited, encapsulated CC is handled in one-stage. Every attempt should be made to preserve the posterior canal wall, but it must be taken down if there is a significant mastoid extension of CCs. At present, we are following the same technique.

As the stage of CC advanced, the area of its invasion could be enlarged, which should result in a higher risk of the CC residual. Considering that CC is usually discovered in its advanced stages (stages 3 and 4), the establishment of a screening program including otoscopic and CT examinations and hearing tests for early CC diagnosis should be required.

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THE INFLUENCE OF MIDDLE EAR INFECTIONS ON CHARACTERISTICS OF CONGENITAL CHOLESTEATOMA

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Introduction

In general, the clinical diagnosis of congenital cholesteatoma (CC) is made based upon the certain criteria: presence of cholesteatoma behind an intact tympanic membrane and operative findings without any demonstrable connection with the external auditory canal.[1] However, if there is middle-ear infection (MEI) or a history of otitis media, it sometimes might be difficult to diagnose because of the high incidence of acute otitis media in children. The aim of this paper is to discuss the location of CC and the influence of MEI on the characteristics of CC.

Method

We investigated 46 cases of cholesteatoma, which had no demonstrable connection with the external auditory canal and which had been confirmed by surgery. Mean age was 10.6 ± 11.4 yrs; gender: 14 female and 32 male; affected side: 30 right ear and 16 left ear. All cases were unilateral. The location of CC in the quadrant and the influence of MEI on the location of CC were discussed.

Results

Applying the Potsic staging system,[2] 21% was single-quadrant CC (stage I), 7% was multiple-quadrant CC (stage II), 66% was ossicles CC (stage III), and 12% was mastoid CC. The locations of CC were divided into four categories, that is, ASQ (antero-superior quadrant), AIQ (antero-inferior quadrant), PSQ (postero-superior quadrant), PIQ (postero-inferior quadrant) and mastoid extension. Based upon the above-mentioned categories, 22% occurred in the ASQ, 0% in the AIQ, 65% in the PSQ, 0% in the PIQ and 13% in the mastoid extension (Fig. 1). Changes of CC location by age were investigated (Table 1). PSQ CC tends to increase with age, whereas ASQ tends to decrease. In addition to that, there were some ASQ CC cases which disappeared with age.

There were 20 cases of CC with MEI or the history of otitis media, in which the mean age was lower and PSQ+ASQ and mastoid extension occurred more frequently; and 26 cases of CC without any history of otitis media in which the mean age was two times higher and ASQ occurred frequently. Moreover, the mastoid air cells were sclerotic, ossicular involvement and ‘closed-type’ cholesteatoma were prominent, and the size of the CC was larger in the CC cases with MEI or the history of otitis media group.

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Cholesteatoma and Ear Surgery – An Update, pp. 147-148
Edited by Haruo Takahashi

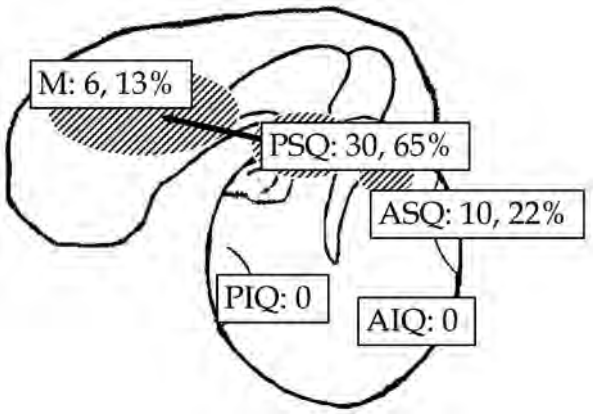

Fig. 1. Location of CC. PSQ CC occurred five times more than ASQ CC.

Table 1. Changes of CC Location by age. PSQ CC tends to increase with age, whereas ASQ tends to decrease.

	3-4 yrs n = 7	5-6 yrs n = 18	7-14 yrs n = 15	< 14 yrs n = 6
PSQ	50%	54%	80%	100% P = 0.038
ASQ	33%	21%	10%	0% P = 0.038
P+A	17%	25%	10%	0%
(M)	(8%)	(13%)	(10%)	(27%)

Conclusion

PSQ CC occurred five times more than ASQ CC. As age increased, PSQ CC increased and ASQ CC decreased. Some of the ASQ CC had disappeared during the observation.

As for the influence of MEI on the characteristics of CC, in the group of no MEI, the patients' ages were higher, the mastoid air cells were significantly more well-pneumatized and ossicular involvements were less noticed. Additionally, open-type CC and small PSQ CC diagnosed as middle-ear anomalies prior to surgery were clearly recognized in this group. PSQ CC tends to expand and ASQ CC tends to shrink or sometimes disappear. It appears that in the presence of MEI CC tends to be larger and closed type cholesteatoma, and frequently erodes IS portion at a younger age.

In conclusion, our study indicates that the presence of MEI affects the location, form, size, extension of the CC and the prognosis of the diseases, and pneumatization of mastoid air cells and ossicular involvement.

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MINI LECTURES

CONSERVATIVE TREATMENT OF CHOLESTEATOMA BY 5-FU OINTMENT

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Introduction

It is well known that middle-ear cholesteatoma is characterized by the presence of a keratinizing epithelium which is believed to have hyper-proliferative properties. In 1985, Smith[1] used 5-Fluorouracil (5-FU) for treating cholesteatoma as an agent suppressing its abnormal proliferation, and reported its clinical effect. 5-FU is a pyrimidine analog developed in 1957.[2] It is an effective inhibitor of both RNA function and the synthesis of thymidylate.[3] 5-FU is recognized as an anti-neoplastic agent effective especially for skin tumors.[4] Wright *et al*.[5] confirmed its efficacy in animal experiments, and Sala *et al*.[6] reconfirmed its clinical efficacy and safety. We have also tried this treatment for cholesteatoma using commercially available 5-FU topical cream, and reported its usefulness.[7]

In this lecture, we introduce: how we perform this 5-FU topical cream therapy, the clinical efficacy of 5-FU topical cream on various types of cholesteatoma, the influence of 5-FU topical cream treatment on inner ear function in guinea pigs and the cell-biological effect of 5-FU cream on middle-ear cholesteatoma.

Method

We use 5-FU topical cream produced by Kyowa Hakko and Kirin Pharma. After cleaning the debris within or on the surface of the cholesteatoma as much as possible, two to three mm^3 of 5% 5-FU topical cream should be meticulously applied on the outer surface of cholesteatoma transmeatally under the microscope (Fig. 1). The application of 5-FU topical cream is done two to five times with an interval of two weeks on an outpatient basis. The cream should not harm healthy skin. Treatment should be continued until the debris disappears.

Results

In the previous study,[7] we investigated the clinical efficacy of commercially available 5-FU topical cream on 50 cases of various types of cholesteatoma. In total, 59% of the cholesteatomas showed good effect, 29% of them showed fair effect, and the effect was poor in the remaining 12%. It was particularly effective in cholesteatomas in the external auditory canal (EAC) (good effect was 84%)(Fig. 2), attic cholesteatomas with an aerated mastoid (good and fair effect was 43% and 36%), and in recurrent type cholesteatoma (good effect was 72%). Particularly in attic type cholesteatoma, a long-term follow up and periodical computed tomography (CT) check up are required for several years after this treatment, because this type of cholesteatoma may have a risk to recur even if it showed a good effect just after treatment. There was no serious side effect observed. No patient recognized hearing loss or tinnitus during or after this treatment, and skin erosion was not seen in any patients. Just in one patient, bone-conduction hearing loss occurred, but the air-conduction threshold

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Edited by Haruo Takahashi

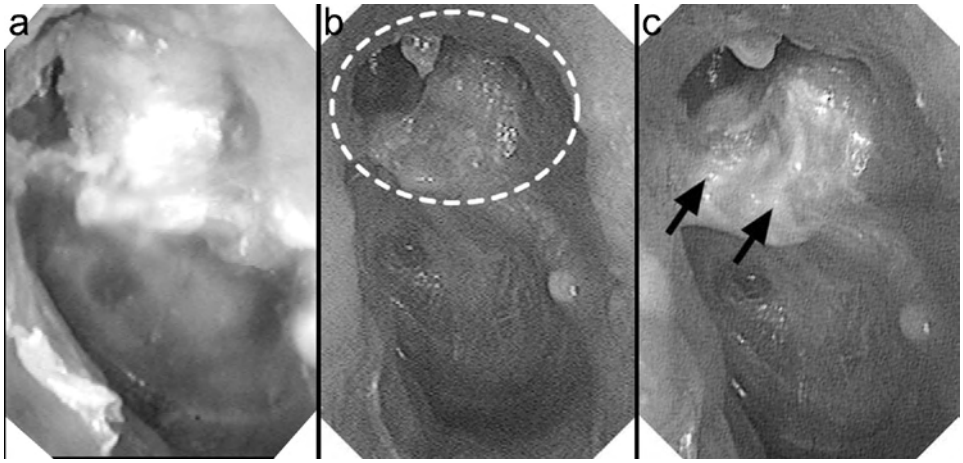

Fig. 1. 5-FU treatment of attic cholesteatoma. a: Step 1, we check the volume of debris, with or without perforation, infection and middle ear effusion before treatment; b: Step 2, cleaning the debris within or on the surface of the cholesteatoma as much as possible. If the patient has inflammation, we use eardrops containing antibiotics, steroids or anti-fungal agents; c: Step 3, 5-FU topical cream is applied exactly on the surface of cholesteatoma. Circle indicates retraction pockets after cleaning the debris. Arrows indicate 5-FU topical cream.

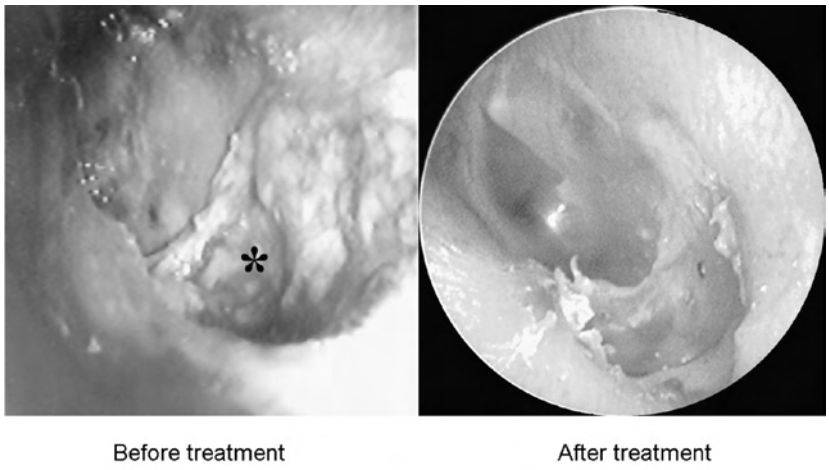

Fig. 2. Ear findings of a representative case with external auditory canal (EAC)-type cholesteatoma showing good effect. Comparing with before treatment, the EAC condition (*) remained clean without debris or infection after treatment, leaving only bone resolution.

remained unchanged and the patient did not complain of a hearing problem. The results of this clinical study indicated that 5-FU treatment is good for the early-stage or EAC-type cholesteatoma, and also for patients who refuse or cannot have surgery for various reasons such as serious underlying diseases, only hearing ear, etc.

The influence of 5-FU topical cream treatment on inner ear function in guinea pigs

One of our colleagues[8] studied the side effects of 5-FU in animal experiments. One week after 5-FU ointment application to the external or middle ear in guinea pigs, the endocochlear DC potential (EP) was measured. When 5-FU ointment was applied on the EAC, there was no significant difference between the EP values of the experimental side and the control side, in other words, inner-ear damage did not occur. When 5-FU ointment was applied on the round window membrane through myringotomy, there was a statistically significant difference between the 5-FU treatment ear and the control ear ($p < 0.05$, Fig. 3). 5-FU ointment application to the external ear appears to be safe but its application to the middle ear may pose some risk of ototoxicity.

The cell-biological effect of topically applied 5-FU on middle ear cholesteatoma

In previous studies, we focused on the possible involvement of both keratinocyte growth factor (KGF) and its receptor (KGFR) in enhanced epithelial cell proliferative activity and recurrence of cholesteatoma,[9] and

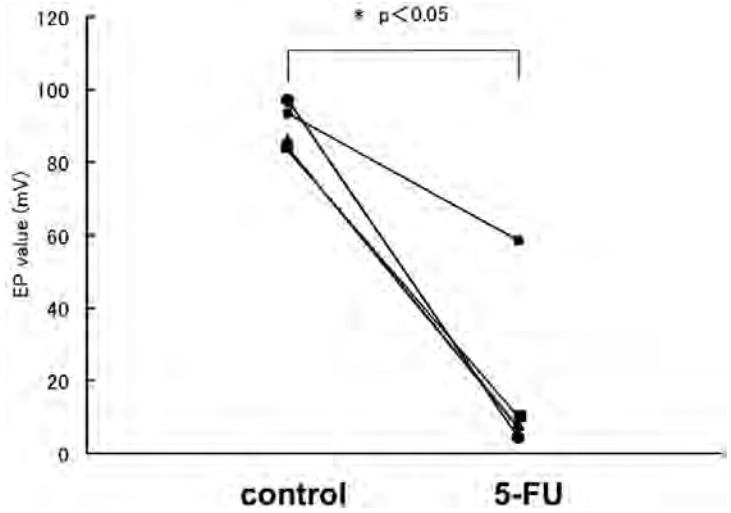

Fig. 3. There was a significant difference between the 5-FU treated ears and control ears ($P < 0.05$). (Reproduced with modification with permission from Iwanaga T *et al*.[8])

we examined the effect of 5-FU on the expression of KGF, KGFR and epithelial cell proliferative activity (Ki-67) in attic cholesteatomas by immunohistochemistry.[10] The study subjects consisted of 77 ears with attic cholesteatoma under 5-FU treatment (n = 12) or not (n = 65). The results showed that the percentage of the KGF-positive cases was significantly lower in the 5-FU group than in the control group (58.3% vs. 90.5%, $p < 0.05$). But the percentage of the KGF-receptor-positive cases in the 5-FU group (50%) was almost the same as that in the controls (60.7%). The Ki-67 labeling index was significantly lower in the 5-FU group than in the control group ($37.5 \pm 0.2\%$ vs. $49.5 \pm 0.2\%$, $t = 2.00$, $p < 0.05$). In the cholesteatoma tissue, 5-FU put on the surface of the epithelium may down regulate the expression of KGF in stromal cells, and may reduce the proliferative activity of epithelial cells through the paracrine action of KGF.

Conclusion

5-FU cream was effective in cholesteatomas particularly in those in the EAC and the attic with an aerated mastoid, and in recurrent-type cholesteatomas. There was no serious side effect observed in this treatment. However, we should use this treatment carefully, and should not put the cream into the middle ear to prevent sensori neural hearing loss. It seems, therefore, recommended to refrain from using it on ears with eardrum perforation. We also found that this agent down-regulated the expression of KGF in stromal cells and reduced the proliferative activity of epithelial cells of cholesteatoma.

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COCHLEAR IMPLANTS AND MUSIC

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Introduction

Recognition of Western music demands high spectral resolution to process pitch cues accurately,[1] but speech sound can be recognized with coarse spectral resolution as long as high temporal resolution is guaranteed.[2] With excellent temporal resolution, cochlear implants (CIs) have been generally successful in enabling users with severe sensorineuronal hearing impairment to attain good speech perception and production.[3] Processing music, on the other hand, had been considered to be far more challenging than speech because of relatively coarse resolution CIs provide. Research on music processing with CIs not only reports levels of perception and production acuity, but also started to reveal benefits of music for users of CIs.

Rhythm processing

Along with pitch processing, rhythm processing is fundamental to music perception and production. CI users can perceive rhythmic patterns at an equivalent level of normally hearing adults.[4-6] Child CI users can reproduce temporal patterning in their sung renditions as accurately as hearing children can.[7]

Pitch processing and music recognition

To judge the direction of pitch change, CI users typically require four semitones or more,[8,9] but the threshold is lower for child CI users when the task is simpler. When required to detect a pitch change in the context of a repeating tone, child CI users can detect change at one semitone.[10]

Melody recognition requires processing of both elementary pitch discrimination and pitch contour. Implanted adults and children seem unable to identify melodies from pitch cues alone.[8,11] In one study, child CI users also showed difficulty in reproducing pitch patterning in their sung renditions.[7] Pitch range was compressed to one third of the range compared to hearing children (means were 76.58 Hz for child CI users and 238.83 Hz for hearing children) and reproduction of directional changes in pitch was at chance levels and significantly lower than hearing children (means were .48 for child CI users and .96 for hearing children).

Enjoyment of music with CI

Despite having difficulty perceiving pitch accurately, most child implant users recognize familiar tunes and enjoy them. From incidental exposure to theme songs of TV programs (anime), child CI users can identify songs in the original commercial recordings (vocal plus instrumental), but not in the instrumental version (identical to the original except for the absence of vocals), nor in the melody-only version.[12] Parents responded to questionnaires on the childrens' music habit. Out of 17 children, all but one child sang at home, but only nine listened to music as a distinct activity. Most of the children who reported to sing, did so dancing and/

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Edited by Haruo Takahashi

or smiling. When tested with pop songs, child and teen CI users identified songs in instrumental version as well as the original version.[10] The children in the two studies gave favorable appraisals of the music even if they were unable to recognize it. Some child CI users may not be appreciating melodic features of music, but they are certainly enjoying rich spectral and rhythmic features of music. In many parts of the world, rhythm plays an central role (*e.g.*, Indian classical music, African drumming). Rhythm actually plays a critical role for young infants when processing music.[13,14]

Exposure to music and enhanced music processing

Processing of spectral information in music is challenging to CI users, but they show improved performance after exposure to music for an extended period of time. CI users showed improved on timbre recognition task after participating in 12-week training on PC designed for enhancing sensitivity to timber perception.[15]

Three teens with implants who participated in the Nakata et al. study[7] agreed to sing six years after their first recordings. Two had taken music lessons at the time of testing (seven and six years of age) and continued for multiple years. The two teens reproduction of directional change in pitch improved tremendously (89% from 69% and 70% from 52%).

Speech prosody as a bridge for gaining music perception and production

Low group performance on pitch perception and production do not mean that CI users are having the same level of difficulty with speech prosody as they do with music. Recent findings on speech prosody processing show CI children can perceive pitch sweep better than expected by previous reports using pitch change detection.[16]

When uttered with positive emotion, features of speech to infants and others with emotional ties are rich in prosodic features that resemble music, such as slow tempo, higher pitch, repetitive and simple pitch contour.[17,18] Infants like to listen to voice with rich prosodic features uttered with positive emotion.[19] Interestingly, six-month-olds prefer audio-visual recordings of their mother's singing over speaking to them.[20]

CI children show benefit of 'musical' features in spoken sentences when identifying the talker. Mothers' speech to both NH and CI infants[21,22] is a typical infant-directed speech which is rich in prosodic features. Until recently, talker identification was considered to be difficult with cochlear implants when speakers are the same gender,[23] but Vongpaisal and coworkers[24] found that child CI users can identify their mother's prosodically rich voice from other speakers, including similar age stranger women's voices. Child CI users could identify their mother's voice even when non-mothers mimicked their mother's voice. Familiarity plays a role in talker identification, but also prosodically rich 'musical' features likely to have played an important role in talker identification with CI.

Research has started to illuminate association between language and music. Music training seems to be associated with enhanced pitch processing abilities[25-27] and higher cognitive functions.[28]

Conclusion

Presumably because hearing individuals do not see the benefit of improving their prosodic features in speech, no study has yet examined the effect of speech prosody training for CI users. Music training and/or extended experience with prosodically rich speech may be a contributing factor common to children with CIs who performed well on pitch perception and production measures. Tremendous efforts and personal sacrifice are involved with CI users who show close to normal music perception and production skills. More research is needed to uncover factors that lead to good music perception and production with CIs.

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GRUPPO OTOLOGICO SESSION

SUBTOTAL PETROSECTOMY IN MIDDLE-EAR AND LATERAL SKULL-BASE SURGERY

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The aim of lateral or subtotal petrosectomy (STP) is to eliminate the middle ear space in order to obtain a regular bony cavity filled with abdominal fat devoid of exposure to the external environment. It involves a double-layer closure of the external auditory canal, a canal-wall-down mastoidectomy with resection of tympanic and ossicular chain remnants and lowering of the facial nerve ridge and Eustachian tube occlusion. The cavity thus created is finally packed with free abdominal fat graft. This procedure has been initially described and used by Fisch since 1988 and was primarily addressed to radically treat pluri-operated discharging ears with compromised hearing.

While adding little or none adjunctive risks for the patients, STP gives definitive advantages in terms of surgical exposure and anatomic control with easy identification of surgical landmarks. In the light of the excellent curative and anatomic long-term results, the subtotal petrosectomy has been adopted for many conditions requiring obliteration of the tympanomastoid system, like temporal bone meningo-encephaloceles, petrous bone fracture with CSF leak and cochlear implantation in complex or unfavorable cases (*i.e.*, malformations, revision surgery, cochlear ossification). STP is also part of skull base surgery basic skills because it may constitute the initial part of a transtemporal (transotic and transcochlear approaches) or combined approach (infratemporal fossa type A and B approaches) in the treatment of skull base lesions. When performing a lateral skull base procedure, STP must be included if a simultaneous cochlear implantation is considered in the pre-operative planning.

The advent of active middle ear implants and bone anchored hearing aids allows for simultaneous or staged hearing rehabilitation in patient undergoing this procedure, if the bone conduction is maintained. This facilitates both the patients and the clinician in the choice of treatment.

The Gruppo Otologico indications for subtotal petrosectomy will be reported, with particular focus on pre-operative clinical and radiologic evaluation, intra-operative technical details, rehabilitation and short- and long-term complications and results. Strategy for radiologic surveillance will also be reported.

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Edited by Haruo Takahashi

EAONO SESSION

DEFINITION OF MASTOID SURGICAL TERMS

THE EAONO SESSION: EAONO GUIDELINE STUDIES ON CHOLESTEATOMA

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Operative terms currently used in ear surgery can be classified according to the anatomic location where the surgical procedure is performed. Most of the ear procedures begin in the mastoid bone. They are not only for infection, but also to access the other regions of the temporal bone such as cochlea or labyrinth.

Surgical procedure for cholesteatoma is defined according to the process on the mastoid bone. However, in the literature, the general term 'mastoidectomy' includes different surgical techniques, and many variations of mastoidectomy with many different names have been proposed, performed and published. As a European Guidelines on Otology Study Group on Otitis Media and Cholesteatoma, we aim to outline the surgical procedures for reporting, publications and follow up.

In the literature, different definitions for simple or cortical mastoidectomy can be found. In the beginning, removing of the mastoid cortex and opening of the infected cavity was performed for acute mastoiditis and it was called 'simple mastoidectomy'. Over time, surgeons realized that complete exenteration of all mastoid cells gives better result and they changed the surgical procedure from removing the mastoid cortex to the complete removal of all mastoid cells. However, sometimes this procedure is incorrectly referred to as 'simple mastoidectomy'. Originally it is used for coalescent mastoiditis, but it is very rarely indicated anymore. Cortical mastoidectomy would be indicated in chronic suppurative otitis media or it provides access for other ear surgeries such as cochlear implantation, labyrinthectomy and so on. 'Cortical mastoidectomy' is the recommended term for this procedure.

Intact-canal-wall mastoidectomy is another closed technique. It has been recommended to avoid the disadvantages of radical mastoidectomy. Originally, Sheehy described this procedure naming intact-canal-wall tympanoplasty with mastoidectomy in his paper published in 1967.[1] Over time, several different terminologies such as 'intact-canal-wall (canal-wall-up complete) mastoidectomy', 'mastoidectomy with tympanoplasty', 'combined-approach tympanoplasty' were used for this technique. Facial recess approach, initially described by Claus Jansen in 1968, is part of the technique most of the time. 'Intact-canal-wall mastoidectomy' is the recommended term for the procedure of complete removal of the air cell system of the mastoid bone leaving the posterior canal wall intact. It may or may not include a facial recess approach. After that hearing can be reconstructed.

Radical mastoidectomy has been defined by two general surgeons based on the surgical principle that a diseased bony cavity should be opened up extensively. Then Zaufal in 1890, described the first radical mastoidectomy. It is a canal-wall-down procedure with exteriorization of the middle ear. No attempt at restoring middle ear function is made. The Eustachian tube is occluded, and the malleus and incus (and possibly the stapes superstructure) are removed. The tympanic membrane remnant is excised, and no graft is placed, leaving the middle ear open. Although it is very rarely used nowadays, the definition is very clear and there is no need for discussion. It may be indicated in situations in which cholesteatoma cannot be completely excised (*e.g.*, cochlear fistula, disease tracking into the petrous apex).

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Most of the confusion over terminology focuses on modified radical mastoidectomy. Frequently, the term ‘modified radical mastoidectomy’ is used interchangeably with canal-wall-down mastoidectomy. Classically, modified radical mastoidectomy refers to the Bondy procedure and in the operation the epitympanum, mastoid antrum and external auditory canal are converted to a common cavity, and tympanic membrane and ossicles are kept to preserve hearing. Originally this operation does not have any reconstructive procedure. However, the term ‘modified radical mastoidectomy’ can be used to describe a canal-wall-down mastoidectomy, even in this procedure; reconstruction of the hearing system is always part of the surgery.

Canal-wall-down mastoidectomy involves removal of the mastoid air cells, saucerization of the cortical edges of the mastoid bone, a complete removal of the superior and posterior canal walls, and a meatoplasty, then tympanoplasty is performed.

The main disadvantage of CWU mastoidectomy is the higher rate of cholesteatoma recurrence. In the open techniques, the rate is lower, however, these techniques have their own disadvantages such as a life-long need for cleaning of the cavity, for avoidance of swimming, possible caloric stimulation and so on. Avoiding these problems, obliteration of the cavity or reconstruction of external auditory canal are the preferred methods. In this case, the technique has been started as an open technique, but the end result is a closed cavity. This technique may define as a closed technique in some papers comparing the rate of recurrent or residual cholesteatoma with the ‘real’ closed technique such as ICW mastoidectomy. For meta-analysis or a correct comparison of scientific papers, the definition of the technique and final situation of the cavity must be clarified.

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INSTRUCTION COURSES

TYMPANOPLASTY THAT WORKS BEST

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Introduction

One of the common sequelae of chronic otitis media is tympanic membrane (TM) perforation, which can cause hearing loss and otorrhea. The two traditional methods for reconstruction of TM perforation have been medial (underlay) or lateral (overlay) graft techniques. In the underlay technique, the graft is placed entirely medial to the remaining TM and annulus. This is perhaps the most common and easiest technique. It is typically used for posterior or central TM perforations. In the overlay technique, the graft is placed lateral to the annulus, and any remaining fibrous middle layer after the squamous layer has been carefully removed. The anterior canal wall is widened with a drill to minimize blunting, and the graft is placed lateral to the remaining TM but medial to the manubrium of malleus to prevent lateralization. The canal skin is placed back as a free graft. Each of these techniques has its advantages and disadvantages.[1,2] We have developed a new medio-lateral graft tympanoplasty technique which seems to be superior to the traditional methods for repairing anterior or subtotal TM perforation.[3,4]

The anterior or subtotal TM perforation is difficult to repair because there is less vascularity than in the posterior tympanic membrane[4] and there is an anterior bony overhang that blocks the vision. Because of reduced vascularity in the anterior tympanic membrane, there is a greater risk of necrosis and re-absorption of the fascia graft.[5] When the medial graft technique is used to repair anterior or subtotal TM perforation, the anterior portion of the fascia graft may fall away, resulting in re-perforation and obliteration of anterior part of the middle-ear cavity.[6] Although the lateral graft technique has a higher success rate for the reconstruction of anterior or subtotal TM perforation, lateralization of graft may occur especially when malleus is absent. These problems have been managed by a variety of surgical techniques, such as the use of Williams's microclip,[8] sandwich-graft tympanoplasty,[6] over-under tympanoplasty,[7] window-shade tympanoplasty,[9] and so on.

During the past 17 years, we have developed and used the medio-lateral graft tympanoplasty for repair of anterior or subtotal TM perforation. In the medio-lateral graft technique, the fascia graft is placed medially to the posterior half of the TM perforation and laterally to the anterior half of the perforation.[3,4] This method is a hybrid of the medial- and lateral-graft techniques that takes advantage of both methods. The purpose of this instruction course is to describe and instruct how to do the medio-lateral graft tympanoplasty for anterior or subtotal TM perforation and medial-graft tympanoplasty for reconstruction of posterior TM perforation.

Materials and methods

Surgical technique

The procedure is usually performed under general anesthesia. Depending on the anatomy and clinical findings, transcanal, endaural, or postauricular approaches are used. A rim of tissue is removed from the perforation edge to de-epithelialize and encourage migration of the mucosal layer and epithelium. Vertical canal incisions are made at the 12- and 6-o'clock positions. The 6-o'clock incision can be extended right up to the annulus.

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Edited by Haruo Takahashi

The 12-o'clock incision is made down to a few millimeters above the annulus close to the short process of malleus to preserve blood supply when anterior canal skin is used as the superiorly based flap. A posterior tympanomeatal flap is elevated, and ossicles are evaluated (Fig. 1A). Mastoidectomy or ossiculoplasty are performed at the appropriate time if needed.

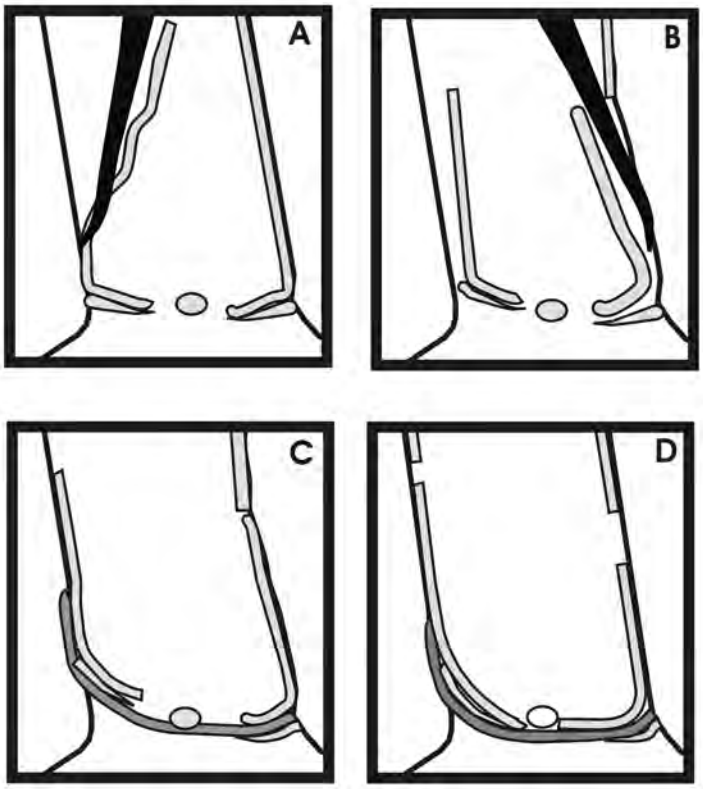

Fig.1. Surgical steps of medio-lateral graft tympanoplasty for anterior or subtotal TM perforation. (A) A posterior tympanomeatal flap is elevated and ossicles are evaluated. (B) Antero-medial canal skin is elevated along with epithelial layer of TM up to anterior half of the perforation. (C) The temporalis fascia is placed medially (underlay) posterior half of the perforation and laterally (overlay) anterior half of the perforation up to the annulus. (D) Antero-medial canal skin is rotated as a superiorly based flap to cover perforation and fascia as a second layer. If necessary, posterior canal skin can be rotated and cover posterior part of graft and perforation especially for the subtotal TM perforation.

Medial graft for posterior TM perforation

In posterior TM perforation, the temporalis fascia is grafted as a medial graft under the tympanic membrane perforation. As shown in Fig. 2, to ensure closure of TM perforation, graft is inserted between manubrium of malleus and TM. To do this the temporalis fascia is harvested, pressed and semi-dried and a slit cut is made. The TM over the manubrium is elevated and one part of the graft is placed between TM and manubrium, and the other part under the TM. The middle ear cavity is packed with Gelfoam soaked in non-ototoxic antibiotic (usually fluoroquinolone) otic drops. The packing is placed as described below.

Medio-lateral graft for anterior or subtotal TM perforation

For anterior or subtotal TM perforation, medio-lateral graft was used. In medio-lateral tympanoplasty, after the tympanomeatal flap is elevated similar to the medial graft technique, a horizontal incision is made in the anterior canal skin with a curved round knife. The distance of the anterior-horizontal canal incision from the anterior annulus should be about the same as or slightly longer than the diameter of the perforation. After the incision, the anterior canal skin is elevated (Fig. 1B), then canalplasty is performed by drilling the anterior bony overhang with diamond burrs and suction irrigator until a full view of the anterior annulus is possible. The antero-medial canal skin flap is elevated up to the annulus or margin of the TM. At the annulus, only the squamous epithelial layer of the TM is carefully elevated to the anterior half of the perforation edge, leaving the anterior annulus intact. The middle ear cavity is packed with Gelfoam soaked in non-ototoxic

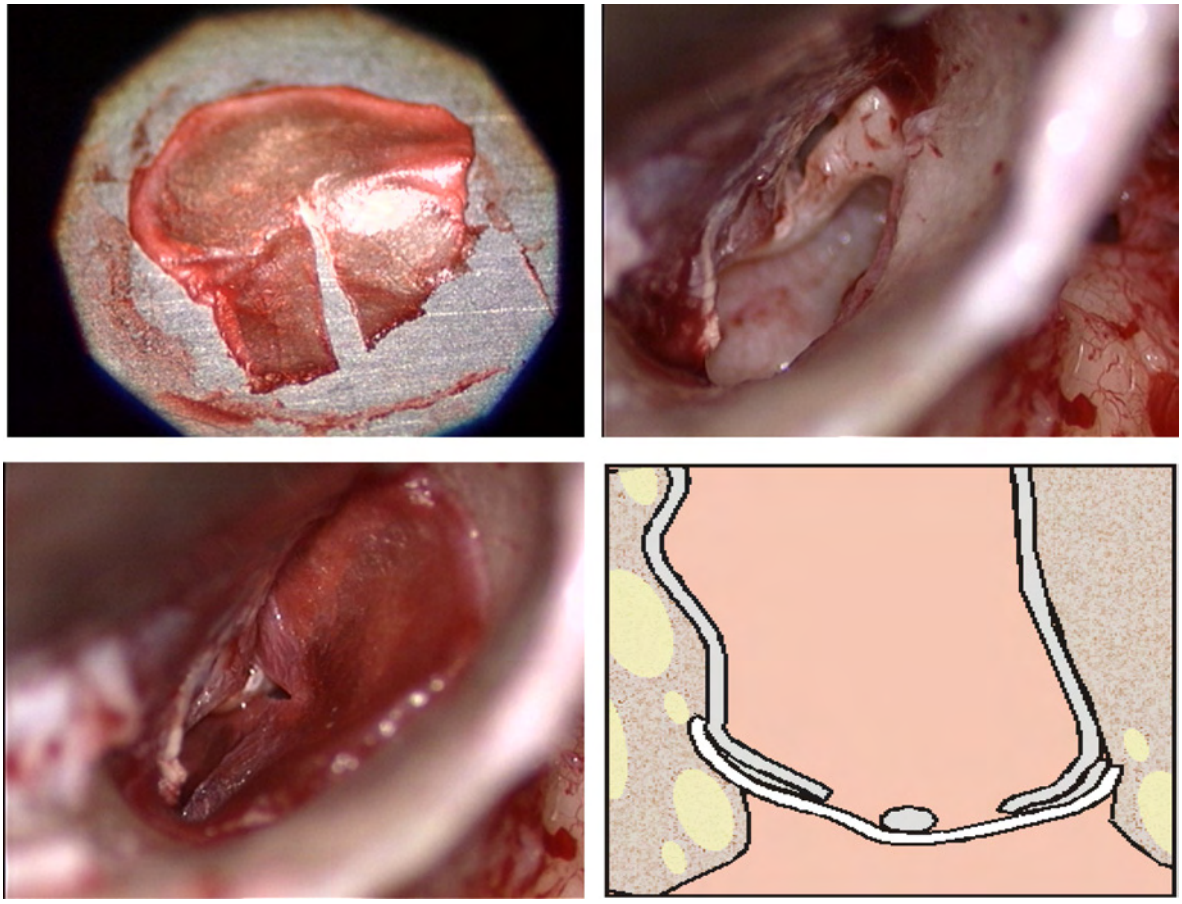

Fig.2. Surgical steps to repair posterior TM perforation with medial graft tympanoplasty. First, the edges of perforation are denuded and the tympanomeatal flap is elevated. From top left, the temporalis fascia is harvested, pressed and semidried and slit cut is made. TM over manubrium is elevated and one part of the graft is placed between TM and manubrium and the other under the TM. Bottom right shows overall scheme of medial (underlay) graft.

antibiotic (usually fluoroquinolone) otic drops. Unlike in the case of usual medial graft technique, the middle-ear packing does not have to be tight because the graft is supported by the intact annulus. In medial-graft tympanoplasty, since the temporalis fascia is grafted medially in relationship to the manubrium of malleus and under the TM perforation and annulus, packing in the middle ear has to be tight to support the graft and to prevent fascia fall-away. In medio-lateral tympanoplasty the temporalis fascia is grafted medially for the posterior half of the perforation and is grafted laterally over the remnant de-epithelialized TM and annulus for the anterior half of the perforation (Fig. 1C). To avoid anterior blunting, the fascia graft is brought only to the anterior sulcus on the annulus, not passing beyond the anterior annular sulcus. As a second layer of closure, antero-medial canal skin is rotated to cover perforation and fascia as a superiorly based flap (Fig. 1D). Antero-lateral canal skin is replaced, and packings are placed. Traditional rosebud packing is inserted by using otosilk strips with a small to medium-sized cotton ball inside, soaked in the antibiotic otic drops. The rest of the ear canal is packed with a gauze strip soaked in antibiotic ointment or Xerofoam gauze. The incision site is closed in the usual manner.

Discussion

Over the years, various techniques have been attempted to improve tympanoplasty results. These include overlay tympanoplasty[10], underlay tympanoplasty,[11] Gelfilm sandwich tympanoplasty,[12] Crowncork tympanoplasty,[13] swinging-door tympanoplasty,[15] sandwich-graft tympanoplasty,[7] window-shade tympanoplasty,[9] and palisade cartilage technique.[16] Among them, underlay and overlay techniques are most commonly used. The advantages of medial (underlay) graft include ease of learning the technique, avoidance of the risk of lateralization and blunting of the anterior sulcus, and high success rate, especially for the posterior perforation. The disadvantages of medial graft are poor visualization of the anterior tympanum, possible anterior graft fall-away when used for anterior perforation, reduction of middle ear space with consequent increased risk of adhesions, and

less suitability for reconstruction of anterior TM perforation.[1,2] The lateral (overlay) graft provides superior exposure, suitable for all perforations, and minimizes reduction of the middle ear space. This technique has a high success rate and has been particularly effective for large, anterior perforations. The disadvantages of lateral graft include anterior blunting, possible lateralization of graft especially with absent malleus, tendency to create more epithelial pearls, need for malleus manipulation, longer healing time, increased operation time, and complexity for repair of small posterior perforations.

One of the most serious complications of the overlay graft techniques is lateralization of the graft. Lateralization of the TM is a condition in which the visible surface of the TM is located either at the bony annular ring or lateral to it and loses contact with the conducting mechanism of the middle ear. Lateralization of the TM may be associated with considerable morbidity, including hearing loss and cholesteatoma. Surgical repair is often necessary for significant underlying disease, but re-establishment of a normal TM can be challenging.[15] Medio-lateral graft tympanoplasty avoids lateralization of the graft by placing the fascia medially to the posterior half of the TM and perforation, as well as the manubrium of the malleus, and laterally to the anterior half of the perforation to prevent lateralization. In our study, there was no lateralization of graft or reconstructed TM.[3,4]

The medio-lateral graft tympanoplasty is a hybrid between medial and lateral graft methods taking advantages of both methods. It has many advantages over traditional medial or lateral graft: (1) prevention of anterior fall-away of the fascia; (2) stability of the graft, like ‘a button in a button hole’; (3) no need for tight Gelfoam packing to support the graft; (4) prevention of lateralization of the graft; (5) better blood supply and faster healing because the anterior canal skin is rotated as a rotational flap rather than a free graft; (6) easier because the epithelial layer of only the anterior half of the TM remnant is elevated, rather than the entire TM; and (7) less malleus manipulation.

Conclusion

The medial tympanoplasty is best suited for repair of the posterior TM perforation with high success rate. The medio-lateral graft method has been developed and used for reconstruction of the large anterior or subtotal TM perforation. Success rate is high (97%), since it takes advantages of both the medial and lateral grafting methods while avoiding their pitfalls.[4] This method should help otologic surgeons to improve the outcome of tympanoplasty for anterior or subtotal TM perforation.

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INTACT CANAL WALL TYMPANOMASTOIDECTOMY FOR CHOLESTEATOMA: TECHNICAL TIPS, TRAPS, PITFALLS AND RESULTS

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Introduction

This instructional course is based on the author's cumulative experience of intact canal wall tympanomastoidectomy in adults and children over a thirty year period.

Method

A critical analysis of the outcome of surgery has enabled an evolution of technique by attempting several different methods of achieving the same end result. An exposition of each stage of this procedure has been possible with a personal opinion of the most satisfactory way of achieving a good surgical outcome. Technical tips for achieving this are discussed in the form of an 'How I do it' presentation. This will include the traps and pitfalls for the unwary embarking on this type of surgery. Reconstructive materials in outer attic wall repair and the effect on the outcome of the use of the oto-endoscope and KTP laser will be revealed and discussed in relation to the evolution of technique over time.

Results

The long-term results of this surgery are presented in 245 patients and an analysis of 543 operations. The residual cholesteatoma rate at the second stage was 11% in the adult group and 28% in the children's group. A third-stage operation was required in 13% of the adults and 25% of the children. The recurrent cholesteatoma rate was 2.3% in adults and 6.6% in children.

Discussion

This study has demonstrated a relatively low residual cholesteatoma rate in adults but a higher rate in children and the recurrent cholesteatoma rate which was low in adults was also higher in children. These results compare favorably with other studies and reflect the difficulties in managing cholesteatoma in children. A personal view of how best to improve the outcome in intact canal wall tympanomastoidectomy has resulted from this study.

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Edited by Haruo Takahashi

MIDDLE-EAR CLEFT GAS DEFICIT DISEASE TO MIDDLE-EAR CLEFT ACQUIRED CHOLESTEATOMA: PATHOGENESIS-ORIENTED MANAGEMENT

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Middle-ear cleft gas deficit disease / atelectasis

Definition

Middle-ear cleft atelectasis is an incomplete expansion of the middle-ear space associated with medial displacement of the eardrum towards the promontory. In most cases, the eardrum is atrophic. Middle-ear cleft atelectasis belongs to chronic otitis media. The clinical picture is a tympanic membrane retraction pocket which consists of an invagination of a weakened tympanic membrane into the tympanic cavity.[1]

The retraction pocket constitutes a distinct clinical entity, the importance of which is determined by three types of criteria: topographical, quantitative and qualitative. The topographical criterion means the location, in the pars flaccida, in the pars tensa, or in both. The quantitative criterion refers to the dimension – is it total, or partial; the depth – is the pocket deep or shallow; does the extent allow to control it under the operative microscope? The qualitative criterion refers to the behavior of the pocket – is the pocket self-cleaning or not; is there a bone erosion of the lenticular process of the incus and/or of the tympanic frame; is the pocket adherent to the middle-ear structures, or not?[2]

Pathogenesis

The pathogenesis of the middle-ear cleft atelectasis is pluri-causal.[3] This is, in fact, no matter of inappropriate scientific curiosity, but rather the sharing of a conviction that there are at least five main causes to be carefully taken into account when therapeutic management of the pocket is undertaken.

The indisputable starting point is the presence of an inflammatory process of the upper-airways mucosa and, more particularly, of the middle-ear cleft mucosa. Once initiated, the process will pursue its natural course which will lead to the picture of a well-determined clinical entity. This is a continuous and as such a long dynamic process.[4,5]

A second cause is the negative pressure, the ‘gas deficit’, in the middle-ear cleft. How does gas deficit happen? The middle-ear cleft is lined by mucosa with an embedded vascular network, which provides the metabolic requirements of the mucosa and also represents a source and sink for trans-mucosal gas exchange between the middle-ear cleft and local mucosal blood.

Gas exchange depends on variations in the middle-ear cleft blood flow. In a similar fashion as alveoli in the lung, the direction of gas exchange is predicted by the differences in partial pressure of the component gases in the middle-ear cleft and in the blood compartment.

When the mucosa is healthy, there is an equal rate of gas exchange because oxygen and nitrogen are absorbed by the mucosa in the same rate as carbon dioxide is expelled. For a given mucosal blood flow, nitrogen is the rate limiting factor of the middle-ear cleft gas loss because it diffuses slower than carbon dioxide and oxygen that equilibrate relatively quickly with venous blood.[6] Inflammatory processes increase both number

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Edited by Haruo Takahashi

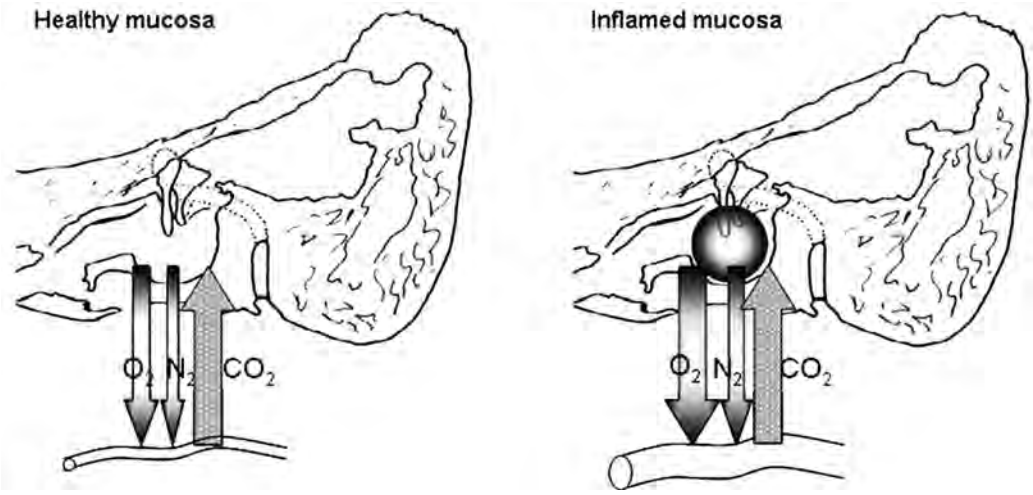

Fig. 1. Gas exchanges between the middle-ear cleft and the blood compartment through healthy (a) and inflamed mucosa (b).

and diameter of the blood vessels in the middle-ear cleft, increasing the blood flow and gas exchanges accordingly.[7] (Fig. 1).

The more inflamed and vascular the mucosa, the greater the rate of gas absorption.

When a light gas deficit, or a small negative pressure, that are normally well compensated, increases into the middle-ear cleft to the extent that it becomes no more compensated, pathology occurs. This constitutes one of the causes of the tympanic membrane retraction pocket formation.

A third cause is the atrophy of the tympanic membrane lamina propria, atrophy of the mesenchymal layer.

General process: The atrophy of the lamina propria never occurs as a sudden or isolated process, but always implies a more or less long-standing involvement. This tissular atrophy is explained by two kinds of mechanisms: biophysical and biochemical.

The bio-physical explanation is based on the visco-elastic properties of the lamina propria.[8] The tympanic membrane lamina propria is a network of load-bearing fibers embedded in a gelatinous matrix. The fibers are cross-linked. When subjected to constant stress, due to 'negative pressure' in the middle-ear cleft, the cross-links gradually break down causing further extension of the material. The volume of soft biological tissue is believed to remain constant under the action of external forces. Therefore, when the tympanic membrane lamina propria is stretched, it will inevitably become thinner.[9]

Under inflammatory conditions, the tympanic membrane gets thicker both at sub-mucosal and sub-epithelial layers. This thickening could be responsible for an increase in constraint forces applied on the tympanic membrane and could lead to a degenerative process of the fibers of the lamina propria. When the atrophy has been installed, the tympanic membrane loses his mechanical armature and a retraction may occur even under the effect of moderate middle-ear cleft pressures.[10]

The bio-chemical explanation is based on the damage on the connective tissue, particularly, the rupture of the di-sulfure bridges, by collagenases, metallo-proteases and cytokines, liberated during the inflammatory process.

Concerning the local process: we know that, in adults, the morphologically weaker areas of the tympanic membrane are the pars flaccida and the postero -superior quadrant of the pars tensa.[3]

The more specific weakness of the pars flaccida is the notch of Rivinus that corresponds to the absence of the tympanic bone at the top of the tympanic frame. At that point, there is also no fibro-cartilaginous ring. Histologically, at this point, the structure of the lamina propria is not clearly organized and, upon dissection, we have even noted congenital defects in these fibrous layers, in 4% of normal adult temporal bones.

With regard to the weakness of the posterior part of the pars tensa, since its early development the posterior arm of the tympanic bone is the most delicate portion of this structure, and is thinner and shorter than the anterior arm. The circular fibers of the lamina propria are more delicate and are seldom seen in this area. The area is thinner (about 60 microns) than near the rim (about 90 microns). The strain must therefore be greater in the thinner parts.

A fourth cause considered is the tympanic membrane stratified squamous epithelium dysfunction. This epithelium is the site of two kinds of specific movement. The centrifugal keratin dispersion process involves the superficial layers, and is particularly well suited to cleaning of the external auditory canal.[11] When disturbed, it provokes an accumulation of keratin into the pocket.

The centripetal epithelial migration process is conspicuous in some pathological cases such as, for example, the healing of tympanic membrane perforations.[12]

Centripetal movement involves the whole stratified squamous layer which lines the perforation.[13] In this case, two conditions are required: the presence of free edges and the presence of a substratum able to give the cells a specific directional impulse. This is the 'contact-guidance' phenomenon, explained by Weiss.[26] These two conditions do not exist when considering a retraction pocket.

An additional contributor is the concomitant association of the local morphological predisposing factors which act as a catalyst agent for the pathogenic mechanism of the tympanic membrane retraction pocket. These local morphological predisposing factors are the unlimited anatomical variants of the temporal bone and their consequences upon the physiology of the middle-ear cleft.[14] They are the result of the dynamic of the organogenesis of the entire temporal bone. This organogenesis is a true 'living puzzle'. Each piece has to develop individually, and at the same time, fuse with the other pieces which are also growing, immersed in unstable surroundings. No more than a minute delay or minuscule modification in the sequence of those programmed events and more or less severe modifications of the normal morphology will be seen. This could generate torsion and traction of the mucous folds of the middle-ear cleft, which would have an effect upon muco-ciliary clearance and pressure variations balance in the middle-ear cleft, associated with nerve and blood supply disturbances and subsequently interactions on the local metabolism.[15] They are particularities or variants which are not considered to be anomalies or pathological phenomena because of the frequency of their occurrence. These variants are tiny morphological modifications, surgical observations, which may sometimes be correlated with certain functional or clinical events: 91% of the external auditory canals that we dissected at the time of tympanoplasty, in cases of chronic otitis media, show protruding lateral posterior bony cristae, as opposed to only 19 in 100 normal temporal bones. We can explain this fact by a slight rotational shifting of the tympanic ring during its early development. Moreover, the fusion of the two arms of the tympanic bone with the other components of the temporal bone takes place during a relatively long period of time and is not synchronous.[16] The definite coupling, slow or rapid, is responsible for minute particularities in the normal morphological development. The concomitant association of local morphological predisposing factors with other causes initiates and triggers the other pathogenic causes, namely: inflammatory process of the upper air ways mucosa, middle-ear cleft 'negative pressure' and both tympanic membrane lamina propria atrophy and stratified squamous epithelium dysfunction.

So, the pathogenesis of the middle-ear cleft atelectasis is really pluri-causal!

Therapeutic management

Therapeutic management of the middle-ear cleft atelectasis involves a triple approach and concerns three entities: firstly, the general curative treatment of the upper airways mucosa inflammatory process, which is particularly important in children; secondly, the prospective curative treatment of the middle-ear cleft pressure variations imbalance; thirdly, the local curative treatment of the tympanic membrane pocket itself. The three areas which have to be taken into consideration simultaneously in the therapeutic management of the atelectatic ear are the upper air ways mucosa; mastoid, tympanic cavity and the fibrocartilaginous Eustachian tube; and the tympanic membrane pocket itself. This therapeutic management of the atelectatic ear is pathogenesis-oriented.

The general curative treatment of the upper airways mucosa inflammatory process

The general curative treatment of the upper airways mucosa inflammatory process must take into consideration the rhino-pharyngeal and sinusal mucosa disease which will be treated by way of antibiotics, anti- inflammatory, and anti- allergic drugs, either *in loco*, by spray, nose-drops, inhalations, aerosols, or generally, mostly orally. Adenoidectomy, or any surgical procedure in the sinus, will be performed, if necessary. In some cases, a stimulation of the immunity is proposed. The pharyngo- laryngal reflux of the gastric content occurs mainly during the night because of the fact that the superior sphincter undergoes a physiological relaxation during nighttime. The gastro-oesophageal and laryngo-pharyngeal reflux will also be treated, by the use of prokinetics drugs which prevent mechanical reflux or other drugs which reduce the chemical aggressiveness of the reflux either forming a protective gel on the surface of the acid gastric liquid – the principle of the 'alginates' – or neutralizing the acid of the stomach – the principle of the 'anti-acids', sealing off the production of acid by the

gastric cells the ‘anti-secretory’ drugs. They contain two families, depending on their respective mechanism of action: Anti H2 and IPP, inhibitor for the protons pump.

Active or passive smoking as well as atmospheric pollution will be suppressed.

The prospective curative treatment of the middle-ear cleft pressure balance loss

Common medical language often evokes the essentials of a ‘well-ventilated middle ear’ while, in fact, a healthy middle-ear cleft is a poorly ventilated, but an intensely perfused middle-ear cleft. The normal balance of pressure variations in the middle-ear cleft depends on the one hand, on the transmucosal gas exchange between the middle-ear cleft and local mucosal blood (the middle-ear cleft includes tympanic cavity and mastoid); and on the other hand on the normal function of the fibro-cartilaginous Eustachian tube.

Treatment of the transmucosal gas exchange disturbances of the middle-ear cleft: tympanic cavity and mastoid gas cells system that are disturbed by the inflammatory process of the middle-ear cleft mucosa

The medical treatment is the same as for the upper airways mucosa, mentioned above. The surgical treatment consists of the curettage of the inflammatory granulation tissue and removing of the pathological post-inflammatory mini-partitioning in the middle-ear cleft. The ideal surgical procedure must acknowledge the need for eradication of all diseased mucosa while preserving the largest surface area possible of healthy mucosa as well as assuring regeneration and restoration of the physiological volume of the gas cell system. Chronic granulation tissue obstructing the aditus ad antrum has been found more frequently in patients with atelectatic ear. It must be removed by curettage. Because of the unhealthy status of the mastoid, opening is essential and permits drainage of the inflammatory secretion, removal of inflammatory granulation mucosa and creation of an enlarged gas reservoir. In contrast, the entire mastoid gas cell surface area is reduced after mastoidectomy, as the fine gas-cell walls are removed. Because the passive physiologic pressure buffer function may be partially governed by the mucosal surface area, it may not be desirable to remove all the gas cells when the mastoid contains healthy cells lined with thin normal mucosa. The posterior wall of the external auditory bony canal has to be preserved or reconstructed because it constitutes an impenetrable barrier giving the mastoid gas cell system the means of performing its function. Simple mastoidectomy is not sufficient. An antro-attico-mastoidectomy, taking care to preserve the scutum and the posterior bony canal wall, and, if necessary, completed with the posterior tympanotomy, must be performed with the aim of restoring the function of gas exchange to the postero-superior part of the middle-ear cleft.[17]

When surgical approaches to the middle-ear cleft are compared and considered it is important to remember that after a canal-wall-up procedure, the mastoid retains its native cuboïdal nitrogen-absorbing epithelium. An inflammatory process increases again both the number and the diameter of the blood vessels; thereby increasing the diffusion of gas. A slight compensated middle-ear cleft gas deficit – negative pressure – increases. Here too, it is no longer compensated and becomes pathological.

The canal-wall-down technique enables the removal of the nitrogen-absorbing mucosa of the mastoid. The new epithelial lining of the mastoid bowl is a stratified keratinizing epithelium. In canal-wall reconstruction, with mastoid obliteration, the removal of the nitrogen-absorbing mastoid epithelium and the obliteration with bone paté, decrease the likelihood of ‘negative pressure’ recurrence.

The mastoid obliteration techniques allow to reduce the function of transmucosal gas absorption in the middle-ear cleft and to reduce, in this way, the recurrence rate of the pathologies that lead to a ‘gas deficit’ in the middle-ear cleft. The main reverse of this surgical technique is the suppression of the buffer effect of this great gaseous area. But, what about the long-term complications of such a technique, including intracranial cholesteatoma and/or abscess. Even if regular controls are performed by accurate imaging of the obliterated mastoid cavity I remain sceptical.

Colleague surgeons too often think according to a mechanistic approach, conceiving the operated middle-ear cleft as permeated by gaseous flows. However, they must think along the lines of a physiologic approach as well. They must conceive the operated middle-ear cleft as a cavity which is covered by a mucosa through which gas diffusion occurs and where each gas nitrogen, oxygen, water vapour, has its own specific diffusion rate that influences the balance of pressure variations in the middle-ear cleft.

Treatment of the fibro-cartilaginous Eustachian-tube dysfunction

Eustachian-tube dysfunction may involve an abnormality at any stage of the dilatation cycle.[18] Eustachian-tube dysfunction is characterized by the inability to equalize imposed pressure differences between the middle-ear cleft and the rhinopharynx. Eustachian tube dysfunction contributes consequently to a middle-ear cleft negative pressure. Eustachian-tube dysfunction is caused by neuro-muscular disorders in 8%, and by mucosal disorders in 92%.

Neuro-muscular disorders are excessive or too low contractions, lack of coordination of one or more muscles involved in the opening, dilating (tensor veli palatine) and closing processes of the fibrocartilaginous Eustachian tube. The neuromuscular disorders of the fibrocartilaginous Eustachian tube are constructively managed by logopedy, by a speech therapist. This therapy is based on the principle of neuro-muscular interaction and on the willpower of the patient. The patients must blow the nose, blow air directly into the ears using the Valsalva maneuver, close the mouth and breath through the nose, and activate the peritubal muscles by stimulating them with various types of exercises.

Mucosal disorders consist of inflammation of the mucosa, that give some obstruction of the tubal lumen, but also disorders of the 'pressure' and 'gas mixture' sensors of the entire system: middle-ear cleft and fibro-cartilaginous Eustachian tube.[19] The mucosal disorders, and particularly the inflammatory process, will be treated by the triple way of medical, surgical and rehabilitative approaches.

The medical approach is just the same as the medical treatment of the inflammatory process of the upper air ways mucosa, explained above. Concerning the surgical approach, at present two surgical procedures are described clinically. Firstly, the laser Eustachian tuboplasty described by Oskar Kujawski and Dennis Poe that consists of an endoscopic transnasal-laser vaporization of inflamed mucosa and cartilage from the luminal posterior wall of the rhinopharyngeal orifice of tube. Indications are extremely restricted. Secondly, the 'balloon dilatation Eustachian tube tuboplasty' described by Holger Suddhoff, which has the great merit to attempt to treat in a non-invasive and so to say easy manner, the functional obstruction, situated at the level of the Eustachian tube isthmus.[i]

In order to manage a functional defect, a rehabilitative approach seems to be more logical and more clinically and scientifically based.

The rehabilitative approach consists of the use of the Kinetube®.[20] The aim of this rehabilitative device, is to restore the normal behavior of the tubal opening, by generating a calibrated positive pressure in the rhinopharynx during swallowing. Kinetube provokes a kind of 'massage' of the 'pressure' and 'gas-mixture' sensors, at the rhino-pharyngeal orifice of the fibrocartilaginous Eustachian tube, that rehabilitate both central and peripheral nervous systems of pressure regulation in the middle-ear cleft, with retro-control between middle-ear cleft mucosa and the muscles of the fibrocartilaginous Eustachian tube.

Local curative treatment of the tympanic membrane retraction pocket

Therapeutic management of the tympanic membrane retraction pocket is a matter of detecting subtle changes and then assigning relative importance to the observed changes in a way that allows, to ear surgeon, to assess the risks and benefits of a proposed surgical procedure, compared with the risks and benefits of continuing approach without any surgical intervention.

Surgical management of the retraction pocket is controversial because the natural course toward cholesteatoma development cannot be predicted with certainty and hearing acuity remains normal until later in the disease course. Consequently, surgery is often delayed until there is clear indication, such as hearing loss or clear cholesteatoma development, but such delay often necessitates more extensive surgery. Because earlier intervention appears to be in the best interest of the patient,

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i. The balloon, once correctly positioned in both the cartilaginous and bony portion of the Eustachian tube, is inflated to a 20-mm length and a three-mm width, to a pressure of ten bars for two minutes. However, the following questions must be considered: Could you imagine that a only mechanic pressure, during two minutes, could solve a chronic inflammatory hypertrophy lasting for long, months and sometimes years? Or cure a long-standing muscular dysfunction? And, in practice, could such a mechanical surgical operation generate scars: fibrosis with the risk to transform a functional obstruction into an anatomic obstruction! Dennis Poe demonstrated that balloon dilatation of the cartilaginous portion of the fibro-cartilaginous Eustachian tube was feasible and appeared to be safe in a cadaver trial. Paul Presti demonstrated that the poly-L-lactide Eustachian tube stent was well tolerated by the test animals and completely resorbed at six months.

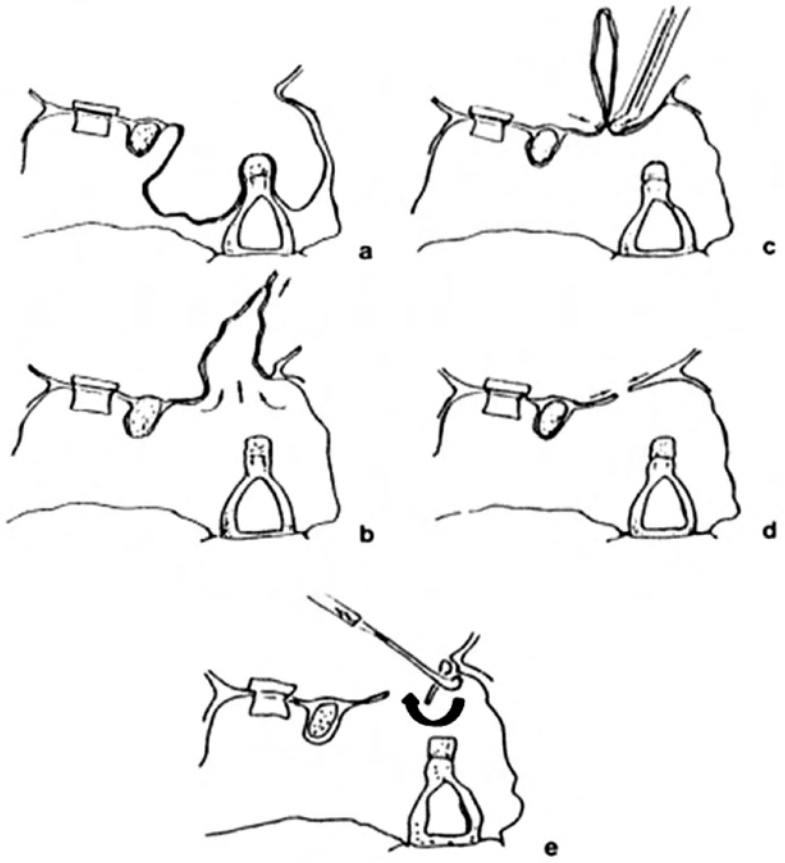

Fig. 2. Curative management of the tympanic membrane retraction pocket, at level 2: surgical steps. (a) Implantation of a ventilating tube through any part of the tympanic membrane that still appears to be covering a ventilated part of the tympanic cavity; (b) Progressive suction of the atelectatic tympanic membrane to convert the retraction pocket into a lateral sac; (c) Pressure followed by: (d) Cutting of the edges of the lateral sac, to prevent eventual recurrence after tube extrusion.

I propose a staging system and a decision scale for therapeutic management of the tympanic membrane retraction pocket.

Local curative treatment of the pocket itself varies according to both the topography of the pocket and the evolutionary character of the pathological process.

Concerning the topography, 30% of the tympanic membrane retraction pockets belong to the pars flaccida and 70% to the pars tensa.

The evolutionary character of the pathological process is given by our classification of the tympanic membrane retraction pockets, based on clinical and tympano-microscopic findings. The quantitative as well as qualitative criteria, considered altogether in an evolutionary pathological character, define three levels of therapeutic management decision.

At the first level, the pocket is safe and stable. The risk for developing cholesteatoma is low. The histological quality of the eardrum does not change with time. Hearing remains normal. The pocket is potentially reversible.

At the second level, the pocket is formed and uncertain. The risk for developing cholesteatoma is not clear. Morphological changes, such as progressively deepening retraction and/or drum hypotrophy, are observed over time; but hearing remains normal, unchanged over time.

At the third level, the pocket is unstable, characterized by adhesion to the surrounding structures, by erosion of the ossicular chain and bony frame, by a non-self-cleaning phenomenon which leads to accumulation of keratin, and by the presence of moist mucous polyps, indicating an additional local irritation.

The risk for developing cholesteatoma is very high. When the pocket is truly destabilized, cholesteatoma starts to evolve. Morphological changes are occurring or have stabilized with symptoms that may include otorrhea, bleeding and discomfort. Conductive hearing loss is usually present.

In case of local curative treatment of the pars tensa pocket, the pocket is the result of a locking of the anterior isthmus of the inter-attico-tympanic diaphragm. The postero-superior quadrant of the pars tensa becomes attracted in the direction of the retro-tympanum.

Level 1: the pocket is safe (44%). Treatment is conservative: regular cleaning of the eardrum and ear canal, by suction under an operative microscope, and, if necessary, with the aid of the endoscope.

Level 2: the pocket is uncertain (43%). We perform the excision of the atelectatic part which has become excessive, and we implant a tympanostomy tube through any part of the membrane that still appears to be covering a place where gas remains, ventilating the tympanic cavity.

The surgical steps are implantation of the tympanostomy tube, progressive suction of the atelectatic membrane to convert the pocket in a lateral sac (Fig. 2), pressure followed by cutting of the edges of the lateral sac, to prevent eventual recurrence after tube extrusion. In some cases, when the reverted sac is too flaccid, it may be necessary to raise the remnants of the tympanic membrane and partially dislocate the annulus from the bony sulcus. Healing is the usual evolution in less than eight days, tympanic membrane becomes again closed. Scarring is just the same as for traumatic perforation.[21] Sometimes, if recurrence occurs, we turn to a CT scan of the mastoid for information concerning the pneumatization.

With a well-pneumatized mastoid, we need only to repeat the precedent procedure:

we add an antro-attico-mastoidectomy , taking care to preserve the scutum and the posterior bony canal wall. If necessary, we complete with a posterior tympanotomy.

Level 3: the pocket is unstable (13%). The fundamental cause is lack of the lamina propria of the drum. For this reason, the lamina propria has to be reinforced by conchal cartilage *e.g.*, perichondrium composite graft), or better, replaced.

Personally, we replace it by a tympanic membrane allograft.[21] Here too, a CT scan informs us about the pneumatization of the mastoid. Large cells: an allograft myringoplasty is sufficient; scanty cells: these call for the opening of the posterior cavities.[ii]

When necessary, we complete the reconstructive surgery with a two-hole ossiculoplasty: for this, the incus is remodeled and fitted in between the handle of the malleus and the mobile stapes; associated or not with a reconstruction of the posterior bony canal wall.[22]

Local curative treatment of the pars FLACCIDA pocket

The pocket is the result of a complete locking of both anterior and posterior isthmi of the inter-attico-tympanic diaphragm. The pars flaccida becomes attracted into the epitympanum.

At Level 1, the pocket is safe and requires a conservative management.

At Level 2, the pocket is uncertain and must undergo the same conservative attitude consisting of a regular cleaning by suction under an operative microscope. Levels 1 and 2 contain 75% of the pars flaccida retractions.

At Level 3, the pocket is unstable and must receive a tympano-ossicular allograft tympanoplasty, with opening of the mastoid.

Middle-ear cleft acquired cholesteatoma

Definition

Middle-ear cleft cholesteatoma consists of the presence of a wrong stratified squamous epithelium in a wrong place: in the middle-ear cleft (Fig. 3). Cholesteatoma is characterized by an accumulation of desquamating keratinized epithelium and an osteolytic power.

Clinical observation indicates the moment when the transformation from tympanic membrane retraction pocket to cholesteatoma occurs. This happens when the pocket becomes fixed to the surrounding structures,

ii. The technique of tympanic membrane allograft implantation consists of both: total excision of the pathological membrane including the annulus. It is called myringectomy and allograft myringoplasty: replacement by a new tympanic membrane. In Belgium, we have a specific legislation which allows the benefit of allografts. In this situation, the tympanic allograft offers a double advantage: natural structure of the lamina propria, which gives to the epithelial cells the fitting directional impulse and rigidity of the membrane, due to the preservation in formaldehyde, which prevents recurrence of retraction.[23]

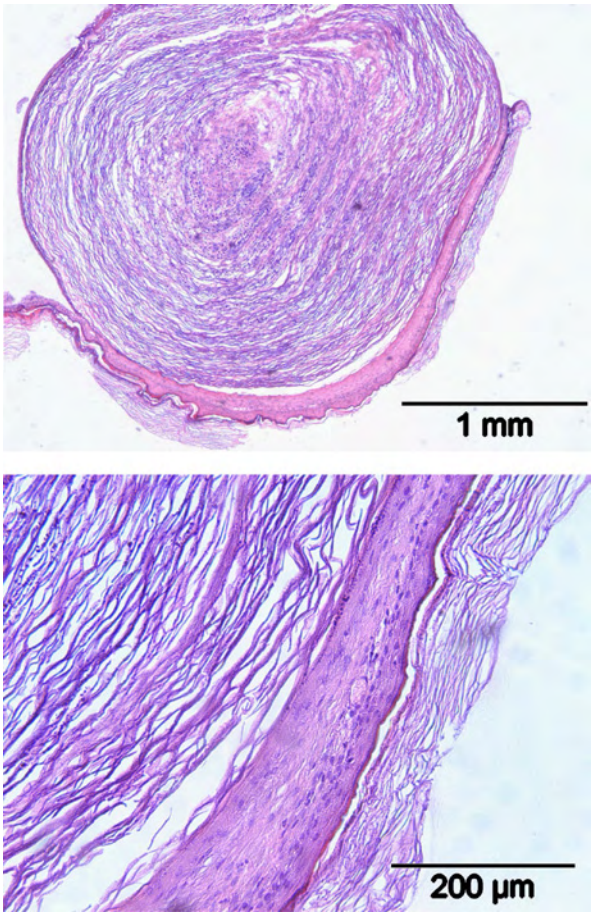

Fig. 3. Histological section through a middle-ear cleft cholesteatoma. (a) accumulation of desquamating keratinized epithelium; (b) The black rectangle points out a region of osteolytic power which is magnified in (3b).

starts to erode the ossicular chain and bony frame, is no longer self-cleaning, which leads to the accumulation of keratin, and shows humid mucous polyps, which indicates the presence of a secondary stimulus, irritating substances, additional local activity, or additional local inflammatory and/or infectious processes.

From tympanic membrane retraction pocket to cholesteatoma[24]

Histological sections of the mucous polyps of a destabilized tympanic membrane retraction pocket show that the granulation tissue contains several inflammatory cells. Two main features occur: the accumulation of keratin, which is the result of the disappearance of the centrifugal keratin dispersion process, and the presence of large Langerhans' cells, which are normally rare in a normal tympanic membrane and which are particularly numerous in cholesteatoma (Fig. 4). As we see in cases of cholesteatoma, Langerhans cells show an increased volume of numerous vacuoles and the loss of dendrites.

However, we know that cholesteatoma is the result of a conflict between stratified squamous and mucous epithelium. A similar conflict exists at the cutaneo-mucous junction of the tympanic perforation. Both situations coexist with a serious localized accumulation of inflammatory cells, which indicates that the cutaneo-mucous junction is not stable and its front can be considered an epidermal defect which is continually being renewed.

From the start of embryogenesis, the epithelium of the bottom of the external auditory canal is pushed forward by the power of aggressive growth.[25] Parallel and concomitantly, this growth forward is regulated by control processes which put a brake on its forceful progression. Loss of the normal regulation of the growth of the squamous stratified epithelium takes place at the bottom of the external auditory canal, due to the secondary presence of an additional local stimulus such as, for example, irritating substances, or an additional local inflammatory and/or infections process, including a papillomavirus.

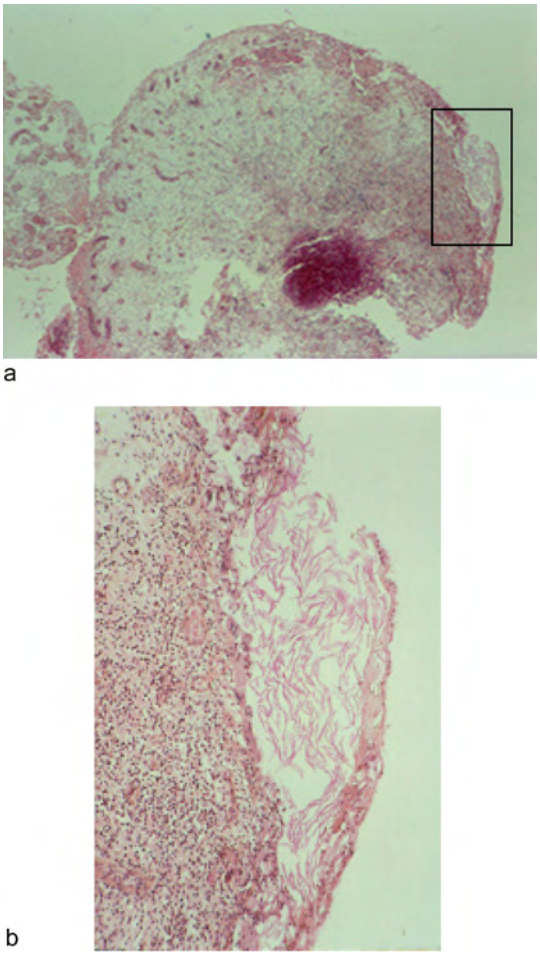

Fig. 4. Histological sections through a mucous polyp of a destabilized tympanic membrane retraction pocket. (a) General view. The black rectangle on the right side points out the accumulation of keratin which is magnified in figure 4b; (b) The magnification of the section through the mucous polyp showed in 4a shows granulation tissue with several inflammatory cells and the accumulation of keratin.

There are two arguments which justify this loss of control of the growth of the squamous stratified epithelium. Firstly, this epithelium develops out of the middle-ear cleft, which is its normal anatomical site, and, consequently, the 'contact-guidance' phenomenon (described by Weiss[26]) does not exist there anymore.

Secondly, due to mucous-epithelial interaction, the inflammatory process produces a self-supporting immunological cycle, in answer to the conflict between cells and tissues.[27] Bacterial proliferation and super-infection of the accumulated keratin debris form a biofilm that lead to chronic infection and epithelial proliferation.[iii]

The latter appears to be influenced by the cytokine-mediated inflammatory response.

Cholesteatoma derives from the epithelium at the bottom of the external auditory canal. This concept is based on the results of experimental works: the similar structure of the stratum corneum and the comparable biology. In the epithelium at both the bottom of the external auditory canal and at the level of the matrix of the cholesteatoma, the presence of identical growth factors, similar reactions to markers of epidermal differentiation, and identical cytokeratin expression, can be seen.

This cytokeratin expression is able to induce hyper-proliferation cell reactions at the level of the normal epithelium at the bottom of the bony canal, and is identical to their expression in the cholesteatoma matrix, contrary to that located in the cartilaginous canal, where their expression is almost non-existent. The stimulating action of the invasion process is the key to the transformation of the tympanic membrane retraction pocket into a cholesteatoma. This process acts as a stimulus to discharge the expression of cytokeratins of cell hyper-proliferation. In other words, the additional local irritation process triggers the keratinocytes at the bottom of the bony canal, leading to cholesteatoma.

iii. A biofilm is a community of micro-organisms encased within a self-produced, extracellular, polymeric substance which is attached to the surface of the micro-organisms.

Therefore, a tympanic membrane retraction pocket is a disease of the middle-ear cleft. Cholesteatoma should be considered as pathology of the squamous stratified epithelium at the bottom of the external auditory canal. This could, however, be two stages in the evolution of the same pathological process.

Therapeutic management

Medical management of cholesteatoma

At the present time, the treatment of cholesteatoma remains surgical. However, when it is proved to be impossible or when it must be delayed or to prepare surgery, the disease may be controlled for some time by repeated cleansing under the operative microscope. Care must be taken to remove all impacted keratin debris.

Hypertrophic granulation tissue may be controlled with a law-concentration solution of trichloroacetic acid carefully applied with a fine probe. Topical ear drops containing antibiotics and, if necessary, corticosteroids are used. Systemic antibiotics, based on culture results, may be necessary occasionally.

Surgical management of cholesteatoma

The modest goal of this part of this paper is to display, on the basis of our experience, the essential principles of the surgical management of cholesteatoma in adults and children.[21] Surgical management of cholesteatoma remains controversial, with the main argument focusing on the surgical approach of the middle-ear cleft; in other words the surgical handling of the posterior bony canal wall.

The canal-wall-up procedure preserves the normal morphology of the ear canal, thus avoiding the necessity for periodic cleaning and the risk of bowl problems. The healing time is reduced. Hearing results may be better. A high cholesteatoma recurrence rate requires a post-operative control imaging or, if necessary, a second-look procedure.

The canal-wall-down procedure creates an open cavity after removal of the posterior bony canal wall. This widely exposes the epitympanum and mastoid, facilitating complete removal of the cholesteatoma sac and diminishing the recurrence rate.

The main disadvantages of this procedure are related to the necessity for periodic cleaning of the mastoid cavity. Subsequent bowl infections can occur, especially when the cavity becomes wet, causing significant lifestyle changes, particularly in the pediatric age group.

Surgical principles

In the surgical management of cholesteatoma, the important issue is not whether the technique should be an open or closed one, but the selection of a technique which allows safe but thorough eradication of the whole cholesteatoma. As far as we are concerned the main basic principles for the eradication of the cholesteatoma consists of absolute adherence to the following points.

Any ear suspected of cholesteatoma should be approached post-auricularly. The mastoid and the extent of the cholesteatomatous sac should be assessed from behind. A large retro-auricular combined approach allowing visualization of the whole middle-ear cleft is advocated in order to be able to totally remove the disease whatever its nature and its location.

The mastoid should be opened in a continuous horizontal plane, from the front of the attic to the tip of the mastoid. The whole mastoid and tympanic cavity should be opened at the same time, with particular attention for the sinus tympani and the anterior epitympanic recess.

The removal of the whole cholesteatoma will always be performed, from the mastoid and the attic towards the external auditory canal. The natural pathways along which the cholesteatoma develops in the middle-ear cleft should be treated in the reverse direction depending on the location of entrance through the tympanic membrane defect. The anterior attic penetration points or perforations essentially involve the attic, the epitympanum, the protympanum and even the whole mesotympanum. The posterior penetration points generally involve the facial recess, the antrum or the whole mastoid gas cell system. Only when the form and size of the cholesteatoma and its extensions are perfectly visualized should the removal be performed, using sponges and a bowl-shaped elevator to sweep the cholesteatoma from the mastoid or attic towards the external auditory canal (Fig. 5). This method allows a very soft and safe elevation, without any risk of breaking or rip

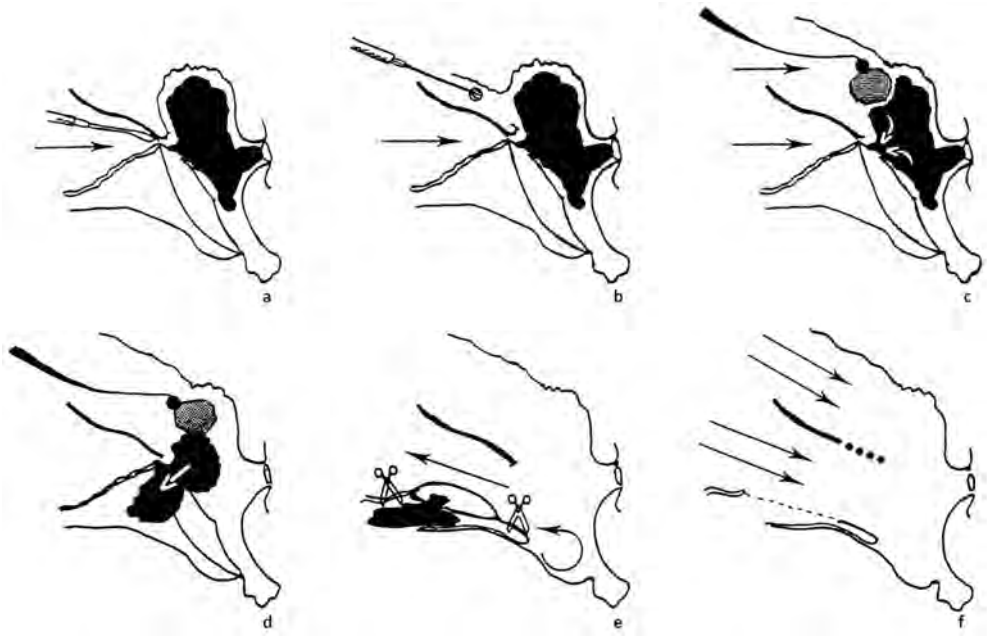

Fig. 5. Successive steps of the surgical management for cholesteatoma. (a) The dissection of the skin of the external auditory canal has to be performed as far as and including the annulus; (b) The mastoid and the extent of the cholesteatomatous sac must be assessed from behind. The whole mastoid and tympanic cavities are opened at the same time so that the cholesteatoma is approached by a double track: external auditory canal and mastoid; (c) Small balls of sponge and a bowl-shaped elevator are used to loosen the cholesteatoma and repress it in reverse direction of the natural pathways along which the cholesteatoma develops; (d) The cholesteatomatous sac is removed from the mastoid and the attic towards the external auditory canal; (e) The group consisting of the cholesteatomatous sac, the drum remnants including the fibrous annulus, the connections between these and the matrix, and the granulation tissue are elevated laterally together with the meatal skin; they are excised and removed in one piece; (f) The tympanic cavity, now completely free of cholesteatoma, is inspected and checked. Attention is turned to a perfect reconstruction of the bony annulus and/or of the posterior wall. The restoration of the mesenchymatous barrier, between the squamous epithelium of the external auditory canal, of ectodermal origin, and the mucous epithelium of the middle-ear cleft of endodermal origin, should always be performed to abolish the risk of recurrence.

ping up the matrix. Once the mass of cholesteatoma has been dissected free from around the cavities, it must be pushed away towards the external canal. When a bulky cholesteatomatous mass hinders the view and/or prevents expulsion, it must be opened and its contents partly aspirated to avoid spontaneous rupture. The cholesteatoma sac, the drum remnants including the fibrous annulus, the connections between these and the matrix, and the granulation tissue should always be elevated laterally together with the meatal skin, excised and removed in one piece.

After eradication of the cholesteatoma, the restoration of the mesenchymatous barrier, between the squamous epithelium of the external auditory canal, of ectodermal origin and the mucous epithelium of the middle-ear cleft of endodermal origin, should always be performed to abolish the risk of recurrence.

Whatever the material used, the tympanic membrane should always be restored during the first stage. When and where necessary, any defect of the bony meatal wall is reconstructed.

Any malleus or incus harboring disease is removed together with the mass of cholesteatoma. Any diseased or suspected ossicle is replaced by a healthy one.

When there is a fistula, we remove the matrix of the cholesteatoma, only at the final time of the operation; afterwards, we seal, on the fistula, a piece of connective tissue, we stick a patch of tympanic allograft with fibrin glue.

Specific cases

Here we will consider the particularities of cholesteatoma in children, ‘attic’ versus ‘tensa’ cholesteatoma and ‘only one hearing ear’.

These principles for surgical management of cholesteatoma remain the same for adults as well for children. The age of the patients is not the most important factor.

The disease most frequently is unilateral, although chronic otitis media often is present in the opposite ear. What important is that there are more likely to be complications in children which demand urgent intervention. Fistulae are more frequent. Facial palsy is also encountered more frequently. Diagnosis and assessment of the extent of childhood cholesteatoma may be difficult because of lack of patient cooperation, small ear canal and meatus, and sometimes equivocal audiometric findings. CT scanning may be useful in establishing the limits of the cholesteatoma, and for medico-legal reasons. Usually, a precise assessment is achieved only by surgical exploration. Cholesteatoma is more aggressive and more invasive in children than in adults. There appears to be an increased incidence of residual and recurrent childhood cholesteatoma after surgical removal, when compared to adult disease. The hypothesis is that the biology of pediatric cholesteatoma may be different, more aggressive. The activity of growth factors normally elaborated in childhood and absent in adult may play a role.

Other contributing factors include: poor fibro cartilaginous Eustachian tube function, poor pneumatisation of the mastoid, poor maturity of the immune system, and frequent otitis media.

In our opinion, pars flaccida and pars tensa cholesteatoma belong to the same pathological process, although their localization and appearance are different. For this reason, the surgical approach is not modified and our therapeutic philosophy remains the same. In cases of pars flaccida cholesteatoma, pneumatization of the mastoid is generally preferable. The cholesteatoma appears in the form of a well-defined cyst, and the prognosis should therefore be better.

In the cases of an only hearing ear, we perform the most conservative management . Sometimes this entails regular review and control with microscopic suction toilet only. During the post-operative stage, the non Echoplanar Diffusion Weighted Magnetic Resonance Imaging (non EPDWI MRI), performed one and five years after the primary surgical event, excludes the presence of residual cholesteatoma in a non-invasive way, avoiding unnecessary second look surgery.[iv]

Conclusion

To summarize my attitude, concerning the surgical management of cholesteatoma, I will quote the following main points:

- Systematic post-auricular approach.
- Large opening of the mastoid gas cells system and the tympanic cavity.
- Repulsion of the cholesteatoma cyst via the natural pathways along which it had developed.
- Reconstruction of the mesenchymatous barrier between the squamous epithelium of the external auditory canal and the mucous epithelium of the middle-ear cleft.

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iv. Semantics about the eventual ‘second steps’: It is not always obvious whether a second operation, second stage or second look should be performed. A second operation may be undertaken by another surgeon in case of recurrent disease. ‘Second stage’ means that the surgical procedure is not completely finished, and that the surgeon has taken the deliberate decision to interrupt the procedure, which will be completed later after a specific period of time. In this case, the surgeon considers the intermediate period to be necessary to improve the condition or stability of the ear, before undertaking reconstruction. The decision for ‘staging’ could be based on pathological conditions and functional reasons. ‘Second look’ may be recommended by the surgeon when the surgical procedure is completely finished. As a precautionary measure, he decides to operate once more to exclude possible residual or recurrent pathologies. Second-stage and second-look procedures are, by definition, carried out by the same surgeon.

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TECHNIQUE TO SIMPLIFY MYRINGOPLASTY

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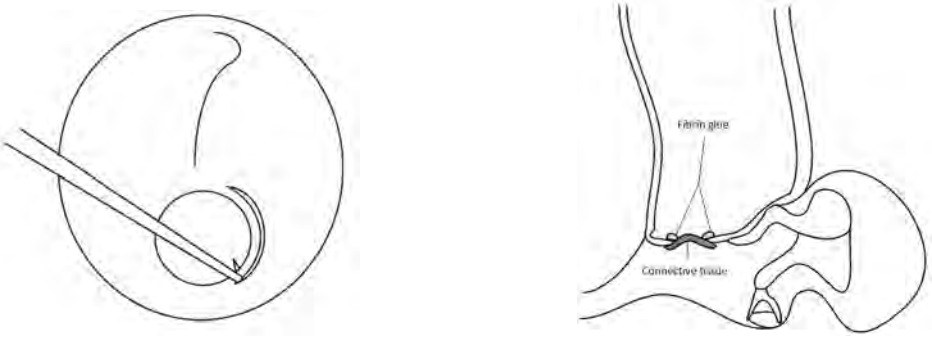
Introduction

Simple underlay myringoplasty (SUM) has been widely performed over the last 23 years in Japan as a less invasive procedure of myringoplasty than conventional methods. SUM has been gradually recognized in the world since an original article in English was published[1] and the detail of the procedure had been introduced into AAO-HNS (American Academy of Otolaryngology-Head and Neck Surgery) for the last six years.

Surgical procedures

A transcanal approach is applied. No skin incision is necessary except to harvest subcutaneous connective tissue for the graft from the retro-auricular region. After the topical anesthesia of the tympanic membrane, the perforation edge is removed for both the debridement and the vascularization to the graft (Fig. 1A). Calcified tissue around the perforation is also removed. When the perforation edge involves the malleus handle, the mucosa behind the handle is removed. The pressed graft is inserted into the tympanic cavity through the perforation, and then the graft is elevated to touch the perforation edge. The graft is fixed to the tympanic membrane with a little fibrin glue (Fig. 1B). Packing is not necessary either in the tympanic cavity or in the external auditory canal. The surgery is performed under local anesthesia except in cases with children because thirty minutes is sufficient to accomplish the surgery for one ear by this method. Although SUM is generally performed under a microscope, an endoscope is sometimes necessary when the anterior part of the perforation edge cannot be treated under the microscope due to a protrusion of the anterior region of the external auditory canal. For the persistent perforation after this method, re-closure is attempted in the outpatient clinic by the same procedure using frozen autologous tissue which has been harvested in the initial surgery.



A B

Fig. 1. Scheme of surgical procedures. A: Removal of the perforation margin. B: Grafting by underlay method.

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Edited by Haruo Takahashi

Results

The rate of initial closure was 478/621 (77.0%). Overall success rate after the re-closure was 595/621 (95.8%). There was no significant difference of the success rate among any size of the perforation or any frequency of otorrhea. There was no serious complication such as sensorineural hearing loss.

Indications

SUM is indicated for the case as follows: (1) Any size of the central dry perforation; (2) No pathology in the tympanic cavity; (3) A sufficient hearing gain after the patch test using thin wet cotton. The case of cholesteatoma or adhesive otitis media is contra-indicated. Because the post-operative hearing deterioration is extremely rare, SUM is also indicated for cases with the only hearing ear or with the same-day surgery for bilateral diseases.[2,3] Additionally, this method is applied to tympanoplasty by the transcanal approach.

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ENDOSCOPIC SURGERY AND ITS VARIOUS OPTIONS

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Objective

The merit of endoscopic-aided ear surgery is that endoscopic observation can give the otosurgeon a wide and clear view of the middle-ear cleft. Detailed observation can avoid an injury of the facial nerve or the inner ear. Moreover, endoscope can show a residual cholesteatoma under the ossicles or in the tympanic sinus where microscopic observation is limited (Figs. 1 and 2).

Therefore, an endoscope should be used more in ear surgery, not only for adults but also for children.

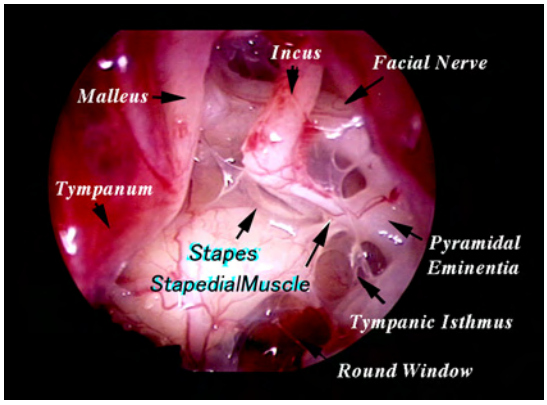

Fig. 1. Endoscopic view of the middle ear. Endoscopic observation is excellent to get a wide and detailed view of the middle-ear cleft. This is a left ear with a perilymph fistel, so the anatomy is normal. With the endoscope, we have a clear and total view of stapes and facial nerve and tympanic sinus in one frame.

Methods, options and cases

Our basic style of ear surgery is that the surgeon holds an endoscope with 2.7 mm diameter and 17 cm length in the left hand and handles surgical tools in the right hand. Both devices are introduced into the middle ear through the external ear canal. Endoaural approach is preferable for minimally invasive surgery, both in adults and children.

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Edited by Haruo Takahashi

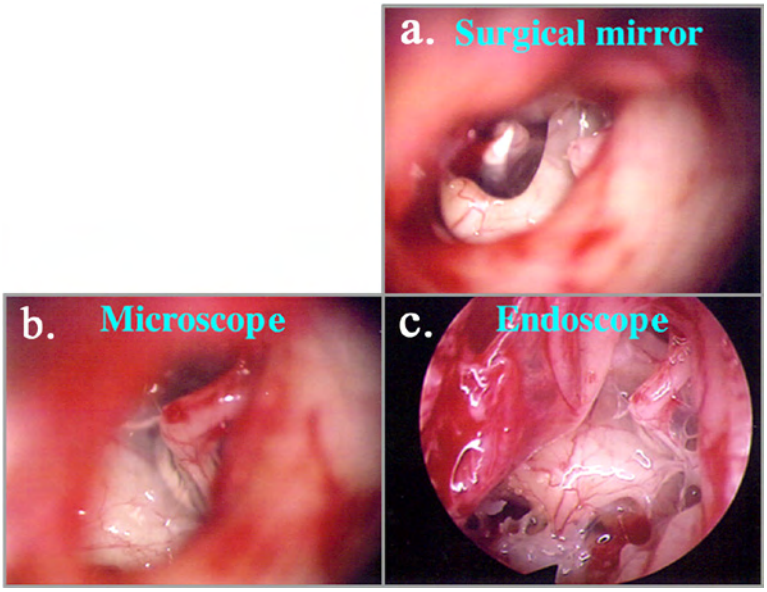

Fig. 2. Comparison of operative views of stapes, all in the same ear. a: surgical-mirror view; b: microscopic view; c: endoscopic view. The operative fields in (a) and (b) were very narrow and small compared with (c).

We have the following endoscopic options besides the basic one: 1) When both hands are necessary for the surgery, the endoscope self-holder (endoarm) is very useful (Fig. 3). The endoarm can sustain the endoscope in the external ear canal and the ear surgeon can use both hands for operating. A two-handed operation is required in cholesteatoma, glomus tumor and schwannoma, because suction and surgical tool must be used at the same time. 2) Fogging at the tip of endoscope might be very annoying. To prevent this problem, we have used the light source of the light emitted diode (LED) because this system produces no heat and consequently no fogging. 3) A high-definition fine visual system is also very useful to give a very bright and clear operative view compared with conventional monitoring (Fig. 4).

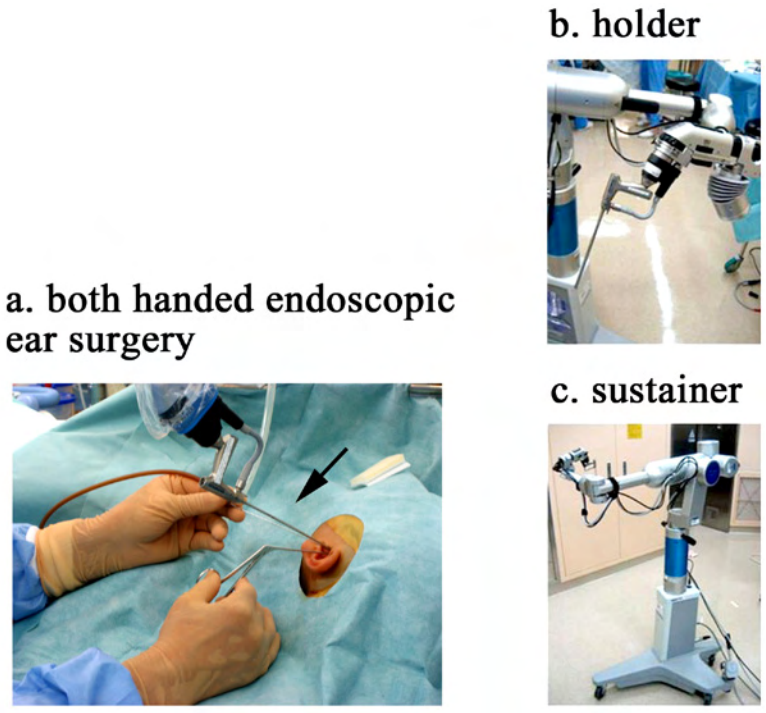

Fig. 3. The endoarm, composed of a specific endoscope holder (b) and sustainer (c). The endoscope is set at the orifice of the external ear canal.

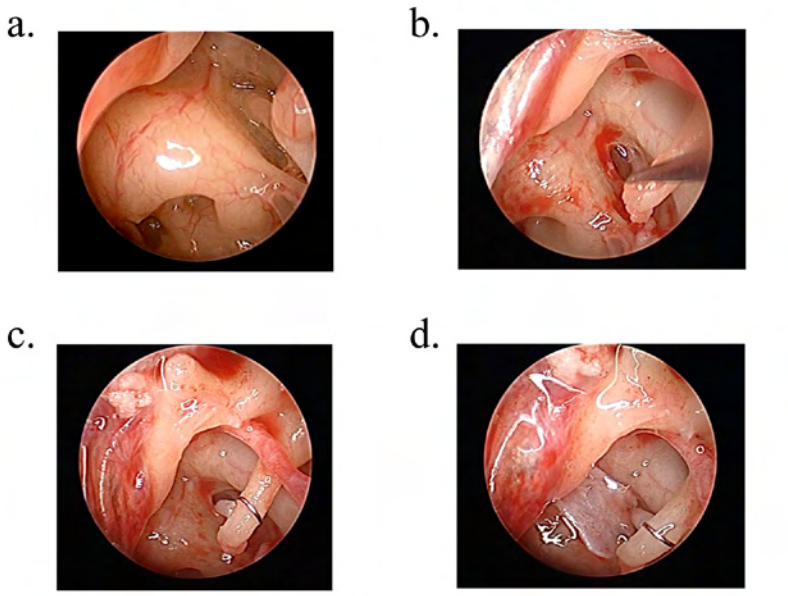

Fig.4. Endoscopic stapes surgery. Stapes surgery is good indication for the endoarm. In clear operative view (a), both hands can be used to remove stapes (b), open the footplate, attach the wire piston (c), and place a fascia (d).

Good indications of endoscopic ear surgery are stapes surgery, tympanoplasty in adults and ossicular anomaly, congenital cholesteatoma in children.

Conclusion

Otosurgery is very dangerous to perform without adequate observation of stapes or facial nerve. Observation with an endoscope is a very useful way to find these structures, and it is possible in adult patients as well as in children (Table 1).

Table 1.

Excellent operative view
• close to ossicles
• wide range and high resolution view
• identify of position of TORP or PORP
Safety operation
• observe middle ear totally
• avoid damage of inner ear and facial nerve
• explore cholesteatoma remnant
Demerits
• surgeon holds camera on the left hand and uses limited tools on the left
• narrow external ear canal
• control of bleeding and mist in the field

Our conclusion is that safe, minimally invasive ear surgery should be performed both in adults and children with an endoscope.

MRI EVALUATION OF MIDDLE-EAR PATHOLOGIES

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Roles of MRI in the evaluation of middle-ear pathologies

In the imaging of middle-ear pathologies, MRI can play an auxiliary role of CT because of its higher tissue contrast. Some diseases can be diagnosed based on their characteristic signal intensities (*e.g.*, cholesterol granuloma). High spatial resolution is needed for the temporal bone imaging. So, a high magnetic field unit such as 3T and a multi-channel head coil can enhance utility of MRI because of the high signal-to-noise ratio and resultant high spatial resolution. The magnetic susceptibility effect is one of major problems of MRI for the application in the temporal bone, especially with higher magnetic field units. Magnetic susceptibility effects result in artificial signal loss, abnormal signal, distortion of images, and so on. So, pulse sequences should be designed to minimize the susceptibility effects and maximize the spatial resolution.

Essential pulse sequences for the temporal bone

Essential pulse sequences include 2D T1- and T2-weighted imaging (T1WI and T2WI), 3D high-resolution heavily T2WI, and post-contrast T1WI. Using a 3D fast spin echo (FSE) or gradient echo (GRE) acquisition, thin slice T1-weighted images can also be obtained. 3D heavily T2WI and T1WI can be estimated in any directions with multiplanar reconstruction (MPR). Diffusion-weighted imaging (DWI) is also important especially for evaluating cholesteatomas. The following sequences were mainly discussed.

3D high-resolution heavily T2WI

Variable sequences of FSE (SPACE, Cube, etc.) and GRE (true FISP, true SSFP, CISS, FIESTA, etc.) can be applied to obtain high-resolution heavily T2WI. FSE is less sensitive to the magnetic susceptibility.[1] Tissue contrast of FSE (SPACE) and true SSFP/CISS may be different.

Post-contrast 3D T1WI

As a high-resolution post-contrast T1WI, 3D GRE sequences have been widely used. GRE, however, is vulnerable to susceptibility effects comparing with SE or FSE sequence. In our experience, the image quality and diagnostic value of 3D SPACE (one of FSE sequence) was superior to 3D FLASH (GRE sequence), especially for lesions of the labyrinth, facial nerve, and tympanic cavity, such as cholesteatoma. This superiority of SPACE was mainly because of its less vulnerability to susceptibility effects (Fig. 1). Signal of vessels is different between FSE and GRE. On the SPACE, most arteries and veins display ‘flow void’ and are associated with faint ghost artifacts along the phase-encoding direction. In contrast, vessels may display hyperintensity and are free from ghost artifacts on the FLASH.

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Edited by Haruo Takahashi

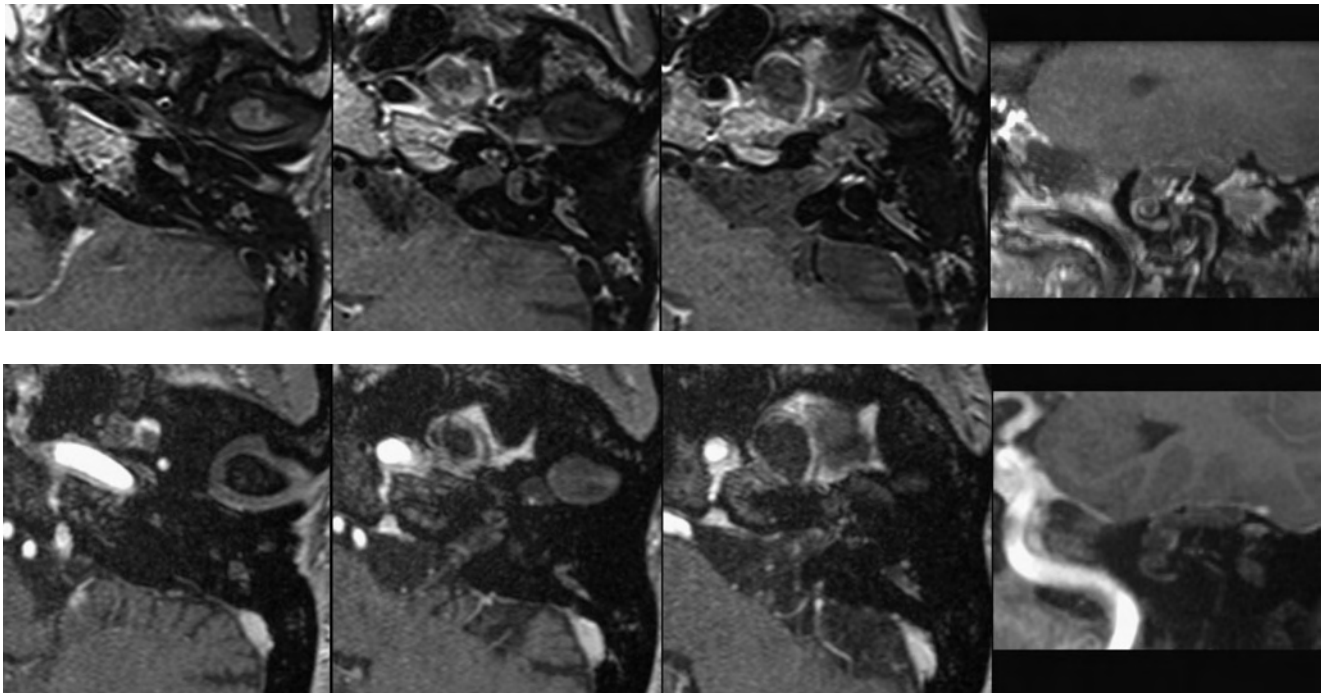

Fig. 1. A patient with cholesteatoma with labyrinthitis. a: post-contrast 3D SPACE. b: post-contrast 3D FLASH. Abnormal enhancement of the cochlea and vestibule is well visualized on the SPACE images. Signal intensity of artery and veins are different on both images.

DWI

Usefulness of DWI in the evaluation of cholesteatomas and tumorous lesions has been widely recognized. Higher SNR obtained by 3T scanner and a multi-channel coil enables thin-slice and high-resolution DWI. Application of parallel imaging and smaller voxel size can reduce susceptibility effects of EPI. EPI-DWI, however, is not free from susceptibility artifacts especially at 3T. So, non-EPI DWI such as single shot FSE and PROPELLER (periodically rotated overlapping parallel lines with enhanced reconstruction) can be more useful.[2]

Signal intensities on MRI

Because T1 and T2 of free water is very long, the signal intensity of tissues on MRI largely depend on their water contents. So, hypointensity on T1WI and hyperintensity on T2WI usually represent increased water content of tissues and are non-specific. On the contrary, hyperintensity on T1WI and/or hypointensity on T2WI may reflect some specific pathological conditions.

Hyperintensity on T1WI

- Paramagnetic substances: hemoglobin products (methemoglobin), melanin, free radicals, manganese, etc.;
- Lipid: lipoma, choristoma such as dermoid, etc.;
- High protein content;
- Calcification in some conditions (surface effect);
- Flow-related enhancement.

Hypointensity on T2WI

- Paramagnetic substance: hemoglobin products (deoxyhemoglobin, methemoglobin, hemosiderin), non-hem iron such as ferritin, melanin, free radicals, copper, etc.;
- Low proton density: gas, calcification, ossification, tissues with high cellularity and/or high nuclear-cytoplasmic ratio, fibrous tissue, etc.;

- High protein content;
- Flow-void.

Fat on MRI

T1 of fatty tissue is short and fat usually displays hyperintensity on T1WI. Fatty bone marrow also displays hyperintensity on T1WI. Bone marrow abnormalities such as osteomyelitis or tumor involvement are more easily diagnosed on MRI compared with CT (Fig. 2).

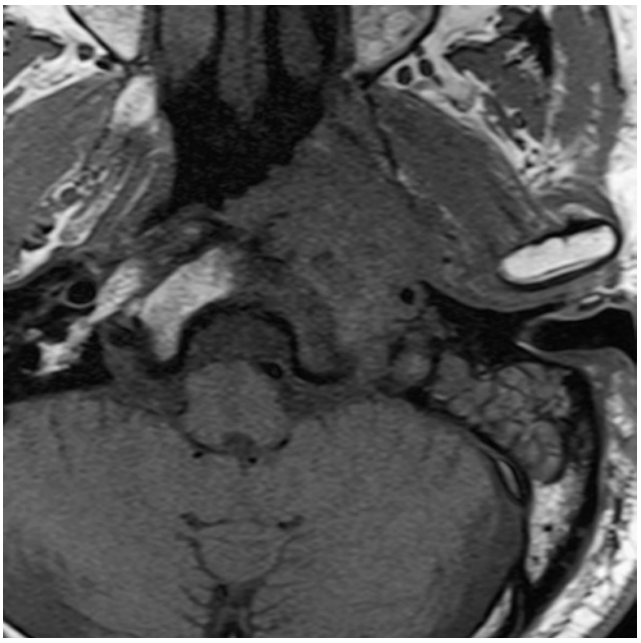

Fig. 2. A patient with malignant (necrotizing) otitis media. T1WI shows hypo-intensity of the bone marrow in the left clivus.

Vascular structures on MRI

Signal intensity of vascular structures depends on pulse sequence, slice thickness, location, flow velocity, and so one. High flow vessels usually display signal loss (flow void) especially on T2WI. Intravascular signal may mimic a tumorous lesion especially at jugular bulb ('pseudolesion').

Representative MRI findings of middle-ear pathologies

Cholesteatoma

Roles of MRI for evaluating a cholesteatoma include: 1) Precise definition of the borders of lesions and demarcation between a cholesteatoma and other pathologies such as cholesterol granuloma, granulation tissue, scar tissue, inflammatory mucosa, and so on; 2) Depiction of the relationship of the lesion to intracranial structures; 3) evaluating complications such as labyrinthitis, facial-nerve palsy and intracranial complications (meningitis, encephalitis, brain abscess, sinus thrombosis, etc.); and 4) Post-operative follow up.

Signal intensity of a cholesteatoma on T1- and T2WI is non-specific. Although a cholesteatoma usually displays hypointense on T1WI and hyperintense on T2WI, it may display hypointensity on T2WI because of low water content (desiccated material). A cholesteatoma rarely shows hyperintensity on T1WI (white epidermoid), probably because of high protein content or high lipid content (mixed triglycerides and no cholesterol). Intensity on heavily T2WI (MR hydrography) is usually lower and more inhomogeneous compared with standard T2WI.

A cholesteatoma does not typically enhance with gadolinium, except at the margin of the lesion. On the contrary, granulation tissues usually display enhancement. However, it is necessary to obtain delayed contrast-enhanced images with a delay of 30-45 minutes after contrast-material administration.

The usefulness of non-EPI (echo planar imaging) DWI for evaluating a cholesteatoma has been widely recognized.[2,3] However, false positive or false negative studies can be seen due to the influence of T2 (T2 shine-through or T2 dark-through). In our experience, apparent diffusion coefficient (ADC) value can differentiate cholesteatomas from other pathological conditions of the middle ear, such as granulation, cholesterol granuloma, inflammatory scar and so on.

In our institute, following imaging protocol has been used for evaluating a cholesteatma; 1) T1WI (SE or 3D FSE); 2) non-EPI DWI; 3) T2WI (FSE); 4) 3D heavily T2WI, 5) 3D T1WI. To obtain delayed post-contrast images, contrast media is injected before T2WI and 3D heavily T2WI (Fig. 3).

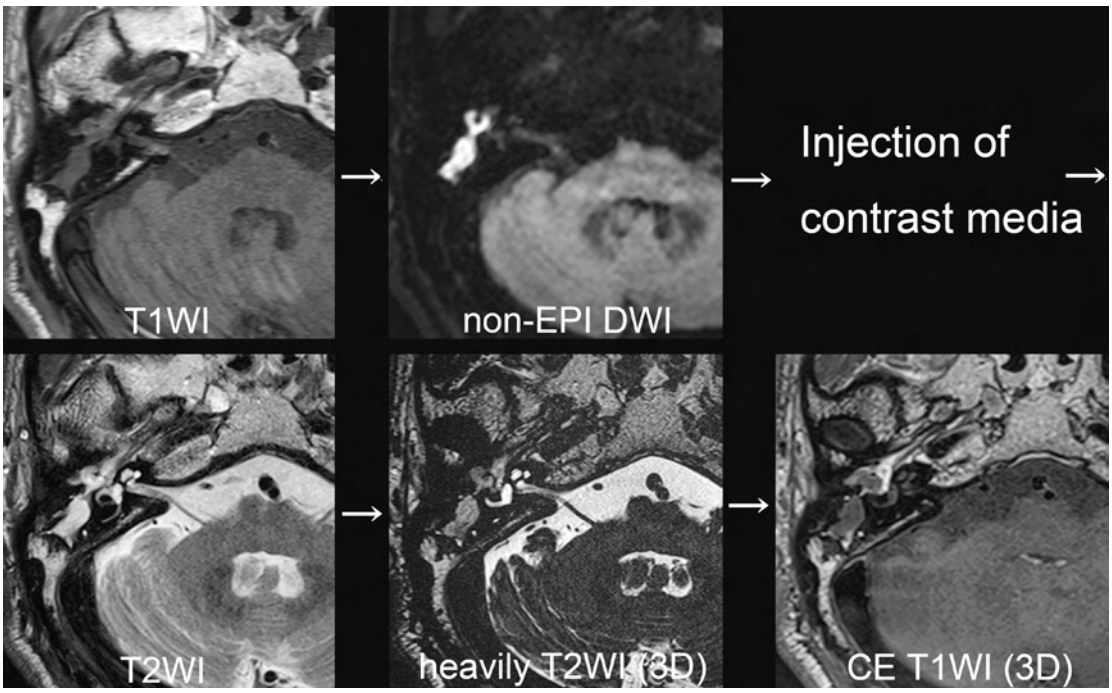

Fig. 3. Protocol for evaluating a cholesteatoma.

Cholesterol granuloma

Cholesterol granulomas arise in aerated spaces such as the mastoid air cells as a result of inadequate ventilation. Hemorrhage in the space without ability to drain leads to inflammatory response. The lesion is usually bright on T1 and T2WI representing blood products especially extracellular methemoglobin.[4] A ring of low signal intensity may confirm the presence of hemosiderin-laden macrophages. However, the signal may be variable depending on the blood products (Fig. 4).

Acute and chronic otitis media

MRI may be useful to estimate following complications.[5]

- Coalescent otomastoiditis: The pneumatic cells coalesce into larger cavities filled with purulent exudates and granulations, resulting in empyema;
- Subperiosteal, bezold, or perisinus abscess;
- Gradenigo's syndrome;
- Labyrinthitis;
- Intracranial complication: epidural abscess, meningitis, encephalitis, brain abscess, dural sinus thrombophlebitis, etc.

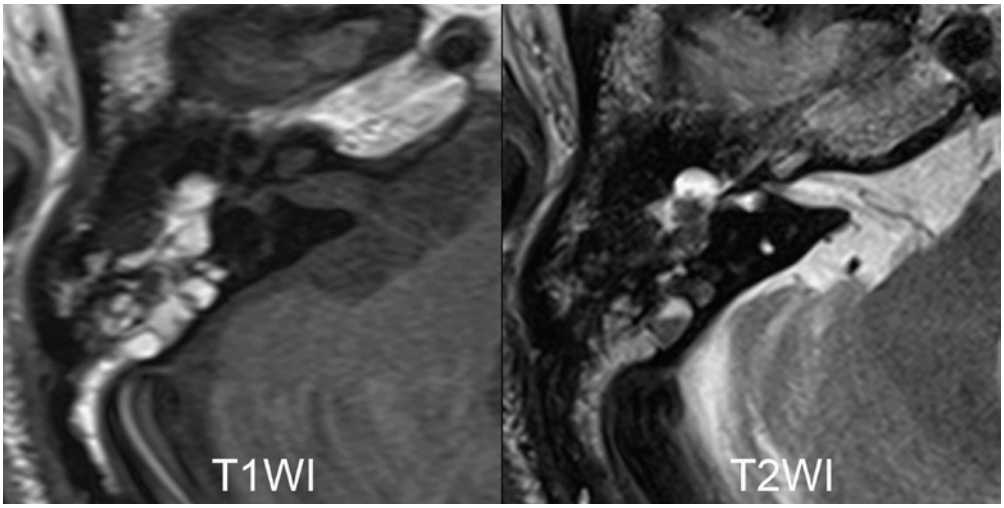

Fig. 4. Cholesterol granuloma. The lesion displays hyperintensity on T1WI. T2WI shows hyper- and hypo-intensity with fluid-fluid levels.

Labyrinthitis

Labyrinthitis can complicate a cholesteatoma or otitis media and may be associated with labyrinthine fistula. MRI plays an important role. Abnormal enhancement can be seen in the acute or subacute phase (Fig. 1). Fibrosis of the labyrinth can be detected with high-resolution heavily T2WI. In the chronic stage of the labyrinthitis, calcification or ossification occurs (labyrinthitis ossificans). CT is better for detecting the condition.

Tumorous lesions of the middle ear

Facial-nerve schwannoma (Fig. 5)

Although the lesion can be seen anywhere along the course of the facial nerve, it is most commonly originated from the geniculate ganglion.[6] Facial-nerve palsy is associated in less than 50% of the cases. Morphology depends on its location. Lesions in the internal auditory canal mimicked acoustic tumors. Lesion in the tympanic or mastoid segment displays ‘sausage-like’ configuration.

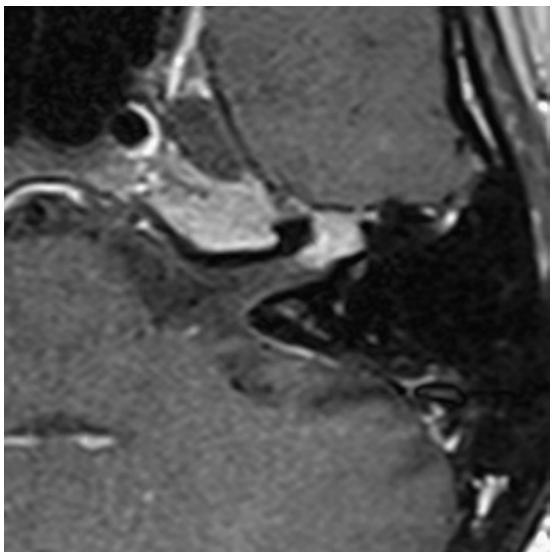

Fig. 5. Facial-nerve schwannoma. Post-contrast T1WI displays enlarged labyrinth segment of the left facial nerve with abnormal enhancement.

Glomus tympanicum tumor, paraganglioma

The lesion usually located at the cochlear promontory and is usually small (millimeters to two cm). Characteristics of a glomus tumor is its hypervascular nature.[7]

Retrotympanic 'vascular' lesions

Different diagnoses include the following pathologies;
- Congenital lesions: aberrant internal carotid artery, persistent stapedial artery, dehiscent jugular bulb;
- Inflammatory lesions: cholesterol granuloma;
- Benign tumors: Glomus tympanicum and jugulare.

Facial-nerve palsy (FNP)

Classic Bell palsy requires no imaging in initial stages. If decompressive surgery is anticipated, however, MR imaging is warranted for excluding lesions causing FNP. Atypical FNP requires search for underlying lesion.

Conclusion

MRI can play an auxiliary role of CT because of its higher tissue contrast and some diseases can be diagnosed based on their characteristic signal intensities.

Pulse sequences should be designed to minimize the susceptibility effects and maximize the spatial resolution.

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POLITZER PRIZE AND JOS PRIZE SESSION

MECHANISM OF BONE DESTRUCTION IN CHOLESTEATOMA

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Introduction

Cholesteatoma destructs bony structures of temporal bone to cause complications such as inner ear dysfunction, facial nerve palsy and so on. However, the mechanism of the bone destructive feature of cholesteatoma remains to be elucidated. It was recently reported that the RANKL (receptor activator of NF-κB ligand)-Th17 system contributed to bone destruction in rheumatoid arthritis.[1] The RANKL-Th17 system was focused on as a possible mechanism of bone destruction in cholesteatoma.[2,3]

Material and method

Subjects were 33 patients with cholesteatoma and three patients with suppurative chronic otitis media (OMC) who were operated in Fukuoka University Hospital. Cholesteatoma was taken with surrounded granulation during surgery. For PCR, materials were snap-frozen using liquid nitrogen and preserved with the freezer of -80°C. Total RNA was obtained using TRIzol reagent (Invitrogen®) and quantitated. First-strand cDNA synthesis was done using SuperScript II (Invitrogen®). Real-time PCR of cDNA products was performed. Values were normalized by relative quantification. Values obtained were expressed as fold changes of gene expression. For flowcytometry, fresh materials were prepared as single cell and stained by antibodies. FACSCanto™II flowcytometer and FACSDiva™ software were employed.

Results

Immunity cytokine expression in cholesteatoma

Active inflammation was assessed by a clinical scoring system focused on three points: state of granulation (0: scar, 1; mild; 2: moderate; 3: marked); pre-operative ear discharge (0: none; 1: wet; 2: seromucous; 3: purulent); and cellular infiltration by pathohistological findings (0: none/few; 1: mild; 2: moderate; 3: severe). Active signs of granulation were evaluated by extent of redness, swelling and easy bleeding.

The value of tumor necrosis factor alpha (TNFα) was measured by PCR in order to compare the clinical score. The relative ratio of TNFα/GADPH was a hundred times larger than when the clinical score was equal or higher than three. Thereafter, we classified the inflammation phase by TNFα value of 10^2. Since macrophage is reported to secrete TNFα, macrophage might be activated in a severe inflammation phase of cholesteatoma.

Acquired immunity in cholesteatoma was investigated by flowcytometry. Cells were gated by T-cell receptor beta (TCR-β). Two populations with cell surface markers CD4 and CD8 were noted (data not shown). CD69 is a T cell early activate marker. It was expressed only in CD4-positive cells. It indicated that helper T cells (Th cell) were activated as acquired immunity in cholesteatoma.

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Cholesteatoma and Ear Surgery – An Update, pp. 203-205
Edited by Haruo Takahashi

Cytokine expression in cholesteatoma was investigated by real-time PCR. Interferon gamma (IFNγ), IL-23 and IL-17 were expressed more in the TNFα high group than in those in the low group. IFNγ enhances activation of macrophage which secretes IL-23. IL-23 induces differentiation of Th17 which releases IL-17.

Bone destruction-related substances in cholesteatoma

Bone destruction related substances were investigated in severe inflammatory cholesteatoma by real-time PCR. RANKL and one of the proteins of type I matrix metalloproteinase (MMP-1)[2] were found significantly more in the TNFα-high group. This indicated that bone was being destructed in the severe inflammatory phase of cholesteatoma. By flowcytometry, RANKL was found only in CD4-positive cells. This may suggest that Th17 cells contain RANKL.

Specificity of the RANKL-Th17 system in cholesteatoma

Relative mRNA expression of various substances was compared between cholesteatoma of the TNFα-high group and granulation of suppurative chronic otitis media. The value of RANKL was higher in cholesteatoma than that of OMC. It is indicated that RANKL-Th17 system is induced more specifically in cholesteatoma than in other otitis media.

Discussion

Figure 1 shows a hypothesis of bone destruction in cholesteatoma from the present results. Once inflammation occurred, macrophage was activated as an innate immune system in the severe inflammatory phase of cholesteatoma and secretion of IL-23 and IFNγ. IL-23 from activated-macrophage induces differentiation of Th0 to Th17. Activated Th17 cell expresses bone destructive factor, RANKL, on itself, and releases IL-17. Both RANKL and IL 17 involve bone destruction in a certain way.[4] Resulting from bone destruction, MMP-1 can be detected.[5] Since the value of RANKL was higher in cholesteatoma than that of OMC, the RANKL-Th17 system is more activated in cholesteatoma. Further studies are necessary to investigate why cholesteatoma predominantly induces the RANKL-Th17 system compared to other otitis media. There is a possibility that an inhibitory agent against the RANKL-Th17 system suppresses the bone destruction feature of cholesteatoma.

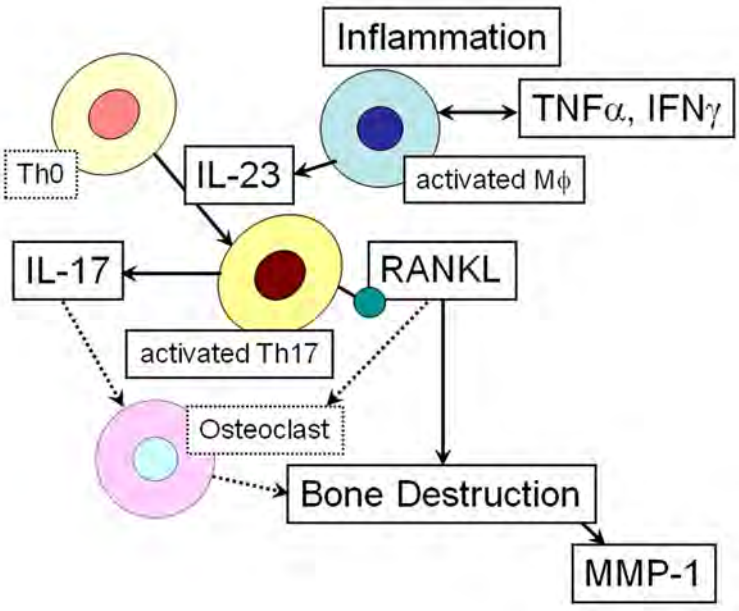

Fig. 1. Hypothesis of bone destruction in cholesteatoma. Mφ: macrophage.

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SRF AND KI-67 EXPRESSION IN ACQUIRED CHOLESTEATOMA, EXTERNAL AUDITORY CANAL SKIN AND THORACIC SKIN

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Introduction

Cholesteatoma is defined by a keratinizing squamous epithelium in the middle ear cavities. Although cholesteatoma is a benign disease, it can invade neighboring tissues and often recur even if surgical resection is considered to be complete. Cholesteatoma is traditionally classified as either acquired, essentially due to a chronic otitis process, or congenital. The origin of acquired cholesteatoma (AC) remains under discussion. Among the various advanced theories, the most probable one considers the migration of epithelial cells from the external auditory canal as the origin of the pathology. This migration can either start from the margins of a tympanic perforation, or from the retraction of the tympanic membrane.[1,2] This hypothesis is based on two observations: Firstly, the epithelium of the external auditory canal skin (EACS) presents a histological structure similar to that of cholesteatoma. In addition, the keratinocytes of the EACS exhibit particular kinetic properties that allow them, in the absence of underlying pathological conditions, to move laterally towards the pinna (lateral keratinization).[3] The particularly aggressive behavior of AC can be explained – at least partially – by disorders in growth regulation of keratinocytes. They exhibit in particular a high rate of proliferation.[2]

Serum response factor (SRF) is a transcription factor involved in the regulation of numerous genes mostly implicated in proliferation and migration of several cell types, in the control of cell cycle, in apoptosis and in cytoskeleton regulation. It has recently been involved in the pathogenesis of hyperproliferative skin disease.[4] AC is precisely characterized by hyperproliferative keratinocytes.

The aim of this study was to investigate the potential role of SRF in the pathogenesis of AC. For this, its expression was first studied in AC, EACS without middle ear associated disease and in thoracic skin. The interest of comparing these two types of epidermis is to study an epidermis with a classical keratinization (thoracic skin) in comparison with an epidermis with a lateral keratinization (thoracic skin). Also, we studied SRF expression in EACS of patients suffering from AC and in EACS of patients suffering from non-cholesteatomatous chronic otitis media (NC-COM).

Materials and methods

Histopathologic and clinical data

This study was approved by the ethical committee of the Erasmus University Hospital (ref P2010/068). For the first part of this study, 30 AC (16 male, 14 female, average age 47.2 years) were obtained in adult patients immediately after ear surgery. All AC were subjected to the standard diagnosis routine in our department of Pathology. Specimens of normal EACS and thoracic skin (n = 9) obtained during autopsy served as controls. All autopsies were performed within a 24 hours postmortem delay to ensure a good preservation and staining of tissue antigens. For the second part of this study, samples of EACS were collected during middle-ear

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Cholesteatoma and Ear Surgery – An Update, pp. 207-210
Edited by Haruo Takahashi

surgery. Ten specimens of EACS of patients with AC and ten specimens of EACS of patients with NC-COM were obtained.

All specimens were immediately fixed in 4% formaldehyde and embedded in paraffin. Sections were cut at a thickness of five µm and processed for hematoxylin-eosin staining using routine protocols.

Immunohistochemistry

Immunohistochemistry was performed on five-µm-thick sections mounted on silane-coated glass slides. First, the dewaxed tissue sections were subjected to microwave pretreatment (2 x 5 min, 850 W) in a citrate buffer (pH = 6). The sections were then incubated with a solution of hydrogen peroxide to block the endogenous peroxidase activity, and rinsed in a phosphate-buffered saline. Samples were then exposed at room temperature to solutions of the specific primary antibody against SRF (G-20, polyclonal, Santacruz) and Ki-67 (MIB-1, monoclonal, Dako), and then, after rinsing, to the corresponding biotinylated secondary antibody. After another rinsing, the samples were incubated with the avidin-biotin-peroxidase complex (Avidin/Biotin blocking kit, Vector, CA). Finally, incubation with the chromogenic substrates containing diaminobenzine and hydrogen peroxide revealed the antibodies. A counterstaining with hematoxylin completed the preparation.

Evaluation

For both antibodies, we calculated the ratio between the number of labeled nuclei and the total number of nuclei within the basal (first cellular layer) and suprabasal layer of the epithelium. The entire slide was analyzed using binocular microscopy at a 400 x magnification. All the statistical analyses were carried out using Statistica (Statsoft, Tulsa, OK).

Results

Evaluation of SRF and Ki-67 in AC, EACS without middle ear associated pathology and thoracic skin (Fig. 1)

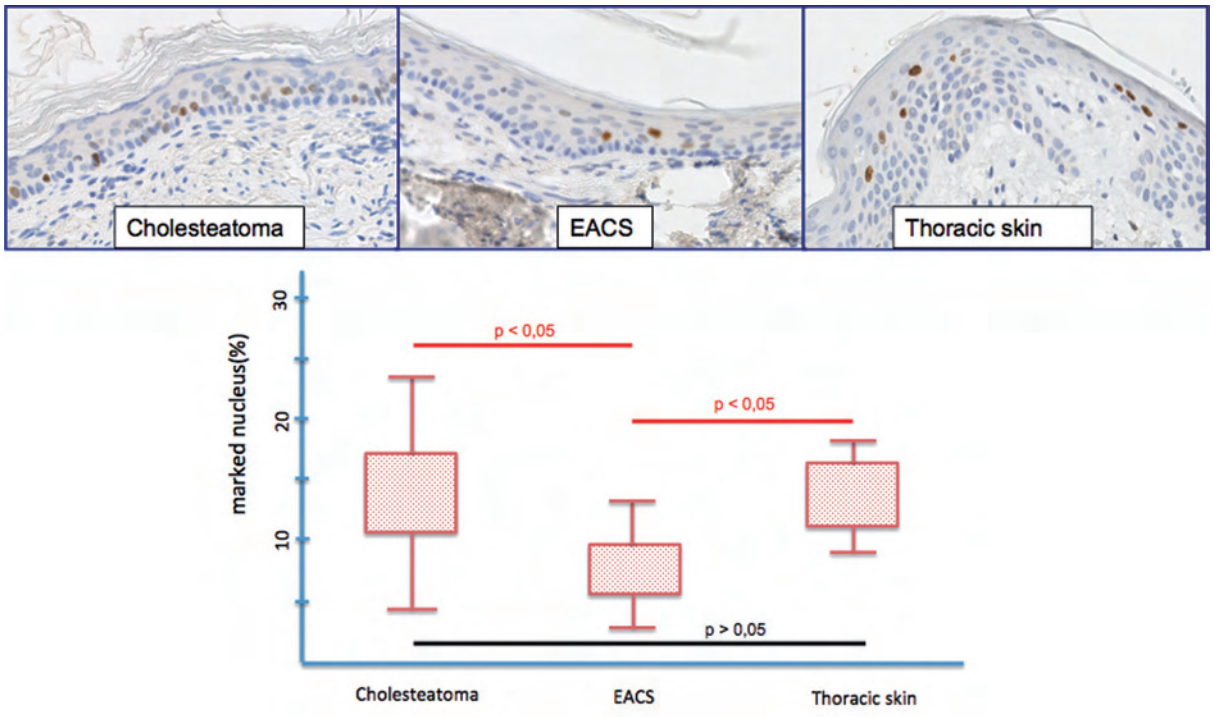

Fig. 1. Ki-67 epithelial expression (% of marked nuclei) in AC, EACS and Thoracic skin.

Ki-67 exhibits as expected a higher rate of labeled nuclei in AC compared to EACS ($p < 0,01$, Mann-Whitney U test). However, this difference does not appear if we compare AC and thoracic skin ($p > 0,05$).

SRF exhibits a nuclear and cytoplasmic staining in all epithelia. SRF is less expressed in EACS in comparison to AC and thoracic skin ($p < 0.01$). Moreover, the expression of SRF between the epithelial layers exhibits some particular feature, with a preferentially suprabasal staining in AC ($p < 0.001$, Wilcoxon signed-rank test) and a larger basal expression in thoracic skin ($p < 0.01$).

Evaluation of SRF in EACS of patients with AC and in EACS of patients with NC-COM (Fig. 2)

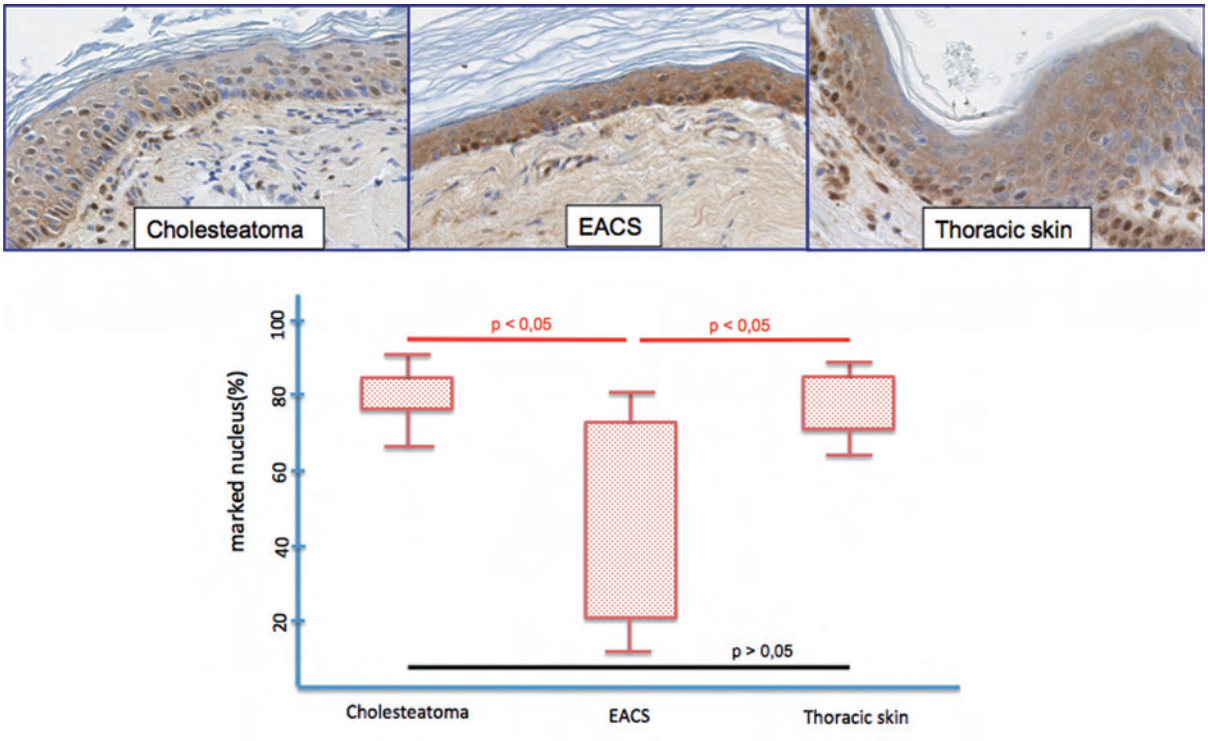

Fig. 2. SRF epithelial expression (% of marked nuclei) in AC, EACS and Thoracic skin.

EACS of patients with AC exhibits a preferentially suprabasal staining compared to EACS of patients with NC-COM ($p < 0.01$, Mann-Whitney U test). Unlike the suprabasal layer, there is no difference in the SRF expression pattern of the basal layer.

Conclusion

Besides the interest of studying the potential role of SRF in the pathogenesis of cholesteatoma, this study has the benefit of being the first comparative study of the EACS in two different middle ear pathological conditions (in AC and in NC-COM). The data reported in this study suggest that the changes of keratinocytes proliferative activity in AC could be linked to the shift of the SRF expression from the basal to the suprabasal layer. Because SRF is an important regulator of cell migration and actin cytoskeleton,[5] an assumption would be that potential change of expression observed within epidermal layers can alter the normal movements of keratinocytes. Moreover, the fact that this expression pattern is found in EACS of patients with AC but not in EACS of patients with NC-COM constitutes an argument in favor of the participation of the EACS in the development of AC.

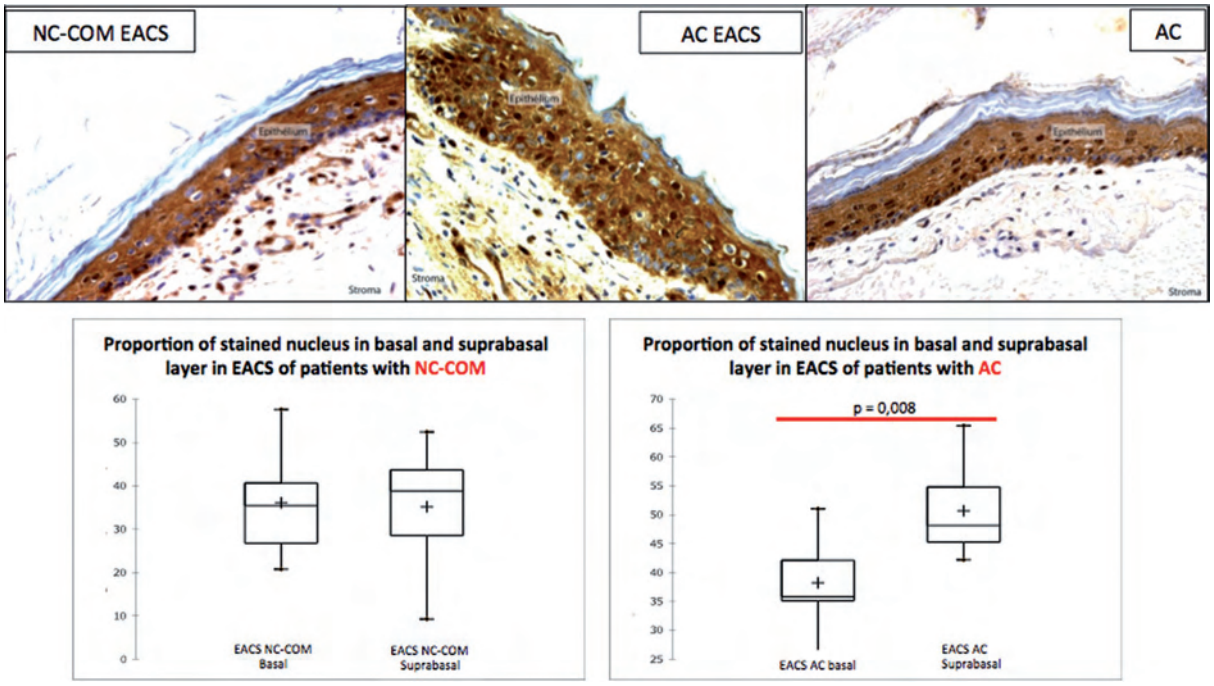

Fig. 3. SRF epithelial expression in EACS of patients with AC an NC-COM.

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THE ROLE OF JAK-STAT SIGNALING PATHWAY IN CHOLESTEATOMA: AN IMMUNOHISTOCHEMICAL ASSAY

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Introduction

Histologically**,** the epithelium of cholesteatoma is similar to the epidermis of the skin, however, uncontrolled keratinocyte hyperproliferation is one of the major differences.[1] In literature, the relationship between cytokines and uncontrolled keratinocyte hyperproliferation in cholesteatoma has been demonstrated in several studies.[2-7] The biological functions of cytokines such as cell activation, proliferation, differentiation, and survival or apoptosis are triggered by signal transduction cascades. One of the major cellular signalling pathways for cytokines is Janus tyrosine kinase (Jak)/signal transducers and activators of transcription terminators (Stat) signaling pathway. In addition, any perturbation in Jak/Stat signaling pathway may predispose dysregulation of cellular proliferation, differentiation, and apoptosis, which processes also have a significant part in the pathogenesis of cholesteatoma.

Patients and methods

Skin and cholesteatoma specimens were obtained from ten patients who underwent otological surgeries for chronic otitis media with cholesteatoma (n = 10). Skin specimens were obtained from external acoustic meatus after surgical incision. Both skin and cholesteatoma specimens were collected for histopathological and immunohistochemical examination.

The intensity of immunostaining for each parameter (Jak1, Jak2, Jak3, Stat1, Stat2, Stat3, Stat4, and Stat5) was graded as: (i) no staining: 0; (ii) weakly stained: 1; (iii) moderately stained: 2; and (iv) strongly stained: 3. In addition, H-score measurement [$\Sigma n\,(i+1)$; n: percentage of immunostained cells, i: intensity of immunostained cells] was performed for cholesteatoma and skin specimens individually.

Results

In cholesteatoma, Jak1, Jak2, Jak3, Stat1 and Stat3 immunoreactivities were absent; on the other hand, Jak1, Jak2, Jak3, Stat1 and Stat3 immunoreactivities were detected in skin specimens. In addition, the comparison of cholesteatoma and skin demonstrated a statistically significant difference for Jak1, Jak2, Jak3, Stat1 and Stat3. Stat2, Stat4 and Stat5 immunoreactivities were detected in both cholesteatoma (mean values of 172.8,

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Edited by Haruo Takahashi

166.7 and 120.0) and skin (mean values of 400.0, 284.9 and 292.0) specimens. However, the differences between the H-scores of cholesteatoma and skin were statistically significant for Stat1, Stat4 and Stat5.

Discussion

In this study, absence of Jak1, Jak2 and Jak3 immunoreactivities in cholesteatoma was determined; in contrast, Jak1 (H-score: 149), Jak2 (H-score: 289) and Jak3 (H-score:32) immunoreactivities were detected in skin. It was demonstrated that Jak3-deficient mice had a high apoptotic index and Jak3 provides survival signals, in the light of this knowledge we believe that absence of Jak kinases in cholesteatoma may augment the apoptotic processes in keratinocytes which also have a behavior of uncontrolled hyperproliferation. A statistically significant difference between Stat1 and Stat5 immunoreactivity of cholesteatoma and skin was determined ($p = 0.0005$, $p < 0.0001$). Stat1 is a negative regulator of cell growth and have an proapoptotic effect[8] Stat5 is an important regulator protein for hyperproliferation and differentiation.[9] Ultimately, we believe that the absence or deficiency in the members of Jak/Stat signaling pathway may trigger the uncontrolled hyperproliferation, migration of keratinocytes and apoptotic processes in cholesteatoma.

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SPONSORED SEMINARS

CONE-BEAM CT IMAGES OF A SCUTUM DEFECT AS A TOOL FOR PRE-OPERATIVE ASSESSMENTS

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Introduction

It is well known that Prussak's orifice is the most frequent site of attic retraction pocket formation. The bony wall around Prussak's orifice, the tympanic scute, progressively becomes eroded, resulting in a scutum defect.

Given the clinical importance of Prussak's orifice and scutum bone defects, we developed a way to measure their sizes using a cone-beam computed tomography (CBCT). The previous report demonstrated that the width and height of the scutum defect were significantly greater in ears with an attic cholesteatoma than in normal ears or ears with chronic otitis media, regardless the stage of the cholesteatoma.[1] The width of Prussak's orifice was not closely related to the cholesteatoma stage. However, the height of Prussak's orifice was significantly greater in stage-II cholesteatomas compared with stage I.

In our study, we compared the scutum defect among pre-operative eardrum findings, the volume-rendered images which are three-dimensional (3D) images and the operative findings in the same cases, and described how the image provides useful information.

Materials and methods

Subjects and operations

The study included two ears of two patients with extended middle-ear cholesteatoma who underwent staged canal-up tympanoplasty in the following manner.

In the first stage of the operation, the cholesteatoma was removed with a combined transmastoid and transcanal approach. After inserting a Silastic sheet as support, we reconstructed the tympanic scute with bone paté, and the eardrum with fascia.[2] One year later, the second stage of the operation was carried out. We detached the skin of the external ear canal in order to view the reconstructed tympanic scute, and enforced the scute with the sliced cartilage.

Imagings

A cone-beam CT device (3D Accuitomo 170, manufactured by J. Morita Manufacturing Corp., Kyoto, Japan, Resolution: 0.08 mm) was used. An image of the temporal bone was made with the volume-rendering technique.[3] Next, the image of the lower area on the external ear canal was removed using a cutting tool. In this way, a volume-rendered image of the scutum defect was obtained. Thereafter, the volume-rendered image was rotated to an optimal view.

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Results

At the assessment before the first-stage operation, the scutum defect was sometimes hidden by the skin of the external ear canal (Fig. 1). However, a large bone defect was identified on the volume-rendered image. The size and shape determined from the image were correct, as confirmed in the operative finding. At the assessment before the second-stage operation, the eardrum had clearly healed, and the volume-rendered image showed that the defect was completely reconstructed. The defect had formed completely in the operative finding in one case. In the other case, the volume-rendered image displayed that a bone defect remained although the eardrum had clearly healed. The results suggested that the bone paté had been absorbed, but the scutum plasty of the first-stage operation had not succeeded.

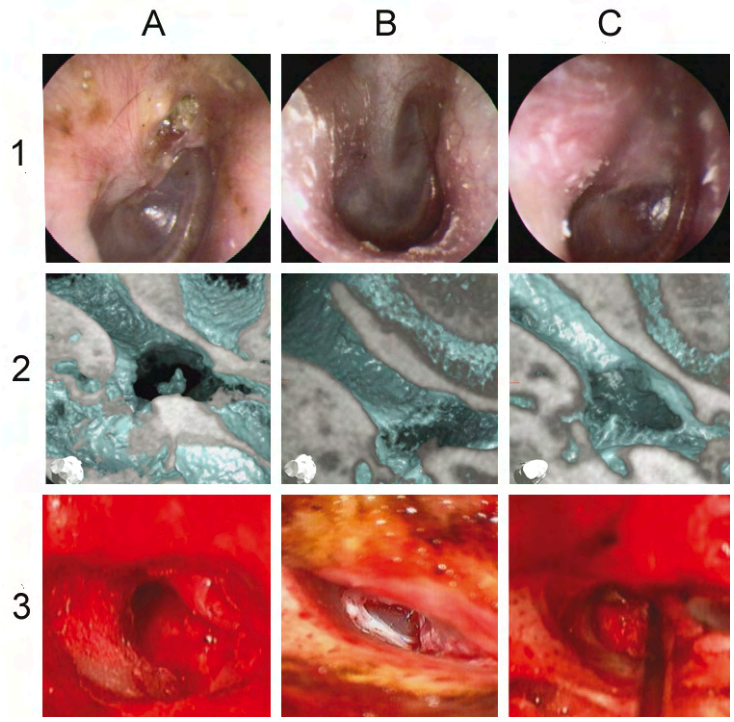

Fig. 1. Scutum defect in staged canal-up tympanoplasty. A large bone defect was identified on the volume-rendered image, although the defect was hidden by the skin of the external ear canal (A1,2). Even though the eardrum had clearly healed (B1,C1), a bone defect was reconstructed completely or remained (B2,C2). The size and shape determined from the images were correct, as confirmed in the operative findings (2, 3). A: a case at the first-stage operation; B: a case with complete scutum plasty at the second-stage operation; C: a case with incomplete scutum plasty at the second-stage operation; 1: pre-operative eardrum findings; 2: volume rendered images; 3: operative findings.

Discussion

CT is a crucial tool for both pre-operative and post-operative assessments. The volume-rendered image is a 3D-CT image called the 3D view or virtual endoscopy in previous reports.[4,5] The volume-rendering technique was introduced in 1998.[3] In otology, Klingebiel *et al*.[4] described the use of 3D images created by multi-slice CT in 2001. After that, Martin *et al*.[5] reported that this technique was more valuable than two-dimensional (2D) CT for the diagnosis of ossicle and ossicular prosthesis dislocation.

CBCT technology gives a high-resolution view of the temporal bone from any desired direction with very little radiation hazard and no metal artifact.[6-9] Previous studies were performed with 2D-CBCT images of the position of the ossicular prostheses or cochlear implantation.

The study demonstrated that the volume-rendered images correctly predicted operative findings of the scutum defect. This technique was particularly useful for a case in which the bone defect was hidden behind the skin of the external ear canal, and it provides valuable information for planning operations.

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FREE PAPERS

PRE-OPERATIVE MIDDLE-EAR AERATION PREDICTING LONG-TERM AERATION IN STAGED CANAL-UP TYMPANOPLASTY

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Introduction

Post-operative middle-ear aeration must be important for the prevention of recurring cholesteatomas and good surgical outcomes. Based on this assumption, we have been developing the staged canal-up tympanoplasty (SCUT) for cholesteatoma.[1] Since 1995, we have performed scutumplasty using bone paté and silastic sheets in the first-stage operation.[2] The incidence of retraction pocket development in the second operation was reduced significantly from 20% to 6%. Since 1997, we have performed mastoid cortex plasty using bone paté to prevent soft tissue ingrowth into the mastoid cavity.[3,4] This facilitated aeration of the mastoid cavity significantly. Our recent report demonstrated that a total mastoid obliteration technique for SCUT effectively controls cholesteatomas arising in poorly ventilated middle ears because of restoring good tympanic aeration.[5]

Here, we investigated the change in the middle-ear aeration to determine whether the aeration improved with these procedures, continued for the long term.

Materials and methods

Subjects

The study included 37 ears of 37 patients with extended middle-ear cholesteatoma who underwent SCUT. They were followed up more than three years.

Operations

In the first stage of the operation, the cholesteatoma was removed with a combined transmastoid and transcanal approach.[2] After inserting a silastic sheet as support, we reconstructed the tympanic scute with bone paté, and the eardrum with fascia. One year later, at the second stage of the operation, ossicloplasty was carried out after removal of residual cholesteatoma. Then the tympanic scute was enforced with the sliced cartilage, and the obliteration or cortex plasty was performed based on the assessment of middle-ear aeration.[2,5]

Grades of middle-ear aerations

Computed tomography (CT) was carried out before the first- and second-stage operation, and every one year after the operation. In the study, aeration of the tympanum was assessed from pre-operative and most recent CTs and was graded. Grade 0: no aeration in the tympanic cavity; Grade 1: only the mesotympanum was

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Edited by Haruo Takahashi

aerated; Grade 2: the entire tympanic cavity including attic was aerated; Grade 3: the tympanic and mastoid cavities were aerated.[4]

Results

Figure 1 shows the change in middle-ear aeration. Twenty-seven of the 37 ears (73%) improved the aeration with the staged operation. All ears with a grade-3 middle-ear aeration at the second-stage operation maintained aeration for a long time. Twelve of 16 ears (75%) with grade 2 also kept aeration. Four of 16 ears grade 2, three of nine ears grade 1, and one ear grade 0 was improved from the second-stage operation to the latest follow up. On the other hand, the ears with grade-2 and grade-3 middle-ear aeration at the first stage maintained the same grade for a long time. Deep retraction pocket was found in two ears with grade 1 or 0 aeration.

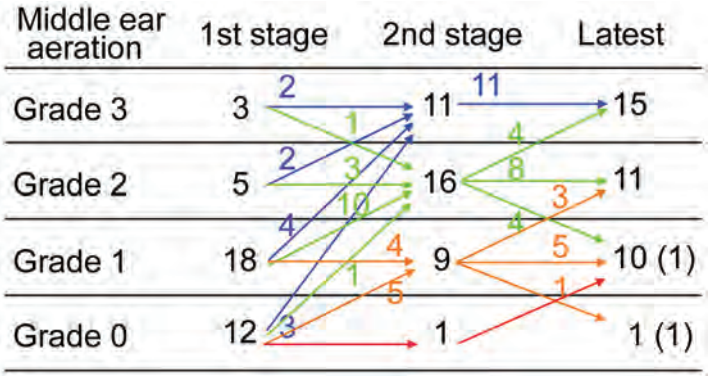

Fig.1. Change of middle ear aeration. Black numbers: numbers of ears of each grade and at each time point. Colored numbers: numbers of changing aeration indicated by arrows. ( ): numbers of recurrent case.

Discussion

Middle-ear aeration is well known to play an important role in prevention of recurrent cholesteatomas and sound conduction. To secure both tympanic and mastoid aerations or at least tympanic aeration, we modified the surgical procedure in the SCUT such as the scutum plasty, cortex plasty and obliteration. Accordingly, these techniques facilitated aeration significantly.[4,5]

The study revealed the middle-ear aeration improvement maintained for a long period of time, suggesting that the techniques were valuable for long-term outcomes of aeration. On the contrary, because well-aerated middle ears (grade 3) at the assessment before the first stage operation maintained aeration for a long period of time, we also noted that we can perform a one-stage operation if we eliminate the residual cholesteatoma and mucosal inflammatory disease.

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OPEN TECHNIQUE – INDICATIONS AND RESULTS

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Introduction

The surgical treatment goals for cholesteatoma are complete disease eradication, achievement of a dry and self-cleaning ear, creation of anatomic conditions that prevent recurrence, preservation of hearing, and avoidance of routine revision surgery.[1-3] There are two basic surgical approaches to treating cholesteatoma: open technique and closed technique. When marked attic erosion is present in cholesteatoma, especially in adults, we perform the open technique to avoid cholesteatoma recurrence. Other major indications of open technique in our institution are: cholesteatoma in cases of only hearing ear, advanced age, poor general condition, extensive canal-wall erosion, recurrence after the canal-wall-up tympanoplasty, profound sensory neural hearing loss, contracted mastoid, large labyrinthine fistula, and far anterior sigmoid sinus or low dura.[4] Extensive pneumatization of the temporal bone is the contra-indication. To avoid post-operative complications in the open technique, it is very important to create an ideal cavity in the first surgery by following correct procedures such as sufficient saucerization of the cavity and appropriate meatoplasty.

Methods

Seven hundred and ninety-nine patients were operated by open technique at the Gruppo Otologico from 1994 to 2005. Pre-operative symptoms, surgical complications, and hearing results were evaluated.

Results

Major pre-operative symptoms were hearing loss (67.2%), otorrhea (60.5%), tinnitus (18.7%), and vertigo (14.0%). There were no post-operative symptoms in 82.9%. Recurrent otorrhea (10.0%), perforation (3.0%), atelectasis (1.7%), recurrent cholesteatoma (1.3%), and stenosis of external auditory canal (1.1%) were detected and other complications were seen in less than 1%. Complications of otorrhea, infection, and vertigo were temporary. On almost half of all patients staged operation was performed, and 3.2% of the patients had residual cholesteatoma at second surgery. Out of 799, 528 patients were followed up more than one year at our institution, and underwent post-operative audiometry. The mean follow up was 60.5 months (range, 12 to 182 months). The pre-operative ABG was within 20 dB in 15.5% cases, but improved to 45.2% post-operatively (Fig. 1).

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Edited by Haruo Takahashi

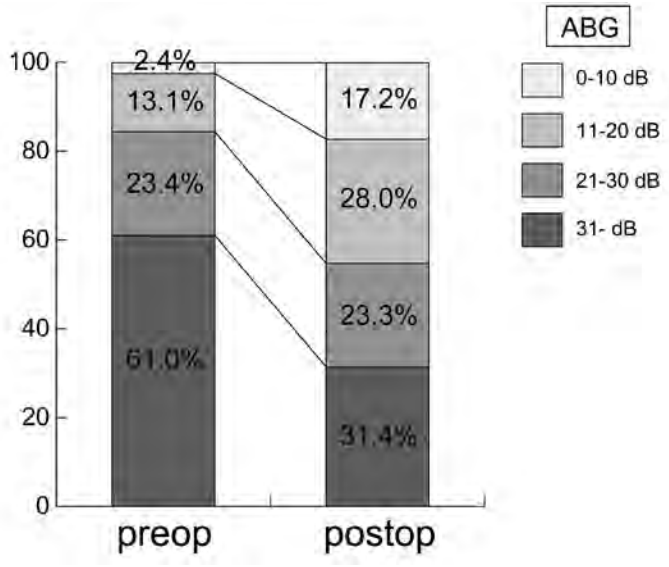

Fig. 1. Hearing result. Pre-operative ABG was within 20 dB in 15.5% cases, but improved to 45.2% post-operatively.

Table 1. Postoperative complications in 539 patients.

NONE: 447 (82.9%)			
otorrehea	54 (10.0%)	vertigo	5 (0.9%)
perforation	16 (3.0%)	SNHL	5 (0.9%)
atelectasis	9 (1.7%)	ret. pocket	4 (0.7%)
recurrence	7 (3.2%)	granulation	3 (0.6%)
EAC stenosis	6 (1.1%)	tinnitus	3 (0.6%)
infection	5 (0.9%)	facial palsy	1 (0.2%)

Conclusion

Open technique is an appropriate treatment for acquired cholesteatoma. When performed well, the rate of recurrent cholesteatoma is very low. But poorly performed open technique leads to failures and complications.

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FUNCTIONAL SURGERY FOR PEDIATRIC CHOLESTEATOMA: A 25-YEAR LONGITUDINAL STUDY

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Introduction

There has been a trend of less radical surgery with functional preservation that was being applied in many fields. For example, functional endoscopic sinus surgery has been applied in treating chronic rhinosinusitis, and transoral laser microsurgery was appliedfor laryngeal and hypopharyngeal cancers so as to avoid total laryngectomy.

Since 1986, Dr. Lien has performed a functional surgery for pediatric cholesteatoma: tailor-made tympano-mastoidectomy with cartilage reconstruction (TTCR). This technique emphasizes the importance of preserving uninvolved, functional mastoid air cells, and anatomical reconstruction with cartilage, which may facilitate mastoid aeration and avoid cavity problems.

The purpose of the study was to evaluate the long-term outcomes after TTCR for pediatric cholesteatoma and to analyze the impact of the applied statistical method on the recurrence rate.

Methods

From 1986 to 2011, we retrospectively screened a consecutive series of children (18 years or younger) with acquired cholesteatomas after primary TTCR.

The pre-operative conditions were recorded according to the CAO staging system of cholesteatoma proposed by Dr. Lien in 1985.[1]

Outcome assessments

The medical records of all patients were analyzed for the last otologic examination. Data were collected on sex, ear side, age of surgery, operative findings (*i.e.*, cholesteatoma extent, degree of atelectasis, ossicular condition, labyrinthine fistula, and facial nerve dehiscence caused by cholesteatoma), recidivistic disease, and complications.

The anatomical outcomes were determined by the cumulative recurrence rate and recurrence-free survival rate (percentage of ears free from recurrence) at five, ten, 15 and 20 years after the initial surgery.

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Edited by Haruo Takahashi

Surgical techniques

The new surgical technique employed was one-stage TTCR via an anterior approach. An endaural incision was made to expose the external auditory canal (EAC) and temporalis fascia. The fascia was harvested as a graft. After elevation of the tympanomeatal flap from the posterior canal wall, the EAC was enlarged and the scutum was removed, giving direct access to the cholesteatoma.

Retrograde mastoidectomy was applied to remove the cholesteatoma, which extended along the route of involvement, thus creating an atticotomy, attico-antrectomy or attico-antro-mastoidectomy open cavity (Fig. 1). TTCR emphasizes the importance of preserving uninvolved functional middle-ear structures and mastoid air cells, which facilitates post-operative middle-ear ventilation.

Conchal cartilage was harvested to reconstruct the posterior canal wall and maintain the EAC anatomy, avoiding post-operative cavity problems.

Tympanoplasty was then performed to restore hearing.

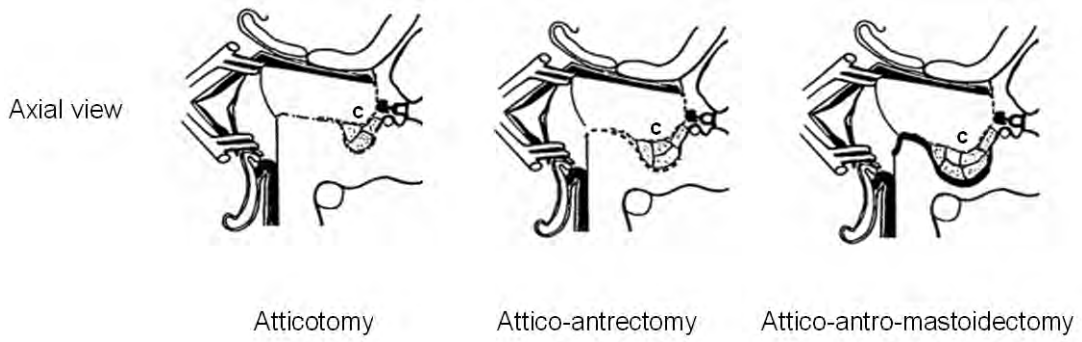

Fig. 1. Progressive retrograde mastoidectomy is individualized by the extent of the cholesteatoma, thus creating three types of tailor-made tympanomastoidectomy with cartilage reconstruction. c: obliterated cartilage in the tailor-made open cavity.

Statistical analysis

The cumulative recurrence rate was calculated by summation of the recurrence rates obtained for each observation year.

However, because the length of the post-operative observation period varied, the true percentage of ears free from recurrence may have been overestimated using the standard rate calculation.[2,3] Event-free survivals were thus determined using the Kaplan-Meier survival analysis.

Statistical comparisons and descriptive statistics were conducted using a commercially available software package, SPSS version 18.0 (SPSS, Inc., Chicago, IL, USA).

Results

The patient characteristics are shown in Table 1. A total of 59 operations were performed on 57 patients. Two patients experienced bilateral cholesteatomas. The mean age was 10.7 ± 4.5 years. The mean follow-up period was 14.1 ± 7.1 years, and 49 (83%) operations had a follow-up period of five years or longer. Dryness, self-cleaning and water-resistance were observed in ears without recidivism.

Cholesteatoma recidivism was observed in seven ears (five recurrent and two residual diseases). The mean detection time was 10.4 years, range 1.9-17.2 years. Five of seven ears (71.4%) with recidivism were detected more than ten years after surgery.

Cumulative recidivism rates calculated by different methods are shown in Table 2. The recidivism-free probabilities at five, ten, 15 and 20 years were 98.2%, 96.2%, 84.3% and 79.6%, respectively. Therefore, the recidivism rates at five, ten, 15 and 20 years as calculated by the Kaplan-Meier method were 1.8%, 3.8%, 15.7% and 20.4%, respectively. The cumulative recidivism rates as calculated by the standard rate method at five, ten, 15 and 20 years were 1.7%, 3.4%, 10.2% and 11.9%, respectively. The recidivism rates at five, ten, 15 and 20 years as calculated by the standard rate method were underestimated compared with those

by Kaplan-Meier analysis. Besides, the difference in recidivism rates between the two calculation methods increased with the follow-up time, from 0.1% to 8.5% between five and 20 years of follow up.

Table 1. Patient characteristics (n = 59).

Variables	*No. (%)*
Sex	
Male	33 (55.9%)
Female	26 (44.1%)
Ear	
Right	37 (62.7%)
Left	22 (37.3%)
Age of surgery (years)	10.69 ± 4.48
Follow-up period (years)	14.10 ± 7.10
Stage of CAO system	
Stage I	13 (22.0%)
Stage II	12 (20.3%)
Stage III	34 (57.6%)
Intact ossicular chain	11 (18.6%)
Labyrinthine fistula	4 (6.8%)
Facial nerve dehiscence	14 (23.7%)

Table 2. Cumulative recidivism rates calculated by different methods.

Observation time (year)	*Cumulative cases of recidivism*	*Recidivism-free probability (Life table, %)*	*Recidivism rate by Kaplan-Meier analysis (1-life table, %)*	*Recidivism rate by standard rate calculation (%)*	*Difference between two methods (%)*
5	1	98.2%	1.8%	1.7%	0.1%
10	2	96.2%	3.8%	3.4%	0.4%
15	6	84.3%	15.7%	10.2%	5.5%
20	7	79.6%	20.4%	11.9%	8.5%

Conclusions

The long-term outcomes in this study show that the functional surgery, TTCR, is an alternative and valid therapeutic option for children with cholesteatoma, allowing for higher rates of recurrence-free survival and cavity problem control, facilitating easy postoperative care for children.

The Kaplan-Meier survival analysis method should be used when discussing recidivism rates. The standard rate calculation method should be used only in cases where there are no censored data.

Due to late recurrences, the importance of long-term follow up cannot be overemphasized. We recommend that parents ensure that their children receive follow up periodically until they reach adulthood.

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CHOLESTEATOMA IN CHILDREN: CLINICAL PARTICULARITIES AND THERAPEUTIC OUTCOMES OF 57 CASES

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Introduction

The term 'cholesteatoma' is attributed to Johannes Müller in 1838. Radicals 'chole' and 'steat' refer to the cholesterol crystals and fatty acids found within this tumor, the suffix 'oma' to any neo-formation tissue characterizing a tumoral appearance.[1-4]

Poorly named, cholesteatoma of the middle ear is defined by the abnormal presence in the cavities of the middle ear of a keratinizing squamous epithelium (skin) endowed with a potential for scaling, migration and erosion.

This epidermis, except in cases of congenital cholesteatoma, always originates from the deep part of external acoustic canal (EAC).[5-8] According to Gray it is 'wrong skin in the wrong place'.[1,9-11]

Within this debate, we have considered, for many years, that cholesteatoma in the child, compared to that in the adult, is special. This feature goes for everything from diagnosis to therapeutic management and it would be related to the specific characteristics of the child itself. Cholesteatoma in this age is characterized by:

- An extremely difficult diagnosis. Hearing loss is usually unnoticed and otoscopic examination is often difficult due to the pusillanimity of the child.
- The occurrence of a pneumatized mastoid which is a subject of much debate.[12-16]
- A high aggression, responsible for a larger extension in the cavities of the middle ear and a greater number of complications.
- The difficulty of surgical treatment. In fact, even a drastic well-performed procedure will not restore the ear to normal once and for all. In addition, despite the surgeon's experience, cholesteatoma in children has a high tendency to recur (recurrence and residual).[17-19]

In this study of 57 cases of acquired cholesteatoma of the middle ear of the child followed up at our department, clinical and paraclinical data and therapeutic modalities have been carefully recorded and analyzed. We tried to highlight the elements of choice of a therapeutic strategy and to chart the pathway for a preventive approach covering both pre-cholesteatoma states and the cholesteatoma itself, once declared.

Materials and methods

This is a longitudinal-type study on 57 cases of acquired cholesteatoma of the middle ear of children recruited and followed at the ENT department of Ferhat Abbas University of Setif (Algeria) between January 2004 and December 2008.

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Cholesteatoma and Ear Surgery – An Update, pp. 229-236
Edited by Haruo Takahashi

The study population is represented by 53 children whose age cut-off point is 16 years at diagnosis. The aim of this work is the study of clinical and paraclinical particularities, therapeutic approach and the evolution of this disease in the pediatric population.

Were carefully recorded: age, gender, history, reasons for consultation, examination of both ears, CT scan, the audiometric assessment, the localization and extension of the cholesteatoma, assessment of the lesions, and the surgical procedures and results.

In short, we had to deal with 389 variables. These data were recorded, entered and analyzed using the IBM software SPSS 11.5 (SPSS: Statistical Package for the Social Sciences).

Results

The average age is 12 years ranging from four to 15 years with a peak incidence at age 13. In fact, 77% of children were under 11 when they underwent the first surgical treatment. There were 32 boys and 25 girls (sex ratio 1.2).

In addition, low socio-economic status, unemployed father, large families with many dependent children, a mostly rural residence far from the health care centers are all factors identified in our study and characterize much of our cases. Personal otological history was often reported including either acute otitis media (49.1%) or serous otitis media (22.8%).

The time from the onset of the first clinical signs to the date of the treatment (S1C1) is also very important, but very difficult to specify. The results are impressive, showing extremes ranging from 32 to 131 months, therefore an average of 75 months. This result confirms, once again, the insidious development of cholesteatoma or sometimes even the carelessness of the parents.

As for reasons for consultation, the first sign is largely dominated by the fetid otorrhea (96.5%) characteristic of the disease. Hearing loss is well behind (66.7%). Cophosis was not detected in any case, neither in the ear to be operated nor in the controlateral ear. Indeed, only one case had a bone-conduction hearing loss at 40 dB pre-operatively. The mean pre-operative air-bone gap is 27.1 dB.

Otorragie, an alarming sign motivating an urgent consultation, is noted in more than 38% of cases and is often the revelation of a polyp occupying the ear canal, characterized by the suffering of the lining of the tympanic cavity (Fig. 1).

At other times, cholesteatoma was revealed immediately by a complication. This situation applied to one out of four children (14 cases; 24.6%). The most common complication was acute mastoiditis (15.8%) with different modes of externalization, retro-auricular exteriorization was even the most frequent complication. Dizziness was noticed in 3.5% and facial palsy also in 3.5%, which regressed dramatically after medical treatment and surgery. We also noted a case of meningitis in a girl of four years, and a lateral sinus thrombosis and subdural abscess in the posterior fossa in a boy of 14.

After physical examination, no differences in the affected side were noted. There were more lesions of the pars tensa than of the pars flaccida, and these lesions were the cause of cholesteatoma in 65% of cases.

In more than 70% of cases the controlateral ear was diseased, with cholesteatoma in seven cases (a rate of 12.2%). We also noted the existence of an ear infection with effusion and/or tympanic retraction in over 36% of cases, a considerable figure that raises interest for a preventive action.

A CT scan remains the radiological examination of choice. A CT-scan was made pre-operatively in 84% of cases and in only 14% of cases during post-operative monitoring. It enabled us to study the anatomy of the middle ear cavities (a good pneumatization of the mastoid was detected in more than 61% of cases) and to make an assessment of the lesions of cholesteatoma (localization, extension and damage caused by it)(Fig. 2).

All patients were operated on first-hand. Overall, 81 operations were performed. Of these, 34 were performed using the closed technique (59.6%) and 23 using the open technique (40.4%). Nineteen cases, however, required a second or third time of revision surgery, and in five cases the closed technique of the first time has been changed into a canal-wall-down (CWD) technique.

Whenever possible, the closed technique was always attempted first. However, for particular situations, the open technique was used. These situations were either anatomical (mastoid eburnation and/or prolapses of the meninges or the lateral sinus: two cases), or clinical (complications: 11 cases), or even an important extension of cholesteatoma removal of which was considered difficult (nine cases). There was also the risk of patients not following up due to low socio-economic level or remoteness from health facilities (one case).

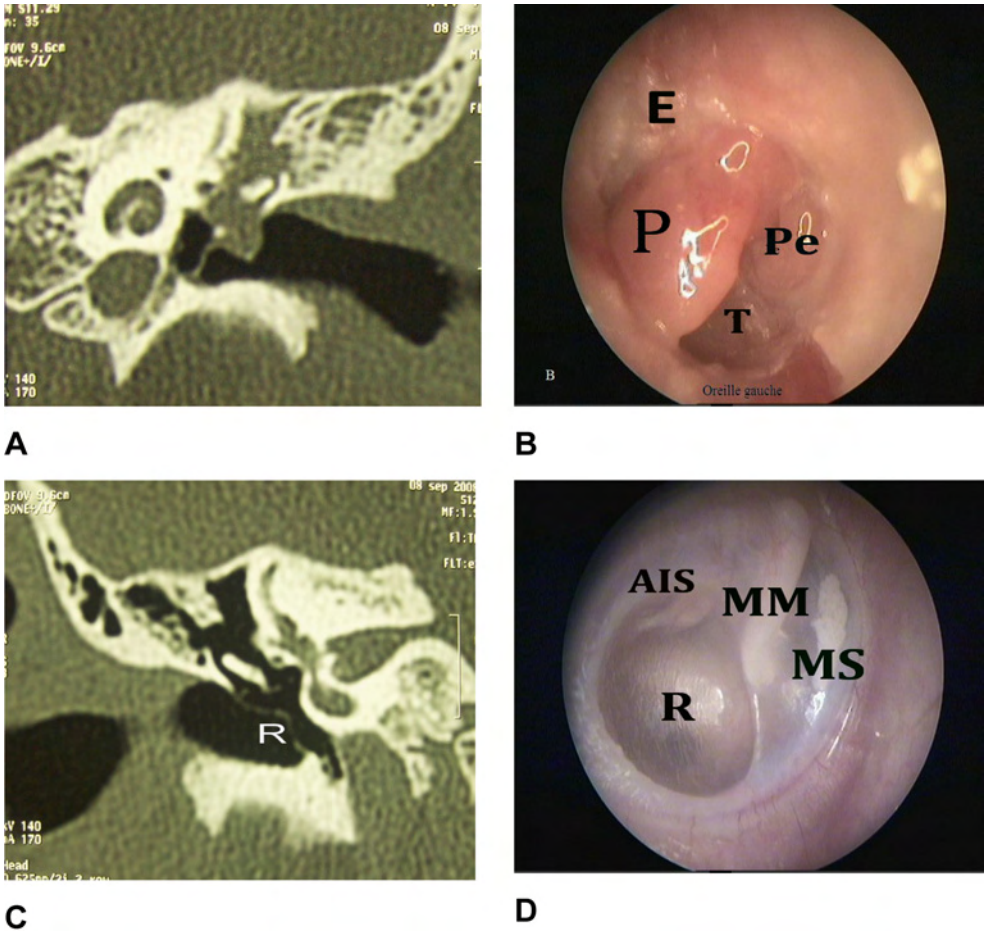

Fig. 1. Polyp of the external auditory canal of a left ear. A. Coronal cut of CT scan of the left ear, lysis of the descending branch of the uncus with prollabing aspect of tissue mass through the tympanic membrane into the lumen of the external acoustic meatus. Note the denudation of the tympanic portion of facial canal and that of the lateral semicircular canal. B. Oto-endoscopic aspect with rigid optic 0, showing the polyp at the bottom of the external acoustic meatus. C. Coronal cut of CT scan of the right ear, in the same child. Integrity of the ossicular chain, good ventilation of the middle ear cavities. Note the invagination of the pars tensa which is visible on clinical examination. D. Oto-endoscopic aspect with rigid optic 0, showing the poster superior retraction of the pars tensa which remains controllable and self-cleaning. E. Part of the cupular epitympanum. P: Polyp; T: Eardrum; Pe: Perforation of the posterior superior part of the pars tensa; MAE: External acoustic meatus; CSCL: lateral semicircular canal; CT: computed tomography; AIS: incudostapedical joint; R: Retraction; MS: myringosclerosis; MM: Handle of malleus.

The closed technique is a masto-antro-atticotomy transmastoid with transcanal atticotomy with a dual approach: through the duct, and transmastoid. The posterior tympanotomy was not done systematically, but it was performed in 67.6% of cases in response to the imperatives of excision of cholesteatoma. A second operation was performed in 15 cases among 34 operated by closed technique; this represents 44.1%, with an average period of 14.9 months. A third operation was necessary in two cases.

Ossiculoplasty was done whenever possible during the same operation, using the ossicles of the patient, usually the incus. In the absence of these materials or if they were embedded in cholesteatoma, the cortical mastoid bone was used instead. The cartilage (conchal or tragal) was not only used for reconstruction, but also doubled to ensure sound transmission in the open technique.

Over an average period of four years (three to five years) the overall recurrence rate (recurrence and residual) was approximately 45.6%, with more recurrence after the closed technique than after the open technique.

The mean post-operative air-bone gap was 23.3; this means an average gain of 3.9 dB compared to the average pre-operative air-bone gap of 27.1 dB. The difference is statistically significant at Khi square test ($p < 0.001$). A better functional outcome is obtained with ICW than with CWD and with an intact stapes than with a destroyed stapes.

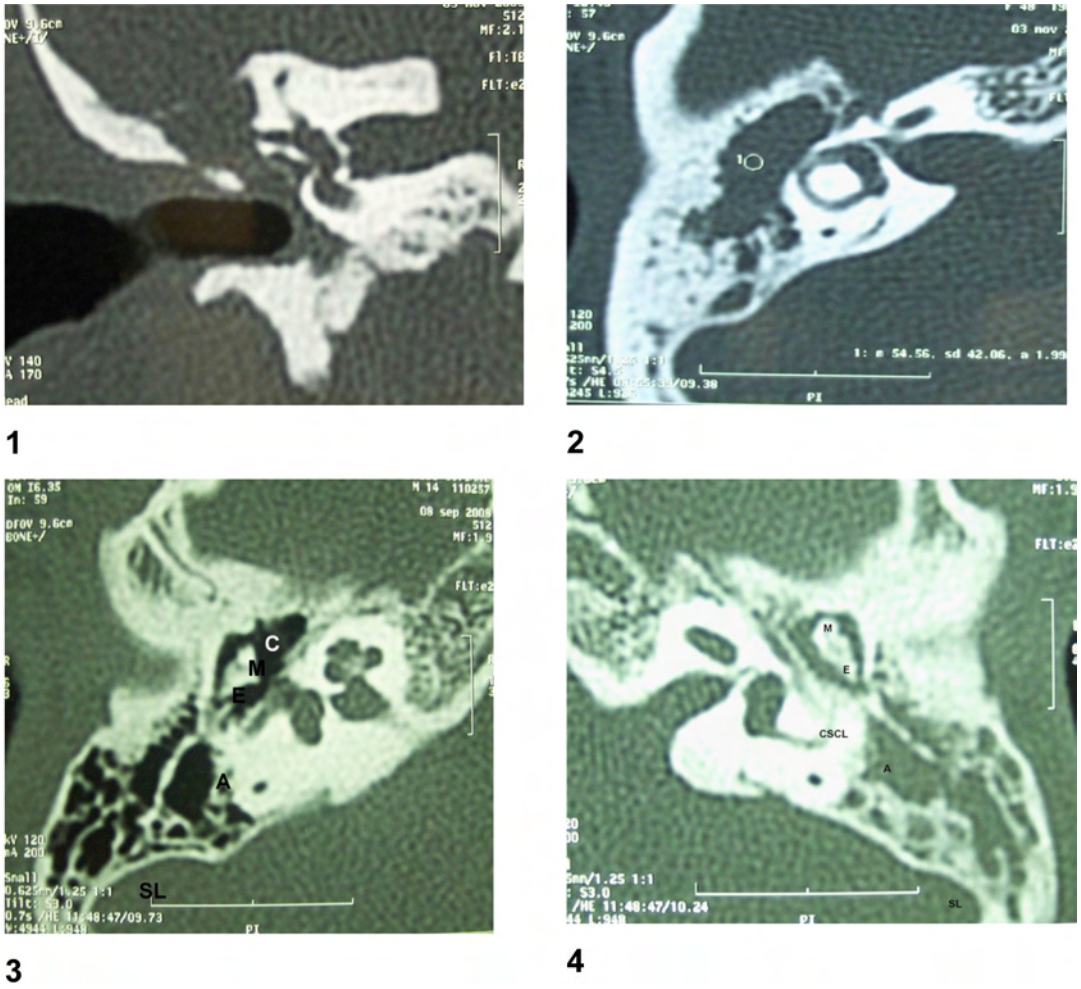

Fig. 2. Some aspects of the different lesions observed CT. 1 and 2. Coronal and axial CT scan cut respectively, a right ear showing a cholesteatoma with extensive denudation of the lateral semicircular canal, the denudation of the meninges of the middle cerebral fossa. lysis of the scutum and the ossicles chain. Note also the denudation of the second portion of the facial canal. 3 and 4. CT of both ears of the same patient, we note the importance of pneumatization of both mastoids: right without cholesteatoma and left side with cholesteatoma, which was extended and digitiform at the operation. A: antrum; SL: Lateral sinus; CSCL: lateral semicircular canal; V: Vestibule; M: malleus; E: incus; C: tympanic cavity.

Discussion

G.J. Duverney (1648-1730) reported in 1683 local accumulation of steatoma in and around the skull base.[20] Cruvelhier in 1829 named this a pearly tumor and it was Johannes Müller (1838) who named it cholesteatoma.[2-4]

Cholesteatoma was and is until today a subject of endless controversy mainly concerning its etiopathogenic mechanisms and modalities of surgical management.

In the literature, cholesteatoma in the child is often treated separately and considered special compared to that of adults. Our study enabled us to appreciate its clinical features, laboratory findings, evolution and the terms of its therapeutic management.

This is a disease of the second childhood occurring in a particular surroundings influenced by Eustachian-tube dysfunction and the upper respiratory tract infections very common at this age. Low socio-economic status and remoteness of health facilities were also reported.[21,22]

The average age is even higher than the upper age limit set is high.[18,23-26] The male predominance is marked and emphasized by all (Selmane, Yahi, Zribi).[26-28]

All authors agree that the first clinical sign is the fetid otorrhea (96.5%) which is very characteristic and immediately attracts attention towards cholesteatoma especially in children.[15,26,28-30] Hearing loss is well behind (66.7%) and is often unnoticed for two reasons: firstly, this is often unilateral and secondly, the child cannot express this symptom.

Cholesteatoma revealed by a complication was noted in one child out of four. The most common complication was acute mastoiditis exteriorized in the post-auricular region. In the literature, this rate varies from 0 to 52% with relatively low rates in the so-called industrialized countries: 0 for Gillot and Tos,[30] 4.5% for Darrouzet *et al*.[14] and higher rates elsewhere: 27% Prescott *et al*. in South Africa,[25] 33% for Rachidi in Morocco[28] and 46% for Cruz in Brazil.[30]

In Algeria, more specifically in its capital Algiers, Selmane[26] and Yahi[27]noted rates of about 10.4% and 12.7%, respectively, while Benchaoui in Constantine (300 km east of Algiers) in her thesis on complications of cholesteatoma in all ages, noted rates between 47.9 and 52.1%.[31] Thus, it seems quite clear that the development level of a country's socio-economical conditions, remoteness from health facilities and level of education and awareness are very influential in the increase of the incidence of this disease, but also in when this disease makes its first appearance by a complication.

In terms of otoscopic data, the postero-superior quadrant of the pars tensa is the most common starting point of cholesteatoma found in both the study of our series (64.9%) and the literature (83% for Darrouzet,[14] 65.5% for Mishiro,[17] 43.6% for Schilder *et al*.[32]).

We encountered many difficulties in interpreting these data due to a significant inhomogeneity between different authors linked to the following points: 1) The lack of a clear boundary between retraction pocket in the pre-cholesteatoma state and cholesteatoma; 2) Each author related his own specific otoscopic aspects to a physiopathologic mechanism.

The status of the controlateral ear is very important to clarify. In fact, otitis media with effusion and tympanic retraction were noted respectively in 17.5 and 19.2% of cases in our study. Darrouzet noted 12 and 17%. For Lerosey, however, it was six and 25% of cases.[14,16] Bilateral cholesteatoma was found in 12.8% of our cases. In the literature it varies from 0.8% for Selmane[26] to 18.5% for Prescott.[25] For Schilder it is bilateral in 15% of cases.[30] This bilateral involvement further complicates the therapeutic management.

The treatment of cholesteatoma remains exclusively surgical because the main goal is total eradication of the disease. The second goal, once overlooked, has become as important as the first one: preservation and even restoration of hearing.

There is still no consensus on the question what to use: closed technique or open technique. The current trend in the literature is to respect the normal architecture of the middle ear, which implies a closed technique. Although some authors have performed the open technique, in their studies we always find arguments against the choice of this technique. For many others, the attitude is rather eclectic, depending on each separate case (Charachon, Darrouzet, Lerosey, Sanna *et al*., Tos, Smyth).[12,14,16,18,30,33] Indeed, each technique has advantages and disadvantages. In our department, a closed technique was always attempted first. The current trend especially in children is a closed technique with mastoid obliteration.

In the vast majority of cases cholesteatoma is digitiform: 70% in our series, 60% for Yahi[27] and 51% for Darrouzet.[14] It is also very extensive, despite numerous classifications used in the literature (Sanna and Zini, Selmane, Saleh and Mills).[18,26,30] In our study, we noticed that some anatomical regions were more involved than others, *e.g*., the epitympanum with three compartments (the anterior half the time, the lateral in more than two thirds of cases, but especially in the posterior in over 98% of cases); followed by the aditus which is almost always affected by cholesteatoma, while the antrum was affected in 87.7% of cases. The rest of the mastoid air cells was infected in 49.1% of cases.

It should be noted that cholesteatoma in children is known to affect especially the atrium,[14,34,35] and is difficult to remove because it invades the tympanic cavity, particularly the retrotympanum. Indeed, the sinus tympani was invaded in 73.7% of our cases and the facial recess in 89.5% of cases.

In our series, in cases in which the ossicular chain was intact, the ossicle most affected was the incus, particularly its long process (72% lysis). In the literature an unbroken chain was observed in different percentages from one study to another. This rate varies from 9.7% (Silvola *et al*.) to 67% (Stangerup *et al*.).[25,29,36-41] Actually, it is higher in the European studies than in other studies, which suggests a rapid diagnosis and especially an early treatment of the disease before it destroys the ossicles.

Concerning function and reconstruction, the most interesting lesion to note is that of the superstructure of the stapes, 40.4% in our study. It was more than 76% for Prescott *et al*.[25] in South Africa and only 15% for Stangerup *et al*.[41] This will determine the ossicular reconstruction and predict the functional outcome.

The destruction of the ossicular chain associated with the lesion of the inner ear, especially the lateral semicircular canal (10.5% of cases) and that of the facial canal (19.3%) all show an aggressive cholesteatoma and also a long-lasting evolution of this disease (Fig. 3).

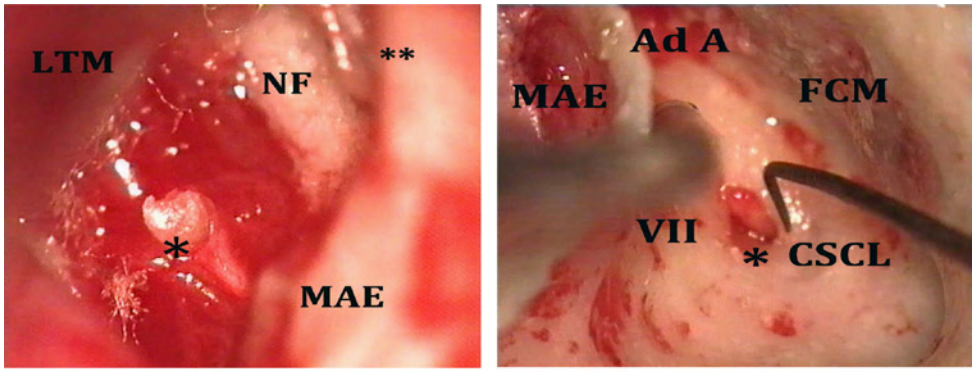

Fig. 3. Around the cholesteatoma. A. Epitympanic cholesteatoma with spontaneous atticotomy in a left ear, the stapes is present alone, there is also a lysis of its head (*). Enlargement of a spontaneous atticotomy (**) has led to better control of the extension of cholesteatoma, there is a denudation of the entire second portion of the facial nerve (NF). B. Erosion of the lateral semicircular canal without opening the membranous labyrinth (*): here the matrix of cholesteatoma is dissected. MAE: External acoustic meatus; LTM: Ttympano-meatal flap; NF, VII: Facial nerve; CSCL: Lateral semicirculair canal; FCM: Middle cranail fossa; Ad A: Aditus.

Ossiculoplasty was performed in 63 cases of 81 cases operated (any technique and time combined) of which 82.5% of cases at the first time. In almost all our cases, we had to use only autologous material because of the unavailability of ossicular prostheses. Actually, we prefer the ossicles of the patients (incus, malleus) if they can be used, otherwise the mastoid cortical bone or the cartilage can be shaped as needed.

Audiometric results: the mean post-operative air bone gap was 23.2 dB, indicating an overall improvement of 3.9 dB compared with the mean pre-operative air-bone gap of 27.1 dB. The difference is statistically significant at Khi square test ($p < 0.001$).

With the closed technique, the average post-operative air-bone gap is 19.3 dB, while it is in the range of 31.4 dB in the open technique. Similarly, with an intact stapes the mean post-operative air bone gap is 21.1 dB, while it is in the range of 27.3 dB when the stapes was destroyed (all techniques). We note, in light of these data and those of the literature, the hearing result is better with the closed technique than with an open technique and also with an intact incus as compared to a destructed one.[14,16,23,26,32,34,37,40-45]

Anatomical result: whatever the procedure, open or closed, zero risk does not exist; a residual or recurrent cholesteatoma is found with various techniques, according to the study and the authors.

Residual cholesteatoma is newly formed from a matrix left in place voluntarily or involuntarily, while recidivism is a treatment failure which results in the formation of a new cholesteatoma, often on a retraction pocket, *e.g.*, tubal dysfunction frequently reported in children.

Some authors[19,34,37-42] report about recurrence without identifying whether it is residual or recurrence. Silvola uses the term re-cholesteatoma,[37-39] others calculate the rate of recurrence and residual from the total number of operated ears, regardless the technique used. Finally there are those who report only the results of the revised ears. This caused a certain inhomogeneity in the comparison of results.

In our series, the overall recurrence (recurrence and residual) was 45.6% of cases (calculated by the total number of series); residual cholesteatoma in 26.3% and recurrent cholesteatoma in 21%.

In recent publications,[14-17,23,24,34] rates of residual cholesteatoma vary from 3.2 to 64%, while those of true recurrence vary from 8 to 30%. The risk of recurrence (residual recurrence) requires prolonged monitoring. If the emergence of a residual cholesteatoma is exceptional beyond five years,[14,40-42,46] recurrence may be more insidious and late,[19,46,47] delayed recurrences after five years represent 30% of cases,[42,46] whatever the technique performed. Yahi, in her study, found cases of recurrence at five and eight years after surgery,[47] Austin up to 15 years.[41]

Schraff found that 90% of residual cholesteatomas occur during the first five years[42] and Mishiro noted a recurrence rate even higher than the post-operative follow up is prolonged.[17]

While it is true that there is no consensus for the schedule of monitoring or its duration, the majority agrees that monitoring should be at least ten years,[16,37] regardless the technical procedure used.

For others (Ayache, Schilder, Kaylie)[11,32,48] this monitoring should be for life because the mystery of the etiopathogeny of this disease is still not resolved, also the monitoring of hearing is mandatory. Magnan noted that a new surgery after a gap of ten years is not seldom (6% of cases).[46]

To sum up, the risk of recurrence is always present, especially in children, due to the particularity of cholesteatoma in this age (digitiform, extensive, destructive) and the particularity of the child itself (particular immunity, adaptative disease, tubal dysfunction). This means life-long monitoring is mandatory, if not indispensable, at least annually. This monitoring is done at the clinic (otoscopic examination under a binocular microscope and oto-endoscope). Radiology is becoming indispensable, especially the new MRI techniques.

Conclusion

Cholesteatoma in the child is special. The large clinical latency and its misdiagnosis complicate not only the task of the surgeon, but also the prognosis of this threatening disease with a high potential of recurrence whatever the technique used.

The best anatomical and functional results are obtained with the closed technique, but this does not eliminate the high rates of recurrence. In fact, prevention is crucial. Efforts should be focused not only on the diagnosis and management of pre-cholesteatoma, but also on the cholesteatoma itself once diagnosed. This may reduce damage in the cavities of the middle ear. In other words: 'Early diagnosis and treatment provide the best opportunity for eradication'.[1]

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CLINICAL CHARACTERS OF PATIENTS WITH EXTERNAL AUDITORY CANAL CHOLESTEATOMA SURGERY

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Introduction

External auditory canal cholesteatoma (EACC) is relatively rare in comparison to middle-ear cholesteatoma. Patients with EACC often present with otorrhea and otalgia due to bone erosion, but many cases can be remarkably unnoticed. If the bone erosion progresses, serious complications, like facial palsy, may occur. Accordingly, the surgical indication should be decided upon taking into consideration the patient's age, severity of symptoms, extent of bone destruction, and compliance of cleaning in the outpatient clinic.

EACC is classified according to the cause. EACC of unknown origin has been classified as primary EACC, and EACC due to a known cause has been classified as secondary EACC. Moreover, primary EACC has been divided into strict primary EACC, which shows an accumulation of keratin debris and bone destruction in a localized area of the external auditory canal, and keratosis obturans, which shows accumulation of keratin debris and bone erosion. These two diseases are differentiated from each other by pathophysiology and clinical features. Secondary EACC follows after injury or congenital auditory canal stenosis.

In this study, we investigate the clinical features of patients undergoing surgery for strict primary EACC and the results of surgery.

Methods

The clinical records of all patients diagnosed with primary EACC undergoing surgery in our hospital from 2002 to 2011 were retrospectively reviewed. We evaluated their clinical characteristics, including gender, age, symptoms, complications, invaded area, and the results of surgery from their charts.

Results

There were seven males and five females. All of them had unilateral lesions. The mean age was 29.6 years, with a range of 10 to 63 years. The patients were mostly teenagers (Fig. 1). The most common symptom was otalgia, which occurred in eight cases. The other symptoms were hearing loss, otorrhea, and ear fullness, which occurred in seven, six, and four cases, respectively. Facial-nerve palsy was found in only one case. The most common complaint which was the reason why they underwent surgery was otalgia, accounting for six of 12 cases (Fig. 2). Two cases required urgent surgery because of uncontrollable otalgia. The most common area invaded by EACC was the inferior canal wall (Table 1). EACC extended into the mastoid cavity in only one case, which was treated with tympanoplasty with mastoid obliteration using bone chips and pedicled periostium flap. The vertical portion of the facial nerve was exposed in four ears intra-operatively. In one case where facial palsy was found before surgery, facial movement could not be elicited by electrostimulation. Tympanic cavity invasion was found in only one case. In this case, the inferior part of the tympanic

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Edited by Haruo Takahashi

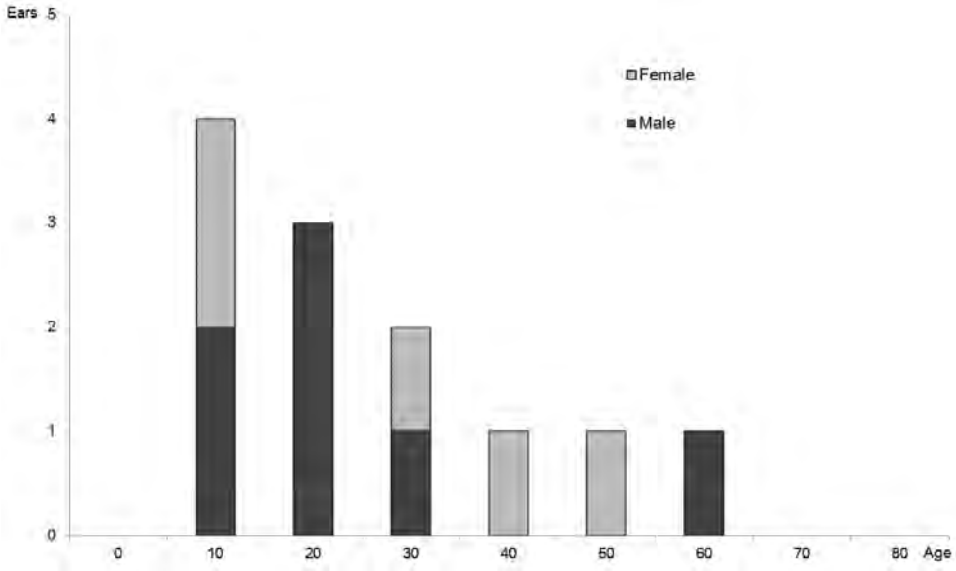

Fig. 1. Patients' age and gender. There were seven males and five females and all of them had unilateral lesions. The mean age was 29.6 years, ranging from ten to 63 years.

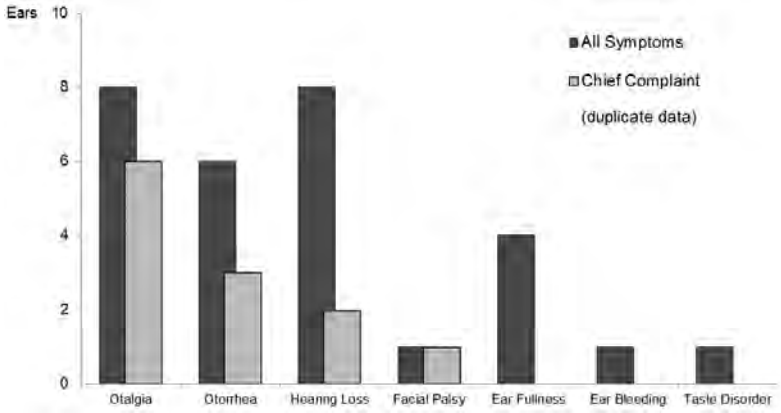

Fig. 2. Distribution of symptoms.

Table 1. Invaded area by EACC.

	Anterior	Antero-Inferior	Inferior	Postero-Inferior	Posterior	Posterior-Superior	Superior	Anterio-Superior	Multiple
EACC (n=12)	0	1	5	2	1	1	0	0	2

membrane was adhered to the promontory. Tympanic membrane perforation was also seen in two cases. Canalplasty was performed in all cases, and enlargement of the external auditory canal was performed in six cases. Tympanoplasty type 1 was also performed in three cases with tympanic membrane abnormalities. In one case of extended invasion into the mastoid cavity, mastoid obliteration using bone chips and a pedicled periosteal flap were also performed. After one year follow up of surgery, ten cases obtained a self-cleaning auditory canal without maintenance. In only one case, residual cholesteatoma arose under the generated canal and repeat surgery was needed. One case dropped out before within one year of follow up. The case with facial nerve palsy completely recovered within a few months post-operatively.

Discussion

The cases of EACC that require surgery are relatively rare. We have applied surgery mainly to young people. We have considered that as patients' age increases, they become less suitable for surgery. In the decision-making process before surgery for EACC, age may be a strong factor in deciding who can undergo this surgery. There were no differences in the gender of patients for EACC surgery.[1] Our results were consistent with previous reports of gender distribution of EACC.

The most common symptoms are otalgia and otorrhea. Otalgia is usually a chronic, dull pain, but sometimes becomes uncontrollably severe and needs urgent surgery.[2,3] We encountered two patients with severe otalgia, and performed urgent surgery. Facial palsy is rare, however, urgent surgery will help the patient to recover completely.

In our patients, the most common area invaded by EACC was the inferior wall. In the literature, a lower migratory rate of epithelium in the inferior canal wall has been described.[4] It may influence the migration speed of the external auditory canal skin. Tympanic-membrane perforation was also seen in two cases. In one case, the history of tympanic-membrane perforation was unclear, and EACC and tympanic-membrane perforation were incidentally found together. However, in another case, EACC arose first, then myringitis occurred in the follow-up period of EACC, and, finally, tympanic-membrane perforation occurred. Although there may be no relationship between EACC and tympanic-membrane perforation, chronic inflammation or skin migration may have an influence on both conditions.

The results of surgery after one year follow up were good in this study. Although we do need a longer follow-up period, we can apply surgery more frequently for young people as well as elderly people to improve their quality of life by creating a self-cleaning external auditory canal that does not require any maintenance.

Conclusion

The cases of EACC that require surgery are relatively rare. The cases with consistent otalgia, otorrhea, and facial nerve palsy and the cases with progressive extension to the middle-ear cavity should be indicated for surgery. However, it is necessary to consider the patient's status and compliance for cleaning in an outpatient clinic. We have applied surgery mainly to young people with uncontrollable symptoms and reached excellent results with surgery for EACC. We conclude that it should be applied more frequently, also on elderly people.

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APPLICATION OF COMPUTED TOMOGRAPHY SCAN FUSION WITH DIFFUSION-WEIGHTED ECHO-PLANNER MAGNETIC RESONANCE IMAGING FOR CHOLESTEATOMA IN CHILDREN

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Otitis media with discharge refractory to medical treatment is occasionally encountered, even in this era of broadly active antibiotics. Aural discharge can be caused by congenital or acquired cholesteatoma, which is difficult to diagnose at the first visit. The incidence of cholesteatoma is < 5% in such children.

In this study, we applied high-resolution CT and DWI (diffusion-weighted image) MRI to identify cholesteatoma in those children with acute otitis media and otitis media with effusion.

DWI-MRI relies on the signal produced by the motion of water molecules and it is one of the T2-weighted image and T2-related shine-through effects.

Limitations of DWI are retraction pocket auto-evacuate keratin debris resulting in a 'dry cholestatoma', which produces no restricted diffusion signal on the DWI-MRI sequence. In addition, air-bone susceptibility artifacts produced with echo-planner imaging (EPI) sequences makes interpretation more difficult.

In this study, a conventional 1.5 Tesla MRI scanner was used. Diagnosis of cholesteatoma was based on the presence of high-intensity signals in the middle ear or mastoid cavity on the DWI-EPI. DWI-MRI and high-resolution CT image were stored on a Ziostation workstation.

Cases presented are pediatric cholesteatoma in patients who visited our outpatient clinic complaining of protracted ear discharge.

We applied DWI-MRI and CT fusion imagery in these cases. Many benefits can be realized by combining these techniques. It can clarify relationships between temporal bone structures and pathology, be of help in diagnosis and precise localization of cholesteatoma, provide useful information for surgical planning, is easy to use in pediatric cases, and when the images are registered, several things can be determined at once. In clinical routine, the CT scan plane is tilted compared with the MRI scan plane to minimize radiation on the eyes, and when we register both images, angle adjustments are needed. Unlike other MRI sequences like T2-WI, anatomical structures such as the facial nerve are unapparent as anatomical index.

Limitations of these images are due to the air-bone susceptibility artifact around the skull base and its blur image; the evaluation of the lesions in supratubal recessus and epitympanic spaces are difficult. In addition, due to the T2-shine-through effect, distinguishing between cholesteatoma and inflammatory water retention lesion can be difficult.

In conclusion, DWI-CT fusion imagery can provide complementary information about temporal bone structures and pathologies. Due to the limitations of DWI which include air-bone susceptibility, blurred images, and difficulties in distinguishing cholesteatoma from inflammatory water retention lesions, evaluation of the extent of the disease can be difficult depending on the site of the lesions.

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Edited by Haruo Takahashi

INTRA-TYMPANIC DEXAMETHASONE INJECTIONS: ARE THEY EFFECTIVE FOR LONG-TERM CONTROL OF VERTIGO?

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Introduction

Menière's disease (MD), characterized by recurrent attacks of vertigo, fluctuating hearing loss (HL) and tinnitus, is a common disease with an incidence of 15-50 per 100,000 population.[1] Intractable MD was referred to a group of patients who are strongly prevented from participating in activities of daily life and interacting with their social environment, due to frequent attacks of vertigo, especially with progressive sensorineural HL, despite various kinds of medical and psychological management.[2]

Since McCabe's original description of autoimmune HL,[3] clinical and laboratorial evidence continues to suggest that MD may be an immune-mediated disorder. The findings of auto-immune response to type-II collagen,[4] elevated levels of IgG circulating immune complexes,[5-7] focal inflammation with intraepithelial invasion by mononuclear cells recognized as 'endolymphatic sacitis' by Danckwardt-Lilliestrom,[8] and the demonstration of auto-antibodies to the endolymphatic sac[9] suggest that auto-immunity plays a major role in the pathophysiology of MD.

This theory of immune involvement has led to the widespread use of steroids as a first-line treatment for MD.[1] Detection of corticosteroid receptors within the inner ear and histological changes to stria vascularis by corticosteroids have evidenced a crucial role of steroids in the inner-ear physiology.

Lohius *et al*.[11] demonstrated that the cellular structure of the stria vascularis underwent atrophy after removal of adrenal steroids, and Rarey[12] has showed that it returned to normal after administration of corticosteroids. As results of these advances, steroids are being used in MD in order to control this immunological reaction. Systemic steroid administration has been proved successful and is one of the current standard treatment options. Oral and intravenous steroid treatment, although very easy to use, has well-known side effects in the long term. According to Nadel[13] these side effects include high susceptibility to infection, diabetes, osteoporosis, peptic ulcer, glucoma, impaired wound healing, myopathy, hypertension, psychological changes, and avascular necrosis of the femoral head. Therefore, they must be used very cautiously and only when there is absolute necessity. Intra-tympanic (IT) therapy for MD has evolved since the introduction of 'titration protocols' for IT gentamicin.[14] The success of such intermittent injections of gentamicin inevitably has led to a similar use of IT steroids. Chandrasekhar[15] demonstrated a significantly higher concentration of dexamethasone in the perilymph after its injection intra-tympanically in a guinea pig model than its level after intravenous dexamethasone. Parnes[16] reported evidence that corticosteroids accumulate in inner ear fluids in much larger concentrations and remain longer after round window perfusion in guinea pig than after oral or intravenous administration. Hamid[17] found high concentrations of dexamethasone in the inner ear liquids with minimal diffusion to the blood. Dexamethasone is one of the most potent corticosteroids, is the longest acting and causes the least sodium retention.[18] After this encouraging findings, it seems reasonable that IT therapy with dexamethasone avoids the side effects associated with systemic steroids, treats only the affected ear, and results in increased level of corticosteroids in the inner ear.

Our current study was constructed to investigate the efficacy of intra-tympanic dexamethasone injection in the form of one single course of injections with or without repeated injections if needed on the long-term control of vertigo and hearing loss in patients with intractable unilateral definite MD.

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Edited by Haruo Takahashi

Patients and methods

A prospective, two-year follow-up study was carried out in the department of ORL & HNS of Tanta University Hospitals, Egypt between January 2008 and November 2011, to assess the efficacy of IT dexamethasone 4 mg/ml on patients with intractable unilateral definite MD. During the study period, 32 patients with a clinical diagnosis of intractable MD were eligible for participating in our clinical trial. The following inclusion criteria were fulfilled by our study group: 1) patients older than 21 years of age; 2) Definite unilateral MD as defined by the 1995 American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) guidelines;[19] 3) Intact tympanic membrane; 4) No intake of oral corticosteroids in the last six months before injection; 5) No previous history of surgery for MD or for chronic suppurative otitis media; 6) Failure of various forms of medical treatment to control the intractable vertigo for at least six months prior to the study; and 7) Pure tone audiometry scores (PTA) better than 70 dB HL.

For eligible patients, a detailed explanation of the study, including risks, complications, and possible benefits was provided and after that a written informed consent was obtained. The present study was approved by the Ethical Review Board of our Faculty of Medicine. Our study group was subjected to thorough history taking to assess their vertiginous spells as regards onset, frequency, duration and severity, in addition to subjective HL, tinnitus and aural fullness during the last six months prior to the IT injection. Afterwards, patients were subjected to general physical evaluation and a complete ear, nose and throat examination. Audiometric evaluation was performed prior to the procedure and during each post-procedure follow-up visit. We used a pure-tone average (PTA) of 0.5, 1, 2 and 4 kHz, because these were the standard frequencies tested in our department. Speech discrimination scores (SDSs) were also recorded.

We used the worst audiogram during the six months prior to the injection as our pre-operative audiogram, while the worst audiogram during the last six months of the follow-up period was considered our postoperative audiogram. A change of ten dB or more in PTA, or 15% or more of SDS as defined by 1995 AAO-HNS, guidelines was considered clinically significant.

Procedure

With the patient supine on a stretcher in the office with the head turned 45°C to the opposite side and the neck fully extended, an Emla cream (Lidocaine 2.5%/ Prilocaine 2-5%, Astra Zeneca Pharmaceuticals) was applied to regionally anesthetize the TM in the antero-superior quadrant of the involved ear. Before injection, one needle hole was made in the anesthetized area for air to escape, followed by another second hole through which dexamethasone in a dose of four mg/ml was injected slowly into the middle ear space using a 25-gauge spinal needle attached to one ml tuberculin-type syringe. The amount injected ranged from 0.4 to 0.6 ml (average 0.5 ml). After the injection, patients were kept supine for 20-30 minutes with the treated ear up and asked not to swallow and stay still. If the initial injection was too uncomfortable, lidocaine 1% without epinephrine was added to subsequent injections at a ratio of 0.1 to 0.9 ml of dexamethasone. The injections were repeated on the next day, and then given weekly for three consecutive weeks. Our protocol entails a total of 5 IT dexamethasone injections in a month. Patients in our study were seen for evaluation and progress notes at three, six, 12, 18 and 24 months intervals. During the study, some cases required reinjection procedures in the form of single or multiple injections if the disease decompensates again (reappearance of the vertiginous spells), or has not shown complete resolution.

Complications, if any, were recorded during the follow-up visits.

All the data from treatment results were processed statistically with the use of SPSS, version 16. We used the mean, standard deviation and chi-square test. The unpaired student t test was used to compare PTAs and SDSS. A p value ≤ 0.05 was assigned for significance.

Results

Thirty-two patients met our inclusion criteria. Eighteen were men and 14 were women. The average age of patients was 46.65 ± 11.1 (26-65 years). IT injections were delivered in 22 right ears and ten left ears. The average duration of illness before treatment was 7.63 ± 2.6 (six months-13 years). Twenty patients (63%) were stage 3, ten patients (31%) stage 2 and only two patients (6%) were stage 1.

Vertigo control

At the end of the two-year follow-up, complete control (Class A) was achieved in 17 patients (53%), and a substantial control (Class B) in seven patients (22%), which means the number of patients who had satisfactory relief or vertigo was 75%. No patient was worse than before treatment. Re-injections in the form of one to a maximum of four were given to seven cases of those who got class A and all seven patients of class B. Before treatment, 97% of our patients were classified as functional levels 3 and 4 (56% and 41% respectively). At the end of the follow-up period, functional level 1 was achieved by 17 patients (53%) and level 2 by 12 patients (38%), with only three patients (9%) having level 1.

During the six months before the treatment, the mean number of vertigo spells per month was 1.47 ± 0.84 (0.2-4). During the follow-up visits, the mean was reduced significantly and reached 0.24 ± 0.32 (0-1) at the end of the two years ($p = 0.001$).

Hearing results

At the end of the follow-up period, eight patients had improvement of their hearing by ten dB or more while only two patients showed deterioration of their hearing by ten dB, and 22 patients stayed the same. The mean pre-injection PTA was 45.71 dB (SD = 0.14), and the mean post-procedure PTA was 41.34 dB (SD = 13.87). The difference was statistically significant ($p = 0.009$). The mean improvement in speech discrimination for the entire group was 1.4%, not quite reaching significance ($p = 0.55$). Along the follow-up duration (two years), none of our patients showed relevant post-treatment complications. No cases of acute otitis media, dead ear or residual perforation were reported.

Discussion

IT steroid injection provides the potential for a near-direct application of corticosteroids to the area of immune dysfunction because the round window has been shown to be permeable to corticosteroids.[16]

The overall improvement of vertigo which was achieved in 75% of our patients with intractable MD (53% complete relief and 22% substantial control) which was sustained for two years, according to 1995 AAO-HNS reporting guidelines, is the most important finding in our study. Our goal would not have been achieved successfully without shifting to the intermittent (as needed) policy of repeated injections. This regiment entails IT dexamethasone injection upon recurrence of vertigo or patients' unease with their symptoms.

This and other research has sparked interest in dealing with this mode of therapy which carries the corticosteroids directly to the inner ear. Sakata first investigated this therapeutic alternative for MD in 1987, and along with Itoh,[20] reported relief of vertigo in 78% and improvement of tinnitus in 74% in their 61 patients after IT dexamethasone injection. None of their patients felt any deleterious side effects. Shea and Ge[21] found 96% relief of vertigo at one year and 76% at two years of follow up. Sennaroglu *et al*.[22] found 17 patients (72%) to have satisfactory relief for their intractable vertigo (42% complete and 30% substantial) when used topical dexamethasone through a ventilation tube. Barrs[23] demonstrated complete control of vertigo in 52% at three months and in 43% at six months. Repeated injections in five patients who had initial control, but later failed, yielded control in three of them. Garduno-Anaya *et al*.[18] reported 100% improvement of vertigo (82% complete and 18% substantial) in his dexamethasone group after two years follow up. Hearing was improved in 35%. Boleas-Aguiree *et al*.[24] have used a Kaplan-Meier time to event statistical method and found that dexamethasone injections were effective in controlling vertigo in 91% of their patients. Repeated injections were needed in more than 50% of the study group during the two-year follow-up. The results of Herraiz *et al*.[25] confirmed the efficacy of IT steroid in control of MD symptoms. After 24 months, the number of vertigo spells was reduced from 4.3 to 0.5; tinnitus relief was achieved in 82% of the patients, and the average improvement in PTA was 8.6 dB. Re-injections of vertigo control were required by 35% of their patients.

Although vertigo control remains the most important goal of treatment in patients with intractable MD, there is also a concomitant interest in the treatment of hearing loss and/or preventing the progressive impairment which usually associates with the disease.

The literature to date is divided, with some articles suggesting improvement and others showing no significant change in the disease course. Shea and Geo[21] initially reported an early 70% improvement, which was later decreased to 35% in patients with long-term follow-up. Silverstein *et al*.[26] reported that hearing improved

in 29% of patients treated in a similar way. Other reports, however, call into question the real efficacy of steroids on hearing. Silverstein updated his original observations with a report on the results of a group of 20 patients with stage-IV MD[27], and concluded that IT steroids injected by his protocol did not significantly improve hearing in patients with MD. Arriaga and Goldman[28] reported on 15 patients and their conclusion was that a single intratympanic steroid injection did not significantly improve hearing in patients with MD.

With the evaluation of the changes in hearing levels obtained throughout our study, we found eight patients (25%) to have a decrease of ten dB or more in their PTA, and only two patients showing deterioration of hearing by the end of the follow-up period. This statistically significant beneficial effect was observed at the 12-months follow-up visit and the maximum benefit was obtained at the two-years visit.

Our results confirm the efficacy of IT steroids in the control of MD symptoms. This success can be explained to some extent by the anti-inflammatory and immunosuppressive action of the dexamethasone on the immunological abnormalities in MD. Another explanation is the adoption of the intermittent (as needed) policy for those who showed failures or those who felt unease with their symptoms. It is unlikely that patients in our series persist with these repeated injections of IT steroids if they do not consider them to have a positive impact on their symptoms. On the other hand, the failure which has been detected in some of our cases with intractable MD during and at the end of follow-up period could be attributed to two reasons: 1) The pathophysiology of their disease does not belong to the immune theory with presence of other factors which share in its eruption; 2) The presence of mucosal bands or fibrous adhesions around or overlying the round window membrane, as Silverstein[29] had found that the round window was partially (17%) or completely covered with obstructing membranes in patients with otherwise normal middle ears.

Quaranta,[30] Green[31] and Silverstein[32] found vertigo improvement in 74% at seven years of follow up, 78% at 14 years, and 71% at 8.3 years respectively. Based on the results of these articles, it cannot be claimed that the success achieved was simply placebo or just the result of the natural history of fluctuation of symptoms in MD.

The high initial response, in addition to the successful long-term resolution seen at the end of the two-year follow up, the significant improvement of hearing, and the absence of complications would seem to give credence to the use of dexamethasone in a dose of four mg/ml given as needed IT injections for cases of intractable MD.

We recommend adopting this regimen as an effective monotherapy in the form of when needed IT injections for those patients who did not find a permanent resolution of their symptoms after a single course of injection.

Conclusion

Our data suggest a definitive role for the use of IT steroids in the treatment of intractable unilateral Menière's disease. At a dose of 4 mg/mL of dexamethasone in the inner ear perfusion therapy by a single course of intratympanic injections given over 3 weeks coupled with repeated injections in some patients, we obtained a complete control of vertigo in 53% of the patients at the end of the follow-up period.

The IT steroid injection of dexamethasone four mg/mL is a promising alternative to more invasive or destructive surgical procedures for patients with intractable MD.

The IT steroid injection is a safe in-office procedure, avoids the side effects that are associated with the use of systemic steroids or aggravation of another pre-existing medical condition, treats only the affected ear, avoids a possible effect on the contralateral normal inner ear, has a better profile and higher concentrations in the perilymph and endolymph than the systemic use, and also has the possibility of avoidance of a more invasive surgical procedure and the costs related to that procedure.

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A CASE OF HYPERPNEUMATIZATION OF THE TEMPORAL BONE PRESENTING WITH VERTIGO

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Introduction

Vertigo is a common symptom encountered in the clinical practice of otolaryngology. There are many causes of vertigo, but it can be difficult to determine the exact etiology. We encountered a case of hyperpneumatization of the temporal bone and surrounding bones which caused thinning of the superior semicircular bony canal. Hyperpneumatization of the temporal bone is a rare condition which induces a variety of symptoms, such as headache, the development of a palpable mass in the occipital region,[1] a feeling of fullness in the temporal region,[2] and conductive hearing loss.[3] Reports concerning vertigo induced by hyperpneumatization of the temporal bone are very scarce. We suspected that the vertigo in the present case was possibly due to the thinning of the superior semicircular bony canal, mimicking superior semicircular canal dehiscence (SSCD) syndrome.

Case report

A 60-year-old woman visited our clinic in March 2011 because of vertigo after airplane take-off. The vertigo occurred a few minutes after take-off and lasted for about 30 minutes. She had experienced this vertigo in two out of three flights. Simultaneously, she had pain in her left lateral neck. There were no other ontological symptoms such as hearing loss or tinnitus. Occasionally, she felt dizzy when she inserted a finger into her left ear canal. She had no Tullio phenomenon. Her past medical history was unremarkable.

Clinical data

The patient was well oriented with normal gait. Physical examination revealed intact tympanic membranes. There were no abnormal neurologic findings. Crepitation was observed in her left neck on a Valsalva maneuver.

Neuro-otological findings

Audiometry on the first visit revealed an average air-bone gap of 15 dB in the left low-tone frequencies (Fig. 1), but there was no air-bone gap in other occasions. Tympanometry was performed twice and both tests revealed Type A for the right ear and the peak shift to the positive pressure for the left ear (Fig. 2). A Eustachian tube test revealed bilateral stenosis. There was no nystagmus on the gaze or positional test. A fistula sign test applying positive and negative pressure into the ear canal was negative. The stabilometry findings were normal. The cervical vestibular-evoked myogenic potential (cVEMP) was hyporesponsive on the left side.

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Edited by Haruo Takahashi

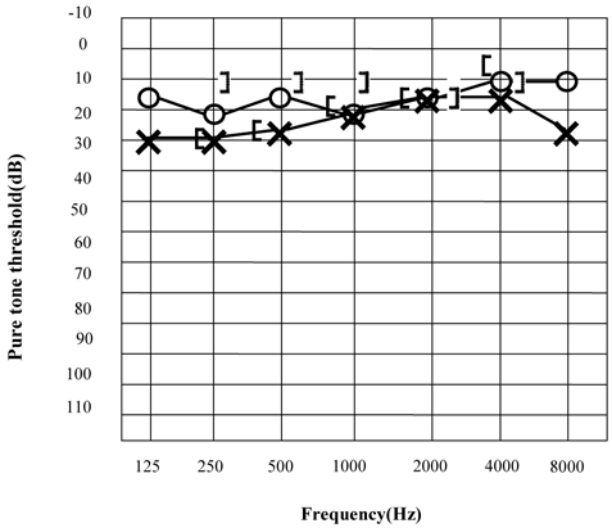

Fig. 1. An audiogram of the first consultation showed an average air-bone gap of 15 dB in the left low tone frequencies.

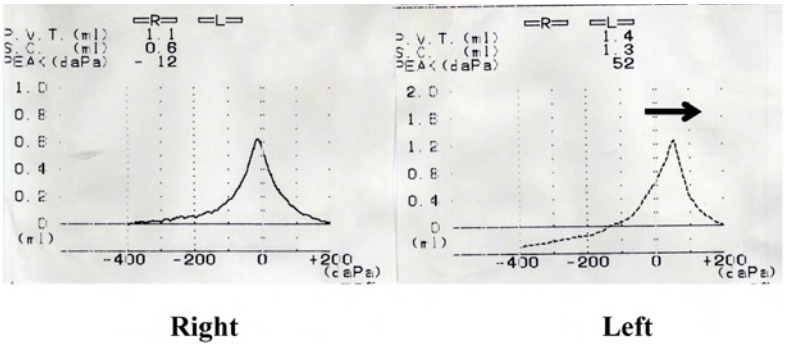

Fig. 2. Tympanometry revealed a Type A tympanogram for the right ear; on the left side, the peak had shifted to the positive pressure side (arrow).

Imaging findings

Computed tomography of the temporal bone revealed hyperpneumatization of the left temporal bone and surrounding bones extending to the sphenoidal sinus and atlanto-occipital joint. The bony wall of the left posterior cranial fossa was extremely thin. Air was noted in the left neck muscles (Fig. 3 a-c). Furthermore, there were well-developed air cells above the left superior semicircular canal (SSC), indicating SSCD (Fig. 4).

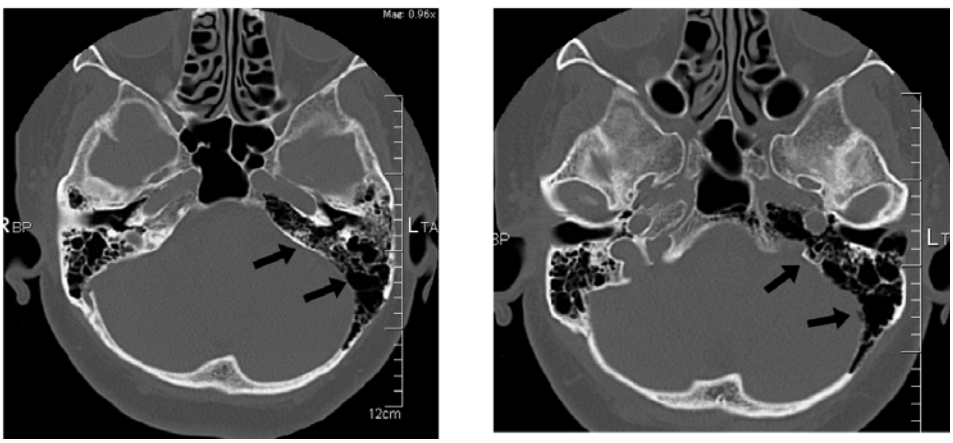

Fig. 3a. Computed tomography showed the hyperpneumatized left temporal bone extending into the petrous apex (arrows).

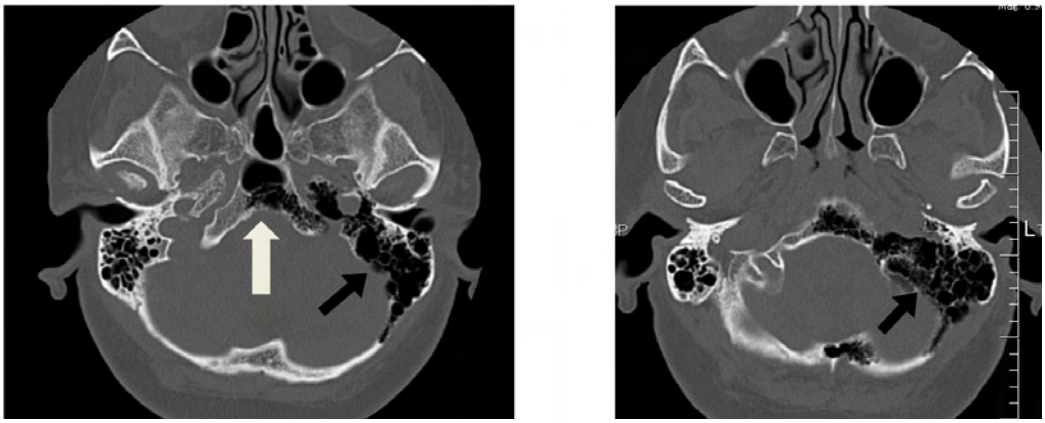

Fig. 3b. The hyperpneumatization extended into the sphenoidal sinus and atlanto-occipital joint (open arrow). Thinning of the posterior cranial fossa bone was observed (closed arrow).

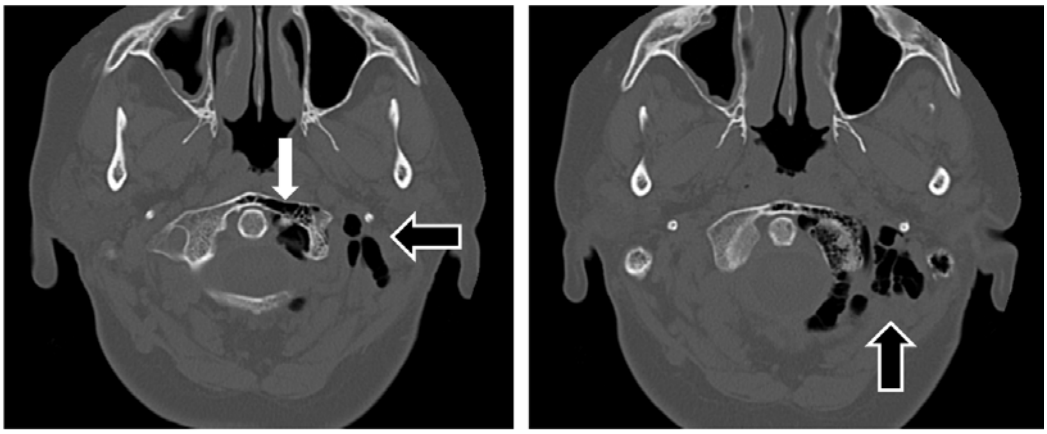

Fig. 3c. Air space in the left neck (closed arrow). A pneumatized atlas bone (open arrow) was observed.

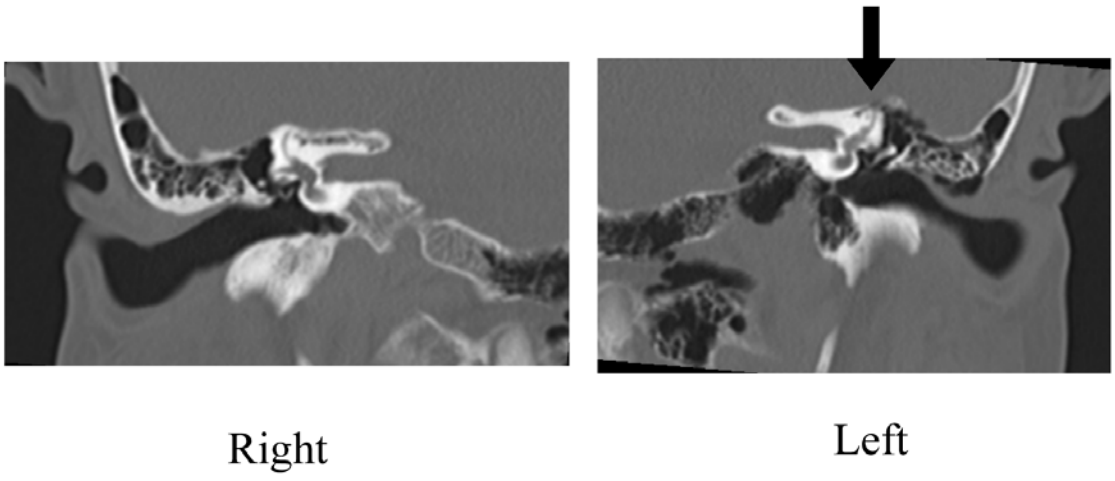

Fig. 4. The hyperpneumatized cells above the left superior semicircular canal. SSCD was suspected (closed arrow).

We diagnosed the condition as hyperpneumatization of the temporal bone and possible SSCD, which induced vertigo at the time of take-off. We advised her in April 2011 to take difenidol hydrochloride and diphenhydramine salicylate, which successfully treated her vertigo. However, despite the successful control of her vertigo, her left lateral neck pain remained. She makes it a practice to take both drugs before each take-off.

Discussion

Hyperpneumatization of the temporal bone is a rare condition with either congenital or acquired etiology. The former is caused by excessive bone resorption during pneumatization or persistent embryological synchondrosis.[4] The latter is known as the 'ball-valve effect': the air enters the middle ear through the Eustachian tube as nasopharyngeal pressure rises during swallowing, coughing or sneezing. This air then becomes trapped by a ball-valve mechanism, which allows the air to enter but not to leave, resulting in hyperpneumatization.[5]

In the present case, we assumed that the hyperpneumatization of the temporal bone and surrounding bones was caused by both congenital and acquired etiologies. The stenosis of the patient's left Eustachian tube may have induced the ball-valve effect. Hyperpneumatization then developed above the left SSC, which eroded the canal and resulted in dehiscence.

We considered that the pressure within the air cells increases after take-off and acts on the SSC through dehiscence, resulting in vertigo. The neck pain was also induced by an increased pressure transmitted into the subcutaneous tissue. The tympanogram curve that was observed to have shifted to the positive pressure side may also have been due to a higher pressure in the pneumatized space, rather than in the ear canal. Charbel *et al*. reported similar results by tympanometry, caused by a middle ear pressure rise with head rotation to one side.[4]

A series of symptoms induced by SSCD was first reported by Minor *et al*. as SSCD syndrome.[6] Patients with SSCD syndrome exhibit symptoms of oscillopsia or vertigo induced by pressure or sound, because the absence of the bony layer covering the SSC creates a third window. Congenital dysostosis, trauma and bone erosion by cerebrospinal fluid pulsation may result in SSCD.[7] SSCD increases the threshold of air conduction due to dispersed energy into the inner ear from the third window effect, and decreases the bone conduction threshold by amplified vibration transmitted to the temporal bone.[8] This mechanism is possibly responsible for the air-bone gap across the low tone frequencies in the present case. However, the audiograms taken on other occasions showed no air-bone gap. This is possibly due to the positive pressure within the air cells that acts on the SSC, thus weakening the third window effect. Tullio phenomenon, positive fistula sign, and enhanced cVEMP are characteristics of SSCD syndrome. In the present case, Tullio phenomenon was absent, a fistula sign test was negative and the cVEMP was hyporesponsive. The positive pressure acting on the SSCD may also be responsible for suppressing these signs and VEMP enhancement.

Martin *et al*. reported the placement of a myringotomy tube for the treatment of hyperpneumatization. Tube placement reduces the air pressure in the pneumatized areas within the skull base and atlas, leading to ossification and new bone formation.[1] In the present case, vertigo was controlled by the use of two anti-vertiginous drugs. Myringotomy tube insertion may also be an option for more secure treatment. The present patient had a risk of infection spreading into the hyperpneumatized air space. We advised her to be aware of the possible development of infection such as acute otitis media.

Conclusion

We encountered a case of a hyperpneumatized temporal bone and surrounding bones. Vertigo developed at the time of take-off in flight. Dehiscence of the SSC was the most likely cause of the vertigo.

Acknowledgements

We are indebted to Mr. Roderick J. Turner, Associate Professor Edward F. Barroga and Professor J. Patrick Barron, Chairman of the Department of International Medical Communications of Tokyo Medical University, for their editorial review of the English manuscript.

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CONGENITAL CHOLESTEATOMA AMONG SCHOOL-AGE CHILDREN IN HEARING SCREENING PROGRAM

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Introduction

Congenital cholesteatoma may occur in different age groups. It can be located in many sites including the cerebellopontine angle, the inner ear, the mastoid, the petrosus apex, the middle ear, the tympanic membrane, the external auditory canal.[1] From the clinical point of view it is very important to detect it as early as possible. Congenital cholesteatoma may lead to destruction of ossicular chain, perforation of the tympanic membrane and destruction of other temporal bone areas. Often patients report to a specialist when the cholesteatoma is large therefore very often the surgery includes reconstruction of the ossicular chain and is connected with considerable risk during and after surgery. Hearing screening in school-age children have been performed in 2008-2011 in Poland. The program included children at the age of seven-12 years old.[2,3] The main aim of the program was to detect hearing disorders which were not observed by the parents or teachers. The paper presents result of the second and third part of the program focused on congenital cholesteatoma detected among children who have not been under supervision of a specialist.

Material and method

The study includes children from Warsaw who underwent hearing screening examination in 2008-2010. This homogenic group includes 27,740 children at the age of 12 years. Data have been collected for three months (April, May, June) from 662 schools in Warsaw.[4] The data were collected by specially trained students, testers and professionals. Sensory Examination Platform™ was used with special data software for the purposes of this program. The platform is constructed on the basis of an efficient, central computer system and portable computers that are equipped with audiometric headphones and a button for the tested person. Computers communicate safely with the central database via the Internet. The Sensory Examination Platform™ (Fig. 1) provides the following examinations and tests:

- Audiometric testing: 'Audiogram'
- Hearing screening test: 'I can hear'™
- Speech screening test: 'I can speak' ™
- Sight screening test: 'I can see' ™
- Audiological survey
- Test 'DDT' – digital dichotic test
- Test 'GDT' – gap detection test

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Edited by Haruo Takahashi

Fig. 1. Sensory examination platform.

In this study we used: Audiometric test, Audiological survey, DDT test and 'I can hear' test.[3] Survey was especially important during this program, because the children with medical history of cholesteatoma were excluded from the analysis (only in part dedicated to that manuscript). Positive results of the test have been obtained by 16.5% of children who afterwards have been examined more profoundly. Of all the tested children, 8.6% gave a positive response in the audiometric test which is very important from the congenital cholesteatoma point of view; 5.6% constituted children with minimum one result on the level 21-30 dB SPL, 1.7% - 31-40 dB SPL, 1.1% - over 41dB SPL. Unilateral hearing disorders were found in 61.1% and bilateral in 38.9% of the positive results. They were referred to Institute of Physiology and Pathology of Hearing or cooperating centers in Poland for further examination. Ninety-two percent of the children from the selected group came for a control visit.

The criteria of positive result were minimum one response on level higher than 20dB SPL in audiometry. Children who came for the control visit were examined by a specialist with micro-otoscopy and video-otoscopy. Forty-three children were found with congenital cholesteatoma (0,046/10000). This included 41 cases of unilateral congenital cholesteatomas and two bilateral cases. Cholesteatomas in all cases have been confirmed surgically.

Results

Our results present that hearing disorders among school age children are a serious problem. Of parents of the children with hearing problems, 65.3% had not observed any problems prior to the examination.[2] Our research confirms that early identification of hearing disorders gives chances to provide adequate treatment earlier and with lower possible complications. The hearing-screening program provides all children with equal chances to develop properly as well as gives better education opportunities what was emphasized in the conclusion of European Union Council.[5]

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TYPE-IV TYMPANOPLASTY FOR PATIENTS WITH SEVERED TENSOR TYMPANI TENDON DURING CHOLESTEATOMA SURGERY

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Introduction

The tensor tympani tendon (TTT) lies between cochleariform process and neck of the malleus. The tensor tympani muscle is connected to the tendon directly.[1] The function of the TTT is to maintain the proper position of the malleus handle and to control excessive lateral movement of the eardrum by retracting and relaxing the malleus handle and eardrum.[2] The TTT sometimes disappears due to cholesteatoma invasion, and is frequently severed to eradicate cholesteatoma in the tympanic cavity to make the anterior part of the eardrum mobile and to aid inspection of the protympanum.[2] If the TTT is severed during cholesteatoma surgery, the eardrum becomes unstable. Generally speaking, in type-IV tympanoplasty, a columella inserted between the footplate of the stapes and the eardrum has a tendency to become unstable. These factors may lead to poor post-operative hearing result, especially in staged tympanoplasty, because the tympanomeatal (TM) flap is repeatedly elevated. Post-operative hearing result of type-IV tympanoplasty for middle-ear cholesteatoma performed in our hospital between 1996 and 2005 was poor (success rate; 52%). During this period, the TTT was frequently severed and the TM flap was repeatedly elevated. Therefore, we performed type-IV ossicular reconstruction without elevation of the TM flap during secondary surgery to improve post-operative hearing results.

Methods

Type IV ossicular reconstruction was performed between 2006 and 2011 without TM flap elevation during secondary surgery in cases with severed TTT during primary surgery. The surgical procedure was as follows: 1) The TM flap was not elevated; 2) Transmastoid approach alone was used; 3) Residual cholesteatoma was eradicated by endoscopy; 4) Ossicular reconstruction was performed using a columella such as autologous ossicle, cortical bone, and TORP (Apaceram®). The distance between the footplate and the malleus handle or eardrum was precisely measured; 5) If possible, ossicular reconstruction of type IV with interposition was selected because then stability of the columella is certain. The postoperative hearing result was considered 'successful' when at least one of the following criteria was satisfied: 1) Post-operative hearing level within 30 dB; 2) Post-operative air-bone gap within 15 dB. 3. Hearing gain over 15 dB.

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Edited by Haruo Takahashi

Results

Post-operative hearing results of type IV with TM flap elevation performed during one-stage and two-stage tympanoplasty between 1996 and 2005 are shown in Table 1. During this period, the TTT was frequently severed. When the TTT was preserved, the success rate was 100%, although ossicular reconstruction was performed during the second stage. On the other hand, when the TTT was severed, the success rate decreased to 53%. In addition, the success rates were similar, even if the TM flap was elevated once or twice. These findings indicate that whether the TTT is severed or preserved is an important factor. During this period, the incidence of staged tympanoplasty in type IV tympanoplasty for cholesteatoma was 72%.

Hearing results after type-IV reconstruction in one-stage surgery alone performed between 2006 and 2011 are shown in Table 2. During surgery, the TM flap was elevated in all cases. The success rate was poor in both severed tendon and preserved tendon groups. These results indicate that post-operative hearing result of type IV in one-stage surgery was poor.

According to preservation or severance of the TTT during primary surgery and elevation or no elevation of the TM flap during secondary surgery, all patients who underwent staged tympanoplasty and re-operation between 2006 and 2011 were divided into four groups. Group 1: severed TTT and with TM flap elevation; Group 2: severed TTT without TM flap elevation; Group 3: preserved TTT with TM flap elevation; Group 4: preserved TTT without TM flap elevation.

Table 3 shows the hearing results after type IV reconstruction (in staged surgery and re-operation) with TM flap elevation during secondary surgery. When the TTT was severed (group 1), the success rate was only 20%. On the other hand, if the TTT was preserved, the success rate was 100% (group 3). There was a significant difference between the two groups ($p < 0.05$).

Table 4 shows hearing results after type-IV and type-V reconstruction (in staged surgery and re-operation) without TM flap elevation during secondary surgery. Type V or stapedectomy with columella was performed in two patients with fixed footplate. In these two groups, the success rates were better: 73% in group 2 and 86% in group 4, respectively.

Table 5 shows the summary of this study. If the TTT was severed, the success rate became better, when the TM flap was not elevated during secondary surgery. In addition, if the TTT was preserved, the success rate was better, even if the TM flap was elevated.

Table 1. Post-operative hearing results of type IV (IV c + IV i) with tympanomeatal flap elevation during one-stage and two-stage tympanoplasty (1996-2005, n = 61).

Tensor tympani Tendon (TTT)	*TM flap elevation (Surgery)*	*Successful*	*Unsuccessful*	*Total*
Preservation	Once (One-stage)	0	0	0
	Twice (Two-stage)	6 (100%)	0	6 (100%)
	Total	6 (100%)	0	6 (100%)
Severance	Once (One-stage)	9 (53%)	8	17 (100%)
	Twice (Two-stage)	20 (53%)	18	38 (100%)
	Total	29 (53%)	26* (47%)	55 (100%)

*Five cases with fixed footplate were included. IV i ; type IV with interposition, IV c ; type IV with columella.

Table 2. Hearing results after type IV (one-stage) tympanoplasty with tympanomeatal (TM) flap elevation (2006-2011, n = 8).

		Successful	*Unsuccessful*	*Total*
Severed tendon with TM flap elevation	IV i	0	0	0
	IV c	1	2	3
	Total	1 (33%)	2 (67%)	3 (100%)
Preserved tendon with TM flap elevation	IV i	2	2	4
	IV c	0	1	1
	Total	2 (40%)	3 (60%)	5 (100%)

IV i ; type IV with interposition, IV c ; type IV with columella.

Table 3. Hearing results after type IV (staged surgery and re-operation) with tympanomeatal (TM) flap elevation (2006-2011, n = 9).

		Successful	*Unsuccessful*	*Total*
Severed tendon	IV i	1 (100%)	0	1 (100%)
with TM flap elevation	IV c	0	4 (100%)	4 (100%)
(Group 1)	Total	1 (20%)	4 (80%)	5 (100%)
Preserved tendon	IV i	4 (100%)	0	4 (100%)
with TM flap elevation	IV c	0	0	0
(Group 3)	Total	4 (100%)*	0	4 (100%)

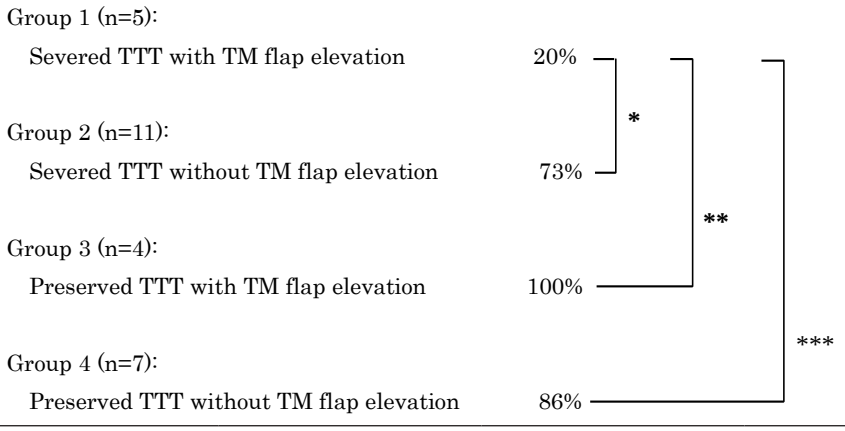
*p <0.05 by Fisher's exact probability test. IV i ; type IV with interposition, IV c ; type IV with columella.

Table 4. Hearing results after type IV& V (staged surgery and re-operation) without tympanomeatal (TM) flap elevation (2006-2011, n = 18).

		Successful	*Unsuccessful*	*Total*
Severed tendon	IV i	2 (100%)	0	2 (100%)
with TM flap elevation	IV c	4 (57%)	3 (43%)	7 (100%)
(Group 2)	V (Stapedectomy)	2 (100%)	0	2 (100%)
	Total	8 (73%)	3 (27%)	11 (100%)
Preserved tendon	IV i	6 (86%)	1 (14%)	7 (100%)
with TM flap elevation	IV c	0	0	0
(Group 4)	Total	6 (86%)	1 (14%)	7 (100%)

*p <0.05 by Fisher's exact probability test. IV i ; type IV with interposition, IV c ; type IV with columella.

Table 5. Post-operative hearing results of type IV& V tympanoplasty during second-stage surgery and re-operation (2006-2011, n = 27).



TTT; tensor tympani tendon, TM flap; tympanomeatal flap.
*p =0.106, ** p =0.047, *** p =0.072 by Fisher's exact probability test

Discussion

The crucial factors that affect the results of tympanoplasty on hearing are presence of the handle of the malleus, the TTT, and the superstructure of the stapes. The handle of the malleus stabilizes the columella and the TTT also stabilizes the columella.[3] However, in type-IV tympanoplasty for cholesteatoma, the TTT is frequently severed and the superstructure of the stapes is already absent. In such conditions, it was recommended, in the second stage of tympanoplasty, that reconstruction of the ossicular chain might be performed through the mastoid without disturbing the skin of the external auditory canal and the position of the new tympanic membrane.[3] Therefore, in patients with severed TTT, if the TM flap is elevated again during secondary surgery, the position of the eardrum becomes more unstable, or so-called lateralization will occur. In addition, if type-IV tympanoplasty is performed during secondary surgery, instability of the columella becomes more serious. From the present study, it was revealed that post-operative hearing results became better if the TM flap was not elevated and ossicular reconstruction was performed by transmastoid approach alone. If the TTT

is severed during primary surgery and the TM flap is not elevated during secondary surgery, lateral movement of the eardrum is minimal. In such cases, when type IV and V tympanoplasty is performed, the columella becomes stable. By these procedures, better hearing results can be expected.

Conclusion

It was concluded that post-operative hearing results was better if the TTT was preserved. When the TTT was severed, hearing result was better in staged surgery and re-operation groups than that in one-stage surgery group. If the TTT is severed during primary surgery, it will be better to perform tympanoplasty type IV and V with columella without TM flap elevation during secondary surgery to obtain better hearing results.

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OSSICULOPLASTY OF SMALL CRUS LONGUM DEFECTS WITH BONE CEMENT

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Introduction

Ossicular erosion is frequently encountered in chronic otitis media (COM) with retraction pockets, atelectasis of the tympanic membrane and cholesteatoma.

The long process of the incus is most commonly affected.[1-3] This condition results in irreversible conductive hearing loss, due to incudostapedial joint (ISJ) discontinuity, which can only be partially recovered by otosurgical reconstruction.[4] In cases of isolated long process erosion, reconstruction of the ossicular chain can either be accomplished by *bypassing* techniques with ossicular replacement or *bridging* techniques with ossicular repair.[1,2,5-7]

Bypassing can be performed with interposition of an autograft or allograft, or by means of a partial ossicular replacement prosthesis (PORP) with a broad range of available biomaterials to chose from, including plastics, ceramics and titanium. Bridging can be performed with different types of angular prosthesis (angular clip prosthesis, Applebaum prosthesis, Plester prosthesis), or as an alternative, by using bone cement such as glass ionomeric cement (GIC) and hydroxyapatite (HA) bone cement.[1,2,6,8,9,10] In cases with smaller defects, bridging with bone cement seems an attractive approach compared to type-II reconstruction by interposition.

The aim of the present study was to describe the hearing results after ossiculoplasty with bone cement in cases with small defects of the long process of the incus.

Materials and methods

The surgical procedure performed in our study was an ossiculoplasty using SerenoCem cement.[5,11] Cement application was performed as the last step of the tympanoplasty procedure to avoid disruption and mobilization of the new ossicular chain. Briefly, diseased or fibrous tissue around the ossicles was removed. Next, mucosa was stripped from the stapes head and the distal crus longum incudis to obtain a bloodless dry bony surface. Gel foam was then placed to avoid inadvertent spillage of cement. Bone cement was prepared and applied by a Rosen needle, with a working time of two to four minutes and a hardening time of five to seven minutes. The result was tested by application of pressure on the malleus. Finally, the tympanomeatal flap was brought back and the ear canal was packed.

The study is a retrospective study of 26 patients operated with SerenoCem cement ossiculoplasty. The inclusion criterion was an isolated smaller crus longum defect with an intact malleus and stapes. The exclusion criteria were tympanosclerosis, inducomallear joint laxity, incus luxation, concurrent otosclerosis, or a follow-up time shorter than one year. The outcome measures were hearing results represented by pre- and postoperative pure-tone-average (PTA) (500, 1000, 2000, and 4000 Hz) and Air-Bone-Gap (ABG) closure as well as post-operative complications.

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Edited by Haruo Takahashi

Results

Twenty-six patients were identified in our database, in whom we performed ossiculoplasty with SerenoCem cement due to small defects of the crus longum. Two patients were excluded because of a follow-up time shorter than one year. Of the 24 patients included, 75% were adults, whereas 25% were children, giving a mean age of 39 years. The patient data included 18 cases of retraction pockets and eight cases of cholesteatoma. The mean follow-up time was 14 months and the mean follow-up rate was 92% (Fig. 1).

We registered the PTA pre- and post-operatively, as well as the improvement in ABG. The graphs shown in Fig. 2 represent the mean pre-operative air- and bone conduction thresholds compared to the mean post-operative thresholds. The pre-operative PTA was 36.8dB HL, whereas the post-operative PTA was 30.4dB HL. Our results showed a statistically significant improvement in mean PTA of 6.2dB HL (Fig. 3), as well as a statistically significant mean ABG closure of 11.1dB HL (Fig. 4), corresponding to a success rate of 77% (AAO-HNS Committee on Hearing and Equilibrium guidelines).

The only post-operative complication observed was an affected sense of taste in two patients (Fig. 5). In one of these patients, the chorda was lost during the operation. In the other patient, however, the chorda was described as intact by the end of the surgical procedure.

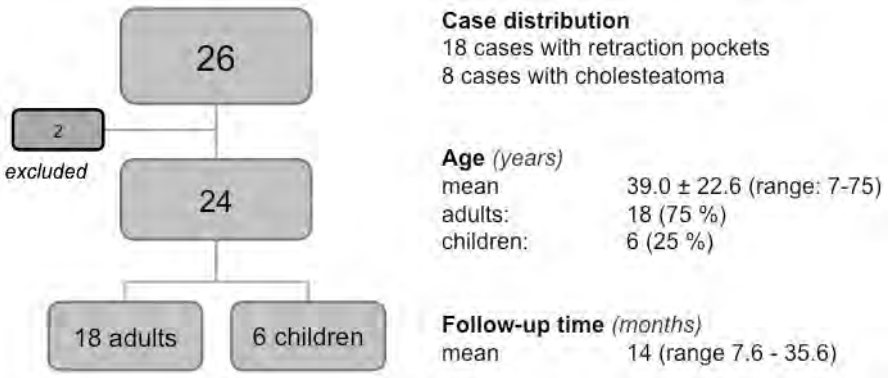


Fig. 1. SereneCem cement ossiculoplasty study (overview).

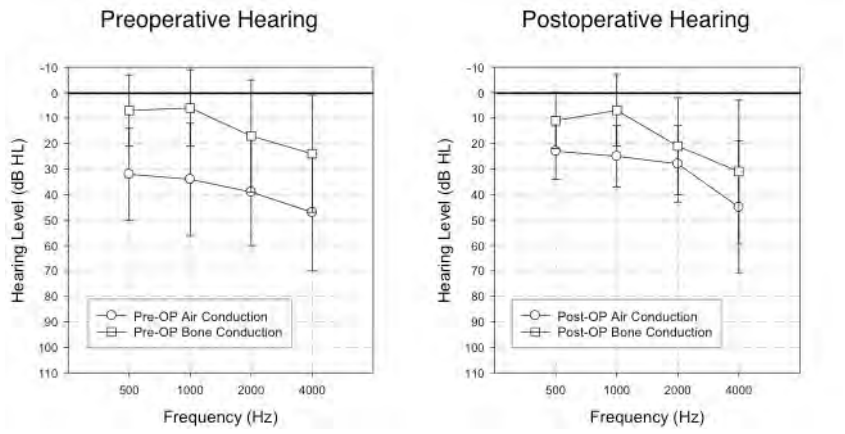
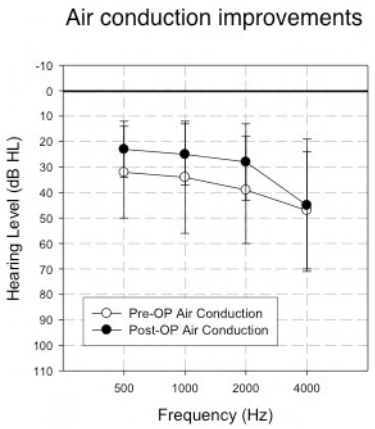


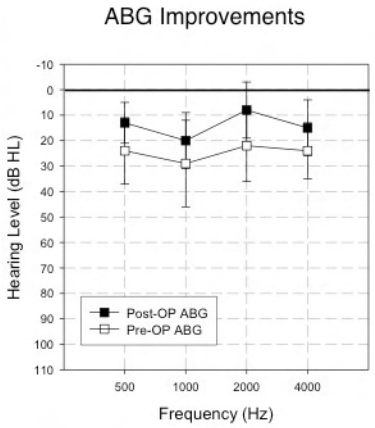
Fig. 2. Pre- and post-operative hearing levels.



	PTA (dB)
Pre-operative	36.8
Post-operative	30.4
Gain	**6.2**
p-value	***0.001***

PTA: Pure tone average

Fig. 3. Air conduction improvement.



	ABG (dB)
Pre-operative	24.8
Post-operative	13.7
Gain	**11.1**
p-value	**0.0003**

Successful ABG closure (< 20dB)* : 77 %

*) AAO-HNS Committee on Hearing and Equilibrium guidelines

ABG: Air bone gap

Fig. 4. Improvements in ABG.

• Facial nerve palsy	0 (0 %)
• Chorda (affected sense of taste)	2 (8 %)
• Bone cement extrusion	0 (0 %)
• TM thickening	0 (0 %)
• Retraction of TM onto the reconstructed ISJ	0 (0 %)
Total	**2 (8 %)**

TM: tympanic membrane, ISJ: incudostapedial joint

Fig. 5. Post-operative complications (n = 24)

Discussion

Erosion of the long process of the incus is the most commonly encountered ossicular chain defect in patients with COM.[1,2] The prosthetic material chosen for reconstruction must be safe, biocompatible, easily handled and applied, capable of efficient sound transmission, and it should not be extruded or resorbed.[2,9,12] Unfortunately, none of the biomaterials presently available meet all of these criteria.

Bypassing techniques with PORPs or incus interposition (type II) are widely used to reconstruct the ossicular chain, but all imply certain disadvantages, including a significant risk of dislocation, followed by deterioration of sound transmission.[2,3,8] Additional risks include prosthesis extrusion, ossicular necrosis, ankylosis to bony surfaces, and implantation cholesteatoma.[1,3,8,10]

In contrast, bridging the ISJ with bone cement restores ossicular chain anatomy, leaving its inherent sound transmission efficacy intact, while eliminating the risk of prosthesis dislocation or extrusion.[1,5,7,8] Furthermore, bone cement ossiculoplasty has the advantage of being a fast, safe and cost-effective procedure.[2,6] Unfortunately, concerns still exist regarding bone cement breakdown or loosening of the re-sculptured incudo-stapedial joint over time.[1,2,9] Moreover, studies have shown that certain bone cements including SerenoCem, exhibit pro-inflammatory properties[13] and are potentially neurotoxic.[2,6,14] There have been no reported cases of toxicity secondary to glass ionomeric cement ossiculoplasty.[2,6] However, contact with the dura, perilymph and neural tissue (facial nerve, chorda tympani, Jacobson's nerve) should be avoided during application.[1,7,9] Postoperative adhesions between the middle-ear mucosa and the tympanic membrane (TM) as well as thickening of the TM have been described.[6]

Hydroxyapatite (HA) bone cements incite minimal inflammation and show better tissue tolerance than GIC's as well as evidence of osseointegration with ossicular bone.[1] Therefore, HA cements with short setting times, such as Otomimix, are preferred materials in the reconstruction of the ossicular chain.[2,3,8]

Almost all bridging techniques using angular prosthesis or bone cement preserve the physiological function of the ossicular chain including the malleoincudial joint, which may play a key role in ambient atmospheric pressure variations.[15]

Although bone cement techniques are faster and much more cost-effective, angular prostheses have the advantage of being easier to remove in case of revision surgery, since bone cement adheres firmly to the ossicles.[15,16]

In our study, we found acceptable short-term hearing results for ossiculoplasty with bone cement of smaller crus longum defects. The results are comparable to those obtained by conventional type II reconstructions by other authors.[1,6,17] Limitations of the study include a relatively small sample size and a short follow-up period.

In conclusion, the present study indicates, that bone cement is a valuable and cost-effective alternative to ossiculoplasty with pre-formed prosthesis for reconstruction of the ossicular chain in patients with smaller crus longum defects. However, extended observation times are warranted to safely evaluate the stability of the re-sculptured incudostapedial joint and to ensure the long-term benefit from this approach.

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DILEMMA IN THE MANAGEMENT OF MASSIVE PETROUS APEX CHOLESTEATOMAS

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Background

Congenital cholesteatoma in the petrous apex is an insidious pathology that often causes sensorineural hearing loss, vertigo, and/or facial nerve paralysis. Although complete removal of the pathology is the goal of treatment, this is sometimes difficult to achieve because of its massive extension and/or anatomical complexity of the petrous apex. In this paper, dilemmas were discussed in managing the massive type of petrous apex cholesteatomas (PAC).

Case report

A 45-year-old female, who had undergone the transpetrosal (middle cranial fossa) surgery for removal of extensive PAC in another hospital seven years before, was referred to our hospital aiming a further surgery for its regrowth. She had already presented with total deafness in the left ear and with the ipsilateral House-Brackmann (H-B) grade-III facial paralysis. High-resolution CT (HRCT) and magnetic-resonance images (MRI) revealed that a huge cystic lesion in the petrous apex, devastating cochlea and labyrinth, extended to involve the internal auditory meatus (IAM) and the internal carotid artery (ICA) completely (Fig. 1). Because she desired to maintain the current facial function because of her occupation, complete removal was first attempted via the transotic approach. After extended mastoidectomy and cochleotomy, the matrices of cholesteatoma were almost removed from the ICA. However, the dura and contents of the IAM could not be identified in the cholesteatoma, and therefore the exteriorization of the lesion was finally carried out (Fig. 2). The facial nerve was left in its original position, and the post-operative grade of facial palsy was thereby unchanged.

Discussion

Congenital cholesteatoma in the petrous apex insidiously grows and sometimes extends to devastate the cochlea and labyrinth and to involve the facial nerve and ICA (Moffat-Smith's massive labyrinthine-apical,[1] Sanna's massive complex[2,3]). Complete removal of cholesteatoma, regardless its size and location, is always a goal of the treatment, but this type of massive PAC is a hard object to control. A transotic approach[3,4] is mainly selected for the case of PAC with profound deafness aiming preservation of the facial nerve, in which the facial nerve remains in position. On the contrary, the transcochlear approach[2,5] provides better access to the petrous apex by a slight sacrifice of the facial nerve, which needs posterior rerouting or tentative transection followed by re-approximation. The latter approach, however, never results in a facial function beyond grade III.

In the present case, cholesteatoma had already destroyed the cochlea and labyrinth and extended far medially to the sphenoid sinus. Furthermore, the IAM was totally involved in the cholesteatoma mass, and so the

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Edited by Haruo Takahashi

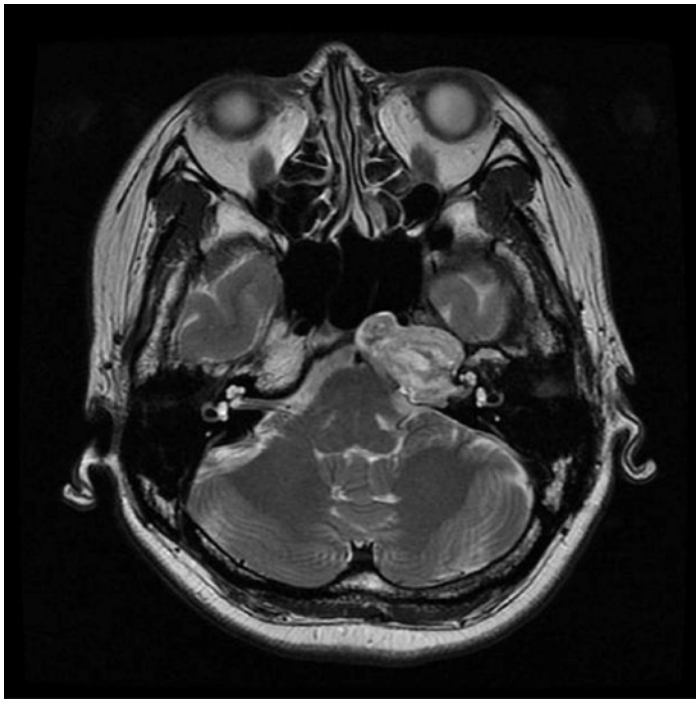

Fig. 1. T2-weighted MRI. A huge cholesteatoma existed in the petrous apex, extending to the sphenoid sinus and involving the internal auditory meatus (IAM) and the internal carotid artery (ICA).

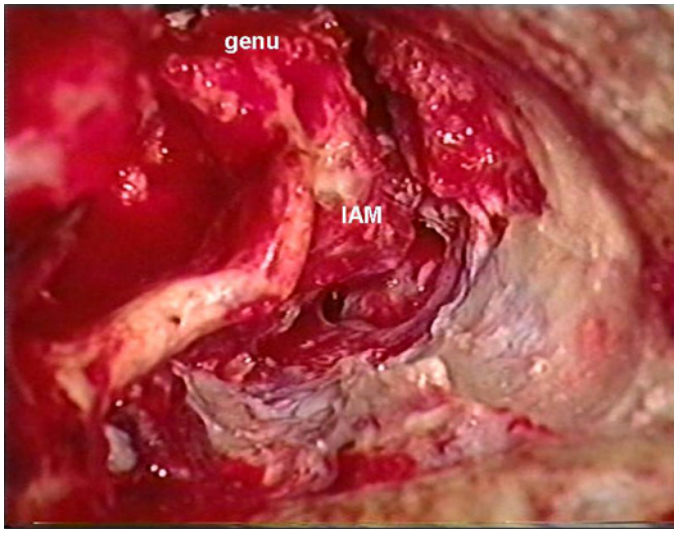

Fig. 2. An intra-operative view after cochleotomy and labyrinthectomy surgery. Transotic approach was selected. The cholesteatoma matrices could not be controlled medially to the IAM.

posterior rerouting of the facial nerve could never be achieved. A fair amount of the cholesteatoma matrices must have been left on the medial wall of the IAM and on the sphenoid sinus wall, even though the transcochlear approach with a tentative transection of the facial nerve was performed. These were the reasons that we believed the exteriorization was the best treatment choice in this case, and that we selected the transotic approach for the facial function surely to be preserved.

Selection of either near-total removal followed by obliteration or exteriorization alone has always been a dilemma in the management of the massive PAC.[6,7] A better solution needs to be pursued in an individual case with PAC.

Conclusion

Massive complex petrous apex cholesteatoma always creates a dilemma and provides us with a surgical challenge. Either transotic or transcochlear approach may have to be selected based on the patient's background and pre-operative facial nerve function.

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RECONSTRUCTION OF THE INCUDOSTAPEDIAL JOINT AFTER REMOVAL OF CHOLESTEATOMA

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Introduction

One of the most common causes of conductive hearing loss (CHL) resulting from chronic inflammation from the middle ear is damaged incudostapedial joint.[1,2] Retraction in the rear quadrants of tympanic membrane, cholesteatoma at the mesotympanum, iatrogenic injuries (very rare) and congenital malformation are direct cases of lack of continuity in the ossicular chain at that part.[3,4] This study presents surgical procedures in ossicular chain reconstruction, especially after the incidence of cholesteatoma.

Material and method

There were 1147 patients with a damaged incudostapedial joint from the World Hearing Center Institute of Physiology and Pathology of Hearing, during the period 2010-2011. In the pre-operative period the conductive component referred to as air-bone gap was in the range of 15-40 dB SPL, measured at 500, 1000, 2000, 4000 Hz. In the group of patients with evident otoscopic features of a damaged incudostapedial joint, but without tympanic membrane perforation or atrophy of the tympanic membrane, in 20% there was no conductive component of hearing loss (tolerance CHL< 10 dB SPL). This was caused by direct adhesion of the tympanic membrane to the suprastructure of the stapes, what ensured the effective transmission of sounds. There were two methods of surgical procedure: 1) Surgical reconstruction of damaged parts of joint with alloplastic material and glass-ionomeric cement (804 surgeries); 2) Reconstruction when there was perforation of the string connecting structure between the tympanic membrane and the inner ear (343 surgeries). Prosthetic ossicular reconstruction procedures (PORP), total ossicular replacement prosthesis (TORP), bone plate, and cartilage flake were used. After removal of cholesteatoma (when present), the surgeon performed restoration of the incus and its connection between the head or the remnant of the suprastructure of the stapes. During surgery, glass-ionomeric cement was prepared. During non-anatomical reconstructions, PORP-columellas were performed with different materials like remnant of incus, glass-ionomeric prosthesis and titan prosthesis. After surgical treatment, the follow-up period was minimum one year with periodical otoscopic and audiologigal control, and functional results were analyzed. Pure-tone audiometry was performed one month, six months, and one year post-operatively. Post-surgical results were obtained by Pure-Tone Audiometry with frequencies 500, 1000, 2000, 4000 Hz.

Intra-operatively there was confirmed damage of the incudostapedial joint:

- 409 (36.5%) – chronic otitis media with retraction of tympanic membrane;
- 388 (34%) – cholesteatoma;

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Edited by Haruo Takahashi

- 63 (5.3%) – injuries of middle ear;
- 34 (3%) – iatrogenic injuries due to otosurgical maneuvers;
- 42 (3.7%) – congenital malformations.

After analysis of patients with the first surgical strategy – reconstruction of incudostapedial joint – among 804 surgeries, sustainable results with reduction of air-bone gap to a level below 5dB SPL were achieved in 97% (786 ears). In nine ears (3%) the hearing result was not sustainable because of disconnection of the connection made with glass-ionomeric cement (eight ears) and extrusion of titan prosthesis (one ear). In many cases it was possible to observe the stapedius muscle reflex after surgery, for example when a reconstruction was performed in which the long process of the incus was connected with the articular disc incudostapedial joint. In nine cases there was re-surgery to reconstruct the connection between incus and stapes. In one case the incus was connected with the perichondrium by myringioplasty.

Of 343 non-anatomical reconstructions, sustainable results were achieved in 237 ears (69%) and no sustainable results were achieved in 106 ears (31%). Lack of good results was caused mainly by dislocation of columella-PORP type in 32 cases (9%) and inability to make a connection with bone plate or cartilage flake in 74 cases (22%). That group of patients underwent resurgery with glass-ionomeric cement (73%) and titan prosthesis (27%) – for example MNP (Kurz). The final results after one year were: the air-bone gap was closed in 91.2% (312 ears); reduction to 10-20 dB SPL in 6.4% (22 ears). Only in 2.4% (8 ears) the results were not satisfactory.

Conclusions

We underline that the surgical issue is to reconstruct the ossicular chain. Sometimes this is not possible, especially in cases of advanced cholesteatoma. It was proven that the ossicular chain could be successfully reconstructed with anatomical reconstruction.[5,6] In our opinion good results not only depend on surgical technique, although this is the most important, but also on other factors like movability of other elements of the ossicular chain and the condition of surrounding tissues and the Eustachian tube in the middle ear. Access to different types of prostheses is also very important. The possibility of adaptation of the prostheses to fit individual patients allows us to expect better results. Developing technologies with various materials allow surgeons to choose adequate materials and techniques to obtain the best results after reconstructive surgery.

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CLINICAL INVESTIGATION OF OSSICULAR ANOMALIES

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Introduction

Ossicular anomalies (OA) are known to cause congenital, non-progressive, and conductive hearing loss. They may occur with a normal tympanic membrane, and many of them are unilateral lesions. Usually, patients are not aware of their hearing loss until testing, because they have no other symptoms. However, we often encounter patients with OA who remember when they first noticed hearing loss.

The purpose of this study was to investigate when the patients first noticed their hearing loss and evaluate the operating methods and post-operative hearing progress of OA treatment.

Materials and methods

Thirty-two patients (35 ears) with OA were diagnosed upon operation in the Tokyo Medical University during the period from 1998 to 2011. There were 15 males and 17 females (age range, six-67 years; mean 26.1 years). The patients were classified according to Funasaka's classification (Fig. 1).[1] This classification divides monofocal OA into three groups. Cases which had more than two types of dysplasia were defined as Multifocal-type anomalies. These cases were also classified into three groups by the time of manifestation: 'infancy', 'enrollment for medical examination' and 'acquired'. In the first two groups, the patients were not aware of their hearing loss. In the last group, they remembered when they lost their hearing.

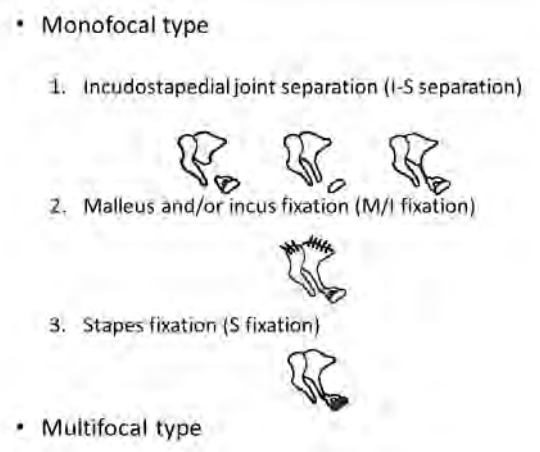

Fig. 1. Funasaka's classification. OA are classified into three groups: I-S separation, M/I fixation, and S fixation. Cases which have more than two types of dysplasia are defined as multifocal type anomalies.

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Edited by Haruo Takahashi

Hearing level was investigated before and after surgery. The air-bone gap (A-B gap) and 2-kHz bone conduction threshold dip (Carhart's notch) were investigated by pure tone audiogram (PTA) before the operation, and the A-B gap was also investigated after the operation.

Operation outcomes were evaluated by the American Academy of Otolaryngology (AAO-HNS) guidelines (1995). In these guidelines, success is defined as when the A-B gap becomes less than 10 dB.

Results

There were 26 ears in the incudostapedial (I-S) joint separation group, one ear in the malleus and/or incus (M/I) fixation group, and four ears in the stapes (S) fixation group (Table 1). Multifocal-type anomalies were seen in the other four ears. There were no differences in OA between the genders. Age was higher in the I-S separation group. There were seven ears with complex anomalies.

Table 1.

		Age	Gender	
			Male	Female
Monofocal (31 ears)	I-S separation (26 ears)	7-67 (mean: 31.2)	12 ears (10 patients)	14 ears (14 patients)
	M/I fixation (1 ear)	6	0	1 ear (1 patients
	S fixation (4 ears)	6-12 (mean: 9.3)	2 ears (2 patients)	2 ears (1 patient)
	Multifocal (4 ears)	6-31 (mean: 15.3)	3 ears (3 patients)	1 ear (1 patient)

The time of manifestation is shown in Table 2. Many of the patients with OA had been diagnosed upon when they first went to school. There were nine ears in the acquired group. Seven out of nine patients in the acquired group belonged to the I-S-separation group. These acquired OA represented 25.7% of all of the OA cases.

Table 2.

	Infancy	Enrollment for medical examination	Acquired	Unknown	Total
I-S separation	13	4	7	2	26
M/I fixation	0	1	0	0	1
S fixation	0	4	0	0	4
Multifocal	1	1	2	0	4
Total (rate %)	14 (40.0)	10 (28.6)	9 (25.7)	2 (5.7)	35 (100.0)

The mean hearing level in each group before and after operation is shown in Figure 2. Before operation, the A-B gap was present across the low-to-high frequencies in the I-S-separation group, and the stiffness curve was seen in the M/I-fixation and S-fixation groups. In multifocal-type anomalies, these hearing defects were combined. Carhart's notch was observed not only in the S-fixation group but also in the other groups. Ossicular reconstruction was performed in the I-S separation and M/I-fixation groups and stapes surgery was performed in the S-fixation group. In one I-S-separation case, bone putty was used to cement the loose the I-S joint because there was not enough space to insert cartilage between the incus and stapes. After operation, decrease of the A-B gap was attained in all groups, but this decrease was less pronounced in multifocal-type anomalies.

The success rate of the operation was 29.4% in the I-S-separation group, 66.7% in the S fixation group, and 29.2% in all cases (Table 3).

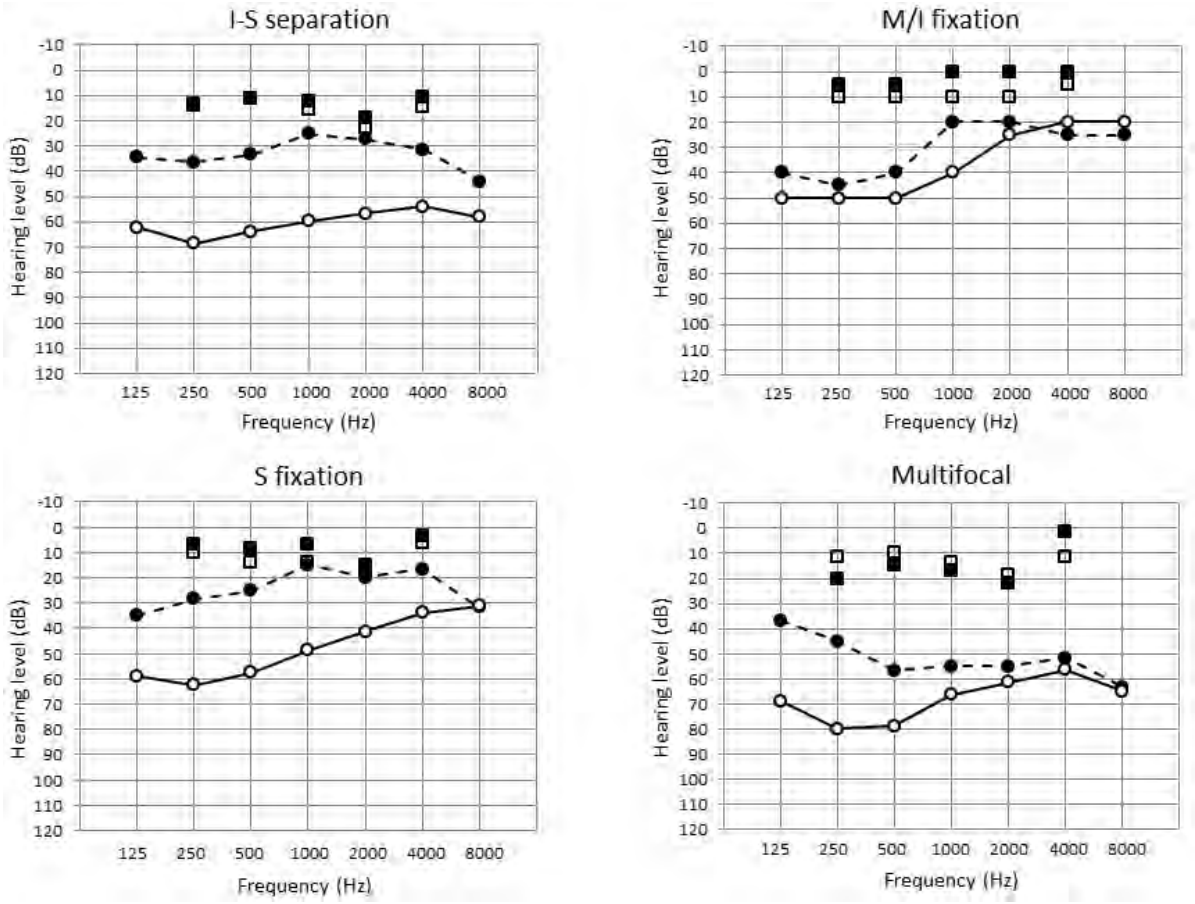

Fig. 2. Pure-tone audiogram. Pre-operative and post-operative hearing levels are shown. Before surgery, an A-B gap was present across the low-to-high frequencies in the I-S separation group, stiffness curve was seen in the M/I fixation and S fixation groups, and both were combined in multifocal type anomalies. In all groups, the A-B gap had improved after surgery, but there was less improvement in Multifocal-type anomalies. White dots: before surgery, black dots: after surgery, round dots: air conduction, square dots: bone conduction.

Table 3.

	A-B gap	I-S separation (17 ears)	M/I fixation (1 ear)	S fixation (3 ears)	Multifocal (3 ears)	total (24 ears)
1)	0 – 10dB	5	0	2	26	7
2)	11 – 20dB	9	0	1	1	10
3)	21 – 30dB	2	1	0	4	5
4)	> 30dB	1	0	0	4	2
Success rate (%)		29.4	0.0	66.7	0.0	29.2

Discussion

Acquired manifestation of hearing loss was seen in 25.7% of patients with OA, and most of these patients were in the I-S-separation group. Therefore, the age was higher in the I-S-separation group. There are two hypotheses regarding the etiology of acquired OA. The first proposes an acquired defect of the ossicular chain, which is thought to be due to the disappearance of existing cholesteatoma[2] or some atrophic changes.[3]. The second hypothesis proposes that acquired OA are due to congenital anomalies and associated acquired pathology. The build up of stiffness of the ossicular chain is involved in this acquired pathology. It was reported that the Valsalva maneuver had been performed to improve hearing in a patient with a loose I-S joint. In this

case, the Valsalva maneuver enhanced the stiffness of the ossicular chain, but the patient started complaining about hearing loss after the Valsalva maneuver became ineffective.[4]

Considering the A-B-gap pattern during PTA measurement, when the A-B gap is present across the low-to-high frequencies, it raises suspicion of I-S-separation-type anomaly. If a stiffness curve can be seen, M/I-fixation- or S-fixation-type anomalies are suspected. In the current study, it was found that Carhart's notch was observed not only in S-fixation-type anomalies but also in other types of OA. It was reported previously that Carhart's notch was not specific for S fixation, and it also could be seen in various conditions of the middle ear.[5] Therefore, the lesions of OA can be inferred from the A-B-gap pattern during PTA analysis.

Surgery was performed with conventional methods in most types of OA. When a conventional operation cannot be performed because of the shape of the ossicular chain, we should consider other methods. For I-S separation, cartilage is useful to bridge and raise the stiffness of the ossicular chain. When there is not enough space to insert cartilage, bone putty can be used as cement instead. To improve hearing, the stiffness of the ossicular chain should be increased, and the ossicles should be kept away from the wall of the tympanic cavity.

Post operatively, complete closure of the A-B gap in OA is difficult, except for S fixation. In the current study, the success rate in the I-S-separation group seemed to be slightly lower than past reports,[6-8] although each criterion used for evaluation is not the same in every report. To improve the success rate for I-S separation, division of new surgical techniques may be necessary.

Conclusion

Acquired OA mainly consists of I-S-separation-type anomalies. The type of OA can be suggested from the A-B-gap pattern of the PTA analysis. Bone putty is useful to cement loose I-S joints.

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HOW WE CORRECT TUBAL DYSFUNCTION

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The Eustachian tube has three functions (aerodynamic-ventilation, evacuation, protection), which depend on anatomical structure, heredity, normal innervation (sympatic, parasympatic, vegetative system, which carry out swallowing and secretion), muco-ciliaris system of mucosa, tubal muscles function, nose and paranasales sinusis. In the beginning of the XIX century many methods of treatment have been described, especially in monographs.[2-4,7-14,18-20,25,44,45,49,50,52,54,55,56]

Over the last 12 years, depending on the causes (inflammation or anatomical character) leading scientists of the world improved techniques as therapeutic methods[2,4,5,6,8, 9,11, 15-19,26,, 29-35, 41-44,51,55,56] and surgical treatment of the Eustachian tube dysfunction (ETD).[8,9,15,20,37-40,41,46,47,48,52,55,56]

According to Bluestone C., many factors are etiology of Eustachian tube dysfunction (ETD) that is why needing complex methods of treatment its diseases.[4,5,6,7,8]

Ours experience more 30 years of the treatment ETD with different etiology elaborate two groups methods: therapeutic complex management and chirurgical treatment.

The first applies for all with ETD in spite of causes (especially inflammation and allergy, upper airways diseases):

- Cleaning of the nasale cavity by endonasal irrigation therapy (eliminating therapy,nasal wash, nasal douche): daily endonasal lavage with assistance of a special device – aqua maris rinsingand with isotonic or hypertonic sea solution (Aqua Maris, Humer and a.o.) during an extended period (three times daily) to remove secretions, congested nose, dust, allergens, harmful substances, biofilms.
- After nasal douch (5-10minutes) local endonasal spray corticosteroid (mometasone furoate or fluticasone furoat) two times daily during 10-12 days in acute process, and one month in the chronic inflammation and 2-4 month in the allergy.
- Antibiotic therapy (macrolide, amoclave, phtorchinolone III generation), antiinflammation therapy (non-steroid-antiinflammation medicines), antileukotriens (montelukast).[30,31]
- Immunocorrection (phyto-engineering medicines-sinupret, imupret, immunotherapy).
- Combined reflexotherapy (acupuncture) according to classic methods (acupuncture, auriculopuncture, vibropressures, vibroacupuncture, electropuncture).

Innervation ET (based on literature, experimental data) shows a close relationship with ET, middle ear cleft and upper respiratory ways (especially ganglion oticom, pl.tympaniy, chorda tympany, pl.pharyngei, baroreceptors).[1,5,7,9,10,16,17,20,26,32,33,45,49,50,53,54,55]

Disorder in balance sympatho-parasympatic systems leads to edema, hyperproduction of the secretion by secretory cells and glands of the mucous membrane of the ET, which leads to disruption of the mucociliary system and consequently to ETD. In addition, disorder of motor innervation MTVP,MLVP and other muscles in the region pharyngeal orifice(n.mandibularis,nn.palatini) disrupts the normal opening act of the ET.

- Therefore, since the 80-ty of last century, we in the most cases of the ETD use periodic sessions of acupuncture in different ways. Combined reflexotherapy (CRT):acupuncture with vibro- or laseropuncture with

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Edited by Haruo Takahashi

auriculomicropuncture (for adult), or electropunctrure with vibropressure (for children),treatment consist with 10-15 sessions. Cycles of the reflexotherapy repeated in 3 and 6 months period. Ours experiences have been confirming, that CRT further of rehabilitation of sympato-parasympatic innevation ET and nasal cavitys and activisated mucociliaris transport in the paranasal sinusis and ET, acts swallowing and open pharyngeal orifice of ET.[29,30,32,33]

- Treatment by hyperbarooxygenisation in the special devices-barocamera, (baroreceptors-stimulation acts swallowing of ET and intratympanic oxygenation). At the beginning XX century, especially in the 40-50 years airmen trained in special chambers to changes in barometric pressure, and it was proved that this way you can activated open of the ET in healthy persones. A with 70-ty years XX century of clinical practice in the cardiology used in therapeutic séance of the Hyperbarooxygenization (HBO). Since the 80-ty years XX century we use treatment by HBO in acute and chronic sensorineural hearing loss, and also at ETD (late and middle degree dysfunction), but after each seances (warning of barootitis) –otomicroscopic control. As shown by ours experience of HBO- treatment not only restore intratympanic pressure and partial pressure of gases, but reduce swelling of the nasal mucosa and ET.[25,37]
- Endonasale ultra-sonopuls-aerosoltherapy whith mucolitics and local corticosteroids[11,12,13,14,16,17,18,21,30,32,34,35,36,43,44,45,46];
- Special exercises-kinesitherapy for tubal muscles by Riu R. and Jacobs A.[3,21,22,23,24,26,27,28,41,51,53], and improve by us (special exercise for ET, pharynx, palate, neck, tongue muscles) and mechanical and vibromassage tubal pharyngeal orifice with special devices.[23,24,26-30]

The second group individual allocation according from causes:

- Complex antiallergy therapy: immunotherapy, spray azelastine(allergodyl) and local (endonasale) corticosteroid (mometazone or fluticasone furoat), antihistamine medecines (levoceterizine, dezloratadine).
- Chirurgical treatment (FESS with rhinoseptoplastic and optical adenotomy particular area of tube pharyngeal orifice and fossa rosenmulleri, polipotomy and vasotomy).[38,39,40]

Materials and results

Functional state of the Eustachian tube had been ivestigating by impedance tympanometry in dynamic (swallowing- test Valsalva,-test Toynbee, – swallowing with drink of water), pneumorhinomanometry, rhinoendoscopie and epipharyngoscopie particulary pharyngeal orifice of the ET (ear salpingoscopie), otomicroscopie (atelectasis of tympan:p.tensa by Sade J.,p.flacida by Tos M.) Grade of ETD determined by C.Bluestone[5-8]: late(1).middle(2),severe(3);acute or chronic.

During 2006-2011years we had been examining 756 patients with different causes: allergic persistence rhinosinusitis(APRS)-105 (ETD-100%), acute rhinosinusitis(ARS)-156 (ETD-97%), ch. rhinosinusitis(CRS)-215 (ETD-98%), ch.rhinosinusitis with polyps(CRSwP)-150 (ETD-100%), deformation septum nasale(DSN)-110 (ETD-78%), ch.adenoiditis II-IIst.(CAT)-64 (ETD-100%).Atelectasis of Middle Ear has been connecting with ETD. Treatment of EDT we are choiced individually for every cases.

After treatment: we are received improving of the functional state of ET in: APRS group-78%, ARS-88%, CRS-56%, CRSwP-46%, DSN-77%, CAT-98%,but without result of the atelectasis of middle ear.

Conclusion

Improvement of the Eustachian Tube function have been receiving only in 46-98% cases in dependence of duration diseases and etiology.

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LOCALIZATION AND DETECTION OF NITRIC OXIDE SYNTHASE ISOFORMS IN HUMAN MIDDLE-EAR CHOLESTEATOMAS: POSSIBLE RELATION TO NITRIC OXIDE PRODUCTION AND INFLAMMATORY BONE ABSORPTION ACTIVITIES

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Introduction

Middle-ear cholesteatoma histologically consists of an epithelial matrix that produces the accumulation of keratin debris and underlying connective tissue layers.[1] The aggressive absorption process of the bone structure adjacent to the matrix is one of the most distinct clinical findings of middle-ear cholesteatoma.[2] We have recently shown that a part of the ligand-receptor system and the cytokine network including the receptor activator of NF-KappaB ligand (RANKL), IL-6, and TGF-beta are deeply involved in the mechanisms through the inflammatory manifestation and bone absorption activities.

In the present study, we focused on possible roles of nitric oxide (NO) in the middle-ear inflammation triggered by cholesteatoma proliferation. Nitric oxide plays a variety of roles in the human upper respiratory airways in relation to airway defense mechanisms, as well as being an inflammatory mediator.[3] We examined the immunohistochemical localization and mRNA expression of RANKL and nitric oxide synthase (NOS) isoforms to understand possible relationship between inflammatory manifestation and bone absorption activities using cell-culture methods.

Methods

Patients' enrollment and tissue preparation

Cholesteatoma tissues were collected from adult patients who had undergone initial surgery for acquired cholesteatoma at the Hiroshima University Hospital.

The degrees of immunohistological expression of inducible NOS (iNOS), endothelial NOS (eNOS), RANKL and Ki-67, a proliferation marker were semi-quantitatively assessed using in vivo pathological specimens. The LSAB™2 system (Dako) was employed as the secondary antibody. The stained cells were observed with a Nikon light microscope and digital photomicrographs were taken for analysis.

The mRNA expression of three NOS isoforms, RANKL, and TGF-beta was quantitatively analyzed by real-time RT-PCR. Cellular RNA was isolated using RNeasy mini kits (Qiagen). For cDNA synthesis, total RNA was reverse-transcribed to cDNA using a High Capacity RNA-to-cDNA kit (Applied Biosystems). Gene expression was measured on a 7300 real-time PCR system using TaqMan Gene Expression Assays. The mRNA levels for targeted genes were normalized to the value of GAPDH by calculating the change in Ct ($^{\Delta}$Ct) for each sample.

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Edited by Haruo Takahashi

Cell culture of cholesteatoma cells

We have successfully established a cell culture of cholesteatoma epithelial cells by elimination of fibroblasts contamination using the trypsinization technique and employment of a defined serum-free medium. Briefly, the cholesteatoma epithelium was dissected from the underlying submucosal tissue, and was then incubated in 0.1% collagenase at 37°C for about one hour. Following cell dissociation and centrifugation, the resulting cell pellet was resuspended in a culture medium and seeded onto 75 mm^2 cell-culture flasks precoated with type-I collagen (Asahi Techno Glass, Tokyo, Japan). The cell culture was maintained in Defined Keratinocyte-Serum-Free Medium (Invitrogen, Tokyo, Japan) with growth supplement and antibiotics. Upon reaching cell confluence, a secondary culture of cholesteatoma cells was performed. The epithelial nature of the cells was confirmed by phase contrast microscopy. In order to standardize for both the period and nature of the cultures, the third generation of cholesteatoma cell cultures from each subject was used.

Results

Immunohistological findings

A positive expression of RANKL was predominantly detected mainly in the basal and para-basal layers of the matrix accompanying severe inflammatory cell infiltration. The Ki-67 LI significantly increased in the group with higher RANKL expression. We also examined the immunohistological localization of iNOS and eNOS in the cholesteatoma matrix. In a patient with minimal inflammation, a positive iNOS immunoreactivity was clearly observed in the cholesteatoma epithelial cells, mainly in their cytoplasm. A weak expression of eNOS isoform was also detected in the matrix. On the other hand, a higher iNOS immunoreactivity tended to be observed in the matrix accompanying severe inflammatory cell infiltration in the submucosal layer.

Real-time RT-PCR analysis of cultured cells

The use of defined Keratinocyte-SFM and collagen-coated dishes has demonstrated superior cell growth of primary cholesteatoma with maintaining epithelial morphology and biological markers. A positive expression of RANKL was detected mainly in the perinuclear cytoplasm of most cultured cells. Ki-67 antigen was also found to be positive in the nuclei of the cultured cells in various degrees. A real-time PCR analysis indicated that the cultured cholesteatoma cells showed distinct expression of TGF-beta and RANKL mRNA. Relatively low, but constitutive levels of NOS isoform mRNA were also noted. Among the three NOS isoforms, iNOS showed higher levels compared to the other two isoforms.

Discussion

Several theories have been proposed to explain the destructive properties of middle-ear cholesteatoma. A higher population of the osteoclast progenitor cell lineage and macrophages was immunohistochemically demonstrated in the cholesteatoma tissue compared with normal auditory canal skin.[4] The balance between osteoblasts and osteoclasts plays a major role in bone formation and resorption. As shown in the present study, the combination of RANK, RANKL, and osteoprotegrin (OPG) is considered to be deeply involved in the disease mechanisms. These results suggest that epithelial proliferative activities and inflammatory manifestation could be possible factors responsible for the markedly increased bone resorption observed in cholesteatoma patients. It has been considered that the activity of cholesteatoma proliferation is greatly affected by difference in micro-environment. Because we found a tendency for the expression of Ki-67 antigen to be augmented in severe inflammatory conditions, we speculate that these factors which often coincide as a result of bacterial inflammation may act to regulate cell proliferation in the cholesteatoma epithelium through paracrine fashion.

In the present study, we hypothesize that NO production in the middle ear has a possible role in some of the clinical features of cholesteatoma such as epithelial hyperproliferation and bone resorption, as is the case for IL-6 and TGF-beta.[5] NO is produced in human respiratory tract broadly by activation of NOS. We found that human cholesteatoma epithelial cells also express two different NOS isoforms. In addition, iNOS immunoreactivity tends to be augmented in the area accompanying increased RANKL expression and severe

inflammatory cell infiltration. RT-PCR analysis of cultured cholesteatoma cells has also shown constitutive iNOS and eNOS expression. Further elucidation of NO synthesis and regulatory mechanisms in the middle ear may imply valuable approaches to assess underlying inflammatory status as well as new treatment modalities to healing processes with restoration of mucociliary clearance abilities.

Acknowledgement

This study was supported in part by Grants-in-Aid for Scientific Research, administered by the Ministry of Education, Science, Sports and Culture, Japan.

The study protocol was approved by the Institutional Review Board at the Hiroshima University School of Medicine.

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EXPERIENCE WITH THE VIBRANT SOUND BRIDGE IMPLANT, INDICATIONS AND RESULTS

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Introduction

Conventional hearing aids are not sufficiently satisfactory or are not well tolerated by a significant number of patients with severe sensorineural or mixed hearing loss[1] because of a chronic, otological pathology in the middle and external ear. Active middle-ear implants (AMEI) could be an excellent option in these cases. The Vibrant Sound Bridge (VSB) is the most extended AMEI system of the last decade.[2-4] This device processes and amplifies sound stimuli through an electrical-magnetic transducer called the Floating Mass Transducer (FMT), which transfers sound vibrations to the cochlea when applied to an element of the tympanic ossicular chain or in the proximity of the windows.

In the last few years, several authors[5,6] have published studies of patients in whom the FMT of the VSB has been placed on the round window (RW) membrane. They have assumed that the impact of this vibration on the basilar membrane is similar to air conduction through the ossicular chain to the oval window.[7-9]

Initially, there are several advantages in placing the FMT on the RW. On this location, the external and middle ear are bypassed. Because of this, for example, this device may be used in ears with sequelae of chronic otitis media (COM), where the function has not been successfully restored. The indication for ears treated with open techniques is especially remarkable since the device can work without air spaces in the middle ear. The objective of the study is to analyze results obtained with the VSB in a group of patients in whom the original purpose was to place the FMT in the RW.

Materials and methods

Population

There has been a retrospective study (from 2007 to present) on the indications and results obtained by individuals implanted at our center with the VSB. Table 1 shows demographic data of the patients studied and the primary pathology on their implanted ear. The follow-up period ranged from five to 64 months (an average of 41.2 months). Eight patients had a history of simple chronic otitis media (COM), four patients had cholesteatomatous COM and one patient suffered from acquired stenosis in the external auditory canal (EAC), associated with tympanosclerosis.

Description of the surgery

In six patients, the implant was placed by means of an already existing radical mastoidectomy. A simple mastoidectomy and a posterior tympanotomy to expose the RW were carried out on the remaining seven patients. To correctly place the FMT on the membrane of the window, the titanium clip designed to place the FMT on the ossicular chain was cut off.

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Edited by Haruo Takahashi

Table 1. Demographic data of the patients included in the study.

Patient	Gender	Age	Date of Implantation	Previous Pathology	Implanted Ear	Previous Surgeries
1	F	53	25.01.2007	Cholesteatomatous COM	Right	Open technique
2	M	59	15.03.2007	Simple COM	Right	Open technique
3	F	55	12.09.2007	Simple COM	Right	Open technique
4	F	31	08.11.2007	External Otitis and stenosis of the EAC	Right	Closed technique
5	M	66	22.11.2007	Simple COM	Left	Closed technique
6	M	59	07.02.2008	Cholesteatomatous COM	Left	Closed technique
7	M	58	21.02.2008	Simple COM	Right	Open technique
8	F	83	26.03.2009	Simple COM	Left	Closed technique
9	F	68	13.08.2009	Simple COM	Left	Closed technique
10	M	47	09.12.2009	Cholesteatomatous COM	Left	Open technique
11	M	55	04.03.2010	Cholesteatomatous COM	Left	Closed technique
12	M	45	28.04.2010	Simple COM	Left	Closed technique
13	M	60	17.11.2011	Simple COM	Right	Closed technique

The edges of the RW were carved until the RW membrane was exposed completely. Afterwards, the *fossula ante fenestram* was shaped to accommodate the FMT in front of the RW membrane. The FMT connection cable was arranged on the walls of the mastoid cavity or the inferior wall of the EAC. The cable and the FMT were handled with non-ferromagnetic microsurgical material. A sheet of fascia temporalis was placed between the FMT and the RW membrane. The correct contact was checked and secured with a wedge of cartilage placed between the FMT lower end and the bone recess drilled in the *fossula ante fenestram*. Moreover, a graft of fascia was used to insulate the *fossula ante fenestram* from the remaining middle ear spaces.

In patient #3, the FMT was placed onto the oval window, since the surgery revealed that the round window was ossified.

A canal was drilled in the bone with a 1.5 mm diamond burr to lodge the device's cable in patients with a history of radical mastoidectomy. The cable's passage through the middle ear spaces was protected with cartilage sheets. The canal was drilled through the facial wall in the first patients. Afterwards, the canal was drilled through the inferior wall of the EAC, at the level of the tympanic bone, following the trajectory towards the tip of the mastoid. For patients undergoing a closed mastoidectomy, the cable was secured with temporal muscle grafts in the mastoid cavity.

Assessments pre- and post-operation

All patients underwent a pre-operatory otological study, including a micro-otoscopy, CT of petrous bones, air and bone-conduction (AC and BC respectively) pure-tone audiometry, and speech-recognition test with bisyllabic words.

After surgery, pure-tone air-conduction thresholds with headsets were tested, without the VSB external component. Pure-tone and speech audiometries using bisyllabic words were performed in free field with the VSB disconnected and connected. The unaided ear was masked using a TDH 39 headset with a NB sound at 70dB SPL. All tests were performed in a soundproof booth.

The VSB audioprocessor (Siemens Signia) has eight frequency bands. It can provide a maximum gain of 70 dB and a maximum output of 113 dB. The initial adjustment was made with the CONNEXX software, referenced to pre-surgery BC thresholds. In subsequent checkups, the gain increased progressively in all eight frequencies up to a comfortable level for the patient. The optimal performance of the system was usually reached three to four months after the first activation.

Statistical procedure

To compare results before and after the surgery, a Student T for related samples has been carried out, using the SPSS 20.0 computer program.

Results

Auditory results

Figure 1 shows the average values of AC and BC thresholds recorded for all patients before and after the surgery. In this case, it illustrates AC thresholds obtained with the VSB post-activation.

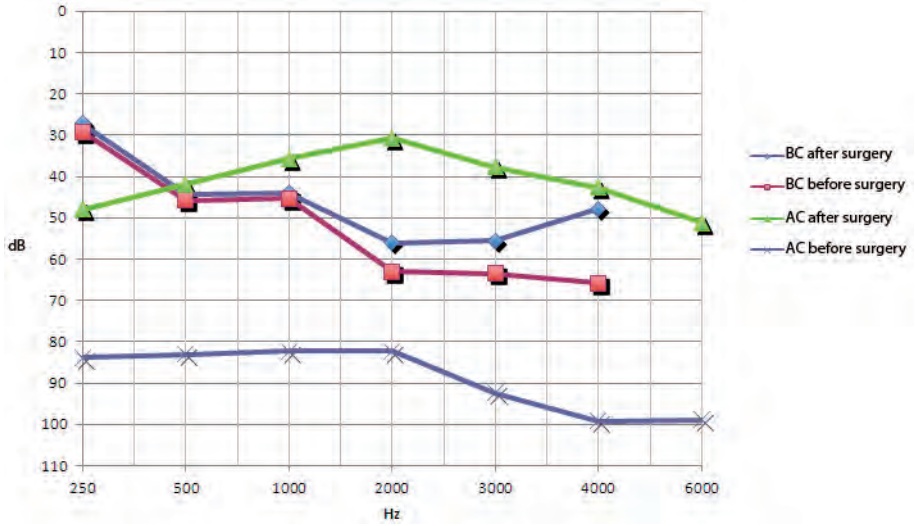

Fig. 1. Mean BC and AC thresholds before and after VSB.

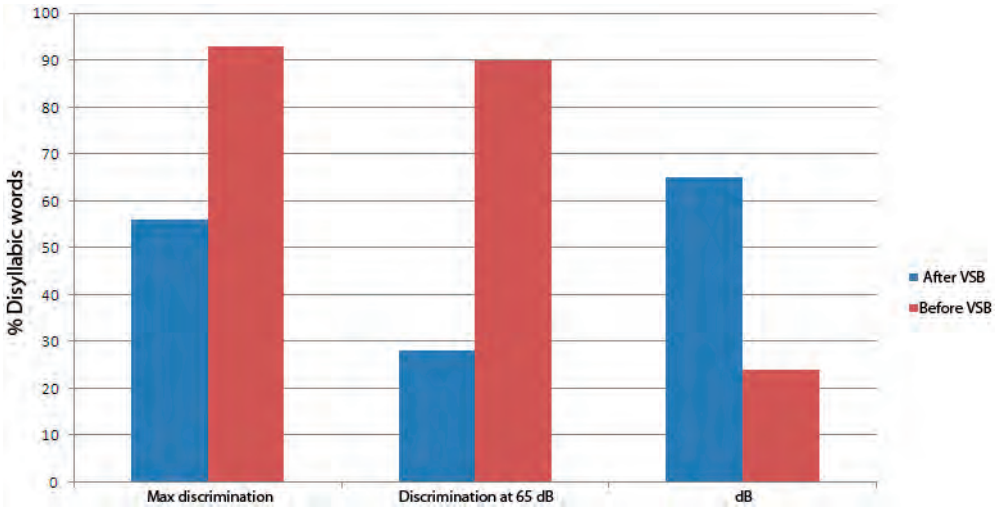

Fig. 2. Mean maximum speech recognition score, speech recognition score and speech detection threshold before and after VSB.

A statistically significant gain ($p < 0.001$) was observed in average tone thresholds of all frequencies studied (0.5-6 kHz) when comparing AC before and after activating the VSB. This gain was more evident in 1, 2 and 4 kHz frequencies. The average auditory gain (Preop AC vs VSB AC) in the frequency range 0.5-6 kHz was 44.07 dB.

All patients had sensorineural hearing loss to some extent. The average AC audiometric performance after activating the device is significantly better than the pre-operatory BC performance in ten out of 13 patients studied. The average gain was 11.3 dB ($p = 0.005$).

When comparing the pre- and post-surgery BC, no statistically significant differences ($p = 0.201$) have been found.

The speech understanding performance with and without the VSB using bisyllabic words is represented in Figure 2. The detection threshold was significaly reduced ($p = 0.012$). The maximum speech recognition score, and the speech recognition score with 65 dB SPL also showed a significant improvement ($p = 0,024$ and $p = 0,004$ respectively) when using the VSB.

Complications

In patient #7, the implant cable was partially exposed in the mastoidectomy cavity two years after implantation. It was covered with free skin graft under local anesthesia. After one year, the cable became exposed again, and, once again, it was covered with cartilage graft. The performance of the device was stable at all times. Patient #8 required a surgical check up under local anesthesia twice (five and 13 months after the operation, respectively). An exploratory tympanotomy revealed the FMT was moved so contact with the RW was not optimal. It was correctly adjusted. The device's performance immediately improved, and this gain was sustained over time.

Patient #6's device failed twice. The metallic cable connecting the FMT to the processor was broken in the second time, although the silicon sheath was intact. The device was explanted, and with the new implantation, the auditory gain of the patient is now similar to the gain acquired with the device before it failed.

Patient #5 did not obtain a satisfactory performance from his device, despite the fact that it was working correctly. The audiological profile included profound hearing loss in the 2, 3 and 4 KHz frequencies and the inability to recognize words. The VSB performance was somewhat beneficial, and the air conduction thresholds reached 70 dB in the free-field-tone audiometry. However, the device did not improve his performance in the speech recognition test. Figures 3 and 4 shows the audiometric results of this patient in the affected ear. Finally, the team decided to place a cochlear implant in the left ear. The results obtained were excellent, as illustrated by the same figures.

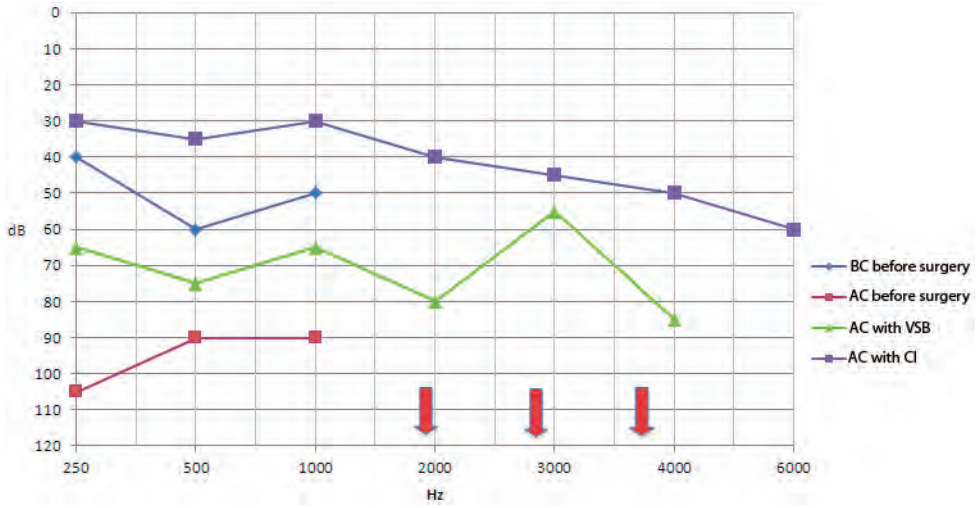

Fig. 3. AC and BC threshold of patient # 5 before BSB, AC threshold with VSB and AC threshold with CI.

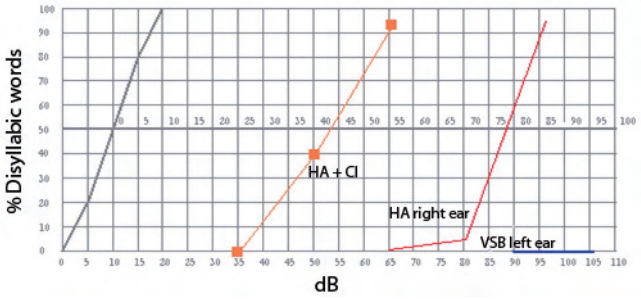

Fig. 4. Speech recognition score of patient # 5 with VSB, with the CI, and with the hearing aid in the contralateral ear.

Discussion

Patients included in this study showed medical or audiological contra-indications to the use of conventional hearing aids or bone-anchored hearing aids. Most patients suffered from severe to profound hearing losses,

some even had hearing loss patterns in some frequencies beyond the recommended criteria set by the VSB implant manufacturing company.

It is important to point out that this study has evidenced an improvement of the audiometric performance beyond expectations, since the air-bone gap was closed in ten out of 13 cases. Other bone-integrated or bone-conduction implants do not replicate this improvement. This may be due to the location of the FMT on the round window, which can convey vibration directly to the fluids of the inner ear. Likewise, the VSB processor may provide additional sound amplification.

This gain above the air-bone gap varied among patients. Several factors may have an impact, such as the anatomy of the round window, the degree of cochlear damage, the stapes mobility or the coupling of the FMT onto the RW. It is obvious that if the total surface of the FMT is not in contact with the RW membrane, the intensity of the sound transmission will be worse. The fixation of the stapes has been linked to poorer audiological results.[6,9]

The audiological results are highly positive. Patients reported a high level of satisfaction in spoken word comprehension. In the group of patients with severe to profound mixed hearing losses, results have been very rewarding, since the evolution of these subjects had not been satisfactory with other treatments (tympanoplasty, hearing aids) and they were rescued by the VSB implantation.

The final audiological results depend on several factors, among others, previously existing cochlear function, the frequency-response curves and the programming of the processor.

Regarding the possible appearance of an acoustic trauma or direct damage to the cochlea, no significant differences have been detected in BC thresholds pre- and post-VSB to date in any of the subjects studied.

The surgery to place a VSB implant on the RW has a medium-high difficulty, although results obtained are on par with the challenge, as already mentioned. There are several characteristics, based on the experience acquired with this group of patients worth mentioning:

- By placing the FMT on the round window membrane, you can use it in ears after a radical mastoidectomy. Therefore, ossicular residues or ventilation of the middle ear spaces are not necessary for its proper functioning. Because of these characteristics, it is very useful for patients with severe sequelae in the context of COM due to tubal dysfunctions.
- Seeking to prevent the exposure of the connecting cable in the mastoidectomy cavities, it is recommended to drill a channel in the inferior area of the pathway, at the level of the tympanic bone, not on the facial wall. Once the cable is placed, it will be covered with bone paté and cartilage. This solution has reported excellent results, and it contributes to reduce the risk of materials fatigue that end up breaking the connecting cable.
- In case the FMT is displaced, it is possible and recommendable to do the check-up surgery under local anesthesia. Aside from being a well-tolerated procedure, it provides immediate intra-operative feedback on the correct positioning of the FMT, by activating the implant during the surgery and gathering the impressions of the patient.

Conclusions

The surgery to implant the VSB requires experience and skill. The FMT must be perfectly coupled onto the RW membrane to attain an optimal performance of the device.

This study suggests the VSB middle-ear implant provides a satisfactory audiological gain in high frequencies to patients with moderate to severe mixed hearing losses. Patients attained greater comfort than with conventional hearing aids.

The VSB is considered to be an appropriate device to be used by patients with BC losses up to 70 dB (a threshold beyond the manufacturing company's recommendation). There must be present hearing levels in all frequencies between 0.5 and 4 KHz, and the maximum speech recognition score must be beyond 60% in the ear chosen for implantation.

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VIBRANT SOUNDBRIDGE MIDDLE-EAR IMPLANTS – LONG-TERM OUTCOMES

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Introduction

Middle-ear implant (MEI) is an option for patients with sensorineural, conductive or mixed hearing loss who are unable to tolerate a conventional hearing aid for hearing rehabilitation. The Vibrant Soundbridge® is a semi-implantable MEI which has been available since 1996. We have used the Medel Vibrant Soundbridge® (VSB) middle-ear implant since 1997 and we report on the long-term outcome.

Method

Retrospective review of 14 patients who had this device fitted between 1997 and 2002.

Results

The etiology of hearing loss was: one sudden idiopathic, eight unknown progressive, four hereditary progressive and one iatrogenic from chemotherapy. There were equal numbers of male and female patients. The mean age at implantation was 46 (28-76 years).

Five of the patients had complications. Two patients needed their transducer re-fixing. One patient had persistent otitis media with effusion. One had wound breakdown and needed revision surgery. The last patient had aural fullness and electrical interference which resolved spontaneously.

At the time of this study, eight patients (57%) were still using their devices. Of the patients who became non-users, two suffered early post-operative hearing deterioration and were early unsuccessful users. Three became non-users due to progressive hearing loss, with one progressing to a cochlear implant. The final patient stopped using the device due to a transducer failure.

The median length of VSB usage was 107 months (range 0-158). Even amongst the current non-users, the average length of time the VS was successfully used was 50 months (0-94 months). Eleven patients had had problems with their devices, with eight needing the external processors changed. However, most patients carried on using their devices after the problems were fixed. As previously noted, only one patient became a non-user though persistent device failure.

Conclusions

The VSB is a viable long-term hearing rehabilitation option for carefully selected patients. Some patients may become non-users through the natural deterioration of their hearing reserve. Notwithstanding some problems, we have a cohort of eight patients who have successfully used their VSB for over ten years.

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Edited by Haruo Takahashi

IMPLANTABLE HEARING AIDS: CANDIDACY CONSIDERATIONS IN PEDIATRIC CHOLESTEATOMA

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Introduction

Tympanic membrane retraction and cholesteatoma are the commonest causes of permanent conductive hearing loss acquired by children in westernized societies. Surgery frequently fails to restore normal hearing after removal of cholesteatoma, usually because of damage to the ossicular chain. Although usually unilateral, the hearing loss may interfere with schooling and socialization. Parents are frequently concerned about this limitation in their child, however the child or adolescent only rarely seems to express any major concern. Consequently most teenagers decline the offer of rehabilitation of their unilateral conductive loss with hearing aids or an extra surgery for ossiculoplasty.

Personal experience suggests that the factors that deter these young people from seeking rehabilitation include: (1) denial that they have a problem that needs fixing; (2) desire not to look different from their peers (e.g. by wearing a hearing aid); (3) desire to avoid surgery. Implantation of an active hearing aid at the time of surgery for cholesteatoma has the potential to overcome all of these objections. This would allow these individuals to experience the benefits of binaural hearing without having an additional surgery. Totally implantable devices, or those with a small external device hidden within the hair and held in place by transcutaneous magnets (rather than a percutaneous fixture) would overcome the cosmetic concerns of aids worn in and around the external ear.

The differing designs of IHA devices influence potential candidacy. Conventionally most are intended to be placed with an intact ear canal wall and several require attachment to the ossicular chain: both may be absent after cholesteatoma surgery. However, although most currently available IHA are not currently licensed for use in children, and were designed for sensorineural or mixed losses rather than the purely conductive loss typical of most children with cholesteatoma, some children could benefit from these devices. The objective of this study is to determine the proportion of children that would be potential candidates for current IHA according to the configuration of devices currently available.

Method

The presenting features of cholesteatoma, surgical outcomes and hearing levels were recorded prospectively in a consecutive series of over 300 cases over a seven year period. This database was reviewed to determine the status of the canal wall and ossicular chain following completion of cholesteatoma surgery. Hearing threshold at the clinic visit closest to 12 months after the last surgery to the affected ear was recorded. Normal hearing was defined as a four tone average air conduction threshold of <30dB HL. Ears with hearing thresholds above this level were considered potential candidates for rehabilitation. Ears with <12 months follow up after their last surgery were excluded in order to exclude all cases in whom planned, staged surgery had not been

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completed. One case of profound congenital sensorineural hearing loss that had been identified through the department's cochlear implant program was also excluded. 27 cases described as "precholesteatoma" were included in this series and were defined by the presence of increasing depth of retraction under the scutum, annulus or ear drum and presence of granulation tissue and or retained keratin within the retraction that was potentially cleanable without surgery.

Cholesteatoma was removed with an intact canal wall approach when practical using endoscopy, KTP laser and cartilage reconstruction as adjuncts. Second stage surgery was planned when removal of disease was piecemeal or extended posterior to the intact canal wall. Prosthetic ossiculoplasty (TORP) was offered at the second stage of canal wall up surgery in cases of absent stapes superstructure, though was often declined by the individual or parents because of concerns arising from the potentially low success rate and possible requirement for surgical revision in the future. IHA and bone anchored hearing aids were not used in this series.

Results

Of the 283 children in the database, 20 had bilateral cholesteatoma (7% of 303 cases). 26 ears were excluded from analysis (1 profound congenital SNHL; 25 cases <12 months follow up). The mean age at surgery of the 277 cases included was 11 years. Cholesteatoma most commonly arose from retraction of the pars tensa (54%) rather than the pars flaccida (24%). Other derivations of cholesteatoma were: congenital (12%), implantation (7%), ear canal (3%).

Of the 277 cases, an intact canal wall was preserved in 209 cases (75%). Of the 68 canal wall down cases, 23 (8% of all cases) had a single CWD procedure and the remainder were revision surgeries following previous ICW or CWD procedures (22 were referred after previous CWD surgery elsewhere). An ICW approach was completed in 82% of all newly diagnosed cases (209 of 252).

After ICW surgery, the ossicular chain was intact in 36 cases (17%) and the stapes superstructure was intact in 100 (48%) cases. Of those with absent stapes superstructure, 17 had a TORP and 25 an incus interposition ossiculoplasty.

A postoperative audiogram around one year after surgery was available for 218 cases. Overall, 120 (55%) ears had hearing loss (i.e. mean 4 tone average air conduction hearing thresholds >30dB HL): 91 (52%) of the ICW cases and 29 (66%) of CWD cases. Of the ICW cases with hearing loss, only 2% had an intact ossicular chain, and 17% had an intact stapes when the malleus or incus were deficient. Hearing loss persisted after ossiculoplasty for absent stapes superstructure in 42% of TORPs and 79% of incus interpositions.

The proportion of children hypothetically eligible for different designs of IHA is shown in Table 1. The applicability of different device requirements is based on examples of currently available IHA, regardless as

Table 1. Table to show the proportion of ears in which different designs of implantable hearing aid may be appropriate, according to ossicular status.

Example of implantable hearing aid	Totally implantable	Ossicular status requirement			Number of ears (n)	% of ICW (n=90)	% total (n=218)
		Stapes	L.p.i.	Incus body			
Carina (1)	Yes	+	+	+	5	6%	2%
Esteem (2)	Yes	+	-	+	22	24%	10%
Soundbridge to ossicle (3)	No	+	+/-	+/-	36	40%	17%
Soundbridge to round window (3)	No	+/-	+/-	+/-	90	100%	41%
Alpha 1 (4)	No	+/-	+/-	+/-	90	100%	41%

1. Otologics GmbH, Heidelberg, Germany
2. Envoy Medical Corporation, Saint Paul, MN
3. MED-EL Elektromedizinische Geräte GmbH, Innsbruck, Austria
4. Sophono, Inc., Boulder, CO

+ = must be present; - = must be absent; +/- = may be present or absent
L.p.i. = long process of incus
n = number of patients ICW, with hearing loss and appropriate ossicular status
% of ICW = proportion of cases of ICW with hearing loss (4 tone average AC >30dB HL) fulfilling ossicular status requirements
% of all cases = proportion of all cholesteatoma cases having ICW, hearing loss and appropriate ossicular status
Proportions assume bone conduction thresholds fulfil audiometric candidacy (one case of profound SNHL excluded from analysis)

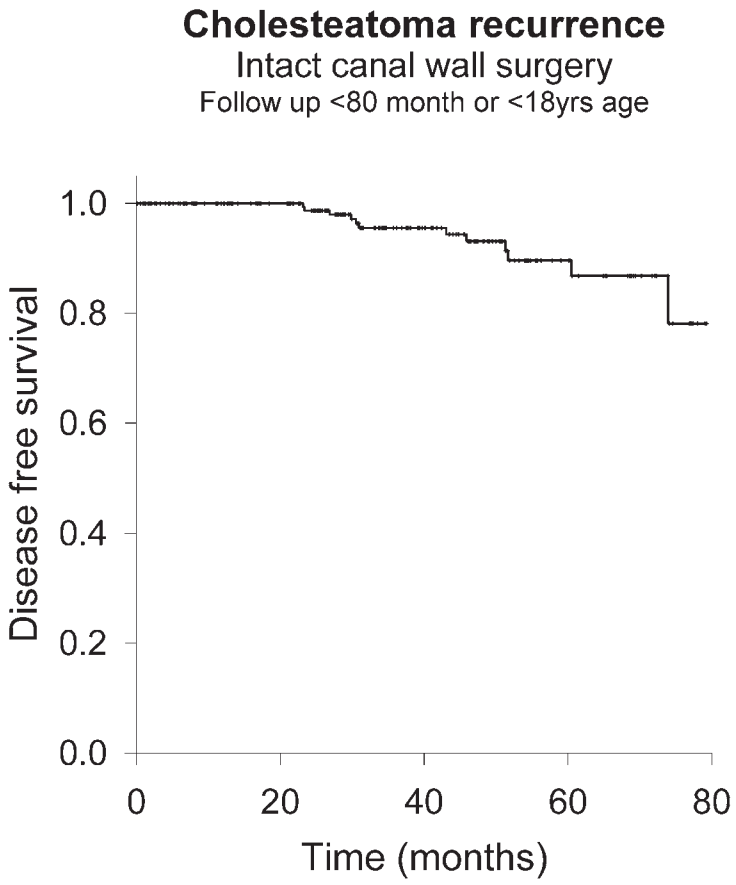

Fig .1. Kaplan Meier survival curve for recurrence of cholesteatoma after intact canal wall surgery. Time in months after first stage of surgery.

to whether they are designed or licensed for use in pediatric conductive hearing loss. It is assumed that an ICW is necessary for most devices, though placement of at least one type of device has been reported with open mastoid cavities.

Overall 25% of all cases were found to have recidivistic cholesteatoma (i.e. recurrent and/or residual disease) with follow up of 12 to 80 months. 107 (51%) ICW cases had a second stage procedure because of identification of recurrent cholesteatoma at follow up or concern about a high risk of residual disease. Recidivism was present in 46% of these cases: 34% had residual disease which was typically a small pearl of cholesteatoma and 22% had recurrent disease, often from retraction around a cartilage scutumplasty. Figure 1 is a Kaplan Meier curve showing the tendency of cholesteatoma to recur at a fairly constant rate for years after surgery. Follow up is not available after age 18 years as care of all cases is transferred to other institutions.

Conclusions

Hearing loss is common after surgery for cholesteatoma in children. Loss of ossicular integrity is the principal cause and this is often not successfully rehabilitated by ossiculoplasty. The high likelihood of hearing loss and good prospect of maintaining the canal wall suggests great potential for benefit from IHA in children. Limitations for pediatric IHA candidacy include:

1. Ossicular status: Good hearing can be anticipated when the stapes is intact; pars tensa cholesteatoma with incus and/or stapes erosion is common. These factors reduce the applicability of IHA devices that require attachment to ossicles.
2. Recidivism: The high risk of recidivism in pediatric cholesteatoma threatens implant survival. While successful removal of residual disease can usually be assured at a second stage of surgery, continued follow

up would be required for recurrent disease that may develop many years later. MRI screening for occult residual disease would be compromised by some devices.

3. Acceptability: Many teenagers do not wish to pursue rehabilitation for moderate unilateral hearing loss. It is conceivable that a totally implantable IHA, or a device with an inconspicuous external component hidden in the hair would prove desirable. Currently available totally implantable devices require ossicular attachment.
4. Expense: The cost utility of IHA for unilateral hearing loss has yet to be established in this population. Provision of binaural hearing at this formative social and educational stage of life may prove to have additional benefits beyond those that these candidates would continue to experience in adult life. The prospect of many decades of benefit will also make QALY measures appear more favorable.

These considerations reduce the number of children with cholesteatoma in whom IHA is appropriate, but there are children (most of them teenagers) with stable outcomes who could potentially benefit from a device that does not require anchorage to the ossicular chain. Ultimately, a long lasting affordable totally implantable hearing aid may prove to be the optimum rehabilitative option for the majority of children following cholesteatoma treatment.

INCIDENCE OF MIDDLE-EAR CHOLESTEATOMA WITH ANALYSIS OF ITS LOCATIONS, EXTENSIONS, AND COMPLICATIONS FROM 1993 TO 2009

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Introduction

Middle-ear (ME) cholesteatoma represents a relatively common and clinically challenging form of chronic otitis media (OM). Formation of a cholesteatoma is associated with atrophic parts in the tympanic membrane, which form the basis of a retraction pocket together with negative ME pressure related to poor function of Eustachian tube and/or the mastoid. Retraction pockets may form in the pars flaccida, in the sinus or in the pars tensa of the TM, and they frequently result in accumulation of cellular debris, which causes recurrent infection with otorrhea and invasion of the ME cavity and neighboring areas with erosion of bony structures.[1-3]

Studies on the incidence rate (IR) of ME cholesteatomas are relatively scarce. In older studies, an IR between 2.8 and 13.2 per 100,000 inhabitants has been reported.[4,5] More studies have investigated the hypotheses that the IR has been decreasing during the last two decades, and that such decrease may be related to a simultaneous increase in the usage of ventilation tubes (VT) during childhood OM. More of these studies have documented a significant decrease in the IR of cholesteatomas together with an increase in the numbers of VT insertions,[6-8] whereas one study finds the IR unchanged.[5]

In our department we also have had the impression of a decrease in the occurrence of ME cholesteatomas referred for surgical treatments over the recent years, but in addition we also have felt that these cases may have been less extensive and with less complications than earlier. If such observations are related to childhood VT insertions, it should be noticeable in our department, since we seem to have the highest IR of VT insertions reported in the literature, amounting to 375 procedures per 10,000 children per year (< 16 years);[9] this IR is around six to seven times higher than comparable countries like Norway and Finland.[10,11] Therefore, the purpose of this study was to determine the IR of primary ME cholesteatomas in our region during the last 17 years including an analysis of their locations, extensions and complications.

Material and methods

The study was based on a longitudinal retrospective survey of primary acquired ME cholesteatoma from January 1, 1993 to December 31, 2009. Information about these surgeries was obtained from our department's otosurgical database, which has been used since 1993 for complete prospective registration of all otosurgical procedures; information was occasionally supplemented by reviewing patient records. The information retrieved contained the number of primary cholesteatoma procedures per year, their locations and local extensions as well as related complications.

Our department is the only one in its specialty serving the North Denmark Region, hence all cholesteatomas were referred to our Department. The North Denmark Region has the merit of a relatively stable population with approximately 600,000 inhabitants; we obtained data about the exact population sizes for all years for

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both adults and children from Denmark's Statistics.[12] Conventional descriptive statistics were used for description of the basic data, whereas the IR of cholesteatoma was investigated by correlation analysis.

Results

Cholesteatoma incidence

Based on the population the *overall* mean annual IR was determined at 8.75 new cases per 100,000 inhabitants (range 4.5 to 12.3) (95% CI: -7.68 to 5.56). The overall annual IR during the study period has been illustrated in Figure 1 displaying a statically significant decrease (p = 0.016, r = 0.33; N = 756).

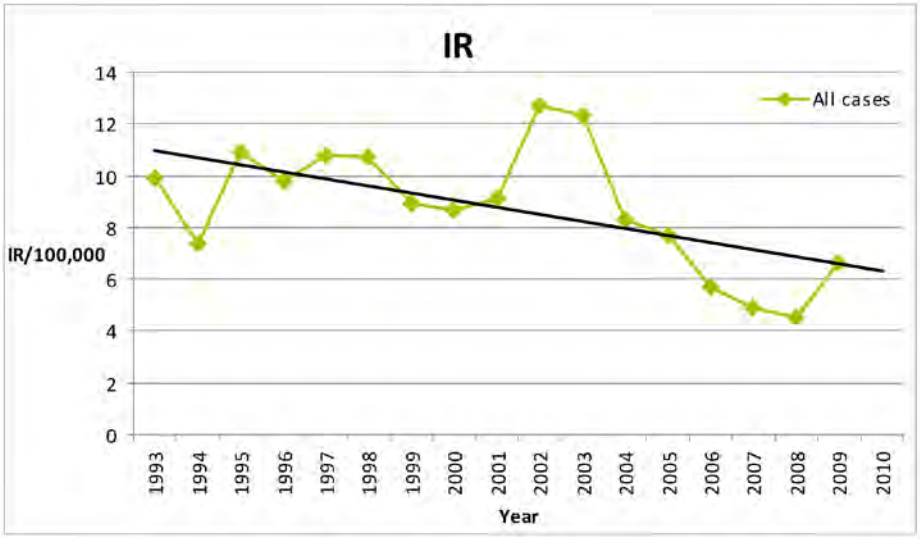

Fig. 1. The *overall* annual incidence rate (IR) per 100,000 inhabitants from 1993 to 2009. The straight line indicates the regression line; correlation analysis shows a statistically significant decrease during the period (p = 0.016, r = 0.33; N = 756).

Locations and extensions

Overall flaccida or attic cholesteatoma was found in 298 cases (39.3%), sinus cholesteatoma in 216 cases (28.8 %), and tensa cholesteatoma in 156 cases (20.6 %). In 85 cases (11.3 %) the origin of the cholesteatoma could not be assessed (mixed cholesteatoma).

The three types of cholesteatomas with well-defined origin showed different regional extensions, so that for instance sinus and tensa cholesteatomas were predominantly restricted to the ME cavity (58% and 63%, respectively), whereas flaccida cholesteatoma showed a more dispersed pattern of growth. These regional extensions have been illustrated in Figure 2. There were no systematic changes in the locations and extensions of the cholesteatomas during the study period.

Complications

Complications encountered before or during surgery comprised of ossicular erosions with affection of the crus longum in 50% of the cases; additional ossicular erosions included the stapes supra-structure in 21% of these. A separate analysis of these erosions showed no systematic variation during the study period. However, various more rare complications were found; these have been outlined in Table 1.

In total 114 of these complications were encountered in 86 cases; thus, the overall frequency of one or more of these complications was 86/756 (11%). The annual numbers of the individual complications were few, and it was not meaningful to investigate these separately; thus, the overall number of complications per year was analyzed during the study period and related to the numbers of surgeries per year; there was a significant decrease over time in the frequency of the overall complications per year (p < 0.05, r = 0.255; N = 756).

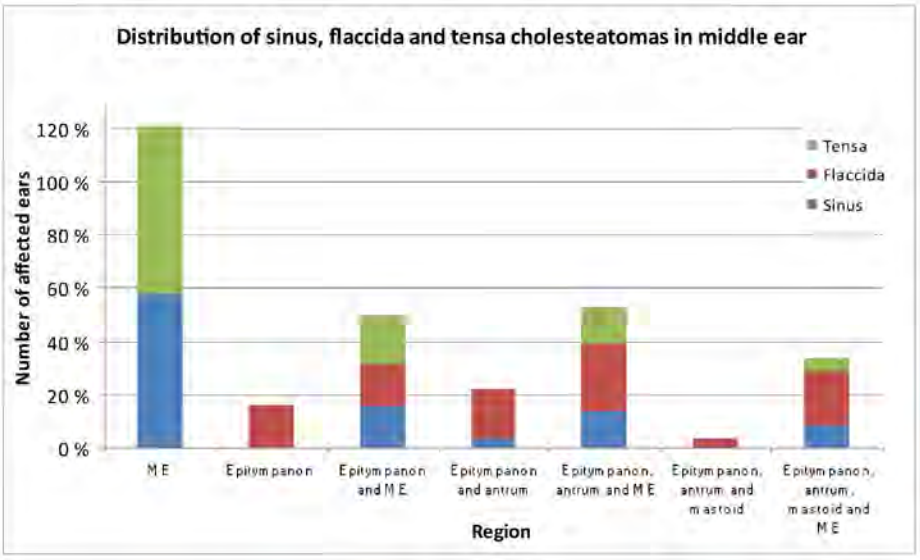

Fig. 2. Distribution of the extensions of sinus, flaccida and tensa cholesteatomas into adjacent regions of the ME and mastoid cavity.

Table 1. Complications of cholesteatomas

Complication type	Cases (%)
Facial nerve or dura exposure	90 (79.9)
Fistula (semicircular canal, cochlea, footplate)	17 (14.7)
Facial paralysis	4 (3.7)
Acute mastoiditis	2 (1.8)
Intracranial abscess and/or meningitis	1 (0.9)
Sinus thrombosis	0 (0)
Deafness	0 (0)
Total	114

The total number of complications appeared in 86 cases; thus, the risk of one or more complications was 86/756 = 0.11.

Discussion

Cholesteatoma incidence and VT's

VT insertion is still one of the most frequently performed procedures in otology. Insertions of VT's often results in permanent TM perforations; other complications include otorrhea, tympanosclerosis, and formation of cholesteatoma.[13,14] In many countries there has been an increasing rate of VT insertions during the last 30 years despite the fact that its justification remains rather controversial, because its long-term effects have not been documented. During the same period more studies have focused on possible changes in the IR of cholesteatomas related to the usage of VT's.

A number of studies have described an increased risk of cholesteatoma arising behind an intact TM or in a perforation of the pars tensa resulting from previous VT insertion; the IR of these secondary developed cholesteatoma are ranging from 0.48 to 1.1%.[15-17] However, other studies have shown a decline in incidence of cholesteatoma formation despite multiple VT insertions.[6-8] Further, some studies have been unable to demonstrate any changes in the IR of cholesteatoma, despite dramatic increases in the use of VT's.[5,18-20]

In a recent, larger epidemiological study, Spillsbury *et al.* reported that insertion of VT at an early age, insertion of subsequent VT without any delay and adenoid removal were related to a reduced rate of cholesteatoma development.[16] Whereas epidemiological results may not prove causal relations, it may still indicate some connections.

In our country we have a very high incidence of VT insertions, around 375 procedures per 10,000 children per year (< 16 years), and at the same time we found a statistically significant decrease in the incidences of cholesteatoma; similar results have been reported by Djurhuus *et al.* [21] The insertion of VT's tends to ensure a

better development of the mastoid,[22] and the mastoid has been shown to play an important part in ME pressure regulation distinct from the Eustachian tube.[23] Thus, VT insertions may contribute to a normal development of the mastoid which may ensure a normal pressure regulation in later life, and thus prevent underpressures resulting in retraction pockets and formation of cholesteatomas. This basic explanation is supported by the epidemiological findings by Spillsbury.[16]

Location and extensions

The majority of cholesteatomas involved the pars flaccida (48%), whereas the tensa (20.6%) and sinus cholesteatoma (28.8%) were less common. Tensa and sinus cholesteatomas presented with a mass in the tympanic cavity and only a few percent had epitympanic spread (Fig. 2). Flaccida cholesteatoma had more tendencies to spread into both the ME and the antrum including the mastoid, and thus, it showed a more dispersed pattern of growth. However, the size of the cholesteatomas as well as their locations has been stable during the study period.

Complications

The most common complication found was erosion of the crus longum in 50% of the cases, followed by additional erosion of the stapes supra-structure in 21%. The ossicular chain pathology did not change during the study period. However, this meant that ossicular chain reconstruction was very often required. In accordance, ossicular destructions have previously been reported to be the most common complication in cholesteatomas.[24] Thus, an intact ossicular chain has been described in only 26% of flaccida cholesteatomas; the most affected part is the crus longum, followed by the incus body and the malleus head.[24]

One of the less frequent complications of ME cholesteatoma is labyrinthine fistula, which has been reported in 3.6-15% of cases.[25,26] A fistula may not show symptoms or signs, but it poses a risk of sensory-neural hearing loss; our findings of 14.7% of cases with complications corresponded overall to 17 among 756 cases, *i.e*., 2.2%. A previous study from our department reported a higher rate of labyrinthine fistula (8.9%) during 1979 to 1990;[27] this may corroborate our overall findings that complications seemed to decrease over the study period. We encountered no cases with deafness before or after surgery. Altogether, we found various complications in only 11% of the cases, and on an overall basis this number showed a significant decline during the study period.

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RECONSTRUCTION OF THE TEMPORAL BONE WITH BONE-MORPHOGENETIC PROTEIN 2 (BMP-2)

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Introduction

Until now, reconstruction of temporal bone defects after tympanoplasty or mastoidectomy has been performed using cartilage, fascia, bone chips, bone putty, and other reconstruction materials. However, each material has its own problems, and there is currently no established method for reconstructing temporal bone. Moreover, current reconstruction methods do not result in physiologically normal bone with remodeling abilities such as bone tissue ossification and absorption. Reconstruction with physiological bone tissue that is capable of bone remodeling would resolve these problems and be the ideal solution.

Bone tissue reconstruction using tissue-engineering techniques is currently receiving attention. The following substances have strong potential for bone formation:

- Bone-morphogenetic protein (BMP)
- Platelet-derived growth factor (PDGF)
- β-tricalcium phosphate (β-TCP)

These bone-forming substances were used in our previous experiment. Of these substances, BMP has the highest ability to form new bone, which is why we used BMP in this experiment.

Experimental method

The test animal is a six-week-old male Hartley white guinea pig weighing 400 to 450 g with normal tympanic membranes. The carrier used was type-I atelocollagen. The animal was injected with BMP. From among the roughly 20 types of BMP, BMP-2, which has the strongest bone-forming power, was used. The BMP-2 concentration was 1.5 mg/cm^3, which in previous studies has been shown to be the optimal concentration. Guinea pig was anesthetized with a pentobarbital injection into the abdominal cavity, and a diamond bur was used to drill a hole in the middle-ear otic capsule of the temporal bone on one side of the face. The carriers were impregnated with the injection drug and embedded, the incision was closed, and the animal was returned to its rearing place. The site where the incision was created was examined with CT scans after certain periods of time. The animal was given isoflurane inhalation anesthesia for CT scan.

CT study

A CT scan was made with a Micro CT for animals that allows scans to be taken of living animal and permits changes to be assessed over time in a single specimen. The LaTheta LCT-200 (Hitachi Aloka Medical, Ltd. Tokyo, Japan) used, enables sensitive imaging with a pixel size of 24 μm. Analysis by 3D software can be used to view any cross-section. Bone-mineral density can be measured directly, and bone-tissue composition can be analyzed.

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Edited by Haruo Takahashi

Results

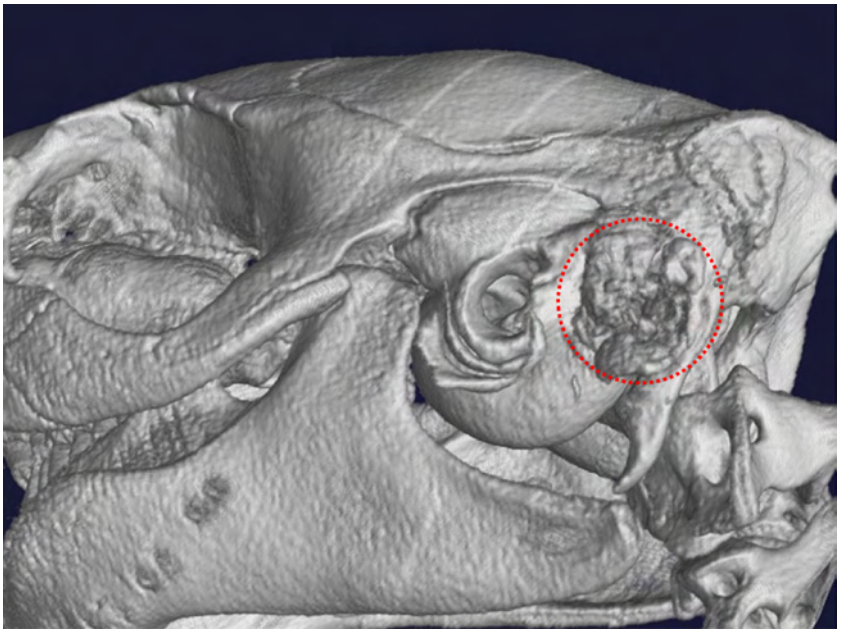


Fig. 1. 3D-CT image of the animal two weeks after BMP injection. Post-incision bone-tissue regeneration identified on the surface of the treatment site.

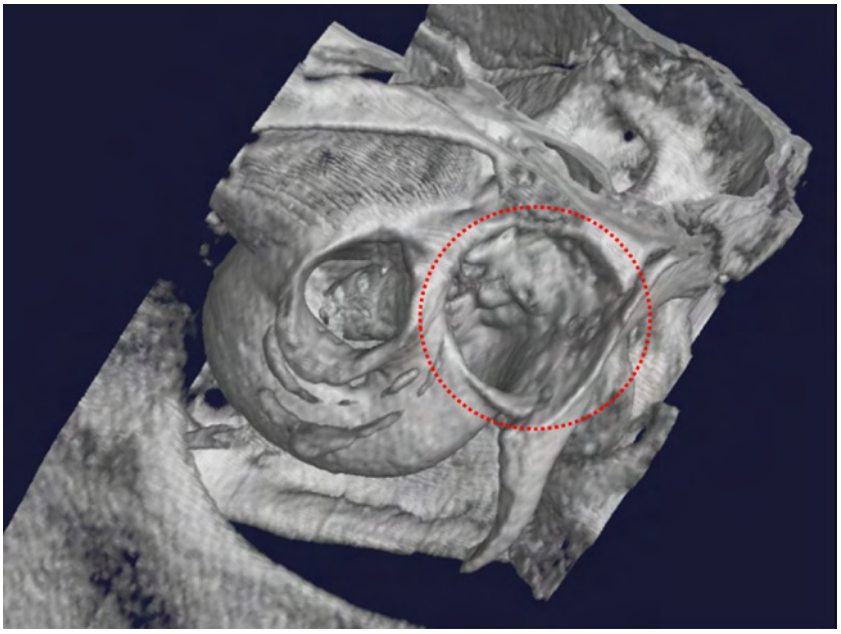


Fig. 2. 3D-CT image of control model (normal saline injection), two weeks after the procedure. There is no post-incision bone regeneration in the treatment site.

*Figures 3 to 6: Bone formation gradually progresses and bone tissue continues to mature. Moreover, composition analysis of the bone tissue shows formation of a pneumatic space in the inner cavity, and an almost normal pneumatic space has been formed in the site one year after the procedure.

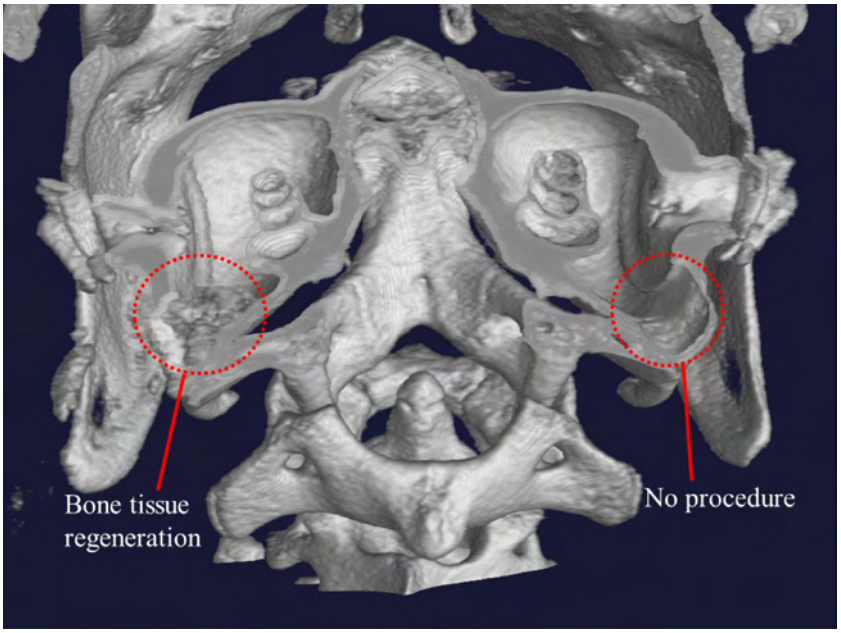


*Fig. 3.** CT images of the bone two weeks after the procedure (3D cross-section, axial image, bone composition analysis image). Observation of this part with an axial view shows that bone-mineral density only increases in the perimeter, and there is almost no increase in bone-mineral content in the center at this point in time. Bone-composition analysis was performed. Green spots show cortical bone and yellow spots show cancellous bone. Green cortical bone is seen in the bone-regeneration site, but no regeneration of bone tissue is seen in the center.

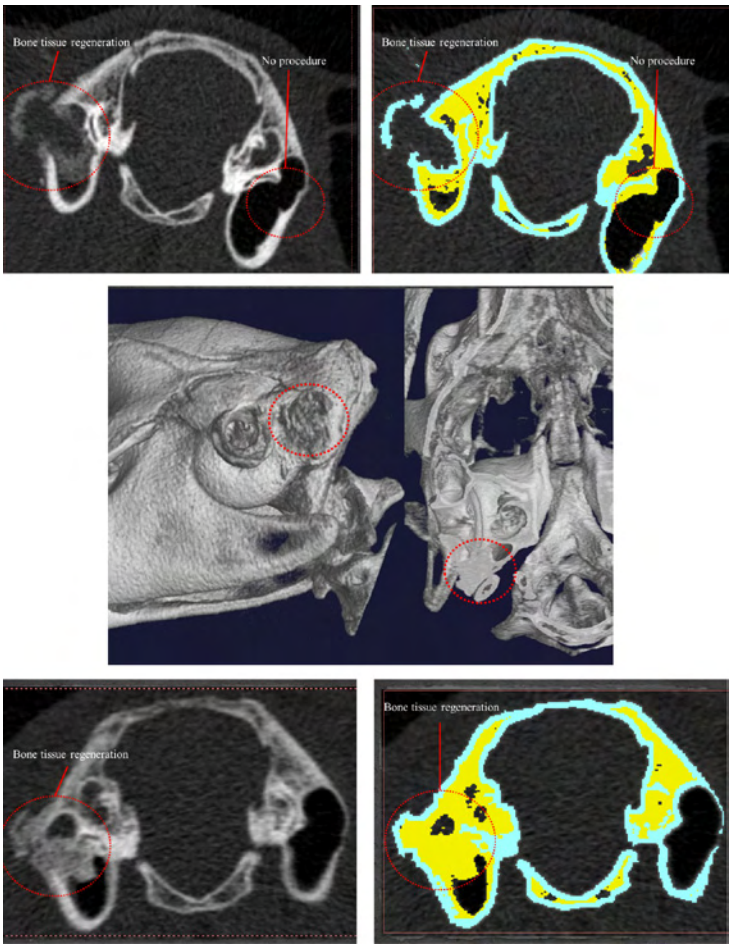


*Fig. 4.** CT images of the bone one month after the procedure (3D lateral view, 3D cross-section, axial image, bone composition analysis image). Further post-incision bone regeneration is seen in the treatment site. Axial views and bone-composition analysis show an increase in bone-mineral density in the center.

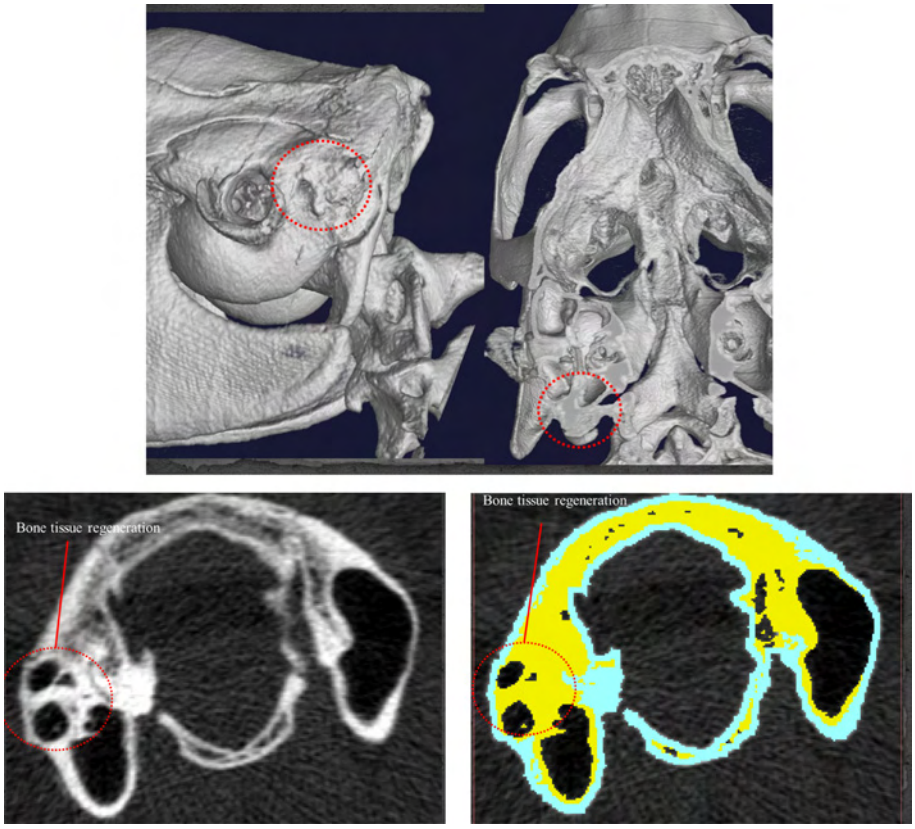


*Fig. 5**. Six months after the procedure. Bone mineral content has increased and the bone tissue is maturing. Pneumatization in the center of the bone-regeneration site in axial views and bone-composition analysis is starting.

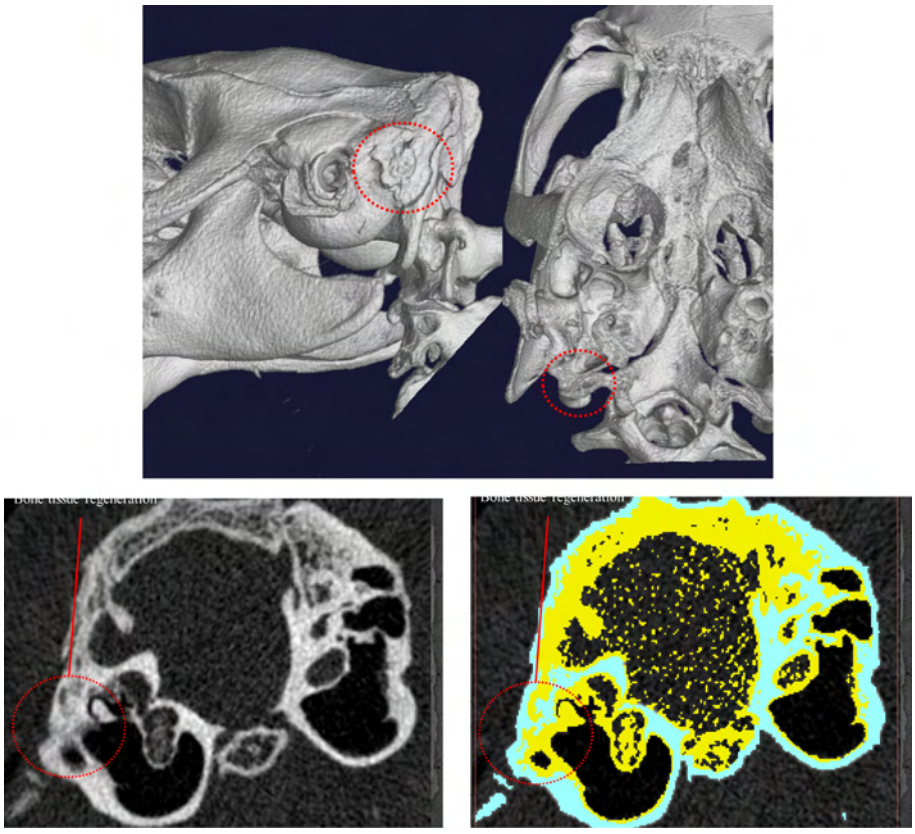
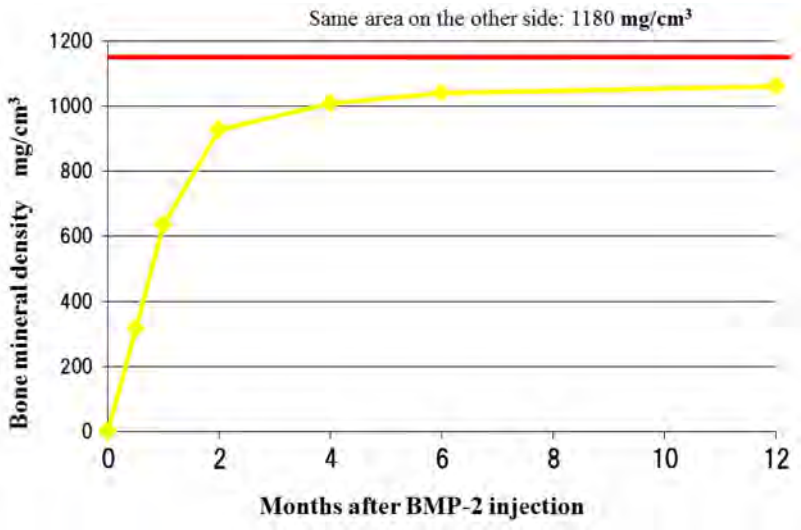


*Fig. 6.** One year after the procedure. Mature-bone-tissue regeneration is identified. Axial views and bone composition analysis show further increase in pneumatization in the center of the bone regeneration site, indicating development of normal temporal bone.

Table 1. Increase of bone-mineral density in the bone regeneration site.



Temporal changes in bone regeneration from BMP-2 treatment were observed, and the increase in bone-mineral density was measured by CT scan in the bone-regeneration site and displayed in a graph. The graph shows that bone-mineral density gradually increases with time. It appears to be approaching 1180 mg/cm^3, which is the density in the untreated area on the other side.

Discussion

Tympanoplasty and mastoidectomy often result in bone destruction due to middle-ear lesions. Alternatively, incision into the bone during surgery often leaves defects in the mastoid cortical bone or ear canal posterior wall. The ossicles may become eroded, leading to a defect and hearing impairment. Until now, temporal bone defects have not received any type of treatment and have been left untreated. However, mastoid cortical bone defects can result in postauricular skin depression after surgery, leaving the site dysmorphic. In addition, many patients may suffer complications of cavity problems for a long time after surgery due to external exposure of the mastoid cavity. The cavity problems are a result of continued otorrhea or excessive earwax in the middle ear cavity that has become immunocompromised. Many such patients must continue receiving periodic treatment from an otolaryngologist for the rest of their lives following middle-ear surgery. Resolving these problems requires some method of reconstruction of bone defects in the posterior ear canal wall and the ossicular chain following cleaning after middle-ear surgery. While there have been reports of using cartilage and bone putty to reconstruct the posterior ear canal wall, both have collection limitations and cannot necessarily be used for all cases. Artificial materials (such as hydroxyapatite) are often used in ossicular-chain reconstruction, but these often cause a foreign-body reaction, and the ossicular prosthesis is naturally expelled. When cartilage is used, sound conduction has been reported to be inefficient. There is therefore a need to develop a new method for reconstructing temporal bone.

In 1965, Urist[1] reported ectopic bone induction in rats by subcutaneous and intramuscular decalcified bone matrix grafting. He suggested that a bone-forming protein factor existed within the bone matrix, and he called it bone morphogenetic protein (BMP). After this discovery, numerous researchers attempted to purify BMP, but it remained unidentified for a long time. In 1988, Wozney *et al.*[2] of the Genetics Institute successfully cloned the DNA of four types of human BMP. The structure of BMP was revealed for the first time, and it can now be artificially produced. About 20 proteins belonging to the BMP family have been found to work not only on bone tissue, but are also involved in morphogenesis of various organs, differentiation, apoptosis, and tissue regeneration. In addition to bone and cartilage tissue, BMPs work on a wide range of organs including nerves, the cardiovascular system, the kidneys, and the digestive tract. Today, BMP is ranked as a cytokine that functions as a signaling protein for morphogenesis, and it has been confirmed to be a protein of the TGF-β superfamily. BMP-2 has the strongest bone-forming power among proteins in the BMP family, and, when placed in the body with a carrier that locally retains BMP-2 and releases it at an appropriate

rate, mesenchymal stem cells are stimulated to differentiate into osteoblasts, and the increasing number of osteoblasts results in rapid formation of bone tissue. Normal bone capable of bone remodeling is then regenerated. Past studies on bone regeneration using BMP-2 have been reported, including a study by Gerhart *et al.*[3] on bone formation of the sheep femur, a study by Mushuler *et al.*[4] on dog spinal fusion surgery, and a study by Seto *et al.*[5] on mandible bone defect repair in monkeys. It is also being clinically used for some spinal diseases in Western countries.

We therefore thought that it may be possible to easily regenerate the ossicles, reconstruct the ossicular chain, and reconstruct mastoid cortical bone defects using BMP-2. Temporal bone is morphologically classified as pneumatic bone because it has an internal air sinus and a special structure with many hollow cavities and cellulae. Since the interior surface of the middle-ear cavity does not come into contact with skin or muscle tissues, undifferentiated mesenchymal cells cannot migrate easily, and bone formation by BMP-2 action is thought to be unfavorable. There are hardly any reports on the question of BMP efficacy in the temporal bone, and this question remained unanswered. We previously reported measurement by soft X-rays after euthanizing the animal, but this method resulted in measurement errors between experiments and subjects. Using a micro-CT device for animals in the present study, inter-experimental and inter-subject errors were removed. This is the first report to describe the use of BMP-2 on the temporal bone, a pneumatic space with an internal sensory organ, and the ability of BMP-2 to promote remodeling of an incision made in the temporal bone so that it once again became a pneumatic space. These results demonstrate the potential usefulness of BMP-2 in otologic surgery.

Conclusions

This study demonstrated that micro-CT devices for animals are very effective for observing bone regeneration.

BMP-2 was shown to have a strong ability to reconstruct bone tissue in temporal bone, and construction of a pneumatic space was confirmed for the first time. These results suggest the potential for clinical use of BMP-2 in otologic surgery.

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THE EFFECT OF TYMPANOSTOMY TUBES ON TYMPANIC MEMBRANE RETRACTION IN CHILDREN WITH CLEFT PALATE

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Introduction

The role of tympanostomy tubes (grommets) in tympanic membrane atelectasis remains controversial.[1] While they are commonly used to relieve negative middle-ear pressure with the intention of alleviating tympanic membrane retraction (TM), concern has been expressed that after extrusion, they may contribute to atelectasis.[2] Children with cleft palate commonly receive tympanostomy tubes because of their predisposition to otitis media with effusion. They are also at higher risk of tympanic membrane retraction: around 44% of their ears have retraction compared with around 10% of ears in the general pediatric population.[3,4] Of more concern, the prevalence of cholesteatoma is around 200 times more common in children with cleft palate.[5] Greater understanding of the relationship between tubes and development of cholesteatoma from progressive retraction may help to lessen the burden of disease for these children.

As tympanostomy tubes are typically placed in the antero-inferior quadrant of the pars tensa, and never placed over the incus in the postero-superior quadrant, we hypothesised that retraction of the anterior or inferior segments of the pars tensa could be a direct consequence of TM atrophy from tube insertion, whereas retraction of the postero-superior retraction would not be. The objectives of this study are to determine: 1) whether anterior or inferior pars tensa retraction is more common after tube insertion; 2) whether postero-superior retraction is less common after tube insertion; 3) whether the severity of retraction correlates with the number of tubes inserted.

Method

Research ethics board approval was given for review of a cross section of our population of children with cleft palate attending a regional tertiary referral center for regular multi-disciplinary review over the course of one year. Prospectively acquired data included age, history of the number of tympanostomy tubes per ear, and digitally captured endoscopic TM images acquired after cerumen clearance. Cases with *in-situ* tubes, perforation, cholesteatoma or previous tympanoplasty were excluded. Cases were also excluded if the otoscopic image was of inadequate quality to allow assessment of the whole TM.

The TM images were analyzed for the presence and location of retraction of the pars tensa, noting whether retractions were present in the anterior and inferior segments or the postero-superior quadrant or both areas. Severity of retraction was based on a system described in Table 1 which was derived from the staging systems described by Sade and the Erasmus group.[6,7]

Analysis was with parametric and non-parametric statistical tests using SPSS software (Systat Software, Inc.).

This sample of children was previously described in an assessment of the reliability of endoscopic assessment of TM retraction.[3]

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Table 1. Staging system used to describe severity of retraction

Stage	*Anterior or inferior retraction*	*Postero-superior retraction*
1	Retraction not in contact with promontory	TM retraction not in contact with promontory, incus or stapes
2	Retraction touching or adherent to promontory	Retraction touching or adherent to incus or promontory
3	Erosion of bone (annulus or promontory)	Erosion of bone (incus, stapes, promontory or annulus)
4	Limits of retraction not visible	Limits of retraction not visible

Results

Two hundred twenty-seven useable TM images were obtained from 143 children with cleft palate. The average age of these children was 13 years with a fairly even distribution of ages across the range of 4-18 years (Figure 1). The severity of tympanic membrane retraction across three different age groups is shown in Figure 2. There was no significant difference in severity of retraction in these different age groups (Spearman rank order correlation). A trend can be seen in Figure 2 suggesting relatively more stage-2 and less type-1 retractions in the youngest age group. Post-hoc comparison to analyze this observation revealed no significant difference (comparison of age with stage 1 versus age with worse retraction; comparison of age with stage 1 versus age with stage 2 retraction; $p > 0.05$, Mann-Whitney Rank Sum Test).

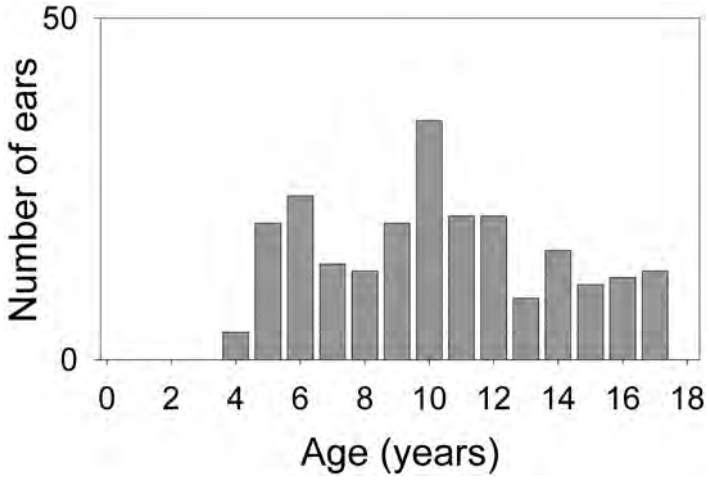

Fig. 1. Histogram showing distribution of study participants by age in years, representing a cross section of the whole population of children aged 5-17 years with cleft palate.

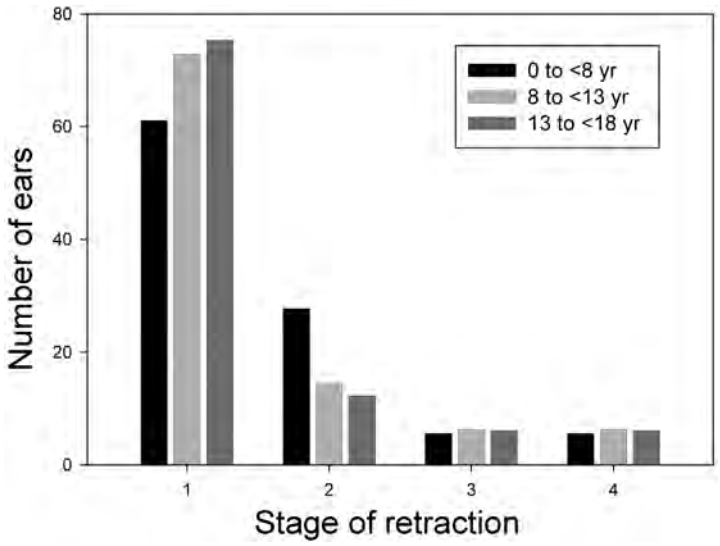

Fig. 2. Histogram showing number of ears at each stage of retraction by age. Stage of retraction is as defined in Table 1.

As shown in Figure 3, 198 (87%) ears had one or more tympanostomy tubes and 99 (44%) had two or more tubes. Fifty-one of 227 (22%) ears demonstrated an anterior or inferior retraction, 32 (14%) a postero-superior retraction, and 12 (5%) had both. To simplify interpretation, the small number of cases with retraction in both areas was excluded from the following analysis. Anterior or inferior retraction occurred in a greater proportion of ears that had received tubes compared with non-tubed ears (*i.e.*, 47 (24%) versus 4 (14%)). Postero-superior retraction was relatively less common in tubed compared with non-tubed ears (*i.e.*, 26 (13%) versus 6 (21%)). These differences were not significant (Fisher exact test). The proportion of cases with retraction at each site plotted by the number of tubes inserted is shown in Figure 4. No correlation was found between severity of retraction, at either site, and number of tubes (Spearman rank order correlation).

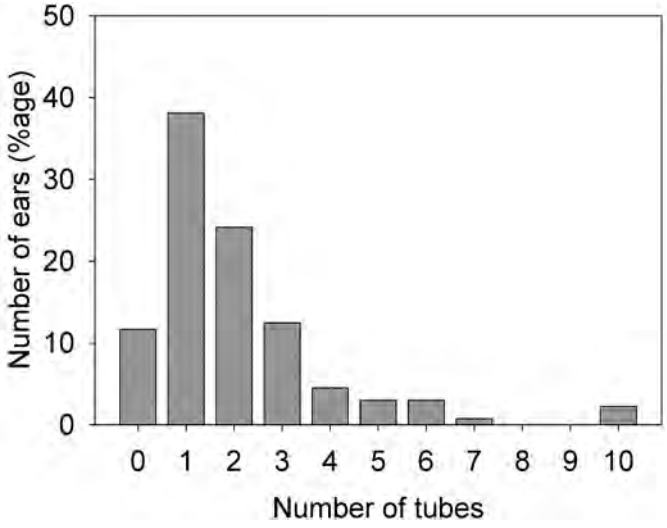

Fig. 3. Histogram showing distribution of number ears by number of tympanostomy tubes inserted.

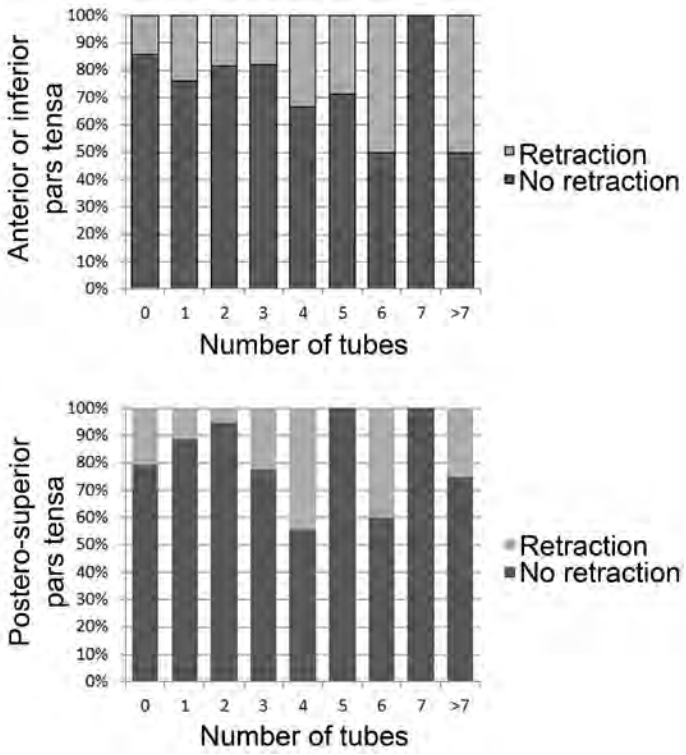

Fig. 4. Proportion of ears with tympanic membrane retraction plotted by number of tubes inserted. Anterior or inferior retraction above; postero-superior retraction below.

Conclusion

The role of tympanostomy tubes in TM retraction remains controversial. While tubes have the potential to aggravate antero-inferior pars tensa retraction by causing TM atrophy, or to alleviate postero-superior retraction by relieving negative middle-ear pressure, this sample was too small to confirm either effect. Power analysis calculation indicates that a sample size of around 500 ears would be required to confirm the trends observed. The validity of the findings in this study have not been confirmed by repeated assessments of the TM images by the same or other observers: but as no significant differences were found with a single assessment, there was not felt to be adequate justification to repeat the observations.

The natural history of tympanic membrane retraction is poorly understood, with some TMs remaining stable for many years, some resolving spontaneously and some, seemingly a small minority, progressing into cholesteatoma. Even though risk factors for cholesteatoma development have been identified, such as that cleft lip and palate gives a higher risk than isolated cleft palate,[5] it is not possible to predict the course or speed of this natural history in any given patient. This makes it difficult to determine the need for treatment in any given patient. It is also not possible to determine whether reported results of treatment, that may be administered to prevent ossicular erosion or cholesteatoma formation, are better than the outcome of no treatment.

In this context, more information about the relationship between tympanostomy tubes and TM retraction would be helpful, particularly for children at risk such as those with cleft palate. Separating how much their tendency to atelectasis and cholesteatoma is prevented or aggravated by the use of tubes from how much is due to their tendency to middle-ear disease (the same issue that leads to tube insertion) will be hard to determine. Well-defined indications for tube insertion, an ethically acceptable alternative to tympanostomy tube insertion (perhaps sub-annular tube placement or hearing aid) and follow up over many years would be prerequisites for prospective study of this relationship with a randomized controlled trial.

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CHOLESTEATOMA IN CHILDREN WITH CLEFT PALATE – DOES ROUTINARY TYMPANOSTOMY MAKE ANY DIFFERENCE?

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Introduction

Reports in the literature establish a higher incidence of otologic abnormalities (cholesteatoma, acute otitis media, retraction, tympanosclerosis, perforation and middle-ear effusion) among cleft palate patients.[1-3] Since ear pathology plays a crucial role in language, speech and educational development, regular otologic care is more than desirable. It is a wide accepted statement that patients with cleft palate have a Eustachian-tube dysfunction due to muscular abnormalities. It seems logical to establish a middle-ear aeration inserting tubes through the tympanic membrane to serve as pressure equalizer.[4] However, recent studies suggest a secondary role of Eustachian tube dysfunction in the genesis of the cholesteatoma[5] and the routine placement of ventilation tubes in cleft-palate patients remains controversial.[6-8] In the current climate of limited health-care funding, evidence of clinical and cost effectiveness is becoming increasingly necessary.

The objective of this study present study was to determine the incidence of ear problems in the cleft palate population and the long-term effectiveness of tympanostomy among these patients.

Methods

Study design

This was a prospective, controlled, open clinical trial conducted between August 1998 and December 2011. The study was approved by the Institutional Board of the Operation Smile Foundation Colombia.

Selection of patients and controls

A cohort of 151 patients born with cleft palate was included in this study; all of them underwent ENT evaluation before primary palatoplasty was performed. According to the reconstructive-surgery protocol, the first surgery should be done after the age of one year, when the child has reached enough weight and maturity of the pharynx tissues. However, in some unfortunate cases, patients from distant rural areas are evaluated as late as eight years old.

Sixty-three patients (126 ears) underwent preventive bilateral tympanostomy at the time of the primary palatoplasty. A Paparella No. 2 or Sheehy-type tube (Medtronic-Xomed, Jacksonville, FL) is placed in a normal area of the membrane, usually through an incision in the anterior-superior quadrant because the epithelial migration is, in this part of the tympanic drum, the slowest to allow a late extrusion of the tube 9. T-type tubes are not used currently due to a higher incidence of persistent perforation and a negative impact in the conductive hearing level that we have seen in a previous group of patients. During a period of at least ten years, a year-base follow up is conducted and a replacement of the tube is done every time we identified the extrusion of the previous. These patients are called the 'Tympanostomy group'.

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Edited by Haruo Takahashi

In 88 patients (176 ears), no surgical treatment for the ear was proposed and a year-base follow up was performed. This group of patients is called the 'Control Group'.

Patients with a history of general or neurological disease were excluded. All the patients belong to the lowest socio-economical levels in Colombia, where a lack of medical resources is evident.

Outcome measures

At the time of the last evaluation (at least ten years follow up), a record of the next information was done:

- *Otoscopy findings:* This information described the oto-microscope evaluation and the information analyzed were: acceptable otoscopy, tympanic retraction, effusion, perforation and cholesteatoma;
- *Hearing Assessment:* Audiometric studies included pure-tone thresholds and SRTs, with special focus in the air-bone gap;
- *Number of acute otitis media episodes:* Parents are previously requested to record every episode of ear infection determined either by the pediatrician, general physician or the otolaryngologist;
- *Number of tubes:* Describes the number of surgical procedures to replace the tube in the tympanostomy-group patients.

After completion of studies in all patients, differences and their significance were determined and calculated among both groups.

Results

Bilateral tympanostomy as a routine procedure was used in 63 patients (38 male and 25 female), ranging in age at the time of the last evaluation from 11 to 19 years, with a mean of 13.1 years. The mean follow up was 11.3 years (range, 10.1 y-13.2 y).

Eighty-eight patients (50 male and 38 female) were used as a control group and no tympanostomy was performed. The age at the end of the study ranged from 11 to 20 years (mean 14.5). The mean follow up was 11.1 years (range, 10.1-12.9 y).

Patients who had been lost to follow up were excluded from the present study.

Each patient in this study underwent microscope examination of the ear and hearing screening at least at the end of the follow up. In only a few patients a CT scan was performed, so those data are not included in the present study.

When describing the appearance of the middle ear, there is a group of patients with an *acceptable otoscopy* encompassing findings of tympanosclerosis and mild retraction (I and II according to the Sáde classification) with no clinical and audiological relevance. In such cases, further treatment was not indicated in the opinion of the author.

Figure 1 shows the results of the otoscopic findings in both groups, suggesting better outcomes in the tympanostomy group. Figure 2 suggest a better long-term outcome in terms of middle-ear function in patients with prophylactic tympanostomy. However, there is a slight but significant difference in the incidence of cholesteatoma, indicating a more favorable outcome in the control group. Figure 3 reveals an important decrease in the number of acute otitis media episodes in cleft-palate patients with a routine tympanostomy, during the first decade of their life. The average number of tubes required in these patients was 2.8 over a ten-years period of time.

Discussion

The face is developed from five processes which fuse with one another. Development disturbances of these processes appear as cleft. These clefts may occur in a wide variety of types, the most common of which is the cleft lip and palate. Complete clefts of the palate may occur on the left or right side or also bilaterally in different degrees of severity; bilateral total cleft is the most extreme form. All these patients suffer an abnormal insertion of the levator and tensor veli palatini muscles into the posterior margin of the hard palate, which are responsible for the Eustachian-tube function. As a consequence, the middle ear is not aerated and a vacuum process begins inciting an inflammatory reaction in the middle ear and tympanic membrane. During

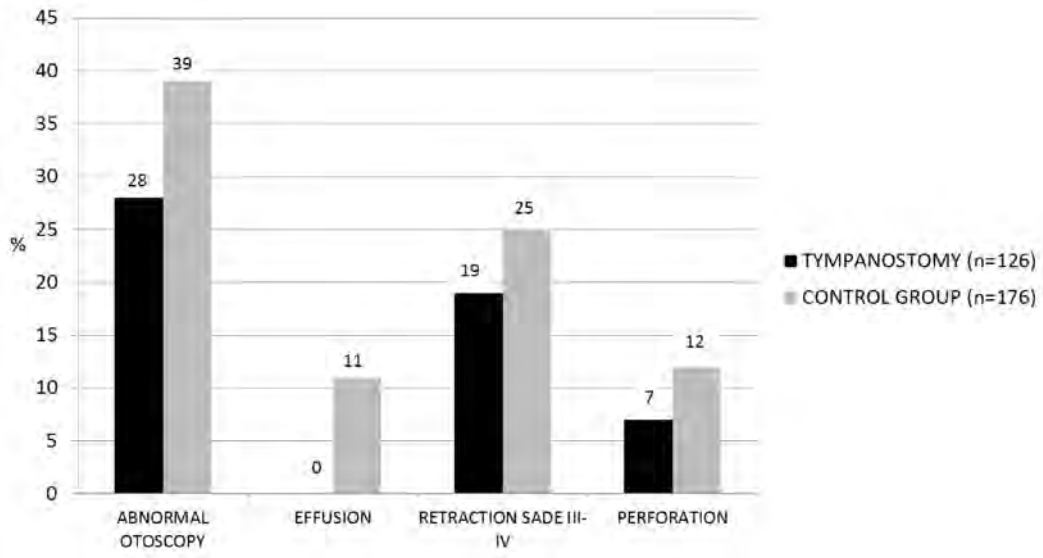

Fig. 1. Percentage of abnormal ear findings.

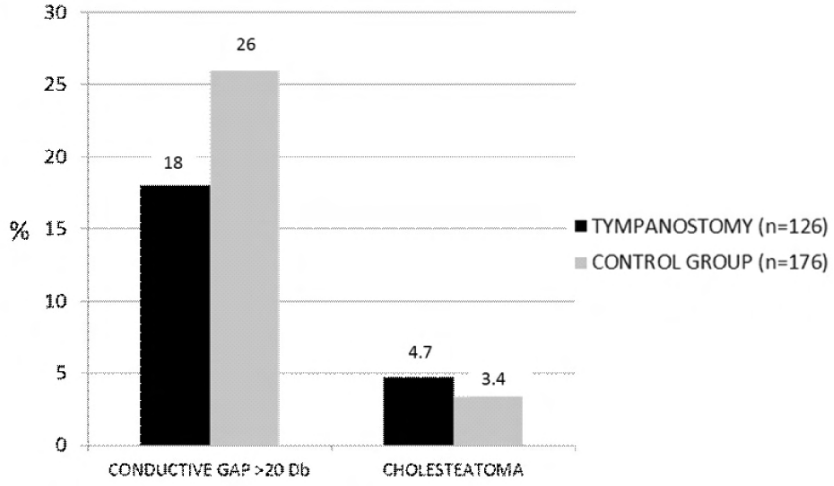

Fig. 2. Air-bone gap and cholesteatoma incidence.

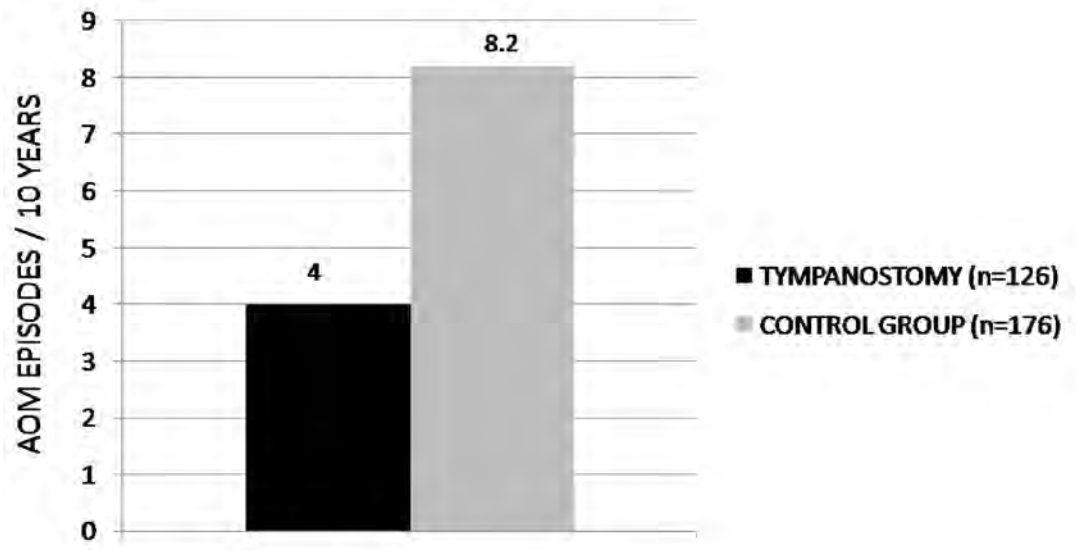

Fig. 3. Number of acute otitis-media episodes during a ten-year period.

the last century, several authors have suggested the role of retraction pockets and secondary bacterial inflammation in the pathogenesis of acquired cholesteatoma.[10,11] In an attempt to aerate the middle ear, tubes are inserted through the tympanic membrane to serve as pressure equalizer and prevent middle-ear pathologies. However, recently, Karmody[12] reviewing the pathogenesis of acquired cholesteatoma, suggested a secondary role of retraction pockets supporting the hypothesis of the medial migration of squamous epithelium from tympanic membrane. Some other authors have reported similar findings that agree with a secondary role of the Eustachian tube dysfunction in the origin of cholesteatomas.[13-15]

Children born with cleft palate are an *in-vivo* and excellent model to observe the long-term consequences of the Eustachian-tube dysfunction. The prevalence of cleft palate in a general population varies from 0.5 to 2.5 in 1000 births.

Our study population is a group of cleft-palate patients observed for the author for more than ten years in order to establish the ear pathologies during their childhood. Another objective of the present study was to evidence the benefits and drawbacks of the routine tympanostomy, with special focus on cholesteatoma.

We found enough clinical evidence to suggest that a tympanostomy in cleft-palate patients, significantly reduces the incidence of middle-ear effusion, tympanic-membrane retraction, perforation and acute otitis media episodes. Our results showed a trend toward reduction of conductive hearing loss in cleft-palate patients with ventilation tubes. However, the incidence of cholesteatoma in our study population was higher in the tympanostomy group, supporting the hypothesis of a secondary role of Eustachian-tube dysfunction in the genesis of cholesteatoma and addressing the question of a possible iatrogenic role of the ventilation tubes.[16]

The author suggests that the ventilation tube placement decision should be taken case by case on the basis of recurrent infection, hearing loss or evidence of abnormalities detected in a regular follow up combined with the surgeon's seasoned judgment, depending on anatomic, pathologic and socio-economic conditions.

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LONG-PALVA-OBLITERATION TECHNIQUE IN CHOLESTEATOMA SURGERY: RESULTS OF A NEW TECHNIQUE

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Introduction

Post-operative care in radical mastoid surgeries is challenging due to many possible problems. These problems include, late cavity epithelialization, wax and epithelium debris impaction, overwhelming infection, and not ideal amplification.[1]

Soft posterior canal wall reconstruction and mastoid exclusion procedures are aimed to overcome these problems.[2] Traditional soft posterior canal wall reconstruction although helpful in some patients, has negative points such as difficult technique and common retraction of the posterior wall. Frequent posterior wall retraction in the absence of large meatoplasty results in difficulties in cavity post-op care.[3]

Mastoid obliteration with hydroxyapatite has some complications and infection and in most of patients does not lead to smaller cavity.[4,5] Bone pate is another option, but in case of post-op infection it resorbs and no obliteration can be achieved.[6]

Here we present a new technique of soft posterior canal wall reconstruction and long term results.

Methods

Cholesteatoma patients undergoing canal-wall-down procedure were included in the study (2000-2011). Seventy-seven patients had good follow up and technique related outcomes could be studied such as achievement of posterior-canal-wall reconstruction, self-cleaning external auditory canal, retraction of canal wall, and flap necrosis.

After retro-auricular skin incision, temporal fascia graft was harvested. In order to elevate the periosteal flap, incisions were made more posterior than usual and a flap of twice the usual length (antero-posteriorly) was elevated (Fig. 1). After canal-wall-down mastoidectomy, a long temporalis fascia graft was inserted beneath the anterior remnant of the tympanic membrane and extended laterally posterior to the vascular strip. The muscle-periosteal flap was then pushed posterior to the fascia to be a soft-tissue support of the posterior canal. If good obliteration was achieved, no further meatoplasty was made.

This study is a retrospective analysis of clinical records, operative notes and audiologic data of patients.

Data from every patient were collected from pre-operative records, operative notes, and post-operative office visits. Statistical Package for Social Sciences (SPSS; version 16.0) was used for the data analysis. Parametric and non-parametric statistical tests were used to compare different variables. The statistical significance was set at $p < .05$ two tailed.

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Edited by Haruo Takahashi

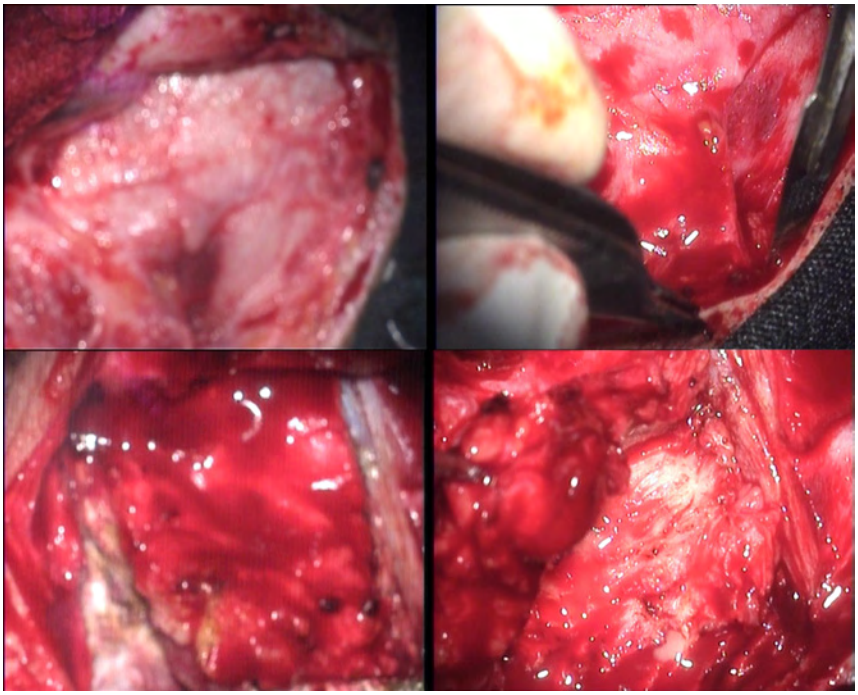

Fig. 1. Harvesting the long palva flap: 1) usual post-auricular approach; 2) elevating posterior skin flap to have access to the periosteal flap; 3) incising flap; 4) elevating flap.

Results

The mean follow-up time was 34 months (range: 10-84). In all of our 77 patients obliteration was possible, but in seven patients (9%) the mucoperiosteal flap could not completely obliterate the mastoid cavity and meatoplasty was performed. In patients with complete obliteration no meatoplasty was necessary.

Four patients (5.2%) had flap necrosis in post-op visits. After out-patient debridement of necrotic muscle, the mastoid cavity enlarged but not to an extent needing revision surgery for meatoplasty.

Ten patients (13%) underwent second-stage surgery for hearing rehabilitation. In these patients, sharp dissection with micro-scissors was used to elevate the flap sharply. None of these cases had flap-related complications. Among patients undergoing ossicular reconstruction, 80% had successful hearing improvement (15 dB decrease in air-bone-gap at least).

Discussion

Concerns regarding post-op problems of the canal-wall-down procedure are the main issue resulting in debates about choosing canal-wall-up or -down procedures. Canal-wall-up mastoidectomy, although it eliminates CWD post-op problems, has a higher rate of recurrent or residual disease.[7] The mastoid-obliteration technique helps the surgeon to benefit from both procedures: a more efficient removal of cholesteatoma and a small post-op external ear canal.

Our results show that the mastoid-obliteration technique by excluding the mastoid cavity from the external ear canal can really help to achieve a self-cleaning, smaller ear canal.

Meatoplasty is performed for patients undergoing canal-wall-down mastoidectomy in order to achieve better ventilation of the cavity (which decreases fungal infections) and makes cavity care and wax removal easier and feasible.[7] However, it does not look natural and patients complain of its cosmetic results. One of benefits of our technique is that it eliminates the need for meatoplasty and has better cosmetic results.

Vertigo due to caloric stimulation of lateral semi-circular canal is not unusual in canal-wall-down patients, especially those who had a LSCC fistula.[8] None of our patients had vertigo during swimming, or equally caloric stimulation. This was because the muscle had covered the LSCC area efficiently.

One of the negative points of our technique is dependance on flap vasculature. In revision cases fibrosis and sacrified perfusion of the flap may lead to flap necrosis. majority of our patients with post-op flap necrosis were revision cases. In conclusion, obliteration of mastoid cavity with a long palva flap seems to be a good option for radical cavity procedures. The main benefits are that meatoplasty is not needed, it provides a better cosmetic result, it does not limit physical activities such as swimming, there is no risk of vertigo due to labyrinthine stimulation by cold water in the cavity, and most importantly, it eliminates the need for long-term cavity care. The negative point is that by using this flap, second-stage surgeries are technically demanding or should be done by the retro-auricular approach, but it is feasible in most of patients.

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THE ROLE OF CHOLESTEATOMA ON THE BONE CONDUCTION THRESHOLDS IN CHRONIC SUPPURATIVE OTITIS MEDIA

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Introduction

Chronic suppurative otitis media (CSOM) is still one of the diseases with high incidence in Indonesia. CSOM incidence based on data from the Ministry of Health in Indonesia in 1994-1996 was 3.8%.[1] The most frequent complication of CSOM is hearing loss.[1,2] There are three types of hearing loss: conductive, sensorineural and mixed hearing loss with degrees ranging from mild to very severe.[1,3] The increasing of the air conduction threshold with a normal bone conduction threshold are the signs of hearing loss in CSOM. In contrast, some researchers reported sensorineural hearing loss (SNHL) in CSOM as a result of a decrease of the cochlear function.[4]

The correlation between SNHL and CSOM still is relatively unknown.[4,5] Gardenghi cited by Kaur[4] reported that 22 (44%) from 50 CSOM patients had a SNHL. Bluvstein reported that 37,5% patients CSOM had a SNHL, and Paparella *et al*. cited by Kaur[4] reported there is increase of SNHL prevalence in 232 CSOM patients. Cusimano *et al*., Levine *et al*. and Paparella *et al*. cited by Tuz[6] reported that the SNHL degree was associated with the duration of the disease, inflammation or the histopathological change of the middle-ear mucosa and cholesteatoma.

The purpose of this study is to determine the effect of cholesteatoma on bone-conduction thresholds in CSOM patients. It will clarify the effect of cholesteatoma on cochlear damage in CSOM patients.

Methods

The design of this study is cross sectional, using the medical record of CSOM patients. The subjects of this study are CSOM patients with and without cholesteatoma who visited the Dr. Saiful Anwar hospital in Malang from January 2006 to December 2009. All subjects underwent mastoidectomy surgery.

The variables measured included pre-operative bone conduction in four frequencies as dependent variables. Data of duration of otorrhea, the destruction of ossicles (incus, maleus, stapes) and the types of CSOM (with and without CSOM) were collected as independent variables. The difference of bone conduction in the types of CSOM and the destruction of ossicles were tested using an independent t-test. A correlation test was used to analyze the correlation between the duration of otorrhea and the mean of bone conduction for all types of CSOM and for each type of CSOM separately.

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Edited by Haruo Takahashi

Results

This study involved 82 CSOM patients: 56% with and 44% without cholesteatoma. The sex ratio in CSOM patients with and without cholesteatoma showed no significant difference (p = 0.83), neither did the mean age in both of types CSOM.

The mean duration of otorrhea complaints in CSOM with cholesteatoma was significantly longer (p < 0.001) than in CSOM without cholesteatoma. The ossicle destruction (maleus, incus, stapes) in CSOM with cholesteatoma was significantly higher (p < 0.001) than in CSOM without cholesteatoma. Bone conduction in patients with CSOM with cholesteatoma was higher in all frequencies and the highest in 4 KHz. The unpaired t-test showed that the mean bone conduction of CSOM with cholesteatoma (mean = 32) was significantly higher (p = 0.003) than without choleasteatoma (mean = 20). (Table 1.)

Table 1. Clinical characteristics.

	Type of CSOM		*Statistic test*
	With cholesteatoma	*Without cholesteatoma*	
Duration of otorrhea	7.6	2.2	< 0.001
Ossicle destruction			
Maleus	40	16	Chi square
Incus	45	19	P < 0.001
Stapes	44	18	
Bone conduction			
Frequency			
500 Hz	29.6	17.6	p = 0.034
1 KHz	28.7	16.2	p = 0.001
2 KHz	33.3	23.9	p = 0.004
4 KHZ	36.2	22.5	p = 0.091
Mean	32	20	p = 0.003

The mean bone conduction was higher in CSOM with destruction of ossicles in maleus (p < 0.001), incus (p = 0.03) and stapes (p = 0.04) than without destruction of ossicles. These results indicate there was correlation between ossicles destruction (maleus, incus, stapes) with the risk of hearing loss as showed by the increasing of the mean bone conduction.

The correlation test also showed that a longer duration of otorrhea meant that a higher bone conduction was found (r = 0.36; p = 0.01). In another analysis, there was no correlation between the duration of otorrhea and bone conduction in CSOM without cholesteatoma (r = -0.14; p = 0.4). A significant correlation between duration of otorrhea and bone conduction was found only in CSOM with cholesteatoma (r = 0.4; p = 0.01).

Discussion

This study identify bone conduction thresholds differences between CSOM patient with (46) and without (36) cholesteatoma. The group of CSOM with cholestatoma had a mean 7.6 years of duration of otorrhea, the group of CSOM without cholestatoma had a mean 2.2 years. In the study conducted by Kaur,[4] the duration of otorrhea in most CSOM patients was less than five years. Cholesteatoma is a medium or good environment of bacterial growth and CSOM with cholestatoma would mean a persistent bacterial infection. On the other hand, the bacterial infection will stimulate the cholesteatoma to secrete several cytokines, such as IL-1α, IL-1ß, IL-6, TNF-α and GM-CSF. These cytokines, particularly TNF-α, play a role in keratinocyte proliferation and differentiation, so that the cholesteatoma will grow faster. The presence of these cytokines will also enhance the inflammatory process resulting in chronic infection which is characterized by an increasing duration of otorrhea.[7-9]

The cholesteatoma and infection have a synergistic effect. Cholesteatoma is a good medium for bacterial growth so the infection becomes persistent. The persistence of bacterial infection stimulates the cholesteatoma to secrete several cytokines that can enhance the growth of cholesteatoma.[7-9] Cytokines TNF-α and IL-1ß, which increase in the chronic infection, can increase the permeability of the round-window membrane. It in-

creases the absorption of toxic inflammatory mediators or materials to the cochlea. The biochemical changes in perilymph and endolymph will damage the outer and inner hair cells in the cochlea that cause SNHL.[9]

Cytokines secreted by cholesteatoma can increase the osteolysis process; which increases the destruction of ossicles. The results of this study also show that the destruction of ossicles in the group of CSOM with cholesteatoma was higher than in the group of CSOM without cholesteatoma. Cholesteatoma can increase osteoclasts through several cytokines. TNF-α causes damage to the bone through the differentiation and maturation of osteoclasts directly and through exposure of the bone matrix indirectly. Other cytokines, such as IL-1, IL-6 and RANKL (receptor activation of NF-κ B ligand), are also found in areas of inflammation and are associated with bone destruction. These cytokines can stimulate differentiation and activation of osteoclast synergistically, thereby increasing the ossicle destruction.[7,10] Cholesteatoma can also damage the lateral semicircular canal and extend into the cochlea. Inflammatory mediators may enter into the cochlea through the opening of the semicircular canal. They can damage outer and inner hair cells and lead to SNHL.[7,8]

The duration of otorrhea in the group with CSOM with cholesteatoma was longer than in those without cholesteatoma. In the study conducted by Kaur[4] they found that the duration of otorrhea was proportional to the increasing incidence of SNHL in CSOM. SNHL in CSOM is due to an increase of biochemical materials in the cochlea through the round window. The structure of the round-window semi-permeable membrane allows the toxic substances to enter into the cochlea. The round window has a groove with a depth of one mm and a diameter of two mm. The surface of this window does not have a ciliated cell. This condition causes the secretion to pile up, restrain, and to be absorbed into the perilymph. There is a significant histological change in the middle ear with otitis media with purulence secretion compared to those without it.[4] This is consistent with the results of this study that show increasing bone conduction is proportional to the duration of otorrhea, especially in CSOM with cholesteatoma.

The conclusion of this study is that CSOM with cholesteatoma increases the bone conduction thresholds. The destruction of ossicles and duration of otorrhea may affect the relationship between cholesteatoma with an increase of the bone conduction thresholds.

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POST-OPERATIVE LONG-TERM HEARING RESULTS IN PATIENTS WHO UNDERWENT OSSICULOPLASTIES

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Introduction

We empirically know that the hearing level of patients changes over time after surgery on their ears. To verify this empirical knowledge, we recently analyzed and investigated the following points: (1) When is the best post-operative hearing obtained? (2) How does post-operative hearing change? (3) Is there any difference in post-operative hearing among operative procedures? (4) What are possible factors for changes in post-operative hearing?

Materials and methods

The study involved 367 ears treated with ossiculoplasty in our hospital from April 2002 to April 2009. Gender and age distributions of the patients were 166 males and 201 females aged six to 79 years for 200 left and 197 right ears. A comparative study of operative procedures involved the following four types: tympano-stapedial interposition with incus (III-c; 127 ears); malleo-stapedial interposition with incus (III-i;120 ears); tympano-stapedial footplate interposition with incus (or cortex bone)(IV-c; 65 ears); and malleo-stapedial footplate interposition with incus (or cortex bone)(IV-i; 55 ears). The underlying disease was cholesteatoma (including congenital cholesteatoma) in 202 ears, chronic otitis media in 92 ears, traumatic ossicular dislocation in 16 ears, tympanosclerosis in 34 ears, ossicular anomaly in 21 ears and others in two ears.

Results

We analyzed the follow-up data for a period of three years after surgery. In cases of III-c procedures, the air conduction hearing level was highest at one year after surgery, although the difference was not statistically significant. After one year, the hearing level in III-c tended to gradually decrease over time. Cases of III-c procedures finally recorded a 2.7 dB elevation in air-conduction hearing threshold. The time course of air-bone gap levels was similar to the time course of air-conduction hearing level. The bone-conduction hearing level rose by 1.9 dB (Fig. 1A). Cases of III-I procedures followed a course similar to that followed by III-c cases. Cases of this procedure also had the highest haring level at one year after surgery, although the difference was not statistically significant. The air-conduction hearing threshold rose by only 1.5 dB, but the bone-conduction threshold rose significantly by 2.5 dB (Fig. 1B). IV-i cases also followed a course similar to cases of III-c and III-i, with the air-conduction threshold rose by 3.2 dB and the bone-conduction threshold by 1.9 dB (Fig. 1C). Unlike the cases having undergone other operative procedures, cases of IV-c recorded the highest air-conduction hearing immediately after surgery. In Cases of IV-c, the bone-conduction threshold

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Cholesteatoma and Ear Surgery – An Update, pp. 325-327
Edited by Haruo Takahashi

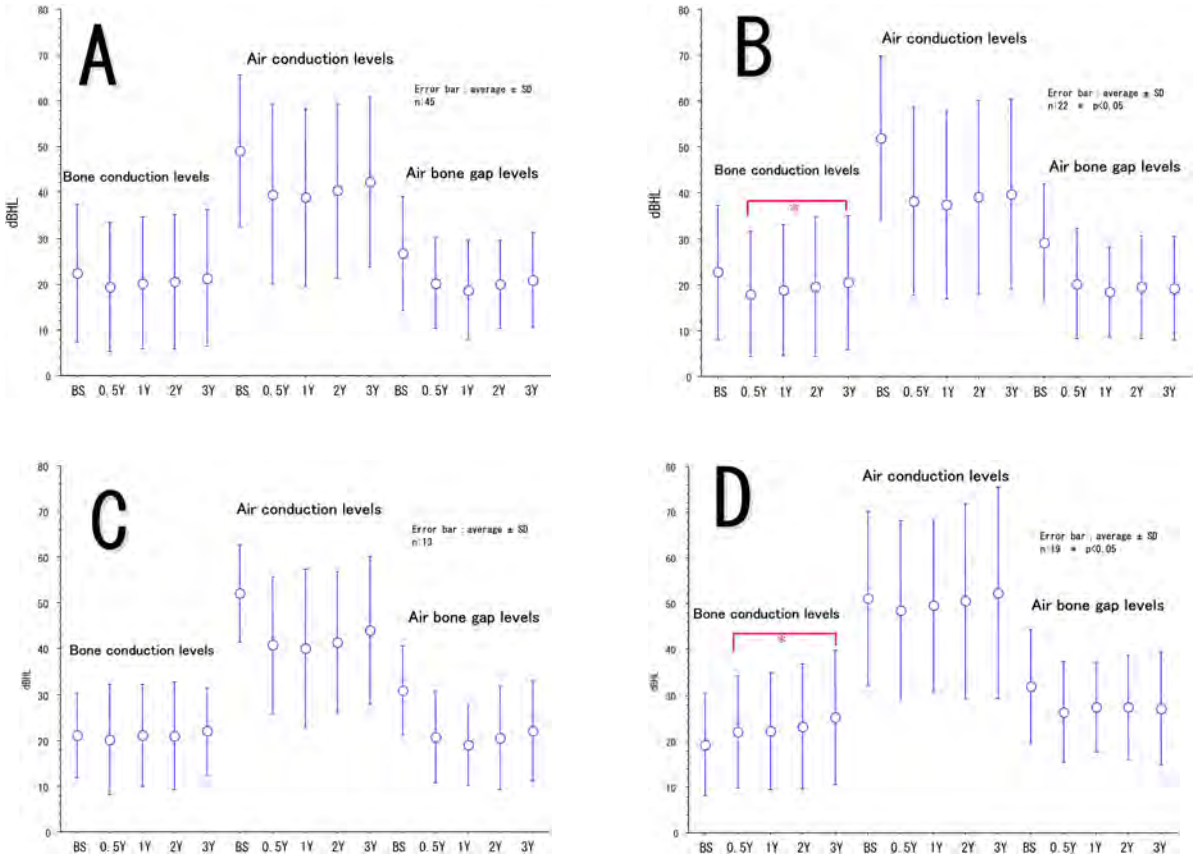

Fig. 1. The time course of bone hearing levels during the three-year period after each operative procedure for ears. A: III-c; B: III-I; C: IV-I; D: IV-c. Graphically, from left to right: the mean bone-conduction hearing; mean air conduction hearing and mean air-bone gap levels immediately before surgery, immediately one to six months) after surgery; and one, two and three years after surgery. The vertical axis indicates the hearing level.

rose significantly by 3.1 dB (Fig. 1D). There was no statistically significant difference in the outcome between any two of the four operative procedures.

Next, we compared six-year post-operative hearing data regardless of operative procedure between two groups divided by age at a cut-off level of age 45. The group aged 45 or younger showed no significant change in air-conduction threshold, bone-conduction threshold or air-bone gap levels even at six years after surgery (Fig. 2A). In the group aged 46 or older, however, significant elevation was recorded in the air-conduction threshold (elevation by eight dB) and in the bone-conduction threshold (elevation by 7.6 dB) during the period from immediately after surgery to six years after surgery. During this period, no significant change was noted in air-bone gap levels (Fig. 2B).

Discussion

As far as the outcome of hearing level after tympanoplasty is concerned, numerous reports comparing the short-term hearing data have been published.[1-5] However, few reports are available with a detailed analysis of long-term course of hearing level.[6-8] In these reports, the success rate often remained unchanged during the five- or six-year period, and the investigators reached the conclusion that the post-operative hearing data were stable.

However, post-operative hearing level was highest at one year after surgery and tended to decrease gradually after that point. Therefore, we cannot rule out that if the patients are followed for longer periods of time, significant changes in hearing level are revealed. When we analyzed changes in bone-conduction threshold, a significant elevation was noted following surgery with procedure III-i or IV-c. According to a past report, the post-operative bone-conduction threshold is usually improved compared to the pre-operative threshold.[9] Also in our study, the bone-conduction threshold improved for a while after surgery, but it aggravated over

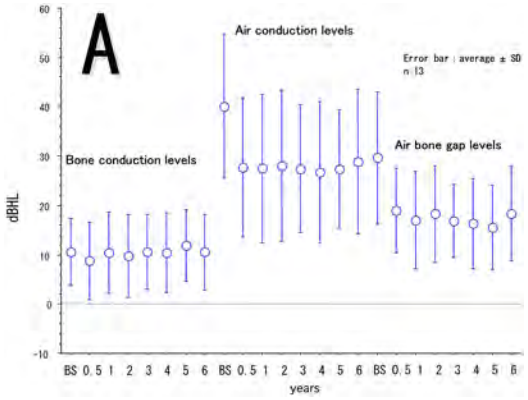

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Fig. 2. The post-operative hearing results divided by age. A: under 45; B: over 46.

time thereafter. To examine whether or not such a change would be associated with age, we analyzed the course of the bone-conduction threshold in relation to age. It was shown that bone-conduction threshold showed age-related changes. During this period, however, no significant change was noted in air-bone gap levels. This indicated that there was no deterioration of hearing associated with the operative manipulation involved in tympanoplasty, but rather that deterioration of air-conduction hearing occurred as the final output because of elevation in bone-conduction threshold. In other words, we may at least say that in elderly patients, age-related aggravation of bone conduction leads to deterioration of air-conduction hearing level over a long period of time after surgery.

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THE ANTERIOR ENDOSCOPIC TRANSNASAL APPROACH TO PETROUS APEX LESIONS

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Introduction

The petrous apex is bounded by the bony labyrinth and internal carotid artery (ICA) anteriorly, the posterior cranial fossa and Dorello's canal (cranial nerve VI) posteriorly, the middle cranial fossa and Meckel's cave superiorly, and the jugular bulb and inferior petrosal sinus inferiorly.[1]

The petrous apex is uncommonly tangled by disease, the involved pathologies are cholesterol granuloma, effusion, petrous apicitis or osteomyelitis, petrous bone cholesteatoma, mucocele, chondromas, chondrosarcomas, arachnoid cysts, vascular malformations, meningiomas and metastases which are extremely rare.[2]

The clinical symptoms of pathologic processes of the petrous apex are often vague but may include diplopia, hearing loss, vertigo, headaches, and facial nerve weakness. Hearing loss and vestibular abnormalities may present as the lesion enlarges and encroaches on the internal auditory canal. Petrous apicitis may present as Gradenigo's syndrome with a sixth cranial nerve palsy.[3,4]

PACG is the most common benign, cystic lesion of unknown etiology. Many theories have been postulated for its formation. The most accepted one is the 'obstruction-vacuum' theory, which entails obstruction of the petrous apex air cell outflow tracts, with consequent development of a vacuum, leading to transudation of blood with hemoglobin into the apex cells and cholesterol liberation that stimulate a foreign-body reaction resulting in granuloma formation.[5]

The diagnosis of a cholesterol granuloma can often be made on the basis of its radiologic appearance, as expansile and erosive mass with well-defined margins on computerized tomography (CT). They have a high intensity on both T1 and T2-weighted magnetic resonance imaging.[6]

There are two plans in managing the petrous apex cholesterol granuloma: the wait-and-see policy and surgical treatment. The first option is suggested to asymptomatic patients who are diagnosed incidentally by imaging and the symptomatic patients that have poor general conditions. These patients are followed by a series of radiologic evaluations with MRI/CT. The surgical treatment is recommended to patients with symptomatic disease and patients having cranial nerve affection.[7]

Several approaches have been proposed in literature according to the situation and expansion of the lesion through the lateral skull base and the transnasal-transsphenoidal route.[9-11] We describe our experience in the treatment of three cases with PACG via a navigation-guided transnasal route in the main university hospital of Alexandria.

Patients and methods

Ethical considerations

A retrospective study was conducted, after the approval of the Institutional Review Board (IRB) of Alexandria University Hospital, on the charts of three patients (two females and one male) with petrous apex cholesterol

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granuloma managed using the endoscopic anterior transsphenoidal approach at the Department of Otorhinolaryngology – Head & Neck Surgery, Alexandria Main University Hospital, from January 2006 to December 2011. An informed consent from all the patients was taken prior to surgery after they were fully informed about the details of the surgical procedure.

Diagnosis and pre-operative evaluation

Careful history and complete otorhinolaryngological examination including endoscopic nasal examination using a Karl Storz 4 mm 0° rigid nasal endoscope was performed in all cases. A thorough physical examination to assess the patient's general condition and to exclude any other associated lesions was also done prior to surgery.

After endoscopic evaluation, radiological imaging studies to localize the defect in the form of multi-detector computed tomography (MDCT) with ultra-thin 1-mm cuts, bone and soft-tissue settings, with multi-planar reconstruction, CT carotid angiography and magnetic-resonance imaging (MRI) high resolution coronal T2-weighted sequences were done. Complete pre-operative routine laboratory investigations were also carried out on all patients.

Surgical technique

The surgical procedure was done under controlled hypotensive general anesthesia. The surgery was performed with the patient lying supine; head slightly extended and rotated 15° to the right. Intra-operatively, the GE InstaTrak™ 3500 electromagnetic navigation system (General Electric Company, GE Healthcare) was used in all cases. The head, face and anterior aspect of the thigh were sterilized and covered with sterile surgical drapes. Nasal decongestant cottonoids impregnated with Oxymetazoline 0.05% and Xylocaine were applied in both nasal cavities for ten minutes before surgery started. We prefer to perform endoscopic transnasal surgery via both nasal fossae using the 'four-hand' technique, in which two surgeons work simultaneously inside the nose after connecting the nasal endoscope to a high-definition video camera and a video monitor.

The surgery was commenced via the endonasal route, sphenoidotomy was performed on both sides as laterally as possible. A posterior septal window was created by removing the posterior part of the bony nasal septum to allow working through both nostrils. In some cases, partial superior turbinectomy and resection of the tail of the middle turbinate were necessary. The optic nerve, the carotid canal buttresses, planum sphenoidal and clival recess were visualized as landmarks.

The petrous apex is situated deep between the horizontal and vertical segments of the petrous ICA. The horizontal portion of the ICA runs parallel and deep to the Eustachian tube and the mandibuler nerve. The ICA turns vertically at the second genu at the level of the clival recess. The vidian artery and its course through the pterygoid canal are useful and reliable landmarks for locating the second genu of the ICA. Large expansile lesions of the petrous apex that expand medially into the sphenoid sinus were easily recognized and create a window for access between the brainstem dura and ICA at the level of the clival recess.

The overlying mucosa is stripped, and the bone overlying the cyst is thinned with a three-mm coarse diamond burr. Drilling started along a vertical plane to avoid injury to the ICA, the bone was thinned out and the rest was removed with a one-mm angled Kerrison rongeur, and the lesion was completely exposed. The cyst wall was opened with a sickle knife, the chocolate content erupting under pressure was evacuated with suctions, curettes, and repeated irrigation. The orifice created was then enlarged as much as possible to establish the necessary aeration and avoid recurrence. There was no need to use any stents in the cyst cavity.

All cases were given an intravenous antibiotic chemoprophylaxis (third-generation cephalosporin) that started on the day of surgery and continued for seven days post-operatively. Patients and parents were given strict instructions to avoid straining or nose blowing for a minimum period of one month and were given stool softeners during this period. The patients were monitored for any CSF leak, signs of meningitis, hemorrhage, neurologic complications. Nasal packing was removed on the second or third post-operative day, and the patient was discharged. The patients were followed up weekly for the first month and then every two weeks for another month. Complete healing usually took place after four to eight weeks. Radiologic follow up was performed at three, six, and 12 months and after that yearly.

Discussion

Cholesterol granulomas are benign, cystic lesions filled with brownish-yellow fluid, lipids and cholesterol crystals. The term granuloma indicates a foreign body, a giant cell inflammatory reaction to blood degradation products. It can be found inside air cells of temporal bone, that is, middle ear, mastoid, and less commonly in PA air cells.[12]

These lesions may remain clinically silent for a long time and become symptomatic if adjacent neurovascular structures become involved. A CT scan is essential to assess bone erosion, and MRI scans should be performed, as it shows high-signal intensity on both T1- and T2-weighted images. Such imaging is therefore essential to assess the feasibility of different surgical approaches as well as to distinguish the lesion from other possible PA pathologies.[13,14]

In literature, several approaches have been proposed by neurosurgeons and neurotologists through the lateral skull base and transnasal-transsphenoidal route.[15,16] However, the choice of surgical approach is based on pre-operative hearing status, location, extension of the lesion, relationship with neurovascular structures, and anatomical variations. Regarding treatment for PACGs; different surgical strategies are used to perform marsupialization of the cyst cavity and establish pneumatization using preformed petrous apex air cell tracts. However, rates of revision surgery for PACG vary from 14% to 16%.[5,8,10]

The most common approaches to use in petrous apex in a hearing ear include transcanal infracochlear, infralabyrinthine, suboccipital, and middle cranial fossa approaches. These procedures require advanced technological skill and can lead to complications associated with intracranial procedures, such as sensorineural hearing loss, vestibular injury, facial nerve damage, cerebrospinal fluid leak, or meningitis.[17-19]

A well-pneumatized sphenoid sinus facilitates identification of important landmarks, especially the course of the internal carotid artery in order to avoid inadvertent injury and complete removal of disease. When there is a medial expansion of the cholesterol granuloma into the sphenoid sinus, the surgery is straightforward with minimal technical difficulty and risk. Also, because of the recent advances in minimally invasive surgery, powered instrumentation and intra-operative image-guidance systems, skull-base endoscopy has led to new treatment options for petrous apex lesions and precise removal of the lesion with minimal surgical risk.[20-22]

Theoretically, stenosis of the opening created with an endoscopic approach can occur. Many surgeons claimed that the use of a Silastic stent to maintain the patency of the drainage tract may helps prevent recurrence of the granuloma.[6-10] We did not use any stenting in our group of cases and no recurrence rate was obtained during a follow-up period of six years. The placement of a stent may be of help when used in lateral approaches away from any possible source of infection, but when using the anterior endonasal approach, the risk of infection and a reaction to the stent as a foreign body is still high. Another point to consider is that the endoscopic approach allows for easy post-operative follow up of the surgical site.

Possible complications of the endoscopic surgery include injury of the neurovascular structure, especially ICA, and cerebrospinal fluid leak. But these can be avoided with adequate surgical technique, knowledge of anatomic relationships, recognition of anatomic variations, and the use of intra-operative navigation. However, a narrow window between the brainstem and ICA has been considered a relative contra-indication for the transsphenoidal approach.[23,24]

Conclusion

This transsphenoidal endoscopic surgical procedure is an appropriate approach for the drainage of cholesterol granulomas involving the petrous apex. It avoids craniotomy and shortens recovery with minimal post-operative symptoms and decreased hospitalization time.

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APPROACHES TO CHOLESTEATOMA WITH AN INTACT OSSICULAR CHAIN: COMBINED USE OF MICROSCOPE, ENDOSCOPE AND LASER

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Introduction

Cholesteatoma surgery focuses on complete removal of the disease in order to obtain a safe, dry ear. Because surgical access to cholesteatoma can be obstructed by the ossicles, traditional approaches remove the incus and head of malleus in order to complete extirpation of the disease. Using an exclusively endoscopic permeatal approach to cholesteatoma, preservation of an intact ossicular chain has been reported in 57% of cases.[1] A combined permeatal and transmastoid canal-wall-up approach with KTP laser but without endoscopy allowed preservation of the intact ossicular chain in 71% of cases.[2]

This report proposes that the combined use of the endoscope and operating microscope though the ear canal and also trans-mastoid when necessary, in conjunction with a fibre-guided laser such as KTP, has the potential to maximize the likelihood of preserving an intact ossicular chain when clearing cholesteatoma, and so improve hearing outcomes. The selection of instruments and approach would be governed by location of the cholesteatoma in relation to the morphology of the ear canal and mastoid, in addition to the preference of the surgeon.

Surgical technique

When considering ossicular preservation, the most challenging location for removal of cholesteatoma is the medial epitympanum.[1] The surgical approaches can be considered in the quadrants demonstrated in Figure 1, divided approximately by the coronal plane of the incudo-malleolar joint and the axial plane under the head of malleus to body of incus.

a. Postero-inferior epitympanum (tympanic isthmus)

An approach through the ear canal (permeatal, end-aural or post-aural) is clearly optimal for the tympanic isthmus (Fig. 1, quadrant *a*). Either microscope or endoscope can be utilized to visualize and remove cholesteatoma from the stapes superstructure and from under the long process of incus. Curettage of the posterior scutum is likely to be necessary. Selection of microscope or endoscope may depend upon whether the surgeon feels a two-handed approach is necessary. Caution with positioning of the tip of the endoscope is paramount when operating close to the stapes.

b. Posterior epitympanum (medial to body of incus)

The space under the body of the incus is difficult to access through the meatus if the stapes and long process of incus are intact. A trans-mastoid approach is therefore needed if the ossicular chain is to be preserved.

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Edited by Haruo Takahashi

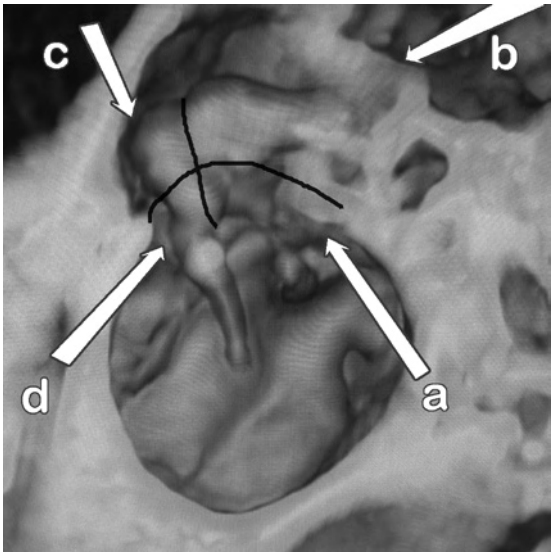

Fig. 1. CT scan of the left ear (Parasagittal plane from multi-planar reconstruction with 3-D rendering). The black cross lies along the axis of the incudo-malleolar joint and along the base of the malleus head and incus body. This divides the medial epitympanum into four quadrants for planning surgical access to cholesteatoma: a. tympanic isthmus via ear canal; b. posterior epitympanum via mastoid; c. anterior epitympanum via ear canal or mastoid; d. supra-tubal recess via ear canal.

Rotation of the patient's head away from the surgeon after drilling of the mastoid back to the sino-dural angle can often provide a clear view of the medial epitympanum with the operating microscope.[3] However, the anteriorly placed sigmoid of a poorly pneumatized mastoid, or medialized oscciular heads can limit microscope access. Selection of an endoscope or microscope for trans-mastoid access to quadrant *b* (Fig. 1) will depend on the morphology of the individual temporal bone as well as the surgeon's preference.

c. Anterior epitympanum

1) Surgical access to the anterior limit of the medial epitympanum, *i.e.*, the space under the head of the malleus, is typically clearer with angled endoscopes than with a microscope. Placement of the endoscope through the ear canal or under the ossicular chain trans-mastoid can be more or less helpful according to tortuosity of the ear canal and medialization of the ossicular heads respectively. In some cases, a combined approach using one route for the endoscope and the other for instrumentation can be effective.

2) The most challenging location for cholesteatoma with an intact ossicular chain is on the antero-medial surface of the malleus head. Combination of all currently available techniques can prove inadequate for a convincing clearance of matrix from this location. A conventional permeatal atticotomy with curette plus or minus drill can provide quite good access. A microscope may or may not be used to open up the approach, but an endoscope is likely to provide optimum visualization for cholesteatoma removal. If the tegmen tympani/floor of the middle fossa is sufficiently high, drilling of the mastoid can be continued toward the root of the zygoma, the cog drilled away and access achieved using a microscope through the mastoid. Placement of a mirror trans-mastoid or canal may improve the permeatal microscope view. The mirror can also be used to reflect the KTP laser beam on the anterior surface of the malleus head with the aim of reducing the likelihood of residual cholesteatoma in this hidden recess.

d) Supra-tubal recess and medial to malleus head

This area (Fig. 1, *d*) is best approached through the ear canal. Clear visualization is not possible with the microscope as a direct view is obstructed by the malleus handle. Angled endoscopes are ideally suited to removal of cholesteatoma from the supra-tubal recess and even the antero-medial epitympanum. Optimal access is provided by elevating the tympanic membrane from off the malleus handle.

Findings

Ethical approval was obtained to search a prospective consecutive database of 256 cases of paediatric cholesteatoma operated on by a single surgeon between 2005 and 2012. Forty-nine ears (19%) were noted to have an intact ossicular chain at the time of surgery. Most cases (94%) with intact ossicles were small cholesteatomas occupying one or two sub-sites within the tympanomastoid system. Cholesteatoma was confined to the mesotympanum alone (including the hypotympanum and retrotympanum) in 33 cases (66%). Cholesteatoma was found in the epitympanum in 16 cases (32%). Extension into the mastoid antrum was only present in three cases (two via the aditus and one via the facial recess).

It was possible to preserve the intact ossicular chain in 47 (96%) of cases. In two cases, the head of the malleus was removed to facilitate removal of cholesteatoma from the anterior epitympanum, leaving the incus *in situ*. A combined approach tympanomastoidectomy was used in one of these cases and atticotomy with cartilage scutumplasty in the other. Both were completed before the introduction of endoscopic dissection or endoscopic laser to the series.

Hearing threshold

Pre- and post-operative audiometry was available for 45 of the cases with intact ossicular chains (inadequate follow up for post-op test in three; soundfield test only available in one young child). Average hearing levels in cases with preserved ossicular chain were unchanged after surgery with median pre- and post-operative air conduction thresholds of 19 dB HL. Thirty-six ears had normal pre-operative hearing levels (*i.e.*, air conduction threshold < 30 dB HL) and 37 had normal post-operative hearing. Hearing deteriorated from normal to abnormal in only two cases (4%) after ossicular preservation surgery (mean 25 dB HL to 35 dB HL air conduction; mean change in bone conduction 9 dB HL). Mean air conduction hearing thresholds in the two cases in which the malleus head was removed deteriorated from 25dB HL to 53dB HL (mean post-op bone conduction 7dB HL).

As shown in Figure 2, the proportion of children with normal hearing was significantly greater in those with intact ossicles, than in those with incomplete ossicular chains.

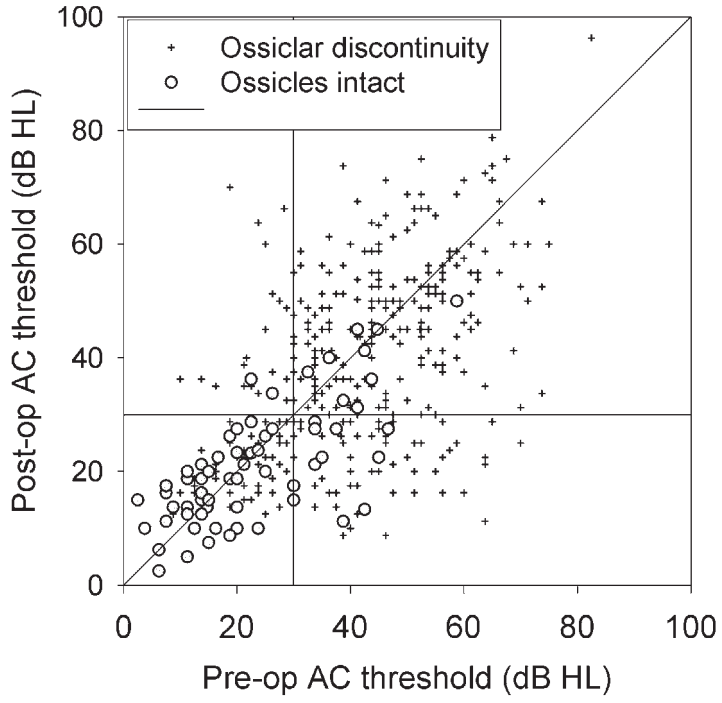

Fig. 2. Hearing thresholds before and after surgery for cholesteatoma. Mean four tone average air conduction thresholds before surgery are plotted against post-operative thresholds. Cases to the left of the vertical line had normal hearing before surgery and cases below the horizontal line have normal hearing after surgery. Hearing improved with surgery in those below the diagonal line and deteriorated with surgery in those above the diagonal line. Ossicular preservation was associated with a greater chance of normal hearing ($p < 0.001$ Chi-square test).

Conclusion

The combination of permeatal and trans-mastoid approaches, and utilization of the operating microscope and endoscopes, facilitate preservation of an intact ossicular chain when removing cholesteatoma. Ossicular preservation can be worthwhile even if the head of the malleus and body of the incus are significantly eroded. Care has to be taken to avoid the sensorineural hearing loss from cochlear trauma that could result from excessive manipulation of the ossicular chain, but this complication was not found in this series or others.[1,2,4] The KTP laser facilitates removal of matrix from the ossicles without mechanical trauma.[2,5] Preservation of the ossicular chain maintains normal hearing thresholds in the vast majority of children with cholesteatoma.

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PREDICTIVE FACTORS INFLUENCING HEARING PRESERVATION AFTER REMOVAL OF ACOUSTIC TUMORS USING MIDDLE FOSSA APPROACH

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Introduction

With the advent of sophisticated microsurgical and anesthetic techniques, morbidity and mortality have dramatically decreased in vestibular schwannoma (VS) surgery.[1] Advances in imaging techniques and audiovestibular testing, which allowed earlier diagnosis, have led most surgeons to attempt two additional goals: conservation of facial function and hearing preservation after complete removal of the tumor.[2,10,11] In these days, the post-operative hearing preservation is becoming a critical issue as tumors are smaller when found.[3,11] Achievement of this depends on patient factors, tumor factors, as well as the surgical technique (translabyrinthine, middle-fossa, and retro-sigmoid approach).[4,7,11] Advantages of the middle-fossa approach compared to the retro-sigmoid approach include the higher rates of hearing preservation, nearly complete exposure of the IAC, and the higher possibility of surgery even if the tumor is involved to the lateral IAC.[5,8] These advantages have made the middle-fossa approach an otolaryngologist-friendly technique.

The aim of this study was to analyze predictive factors that influence post-operative hearing level after removal of VS using middle fossa approach, and to re-evaluate the previous limitations inherent in this approach.

Material and methods

The study participants are 16 patients who received the surgery for removal of acoustic tumor using middle-fossa approach from January 1st 2002 to January 1st 2012. Two groups were hearing preserved group (50 dB and above, n = 8) and unpreserved group (less than 50 dB, n = 8) according to the post-operative hearing level. They were compared retrospectively by means of predictive factors, including the size of the tumor, the presence of extension to cerebellopontine angle (CPA) and the lateral fundus, the presence of tumor adhesion to the facial nerve, the type of nerve originating the tumor, the course of facial nerve, degree of post-operative facial-nerve palsy and the pre-operative hearing level.

Results

The size of the tumor, the presence of extension to CPA and lateral fundus, degree of adhesion between the tumor and facial nerve, the course of facial nerve, and the degree of post-operative facial nerve palsy were not significantly different between the two groups ($p > 0.05$) (Figs. 1 and 2). There was no significant correlation of the length of extension to CPA between two groups either (Fig. 2). Tumors arose most commonly from the superior vestibular nerve, in 56% of patients, followed by the inferior vestibular nerve in 44%.

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Edited by Haruo Takahashi

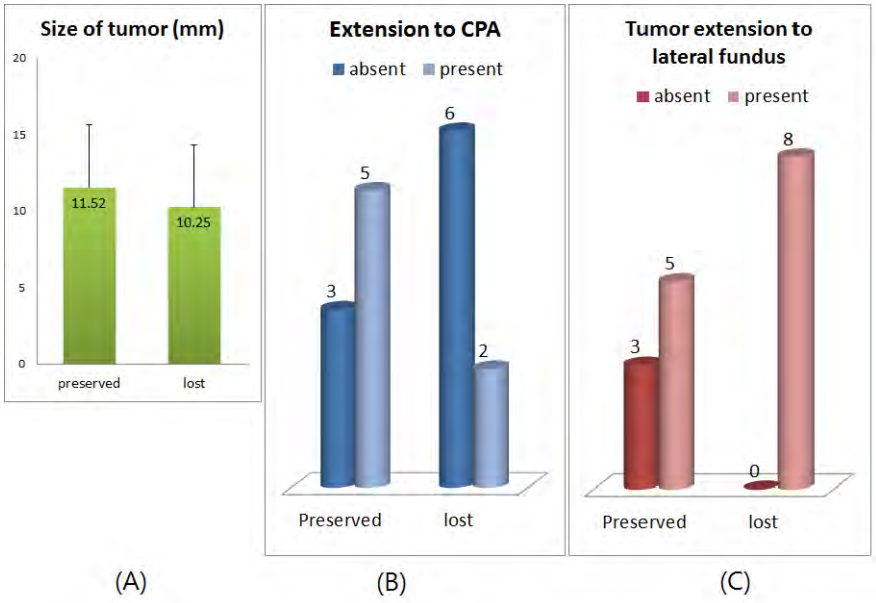

Fig. 1. Comparison of the size of tumor (A) and the presence of extension to cerebellopontine angle (CPA) (B) and lateral fundus (C) between two groups ($p > 0.05$).

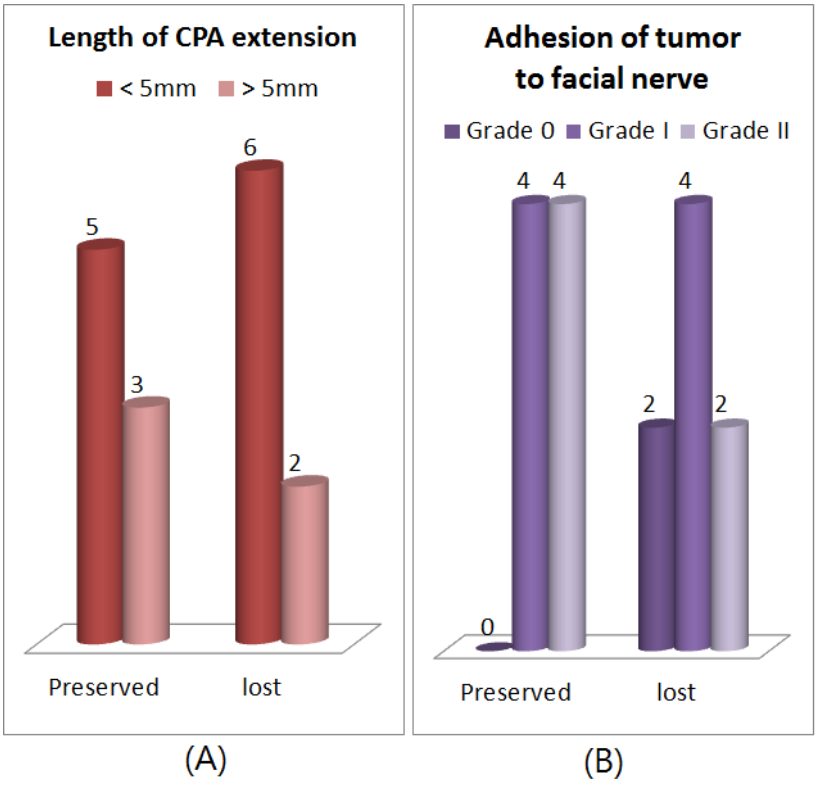

Fig. 2. Comparison of the length of extension to CPA (A) and the degree of adhesion between the tumor and facial nerve (B) between two groups.

Discussion and conclusion

In previous studies, factors that influence the post-operative hearing level after the middle-fossa approach have been pre-operative hearing level and the degree of tumor extension to CPA or IAC lateral fundus.[1,6,11] Better pre-operative hearing level means there is less adhesion between the tumor and the auditory nerve. Likewise,

less extension to the fundus means better surgical field exposure.[9] The extension of the tumor to the lateral fundus was known to be a contra-indication of surgical excision because of the high risk of hearing loss. However, our study indicates that there was no significant correlation between the presence of extension to lateral fundus and the possibility of hearing loss after the surgery. This suggests that the middle-fossa approach may be considered even with the presence of tumor extension to lateral fundus. More cases and standardized operation-procedure-based analysis is needed for a more generalized result to confirm this suggestion.

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CHOLESTEATOMA SURGERY, THE ENDAURAL APPROACH. WALL-DOWN TECHNIQUE WITH OBLITERATION OF PARATYMPANIC SPACES

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Introduction

Unsatisfactory results found even in cases of radical surgery, many patients presenting with cavities, and further problems: all these made us search for a way to minimize the rate of residual cholesteatoma and recurrence of the disease.

From our point of view, the most understandable definition of cholesteatoma was suggested by Jacques Magnan who said at the 2000 Conference on Cholesteatoma and Middle Ear Surgery in Cannes that cholesteatoma is a case of skin in the wrong place. As we know, cholesteatoma can be congenital or acquired and that about 80% of cholesteatomas are developed from retraction pockets. If some piece of the matrix is left after surgery we call it residual cholesteatoma. Recurrent cholesteatoma occurs when all the specific pathogenic mechanisms are repeated (for example negative pressure in the middle ear – retraction pocket – atelectasis) that finally leads to cholesteatoma of external ear canal.

There are some important features which we must mention to understand and justify our approach better. First of all, the balance of middle-ear pressure. The most sensible and clear scheme was published in the book of B. Ars[1] about the chronic otitis media pathogenesis. As such, the anterior-inferior part of the temporal bone is compared with the nose (presence of ciliated respiratory epithelium, mucociliary clearance and so on) and its posterior-superior part is compared with the lungs (alveolar-like structure, rich with superficial vessels). This scheme is very important for a clear understanding of the gas-exchange mechanisms in the middle ear and how its impairment participates in the development of cholesteatoma. In the normal ear, the main gas exchange goes through the blood vessels of the aerated mastoid cavity. In cases of inflammation, the blood flow increases and the pressure of oxygen and nitrogen decreases which leads to an increase of carbon dioxide in the middle-ear cavity and, as a result, to negative pressure. This causes retraction pockets or atelectasis formation, which can be considered as pre-cholesteatoma. The ratio between the volume of the mastoid and the area of the air cells surface in the normal ear is quite large. However, due to inflammation, this ratio decreases, which leads to further reduction of gas exchange. Even several episodes of acute otitis media can lead to a gas-exchange reduction, fibrosis of aerated spaces and predispose to cholesteatoma development. In chronic otitis media with cholesteatoma we usually find a small antrum and poor mastoid pneumatization as a result of inflammation which reduces the middle-ear gas exchange. Recent investigations in molecular biology have revealed the mechanisms of cholesteatoma perimatrix activity which is the main reason of bone resorption.[2,3] The perimatrix is rich in blood vessels. In particular, the perimatrix is active in the attraction of osteoclasts and bone resorption under the cholesteatoma. This means that you can always expect bone resorption of the facial-nerve canal, even fistula of the lateral semicircular canal, or resorption of other structures of the middle ear. The evidence of perimatrix activity can be illustrated by the following case of a patient who underwent radical surgery 20 years ago. The surgeon had left a part of the cholesteatoma matrix in the cavity, presumably concluding that this was just skin. Now we know that this was ‘wrong skin’

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Edited by Haruo Takahashi

and it was active. After 20 years, the patient developed a fistula of the lateral semicircular canal and residual cholesteatoma in the radical cavity.

Surgery

Currently there are two main techniques for cholesteatoma surgery: closed technique (wall up) and open technique (wall down). Advantages of the wall-up technique are better hygienic status and better functional outcome. But there are some disadvantages, namely a possibly narrow surgical field (not always), and the need for long-term follow up. This technique is associated with a higher rate of residual disease (about 20%), a higher rate of recurrent disease (about 13%). It is also time consuming. However, there are some published results which demonstrate zero recurrence and low residual rate after the closed technique.[4,5] It must be noted that these results were achieved with the obliteration of paratympanic spaces. Advantages of the wall-down technique are a lower rate of residuals (about 7%), a lower rate of recurrence (about 5%) and a wide surgical field. Disadvantages include the need for regular cleaning, a higher infection risk, water intolerance, hearing aid fitting problems, vertigo and decreased functional outcome. The post-operative cavity can be considered as a social stigma.

Even stalwarts of the closed technique have, after years of experience, moved to the open technique (wall down) as a better method. The data presented by the Mario Sanna group in Parma in 2010 proved this point of view. But as we mentioned above, the open technique leads to a cavity with all the associated problems. This is why we suggest a modification of the open technique to the closed.

There are several reasons why we uphold the presented approach. The first reason is the conservative tradition. For a long time, the open technique was the only one that was approved for chronic otitis media surgery with cholesteatoma. This is the result of stable conservative traditions, which depend on the educational level. In many cases, surgeons were only taught to perform the open-technique surgery.

The second reason is chimeras in the patient's mind. Very often the patient is scared of ear surgery, because the brain is near, or an incision behind the ear must be made. Often they prefer to avoid such dangerous surgery. So the endaural approach is preferable and more easily accepted by the patient, because the patient is more confident that the surgeon will not perform trepanation.

The third reason is geographic. Large regions and consequently great distances which have to be covered give rise to problems with extensive follow up and second-look surgery. When they are far from central hospitals, surgeons naturally tend to concentrate on saving patients lives and consider the patient's quality of life less important. However, this is not a satisfactory situation.

The fourth reason (the nature of what chronic otitis is) is the creation of the demarcation line around the inflammation, natural obliteration. By and large in chronic ears we find a small antrum and sclerotic mastoid cells. What advantage is there in drilling this solid bone which is not involved in inflammation? As we mentioned earlier, we can usually see the natural obliteration in chronic otitis media with cholesteatoma.

The fifth reason is that we cannot determine the edge of cholesteatoma with a CT scan and in most cases of chronic otitis it is not necessary to open the entire mastoid process. More bone can be left intact. If the surgeon has any doubts during the surgery, he can use an endoscope for inspection of the blind zones.

So taking into consideration the advantages of wall down, obliteration and endaural approach we can do the following.

- *Endaural approach, endaural incision.* It is important not to leave *in-situ* overhangs over the mastoid cavity and high remnants of the external canal posterior wall. But in the case of endaural approach, if you are planning to obliterate the paratympanic spaces it is possible to leave more bone intact with, of course, the obligatory inspection of the blind zones with an endoscope.
- *Drilling, open technique.* Cholesteatoma removing is produced with simultaneous collection of the bone paté. Closing of the tympanic cavity with the chondro-perichondrial flap is performed with simultaneous ossicculoplasty and paratympanic spaces obliteration with bone paté or bioglass and covering it with the chondro-perichondrial flap (Figs. 1 and 2).

We have been following up our patients for some years and then analyzed the results about residue and recurrence of cholesteatoma. So, from 2009 to 2011, we operated 82 ears (80 patients: 29 females and 51 males). Fifty-five operations (67%) were primary surgery, and 27 operations (33%) were revision and re-operations after surgery by other surgeons. The materials used for obliteration were cartilage (30,9%), bone paté (35,8%),

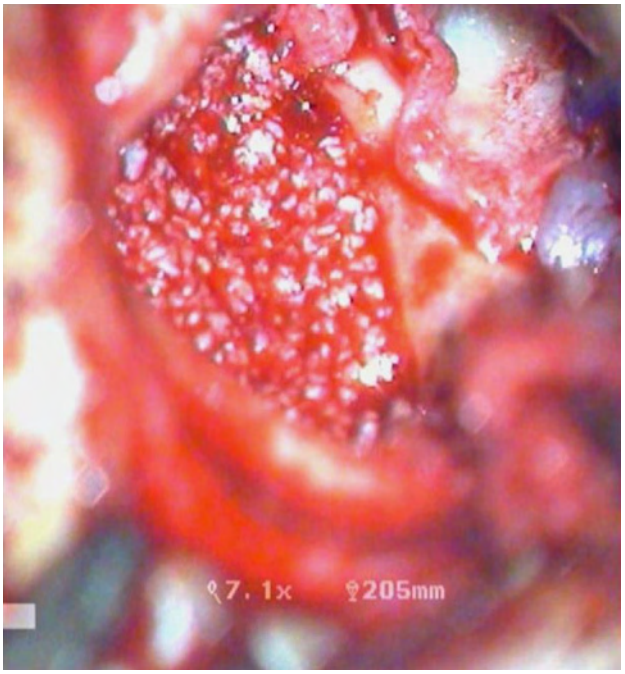

Fig. 1. Wall-down technique with obliteration.

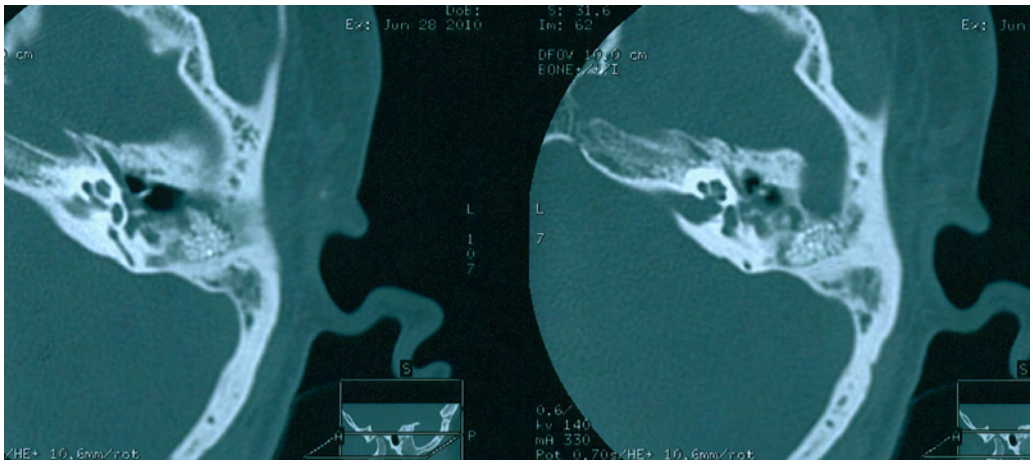

Fig. 2. Obliteration of paratympanic spaces with bioglass.

bioglass (23,5%) and both bone paté and bioglass (9,9%). We did not observe any recurrence. The residue of cholesteatoma was diagnosed in three cases (3,7%).

From 2009 to 2012, the total number of operations was 118 (116 patients: 45 females and 71 males). The primary surgery rate was 65,2%, revisions and re-operations were performed in 34,8%. The materials used for obliteration were cartilage (22,8%), bone paté (38,9%), bioglass (17,7%), both bone paté and bioglass (14,4%), both bone paté and cartilage (6%). There were no cases of recurrence. A residue of cholesteatoma was revealed in seven cases (5,9%).

If we have a big cavity with an opening of the middle and posterior fossa and the exposed facial nerve, how to obliterate it? Of course it is possible to use the soft-tissue flaps. But what if you need to revise such an ear? You will have a lot of problems to revise the scars, so it should be a solid obliteration and you should cover the most dangerous structures with pieces of cartilage. They can be landmarks during revision. If we have a big cavity evident in cases of revision surgery after radical surgery, when it is difficult to get enough bone paté we use the ‘bricks principle’. If something goes wrong you can remove the ‘bricks’ of conserved costal cartilage, correct the problem and use the ‘bricks’ again (Fig. 3).

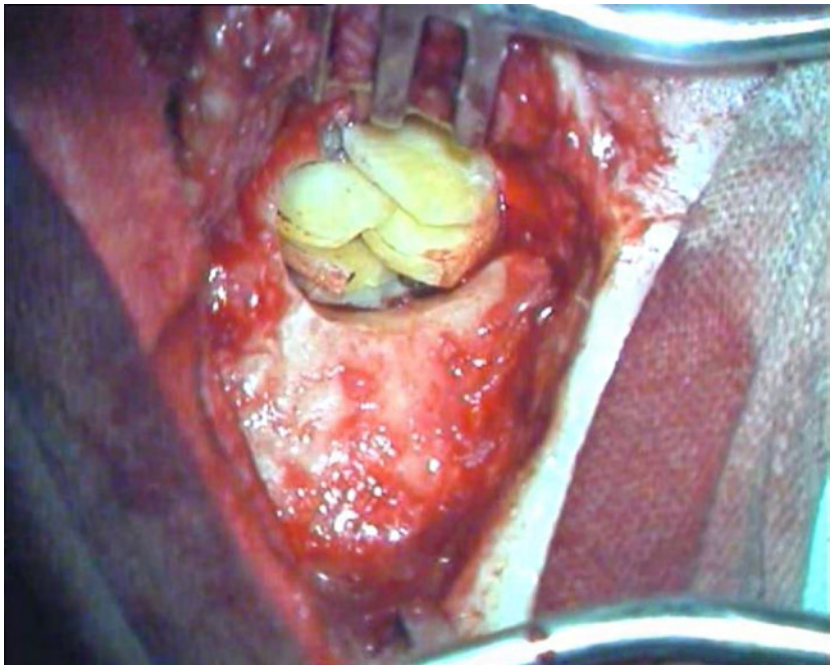

Fig. 3. Obliteration of a big cavity with cartilage ‘bricks’.

Finally, to control the situation and avoid second-look surgery, we use NON-Epi-DWI protocols which are very sensitive in cases of cholesteatoma.

It is important to note that the endaural approach is not a dogma. The choice of approach and technique is up to the surgeon.

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RECONSTRUCTION OF BONY EAR CANAL WALL IN SURGERY FOR CHOLESTEATOMA

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Introduction

In surgery for eradication of middle-ear cholesteatoma, a clear and unobstructed view of the attic or the mastoid antrum is necessary. In order to obtain proper exposure, the posterior superior ear canal wall needs to be removed (canal-wall-down mastoidectomy). In the following cohort of patients, after removal of the ear canal wall, the bony ear canal wall was reconstructed with collagen sponge, autologous bone chips and bone paste in our clinic. The aim of this technique was the reconstruction of the hard ear canal wall and the regeneration of the aerated mastoid cavity.

The status of the reconstructed ear canal wall and aeration of the post-operative mastoid cavity were then investigated.

Materials and methods

Our study included 58 ears of patients with middle-ear cholesteatoma who underwent initial surgery (staged tympanoplasty) in our clinic between April 2002 and March 2012. The patients' age ranged from four to 67 years, and the cohort included 39 male and 19 female patients.

In all ears, a cholesteatoma was eradicated after removal of the ear canal wall at the first stage operation of staged tympanoplasty (Fig. 1a). The bony ear canal wall was then reconstructed with a collagen sponge, autologous bone chips and bone paste. First, a collagen sponge and autologous bone chips were set in the original position of the canal wall (Fig. 1b). Bone paste was placed over the bone chips in order to smoothen the surface of the reconstructed bony canal wall (Fig. 1c). The second-stage operation was performed about one year after the reconstruction of the ear canal wall at the first stage operation.

At the time of the second-stage operation, the status of the reconstructed ear canal wall was classified into three groups based on wall formation: (1) good formation; (2) displaced backward formation; (3) lost formation. In group 1, bone chips and bone paste were fused with the bone of the canal wall, and the bony canal wall was reconstructed well (Fig. 2a). In group 2, bone chips and bone paste were fused to each other, but they were displaced posteriorly (Fig. 2b). In group 3, bone chips and bone paste were partially or totally lost (Fig. 2c).

The aeration of the post-operative mastoid cavity at the time of the second operation was also classified into three groups: (I) totally aerated; (II) partially aerated; (III) non-aerated. In group I, the post-operative mastoid cavity was almost totally aerated. In group II, the post-operative mastoid cavity was aerated mainly in the epitympanic portion. In group III, the mastoid cavity was not aerated at all post-operatively.

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Edited by Haruo Takahashi

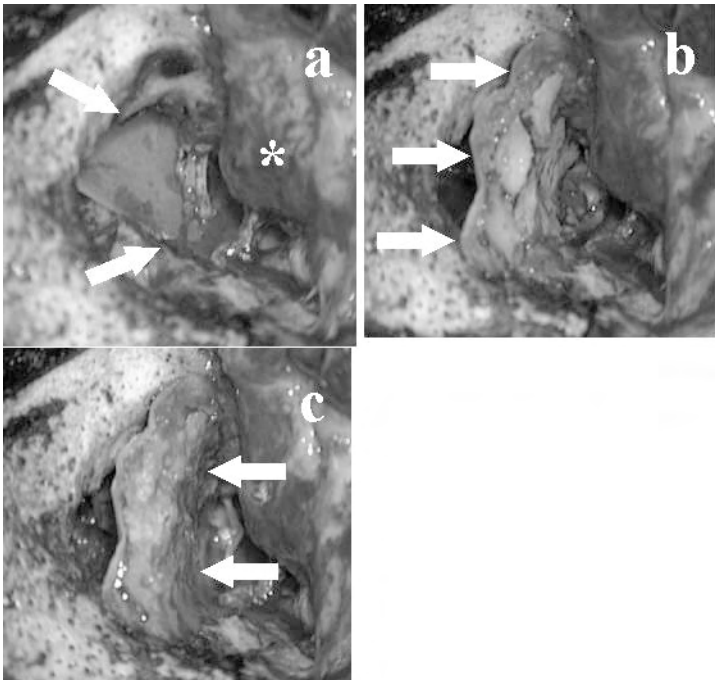

Fig. 1. The technique for reconstruction of the ear canal wall. a. Cholesteatoma has already been eradicated after removal of the ear canal wall (arrow). *The skin of the posterior wall of the external ear canal. b. A collagen sponge (arrow) and autologous bone chips were set in the original position of the canal wall. c. Bone paste (arrow) was placed over the bone chips.

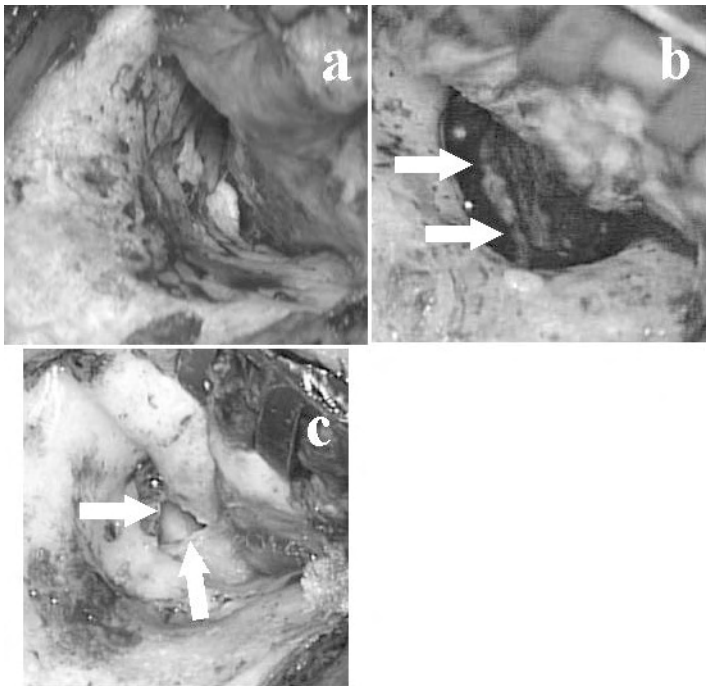

Fig. 2. The status of the reconstructed ear canal wall. a. Good formation: Bone chips and bone paste were fused with the bone of the canal wall, and the bony canal wall was reconstructed with an excellent outcome. b. Displaced backward formation: Bone chips and bone paste were fused to each other, but they showed posterior displacement (arrow). c. Lost formation: Bone chips and bone paste were partially or totally lost (arrow).

Results

Forty-nine (84%) of the 58 ears were classified in group 1. In these ears, the formation of the external ear canal was almost normal. The formation was classified as group 2 in three ears (5%) and as group 3 in six

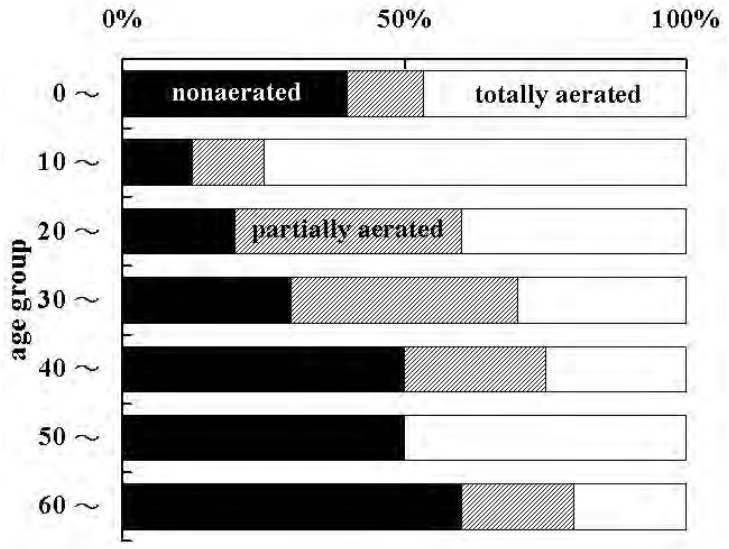

Fig. 3. The aeration of the mastoid cavity according to age group. The mastoid cavity was aerated more often in younger subjects compared to older subjects, except in those under ten years of age. The mastoid cavity was totally aerated (closed rectangle), partially aerated (rectangle with oblique line), and non-aerated (open rectangle).

ears (11%). In group 2, the formation of the external ear canal was slightly or moderately wide. In group 3, a retraction pocket recurred in three ears and cholesteatoma recurred in one ear.

The post-operative mastoid cavity was totally aerated in 25 ears (43%), partially aerated in 12 ears (21%), and not aerated in 21 ears (36%). Figure 3 shows the relationship between the aeration of the post-operative mastoid cavity and the age group. The post-operative mastoid cavity was predominantly aerated in those aged between ten and 30 years, but among patients over 40 years of age, it was not aerated in about half of the ears. The mastoid cavity was aerated in nine out of the 15 ears of those under ten years of age. The mastoid cavity was aerated more often in younger subjects, except in those under ten years of age, compared to older subjects.

Residual cholesteatoma was detected in 18 out of 58 ears (31%). In those under ten years of age, residual cholesteatoma was detected in eight out of 15 ears (53%), while it was detected in ten out of 43 ears (32%) in those over ten years of age.

Discussion

Canal-wall-down mastoidectomy is a superior procedure for the complete eradication of middle-ear cholesteatoma. However, the disadvantages of the open cavity are the necessity of periodical intervention and the possibility of infection (cavity problem). To solve these problems, after removal of posterior ear canal wall, reconstruction of ear canal wall by artificial materials such as titanium[1,2] or hydroxyapatite[3] have been reported in recent years. For restoration of the bony ear canal wall and aeration of the post-operative mastoid cavity, the posterior canal walls were reconstructed by autologous bone chips and bone paste in our clinic.

As a result, bony ear canal walls were well reconstructed and had excellent outcomes. When added together, the 84% ‘good formation’ and 5% ‘displaced backward formation’ groups yielded a clinically acceptable external ear canal in 89% of all reconstructions. In addition, 64% of the post-operative mastoid cavities were aerated. Because of the tissue regenerative ability and the short period of infection, it was thought that the post-operative mastoid cavity was aerated more often in younger subjects. However, in subjects under ten years of age, the aeration of the mastoid cavity was negatively influenced by the Eustachian-tube function.

This technique is useful for the elimination of cholesteatomas and generally yields good post-operative states of the ear canal wall and mastoid cavity.

Conclusion

Bony canal walls were reconstructed well in 49 out of 58 ears by our technique using autologous bone chips and bone paste. The post-operative mastoid cavity was aerated in 37 out of 58 ears. The mastoid cavity was aerated more often in younger subjects than in older subjects (except in those under ten years of age). This technique is useful for the elimination of cholesteatomas and generally yields good post-operative states of the ear canal wall and mastoid cavity.

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NO MORE MASTOID CAVITIES – OBLITERATE THEM ALL

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Introduction

Cholesteatoma surgery has been evolving, with canal-wall-down mastoidectomy, canal-wall-up mastoidectomy, small-cavity mastoidectomy, and obliteration, the aim being eradication of the disease, stopping the recurrence, and minimizing the problems caused by the open cavity. There have been various methods used to obliterate like flaps, free grafts, prosthetic materials, all of which have their own advantages and disadvantages. Generally, obliteration is done only for problem cavities.

Method

This is a retrospective study of 170 patients with cholesteatoma who underwent primary or revision mastoidectomy and had obliteration done between April 2005 and December 2009, with follow up of at least two years.

Patients were reviewed at three weeks, six weeks, three months, six month, and 12 months, and then annually. Following clinical examination, patients have audiograms from month three onwards, and tympanograms done after 12 months. The tympanograms of the operated ear are compared with the patient's other 'normal' ear.

My technique

I use endaural incision with laterally based posterior external auditory ear canal skin flap.

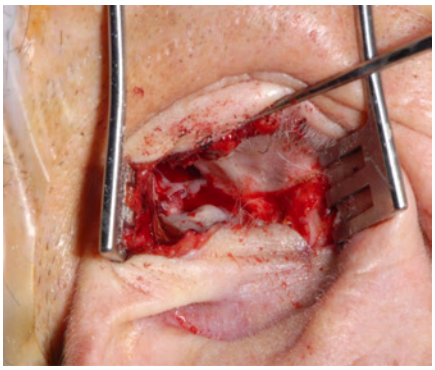

Fig. 1. Skin flap..Stage 1

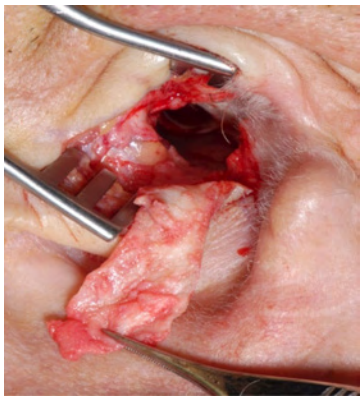

Fig. 2. Skin flap...Stage 2

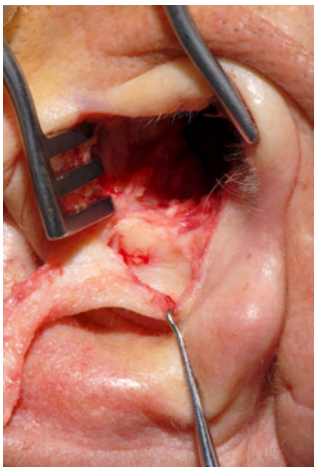

Fig. 3. Conchal cartilage

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Edited by Haruo Takahashi

Mastoidectomy is done by the ‘inside-out’ method, resulting in minimum mastoid cavity. The bony external auditory canal is widened.

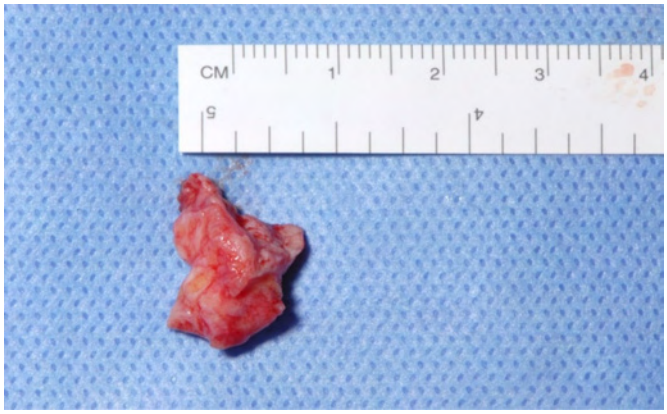
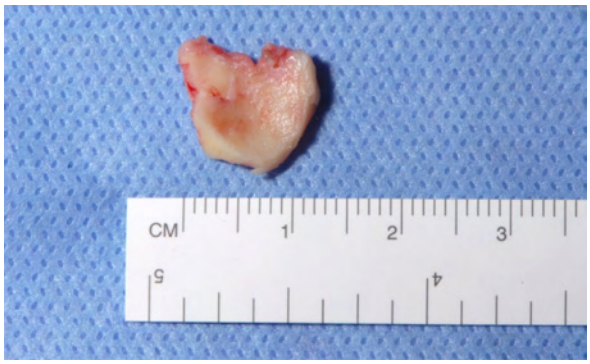


Fig. 4. Soft tissue.



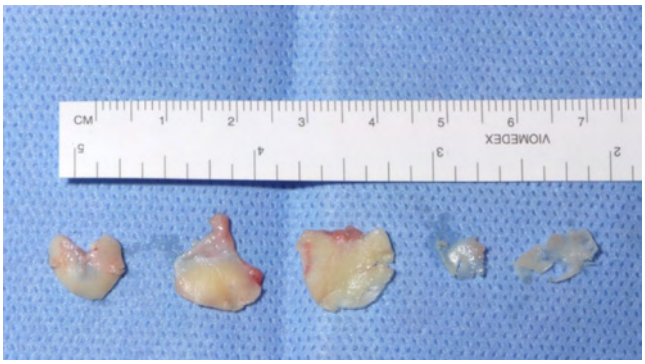


Fig. 5a and 5b. Cartilage and slices.

Conchal cartilage is taken and the soft tissue is removed to expose the mastoid bone and then both are saved in saline for later use. All cholesteatoma is removed meticulously and hidden places like facial recess and sinus tympani, anterior attic, are inspected using an oto-endoscope to confirm complete removal of the disease.

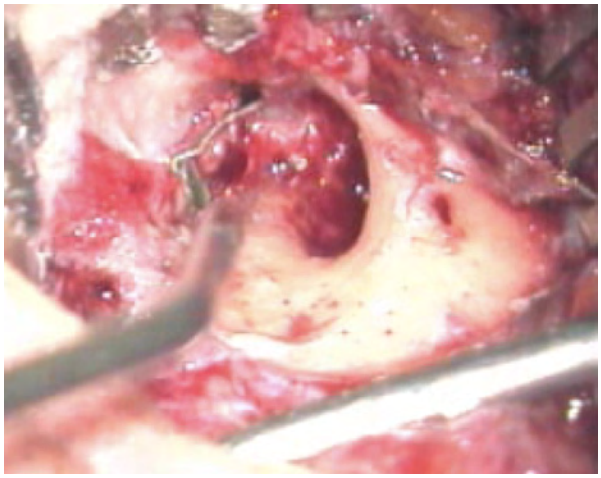


Fig. 6. Cavity at the end of the operation.

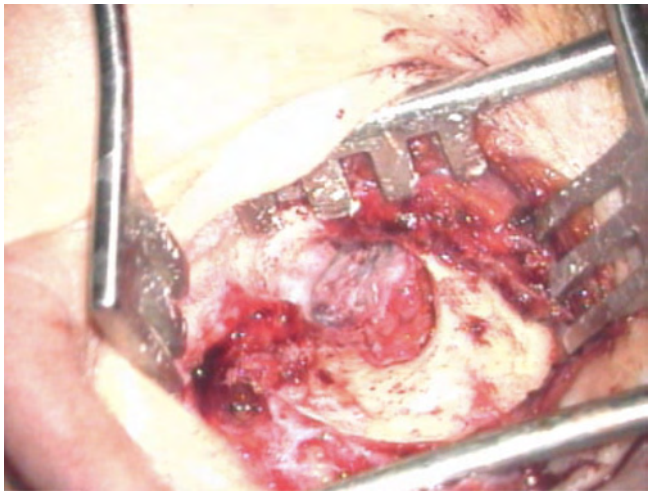


Fig. 7. Obliterated cavity.

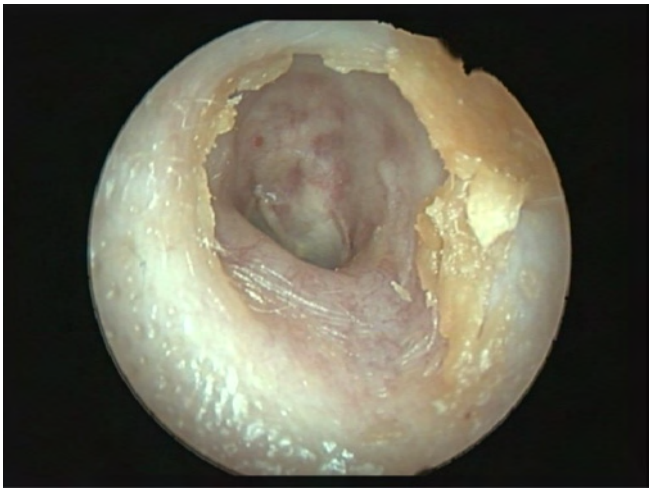


Fig. 8. Two years post-operatively.

The soft tissue removed earlier is saved in saline is used to fill the mastoid cavity. Cartilage slices are used to fill up the attic region and reinforce the tympanic membrane. Cartilage slices can also be used to cover exposed dura, exposed facial nerve, and any fistula in the semi-circular canal. The soft tissue in the mastoid bowl is also covered with cartilage slices, which is then covered with fascia. At the same time the ossicular chain is reconstructed, either with cartilage or a titanium prosthesis. We place the silastic strips in the external auditory canal, which is packed with BIPP impregnated half-inch ribbon gauze. The dressing is changed in three weeks in an outpatient clinic and the second dressing is removed at the six-week mark.

Outcome measures

The ears were assessed by improvements shown in symptoms, clinical examination, audiograms, and tympanograms, and as mentioned the patients are seen at the following intervals: three weeks, six weeks, three months, six months, 12 months, and then annually.

Of the 164 patients included in this series, only 21 failed to continue attending the follow up after various intervals, while the remaining 143 patients, had follow up ranging from two to five years at least.

Results

Of the 170 patients, only 164 are included in this study, as we could not get the notes for the remaining six patients.

Table 1 shows the comparison of my results with the UK Otology Audit Database.

Table 1.

	Local (me)	National (excl. me)
Facial nerve exposed	77 (47%)	280 (18%)
LSC Fistula (take canal fig)	15 (9%)	81 (5^)
Dura exposed	40 (24%)	–
Ossicles	Local (me)	National (excl. me)
Eroded – incus – suprastructure of stapes	139 (85%) 32 (20%)	1008 (65%) 460 (30%)
Fixed	6 (3%)	111 (7%)

It seems that the disease is more advanced in the patients presenting to us, as compared with the National database.

There were no recurrences, and no residual disease seen in any of 164 patients at last review. We have achieved our main aim resulting in dry ears in most of our patients (97 %).

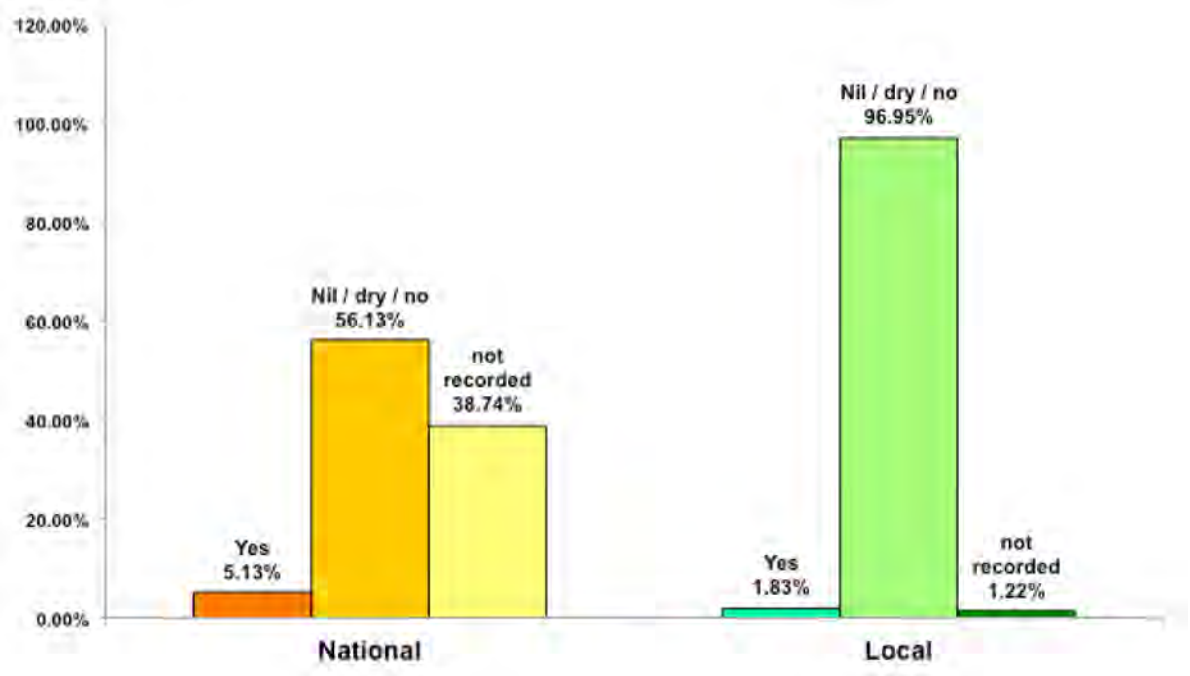


Fig. 9. Outcome comparison of of dry ears with UK Otology Audit Base

Seven patients had granulation tissue up to six months. One child had meatal stenosis. One had infection at three months. One revision mastoid case had post-aural fistula through previous scar. Two patients had epithelial pearls, treated by enucleation.

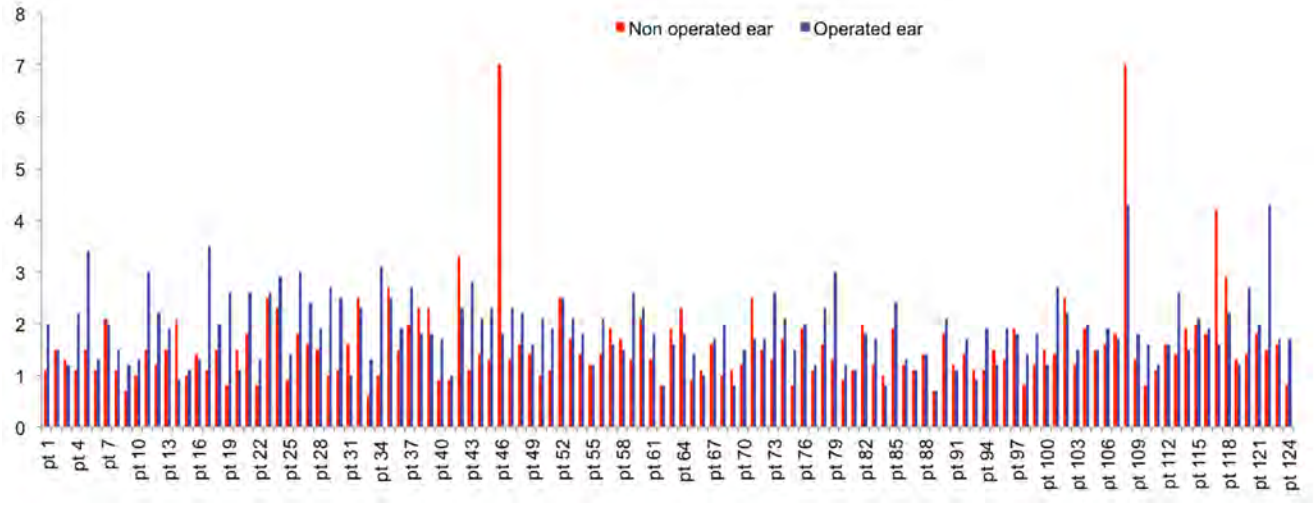


Fig. 10. Tympanogram – Difference between operated and non-operated ears

All the ears have been self-cleaning, while 58.67 % patients had hearing gain. On tympanometry, ten patients had the same volume auditory canal as the non-operated ear, 81 cases had higher value in the operated ear than the non-operated ear, with a range of 0.1 to 2.8 ml, and an average difference of 0.72 ml. Thirty-three cases had lower value in the operated ear than the other ear, but some of these had previously had a mastoid cavity on the other side.

Discussion

Mosher[15] described obliteration in 1911 and T. Palva suggested that mastoid obliteration should be done regularly.[1] Since then there have been various methods tried; with musculoperiosteal flaps, free fat or tissue grafts, cartilage and other prosthetic materials like hydroxyapatite granules and ceramics. As mentioned, each method has its own advantages and disadvantages, for example, prosthetic materials are expensive and may be extruded, while musculoperiosteal flaps and free grafts may shrink in size.

The main objective is removal of all the disease with minimal self-cleaning cavity, thus avoiding accumulation of debris.

The main concern after obliteration is residual disease underneath the obliteration material, so meticulous removal of all the disease is a must. Residual disease is more commonly seen in epitympanum or mesotympanum as shown by Syms[2] and Kang[3] in all types of mastoid surgery. The obliterating material does not hide the attic and middle-ear disease. Several authors have reported minimal or no residual disease.[4-7] In this series there has been no residual disease or recurrence in any case.

The CT scans have been used to monitor residual disease following bone paté or prosthetic material obliterations. Yung found that the residual cholesteatoma were detected through clinical examination while they went undetected by the interval scanning at 12 months.[8] However, it has been shown that it is not as useful with soft tissue obliterations. Vercruysee has stated that the benefits of using diffusion-weighted MRI scans may make them more useful in detecting residual disease in soft-tissue obliterations, given the ‘safe, non-invasive, selective, sensitive and comparatively cheap alternative to exploratory staged surgery’.[9]

To asses the success of obliteration it is essential to assess the volume of the operated ear. The only method we have found in literature to asses the canal volume is described by Ojala, who measured it by filling the bony part of the ear canal with sterile saline solution.[10] Our method of comparing the volume of normal ear with that of the operated ear using tympanometry gives objective evaluation. The drawback of this method is that the operated ear canal is wider which requires the use of different sizes of ear pieces for tympanometry.

Autologous cartilage is useful in reconstructing the mastoid cavity due to its strength and non-absorbant qualities.[11-13]

Reconstruction of hearing is done primarily in every case. Our results have been encouraging with 58% showing hearing gain, which has been comparable with other studies. Furthermore, a study by Dornhoffer showed a significant improvement in quality of life following mastoid obliteration.[14]

Conclusion

Obliteration should become a part of all mastoid surgery. The technique described here is a simple and cost-effective method of achieving a self-cleaning dry ear, using tissues that are normally removed as part of mastoid surgery. Meticulous surgery is shown to reduce the residual disease, while strengthening the tympanic membrane may help in reducing the recurrence, with minimal complications and consistent overall results.

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A COMPARISON BETWEEN NEURAL RESPONSE TELEMETRY VIA COCHLEOSTOMY OR THE ROUND WINDOW APPROACH IN COCHLEAR IMPLANTATION

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Introduction

Brazil is estimated to have about 347,000 deaf individuals, many of them with indications for a cochlear implant (CI). For patients with little cochlear reserve who cannot achieve good sound discrimination even with sound amplification, the CI is one option for their rehabilitation.[1] The CI brings about an improvement in hearing quality and improvements in speech perception and production, rendering a permanent and ascending quality-of-life gain in many aspects – such as self-sufficiency and socialization.[2-5] It is estimated that since the 1970s until today, there are 400 thousand implanted patients.[6]

CI's partially replace the cochlea by turning sound into electrical signals.[7] The survival of enough neural structures in the cochlear nerve allows the transmission of electric stimuli to the cerebral cortex.

The surgical implantation procedure via the transmastoid approach has been well standardized. Cochleostomy was first described in the 1980s.[8] There are two techniques to place CI's: via a cochleostomy, in which the promontory is drilled to fixate the implant, or via the round window (RW). Less drilling is required in the RW technique, thus reducing trauma, loss of perilymph, and bone powder on the tympanic scale.[9] Preservation of residual hearing has been viable and beneficial due to the combination of electrical and acoustic stimulation, but it requires non-traumatic insertion of the electrode to minimize damage to inner ear structures and enable lesser neural tissue degeneration.[6]

There are different ways to perform objective measurements on the auditory nerves of CI users from the electrical stimulation of the auditory system, such as auditory brainstem response (ABR), middle latency response, late potentials, and stapedial reflexes.[1] Neural response telemetry (NRT) is a test used to measure electrically evoked compound action potentials (ECAP) during surgery or post-operatively in implanted patients. This is an important test used to accurately monitor external and internal hardware function, and assess cochlear stimulation through neural responses.[10]

This is a prospective cross-sectional study aimed at comparing neural response telemetry in the immediate post-operative care of 23 patients of both genders with CI placed through cochleostomy or the RW approach to verify whether the choice of implantation procedure produces differences in auditory nerve stimulation.

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Edited by Haruo Takahashi

Materials and methods

This study was approved by the Medical Ethics Committee on Research with Human Subjects and given permit nº 004/2010. This study complied with the standards defined in Resolution 196/96 issued by the Ministry of Health.

Twenty-three patients, seven males and 16 females, were enrolled in this study. Six patients underwent implantation via cochleostomy and 17 via the RW approach. All patients were implanted the same device made by the Cochlear Corporation. The procedures were performed by the same surgeon.

The multi-channel CI's used in this study have 22 electrodes placed on the cochlea. The electrodes are numbered from one to 22, 22 being the one placed more apically. These electrodes were grouped the following way: 1-7 high-frequency sounds, 8-15 mid-frequency sounds, 16-22 low-frequency sounds. This division was needed because during NRT, we could not always get neural responses on one same electrode without changing the assessment parameters, and thus we left it for the software program to randomly choose within the groups which electrodes would be analyzed. Electrodes were split by ranges into high-, mid-, and low-frequency groups for the purposes of statistical calculation, as not all electrodes were analyzed individually.

The surgical technique employed to place CI's consists of the following steps: 1. General anesthesia for pediatric patients and local anesthesia plus sedation for adult patients; 2. Retro-auricular incision of about three cm; 3. Dissection of subcutaneous tissue and muscle plane; 4. Y-shaped periosteal flap; 5. Shift periosteum from skullcap at the site of implantation of the internal unit; 6. Mastoidectomy; 7. Posterior tympanotomy; 8. Perform cochleostomy in the anterior inferior area of the RW in cases where cochleostomy was used as the implantation approach; drill the upper lip of the RW and open it with a probe; 9. Insert electrode beam; 10. Perform neural response telemetry; and 11. Close the planes of muscle and skin tissue using vicryl 3-0.

All patients were discharged on the same day of surgery and had compressive dressings on for two days. Amoxicillin and clavulanic acid were administered for ten days. Implants were activated 30 days after surgery.

The Custom Sound AutoNRT measurement system comprises the following elements: 1. A computer with Windows Vista Home Basic, Intel® Pentium® Dual processor; 2. Software version Custom Sound EP 2,0 (2.0.4.7298) and 3,2 (3.2.3855); 3. Programming interface – POD; 4. Speech processor – Freedom sound processor and headset SPrint; 5. Freedom Implant (Contour Advance). The NRT software was developed by the Engineering Department at the Cochlear Corporation.[11]

A computer equipped with programming interface is used to stimulate specific electrodes inside the cochlea. A series of pulses of information bi-directional communication using encoded radio frequency is transmitted from the Freedom processor interface through an external antenna placed inside a sterile bag placed on the patient's skin above the internal receiver-stimulator. The encoded radio-frequency signal controls the stimulation parameters used to evoke compound action potentials. The internal receiver-stimulator in the Freedom Contour CI is equipped with one amplifier and one analog-digital converter. These additional components allow the voltage recorded on a pair of intra-cochlear electrodes to be amplified, sampled, and transmitted back to the external antenna, and then to the programming interface. These voltages are analyzed and the resulting ECAP wave is shown on a screen and the data can be stored in a computer. The ECAP records show a negative peak (N1) at 0.2-0.4 ms after stimulus onset, followed by a positive peak (P2) at 0.5-0.7 ms after stimulus onset. Response amplitude is measured from N1 to P2 and ranges between 40-2000 μV. Response amplitude varies with current levels between individuals. The parameters used to measure the thresholds on AutoNRT are: search for thresholds starts at 170 CL, at standard intervals of 6 CL for stimulation levels, at a stimulation frequency of 250 Hz.

Results

The patients submitted to implantation through the RW approach were aged between four and 84 years, and had a mean age of 32 years and three months. Patients in the cochleostomy group were aged between four and 54 years, and had a mean age of 19 years.

The Mann-Whitney test was used to statistically treat the samples, as this test allows the comparison of two groups of independent samples of different size. No statistically significant differences were found between implantation procedures as patients were assessed for high-frequency sounds (electrodes 1 to 7) (Table 1).

No statistically significant differences were found between implantation procedures as patients were assessed for mid-frequency sounds (electrodes 8 to 15) (Table 2).

No statistically significant differences were found between implantation procedures as patients were assessed for low frequency sounds, as shown in Table 3.

Table 1. Mean current level values for high-frequency sounds in patients submitted to implantation via cochleostomy and the round window approach.

Implantation approach	n	Mean current levels				Mann-Whitney test
		min-max	media	±	dp	P
Round window	17	110-237	190,44	±	29,28	0,71
Cochleostomy	6	146-239	187,87	±	32,70	

n: number of subjects; min-max: minimum and maximum values; SD: standard deviation; *p*: level of statistical significance. (Source: designed by Hammerschimdt R, Schuch LH.)

Table 2. Mean current level values for mid-frequency sounds in patients submitted to implantation via cochleostomy and the round window approach.

Implantation approach	n	Mean current levels				Mann-Whitney test
		min-max	media	±	dp	P
Round window	17	152-236	192,57	±	22,02	0,23
Cochleostomy	6	161-206	178,58	±	18,51	

n: number of subjects; min-max: minimum and maximum values; SD: standard deviation; *p*: level of statistical significance. (Source: designed by Hammerschimdt R, Schuch LH.)

Table 3. Mean current level values for low frequency sounds in patients submitted to implantation via cochleostomy and the round window approach.

Implantation approach	n	Mean current levels				Mann-Whitney test
		min-max	media	±	dp	P
Round window	17	134-223	183,34	±	25,00	0,19
Cochleostomy	5	143-190	163,89	±	19,32	

n: number of subjects; min-max: minimum and maximum values; SD: standard deviation; *p*: level of statistical significance. (Source: designed by Hammerschimdt R, Schuch LH.)

Discussion

The preservation of the structural tissues of deaf patients is not essential in CI procedures. However, since the introduction of electrical and acoustic stimulation combined in patients with cochlear reserve, the preservation of structural tissue and hearing has become of paramount importance in implantation procedures.[12] The loss of residual hearing is the outcome of a combination of factors, including the approach used in the cochleostomy procedure, the electrode neuronal stimuli, and the location of the cochleostomy.[8] The advent of new electrodes and the increased emphasis given to residual hearing preservation have renewed the interest in using the RW as a portal to place electrodes.[8] When compared to cochleostomy via the promontory, placement through the RW should significantly reduce the number of perforation events during electrode placement and thus the risk of trauma, loss of perilymph, and bone powder in the tympanic scale.[8] Bumps around the edges of the RW may pose difficulties to implant placement and require drilling of the anterior inferior border[8]. Drilling in this area must be done carefully, given its proximity to the opening of the cochlear aqueduct.[8]

Each electrode in the CI has to be programmed so that proper levels of electrical stimulation are provided. The unit used to program electrodes has been arbitrarily chosen and named current level programming units (CL). An important factor concerning cochlear implants is the variation on the current levels needed to elicit hearing for each individual and stimulation channel. Consequently, electrical stimulation parameters must be adjusted on the speech processor for each individual to adapt to specific user needs. This is done through a process called mapping.

A more direct way of measuring cochlear-nerve function is electrically evoked compound action potential (ECAP). ECAP reflects the synchronized triggering of cochlear nerve fibers and, in many ways, carried simi-

larities with wave I on ABR, occurring under 0.5 ms after stimulus onset.[11] Originally, these measurements could be done on human beings only during surgery or through CI's using percutaneous stimulation.

Stapedial reflexes can be measured as a response to electrical stimulation of the cochlea through direct observation of stapedial muscle contractions during surgery or by measuring acoustic impedance on the ear contralateral to the implant. The thresholds of electrically evoked stapedial reflex may be used to estimate C levels; nonetheless, significant variability is present in measurements done intra or inter-subjects. Additionally, according to a number of authors these reflexes cannot be recorded in about 40% of the population.[13,14]

Therefore, NRT is a technique that allows ECAP to be measured directly in implanted patients during and after surgery with greater sensitivity, as it can be done in more than 80% of the assessed individuals. NRT is a valuable tool and can be used to confirm the integrity of the internal device, objectively determine which electrodes can be included in a map, define the best stimulation frequencies and speech encoding strategies, estimate T levels to measure the current levels to induce hearing sensation, and estimate C levels of the maximum sensation intensity accepted by patients, a clinically important variable.[15]

No differences were observed on neural response telemetry between implants placed in the tympanic scale via cochleostomy or the RW. One cochleostomy patient had to be excluded from the analysis of mean values for electrodes 16 to 22 as no neural response was captured for low-frequency sounds.

Karatas *et al*.[8] reported that electrodes placed using the RW approach provided better stimulation when compared to electrodes placed via cochleostomy through the promontory as electrically evoked stapedius reflex thresholds (ESRT) and duration of stimulation were analyzed. In summary, the best response was defined for the shortest response time.

Both CI placement approaches are well established in the literature, cochleostomy being the most frequently used approach today. The choice of surgical approach is based on the preferences and training of the surgeon. There are no significant differences between the two approaches in terms of time of surgery and risk of complications.

This paper presents preliminary results, and no analysis was done on neural stimulation to compare patients in different age ranges. The auditory nerves of children respond better to stimulation than the auditory nerves of older patients. In a future study the groups need to be randomized for their specific ages so that this variable is properly assessed for this criterion.

This paper can be used to support other studies with larger samples, specifically on what concerns the cochleostomy approach and the measurement of all electrodes in the immediate post-operative care period. It is also part of the efforts made to reach better results with sound stimulation and auditory rehabilitation of the countless patients affected by deafness.

Conclusion

The data shown in this preliminary study indicate that there is no significant difference in the acquisition of action potentials from the distal portion of the auditory nerve through neural response telemetry in multichannel CI patients using the implant to elicit stimulation and record responses, whether implantation was done through cochleostomy of the RW approach. Both approaches provide for equal stimulation of the cochlear nerve, and surgeons are free to choose the procedure of their preference.

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TRANSMEATAL APPROACH AND THE OPTIMAL DEVICE FOR MINIMAL INVASIVE SURGERY

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The transmeatal operation is the favorable approach for a small lesion in the mesotympanum, congenital cholesteatoma, malformation of the ossicles, perforation of the ear drum, and also otosclerosis. The advantage of the transmeatal operation is that it involves minimal invasive surgery. The disadvantage of this approach is the limitation of the operation field because of the shape and size of the external auditory canal.

The external auditory canal was measured using temporal CT images of 48 ears of 32 adults. Table 1 shows the width of external auditory canal. The width of the canal was 10.4 mm x 12.4 mm at the external orifice, 8.7 mm x 8.8 mm at the osteochondral junction and 9.1 mm x 9.1 mm at the annuls of the drum. The width of the canal was 4.5 mm x 4.8 mm at the isthmus. The superior-inferior length is always greater than the antero-posterior length.

Table 1. Width of the ear canal measured using temporal CT images.

Diameter	External orifice	Osteochondral junction	Isthmus	Annulus of the drum
Antero-posterior	10.4 mm	8.7 mm	4.5 mm	9.1 mm
Superior-inferior	12.4 mm	8.8 mm	4.8 mm	9.1 mm

The 3D shape of the external canal was reconstructed and cut in round slices. The cross section of the external auditory canal is oval-shaped, not round (Figs. 1 and 2).

We found that the favorable shape of the ear speculum is oval in the cross section in order to use the space more effectively (Fig. 3).

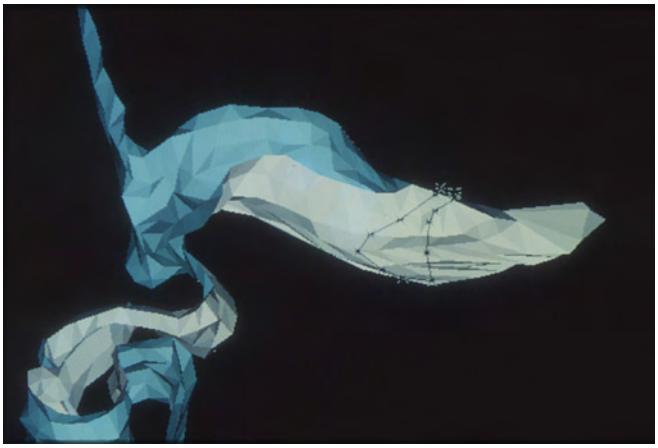

Fig. 1. 3D reconstruction of the external ear using a human temporal bone specimen. The dotted line shows the cutting direction of the ear canal.

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Edited by Haruo Takahashi

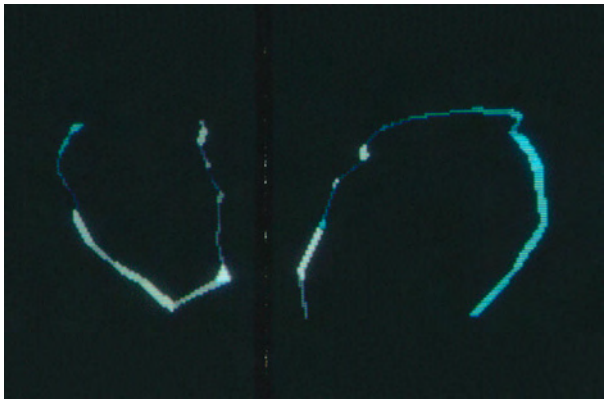

Fig. 2. Cross section of the reconstructed ear canal. The shape is oval, not round.

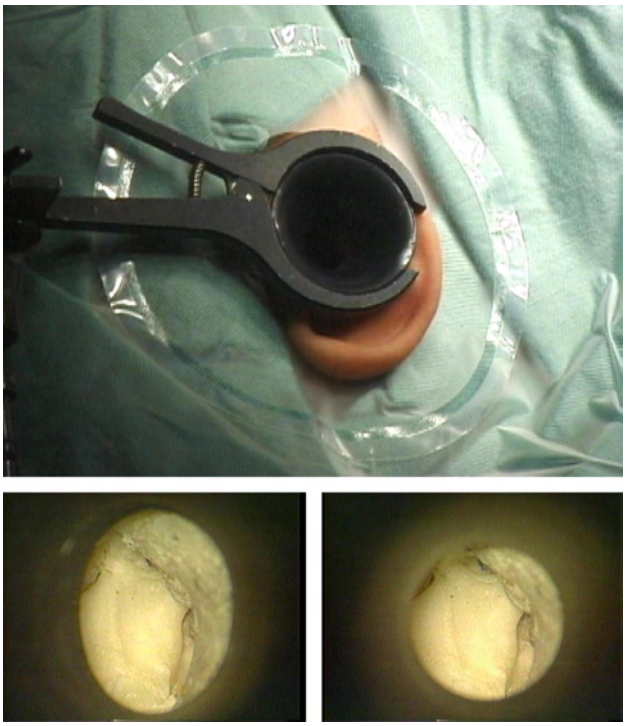

Fig. 3. Top: ear speculum and speculum holder. Bottom left: oval-shaped ear speculum. Bottom right: round-shaped ear speculum.

ANATOMICAL MEASUREMENT OF THE FACIAL RECESS AND THE ROUND WINDOW MEMBRANE USING CONE-BEAM COMPUTED TOMOGRAPHY

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Background and purpose

In Japan, high-resolution computed tomography (HRCT) is routinely used for the pre-operative evaluation of the temporal bone. Recently, cone-beam computed tomography (CBCT) has become available to daily clinical practice. Several studies have shown that CBCT can visualize fine structures of the temporal bone more precisely than conventional CT, both *in vitro* and *in vivo*.[1,2] CBCT has also been used for the post-operative evaluation of cochlear implants to measure the precise insertion length of the electrode array.[3] The surgical anatomy of the round window region is of increasing importance due to the round window surgical approach for hearing devices such as cochlear implants and vibrant sound bridges. The purpose of this study was to measure the width of the facial recess and the round window membrane using CBCT and to evaluate the efficacy of CBCT.

Materials and methods

Materials

Among the patients who underwent CBCT at Kagoshima City Hospital from January to February 2012, 50 ears of 37 patients (aged nine to 78 years; 16 males and 21 females) were selected for the measurements. Thirty ears were excluded because aeration of the tympanic cavities was insufficient to identify the round window membrane at the basal end of the scala tympani.

Technical data of CBCT imaging

CBCT examinations were performed using a 3D Accuitomo F17 (J. Morita Mfg. Corp., Kyoto, Japan). Imaging was performed with a tube current of 8 mA and a tube voltage of 90 kV. During the scan of each subject, 577 raw data-projection images were obtained over 360 degrees. One full rotation took 17.5 seconds. The raw data-projection images were reconstructed using the software i-View (J. Morita Mfg. Corp.) resulting in a voxel size of 0.125 mm. The diameter and the height of the cylinder-shaped reconstruction regions of interest were 60 mm.

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Edited by Haruo Takahashi

Measurement of the facial recess and the round window membrane

The reconstructed images consisted of sagittal, coronal, and axial views, which can be set at a desired angle. The facial recess in a sagittal view is a triangular area composed of the facial canal, chorda tympani nerve and incus buttress. The length of the upper side of this triangle, which is a line parallel to the incus buttress, was defined as the width of the facial recess. The width of the round window membrane was measured at an axial view, in which a basal turn of cochlea and round window niche was visualized. Both distances were measured and the data obtained were evaluated statistically.

Results

The average width of the facial recess was 2.3 mm, ranging from 1.64 mm to 3.03 mm, and the diameter of the round window membrane was 1.6 mm, ranging from 1.23 mm to 1.96 mm. The width of the facial recess and the diameter of the round window membrane showed no statistical correlation with age or gender (Figs. 1 to 3, Spearman's correlation).

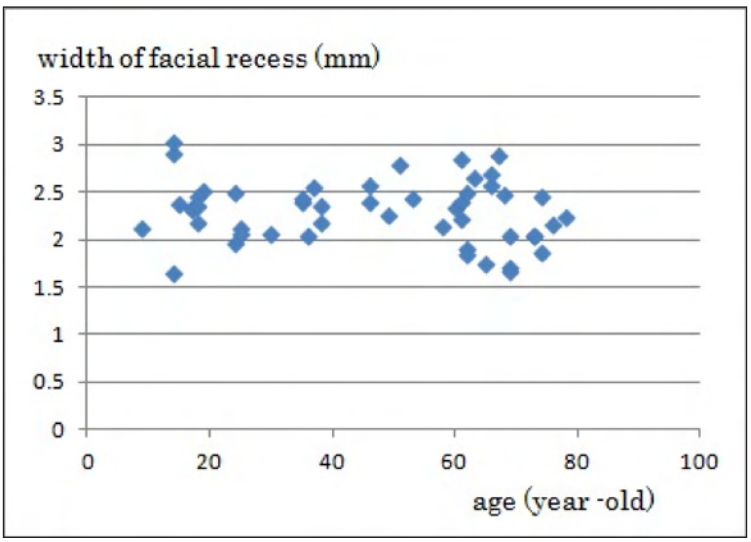

Fig. 1. Width of the facial recess shows no correlation with age.

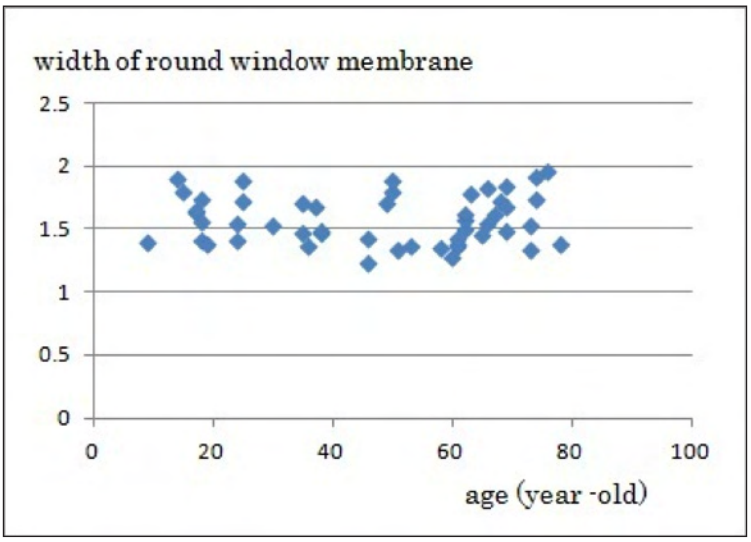

Fig. 2. Width of the round window membrane shows no correlation with age.

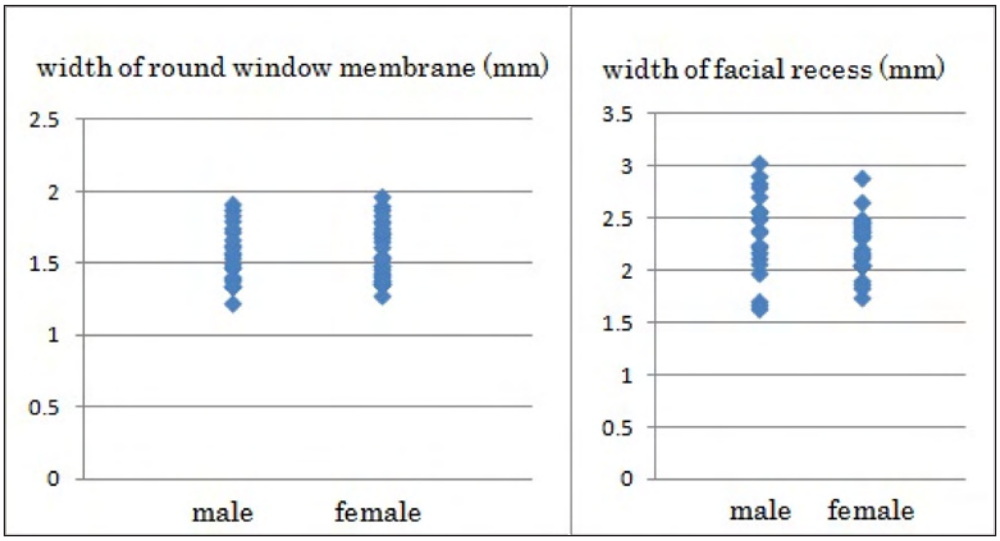

Fig. 3. Width of the facial recess and round window membrane show no correlation with gender.

Discussion

A histological study[4] of the facial recess and the round window membrane showed that the average width of the facial recess and the round window membrane were 4.01 mm and 1.65 mm, respectively. Although the diameter of the round window is consistent with our data, the width of the facial recess is much longer than that observed in our study. The discrepancy between the histological and CBCT studies appears to be due to different anatomical definitions of the facial recess. In the previous histological study, the facial recess was measured between the facial canal and the margin of the tympanic annulus. Thus, the thickness of the external auditory canal was included. In agreement with the previous histological study, our study showed no statistical correlation with age for both measurements. These results indicate that surgical procedures for these anatomical sites should not differ with age.

The opening of the facial recess is described as two or three mm in width in many textbooks of ear surgery or temporal bone dissection. This is supported by the results of our study. However, the range of the opening of the facial recess from 1.64 mm to 3.03 mm suggests that there may be some cases in which the facial recess cannot be opened without injuring the chorda tympani nerve or the facial nerve, even with a burr of 1.5 mm diameter.

Conclusion

In the era of implantable hearing devices, the otologist should be familiar with structures of the middle and inner ear more precisely. For this purpose, CBCT is very convenient and reliable tool for preoperative understanding of the fine structures of the temporal bone.

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PREVENTION OF RECURRENT CHOLESTEATOMA: EXCLUSION TECHNIQUES USING TITANIUM SHEETING

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Introduction

Since its inception by Jansen,[1] intact-canal-wall (ICW) mastoidectomy has become the treatment of choice for attico-mastoid cholesteatomas by many otologists. The technique seeks to restore the diseased ear back to the near normal anatomical and functional state. Compared with canal-wall-down (CWD)/open-cavity surgery, ICW is, however, often perceived as being more at risk to both residual and recurrent disease. This is partly due to flaws of previous ICW drum and canal-wall repair techniques. These left the ear prone to several forms of recurrent sac formation: through a patent or poorly repaired attic defect; via re-collapse of the pars tensa; through necrosis of the external canal wall.[2] Failure to perform routine second stage also leads to residual disease difficulties.

Both ICW and CWD techniques are at risk from drum collapse and residual mesotympanic pearls. CWD encounters open-cavity problems whereas ICW may be troubled by persistent attico-mastoid disease, whether residual or recurrent.

Thus for ICW to be the preferred method, it must be designed to anticipate and overcome these latter problems.

Method

ICW surgery follows several general steps. The first is a trans-canal approach that is the optimal route to the stapes, in order to clear this key site of disease, adhesions, etc. Adequate exposure may necessitate a superficial meatoplasty, an anterior wall drill-back, or a scutum curettage. For the subsequent mastoid and attic exposure the author uses a hairline post-aural incision. A key step is a wide exposure of the zygomatic root that in turn facilitates the best trans-mastoid visualization of the attic. Similar ample exposure is required at second stage to allow re-inspection for small pearls of disease.

Prevention of recurrent disease demands an impenetrable barrier, extending from deep into the zygomatic root, along the plane of the facial nerve into the mastoid tip. To achieve this, fine titanium sheeting (0.125 mm thickness, 99.6% pure, annealed, Goodfellow, http://www.goodfellow.com) is used to line the under-surface of the EAC wall, in part or whole, using an aluminium foil template for precise size and shape

When an attic defect is small, an elliptoid 13-20 mm sheet suffices (Fig. 1). The sheeting may be further supported by a simple U-clip of sheeting for stability.[3] To underlay the entire canal wall, the implant is a hemi-ellipse (divided longitudinally), approximately 30 mm x 13 mm wide, twisted to adapt to the canal wall and is fitted into slots cut into the zygomatic root and the mastoid tip.[3] Any bony-wall defects are repaired with autograft cartilage inlay grafts.[3] The titanium layer supports these grafts *in situ*. The cartilage inlays also prevent biomaterial contact with the canal squamous epithelium. Thinned areas of bony canal wall may be augmented further with either cartilage shavings or with bone pate, especially the area postero-superior to

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Edited by Haruo Takahashi

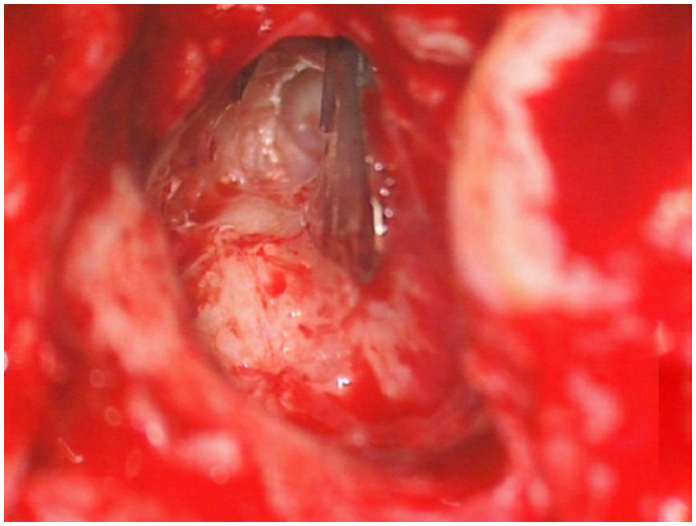

Fig. 1. Closure of a limited attic defect, right side second stage ICW. The sheeting is applied to the underside of the attic defect, to the right, and stabilized by the U-clip (inverted, above). The far anterior attic can be viewed clearly for residual foci.

the pars tensa, where a tortuous canal may be inadvertently excessively thinned during attic access. In larger defect cases, vascular flaps (middle temporal[4] or temporalis fascia variants) may be used to cover the grafts to provide greater viability of the site.

Results

The technique has been used in 185 attico-mastoid cholesteatoma ICW cases (2007-2011 inclusive). Pars-tensa patterns were present in 38%, attic origins in 44%, the remainder (12%) being combined attic and pars tensa collapse cases.[5] Mean follow-up time was 2.5 years.

Cases were complicated by mesotympanic residual disease in 8% and attico-mastoid pearls in 9%. Of these, 30% were smaller than two mm diameter (and thus possibly MRI-invisible) at the 12-month second-stage surgery. Mesotympanic recurrences occurred in 4%. Invaginations penetrated or developed around the reconstructed wall in three cases, corrected at second stage procedures. There were no infections related to the use of the sheeting.

Discussion

Effective ICW surgery counters residual and recurrent disease. Staged surgery is effective in eliminating residual pearls; the author's experience of persisting residual disease beyond a second stage is confined to only anecdotal cases over forty years experience. However, this necessitates adequate surgical access to the attic at both the first and second stages.

To prevent recurrent pockets, the reconstructed canal wall should resist further erosion and display no gaps or slits through which recurrent sacs may invaginate directly, around, beneath, or over the repaired wall. However, the reconstruction must not obstruct review of the previously cholesteatomatous areas. In recent years obliteration has been proposed in several works as an alternative to ICW, but the detection and management of residual (and recurrent) disease remains controversial. Residual pearls are often small and/or slow growing.[6] Obliteration techniques risk these being missed by MRI scans;[7-10] when and how often such scans are required is another difficulty. Revision of ossifying obliteration sites is fraught with problems of distorted landmarks; infection may persist undetected for long periods.

Previous ICW techniques were been troubled by attic retractions if closure of an attic defect was not done routinely, or of closure material was poorly fitted or resorbed. Other problems were related to canal wall resorption.[11-13] Further difficulties occurred when a pars tens collapse 'underflanked' an intact or repaired scutum to enter the attic and beyond.

Other problems arose from the type of wall repairs used. These could be classified as onlay, inlay or underlay techniques. Each had particular difficulties. Onlay repairs were stable, but did not prevent underflanking pockets, and obscured adequate inspection of the deep EAC. Inlay methods were difficult to shape exactly to fit the defect, and were unstable, lacking support. They were also prone to adjacent defects if bony wall resorption occurred. Underlay repairs were likewise unstable, requiring supporting material that obstructed second stage re-inspection. A previously employed hydroxylapatite-plate technique[2] overcame many of these problems, but the plates tended to partially occlude the attic, necessitating thinning or removal at the second stage. Sporadic long-term infections occurred, and the plates were too small to adapt to the largest defects. Grote hydroxylapatite wall implants[14] were cumbersome in this role and fitted poorly into the attic behind an intact wall.

Conversely, the study method stabilizes the inlay grafts, is durable, adaptable to most situations, and permits best possible re-inspection, as the sheeting is applied closely to the canal wall and thus allows best access to the attic (Fig. 2). Titanium sheeting as above is simple to fashion with heavy scissors. It has excellent biomaterial qualities and can be molded to adapt to the curves of the canal. A template affords an exact fit into retaining grooves, promoting fixation and avoiding small chinks around the sheet edges. As the sheeting is applied closely to the horizontal facial nerve, the repair precludes 'underflanking' by pars tensa retractions, further avoided by routine reinforcement of the posterosuperior pars tensa quadrant with a composite graft that also shields the ossiculoplasty site (Fig. 3). The combination provides an effective barrier to invagination.

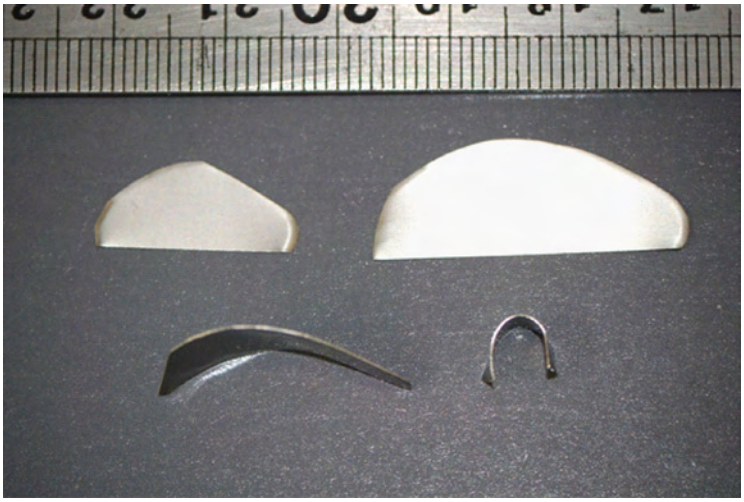

Fig. 2. Titanium sheeting used for wall repair. From top left, clockwise: Section used for limited attic defects; Larger section used to 'underwrap' the entire EAC wall; Section curved to adapt to the EAC wall; U-clip used to stabilize sheeting if required.

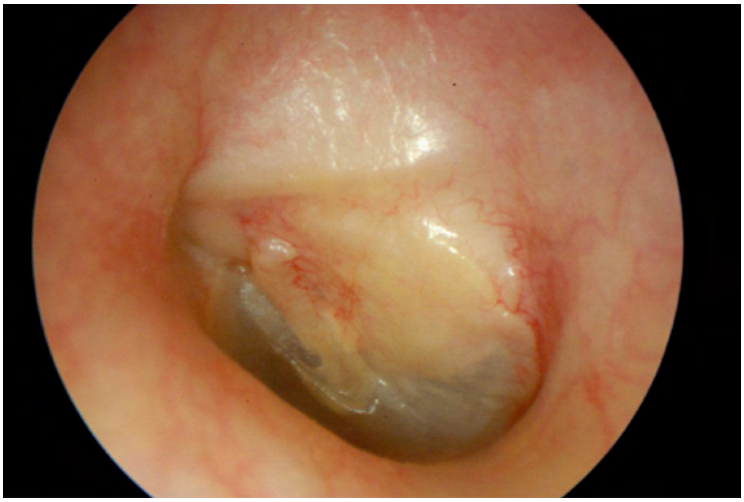

Fig. 3. Attic and drum repair, post-operative appearances of the attic defect repair and the postero-superior pars tensa composite graft.

Conclusion

ICW is the optimal technique to return best anatomical and functional results from cholesteatoma surgery. The method used must adequately anticipate and prevent the problems of residual and recurrent disease.

The above titanium sheeting technique is presented as a relatively simple, but versatile and reliable method of ICW management.

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HRCT-BASED PREDICTION FOR COCHLEAR IMPLANT OUTCOMES OF CASES WITH INNER EAR AND INTERNAL AUDITORY CANAL MALFORMATIONS

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Introduction

Inner ear and internal auditory canal (IAC) malformations account for approximately 20-35% of congenital sensorineural hearing loss[1,2] and an increasing number of children with inner ear and/or IAC malformations underwent cochlear implantation. According to Sennaroglu's classification of inner ear malformations, which is the most widely accepted, the inner ear malformations are divided into labyrinth aplasia, cochlear aplasia, common cavity (CC), incomplete partition type I (IP-I), type II (IP-II), and type III (IP-III), cochlear hypoplasia type I (CH-I), type II (CH-II), and type III (CH-III), and large vestibular aqueduct syndrome (LVAS).[1,3] This classification is essential to investigate the etiology of the inner ear malformations, but with respect to predicting cochlear implant (CI) outcomes, it might not be enough, because it does not include IAC malformations such as narrow IAC (NIAC) and hypoplasia of the bony cochlear nerve canal (HBCNC). These IAC malformations are highly associated with cochlear nerve deficiency (CND), which has a negative impact to CI outcomes.[4,5]

The purpose of this study was to establish a new CT-based categorization which is simple and includes both inner ear and IAC malformations for predicting CI outcomes.

Materials and methods

Between 2004 and 2010, 98 subjects who were under 20 years old underwent cochlear implantation at Kobe City Medical Center General Hospital. Among them, CT revealed that 24 subjects had inner ear and/or IAC malformations at the implanted side.

We evaluated inner ear and IAC malformations at the implanted side based on CT findings. Sennaroglu's classification was used to classify inner ear malformations and the IAC malformations were classified into NIAC and HBCNC. NIAC was diagnosed when the maximum diameter of the IAC was less than 2 mm.[2] The width of the bony cochlear nerve canal (BCNC) was evaluated at the mid-portion between the base of the modiolus of a cochlea and the fundus of the IAC on axial images. When the diameter of the BCNC is less than 1.5 mm, it is diagnosed as HBCNC.[5] CND was diagnosed when a cochlear nerve (CN) appeared smaller than the facial nerve on the parasagittal MR imaging.

We categorized inner ear and IAC malformations into four groups by two criteria: (1) the presence or absence of a bony modiolus in the cochlea; and (2) the diameters of IAC and BCNC. In this categorization, both Group 1 and Group 3 have a bony modiolus in the cochlea, while Group 2 and Group 4 lack this component. Both IAC and BCNC are normal in Group 1 and Group 2, but NIAC or HBCNC was observed in Group 3 and Group 4. Sennaroglu's classification of inner ear malformations clearly discriminates between

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Edited by Haruo Takahashi

the presence and absence of a bony modiolus in the cochlea. According to his classification, a bony modiolus is present in IP-II, CH-III, LVAS, and a normal inner ear, while CC, IP-I, IP-III, CH-I, and CH-II have a cystic cavity without a bony modiolus.[3]

We evaluated CI outcomes by category of auditory performance (CAP) scores,[6] hearing thresholds of pure-tone sounds, infant word speech discrimination scores, and monosyllabic word speech discrimination scores at one to three years after implantation. A subject with 0-4 CAP scores could not even understand common phrases without visual language and, therefore, we defined 5-7 CAP scores as a good CI outcome and 0-4 CAP scores as a poor one.

Results

We categorized our patients based on the two criteria as described above. In this study, there was no case categorized in Group 4. Group 1, Group 2, and Group 3 consisted of 11, 7, and 6 cases, respectively. MR imaging revealed CND in all cases of Group 3.

The post-operative CAP score was equal or over five in all cases of Group 1, but did not exceed four in all of Group 3. In Group 2, the post-operative CAP score was still four in two cases even after three years of CI usage, but reached to five or six in the remaining five cases. As shown in Figure 1, using our new categorization instead of the existing classifications, we can better discriminate between a good and poor outcome.

We examined speech discrimination scores of 22 cases except for two cases of Group 3 whose response to voice was poor. The correct percentage of the closed-set infant word discrimination test was ≥ 80 in all cases of Group 1, while the score ranged from 40 to 60 in tested cases of Group 3. The correct percentage of Group 2 widely varied between cases, ranging from 55 to 100. The open-set monosyllabic word discrimination test is much more difficult than the closed-set infant word discrimination test and, therefore, only 17 of 24 patients, who were over five years old and used their CI for more than two years, underwent this examination. All tested cases of Group 1 and 3 cases of Group 2 could answer correctly in equal or over 80% of accuracy. The correct percentage of the remaining cases, including all tested cases of Group 3, was ≤ 30.

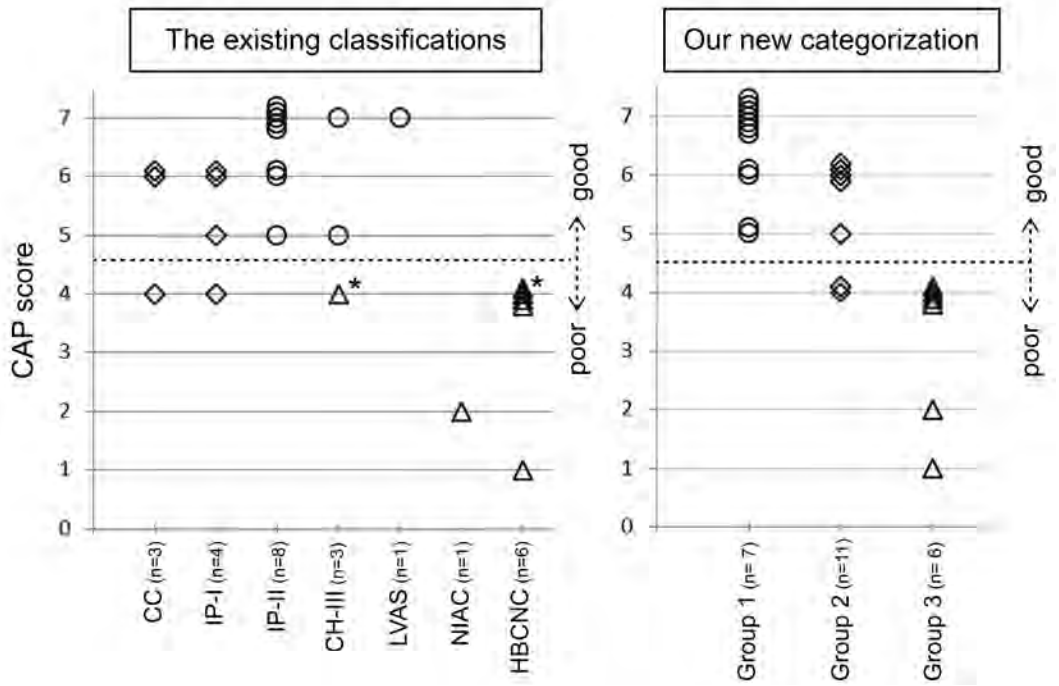

Fig. 1. A. The post-operative CAP score of each type of malformations based on the existing classifications. One case with both CH-III and HBCNC is plotted twice (*). B. The post-operative CAP score of each group of our new categorization. In both graphs, the members of Group 1, Group 2, and Group 3 are represented by a circle, diamond, and triangle, respectively.

Discussion

In this study, we established a new CT-based categorization including both the inner ear and IAC malformations. This categorization is defined by two criteria; (1) the presence or absence of a bony modiolus in the cochlea; and (2) the diameters of IAC and BCNC. We focused on these structures because the bony modiolus contains spiral ganglion cells, the major target of CI-mediated electrical stimulation, and their axons go through BCNC and IAC.

Group 1, which is defined by the presence of a bony modiolus of the cochlea with a normal IAC and BCNC, showed the best CI-aided hearing performance among three groups. The high proportion of post- or peri-lingually deaf cases might also contribute to the high CI outcomes of this group.[7] Group 2 is defined by the absence of a bony modiolus with a normal diameter of IAC. The CAP score and speech discrimination score varied widely between cases in this group, but five out of seven cases could understand common phrases without visual languages. Group 3 is defined by the presence of a bony modiolus in the cochlea with NIAC or HBCNC and their post-operative improvement of hearing performance was limited. Visual languages were necessary for them to understand common phrases even after long usage of their CI. MR imaging revealed CND in all cases of Group 3, which might be responsible for their poor outcomes.

Conclusion

Our new CT-based categorization, which was based on the presence or absence of a bony modiolus in the cochlea and the diameters of IAC and BCNC, was effective in predicting CI outcomes of children with inner ear and/or IAC malformations. The CI outcomes were the best in Group 1, followed by Group 2 and Group 3. All cases of Group 1 showed good CI outcomes and could communicate orally. On the other hand, all cases of Group 3 showed poor CI outcomes and used lip-reading or sign language to understand common phrases. The CI outcomes of Group 2 varied between cases, but many of them showed good CI-aided hearing performance.

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POSTER SESSIONS

A CASE OF CHOLESTEATOMA OCCURRING AFTER COCHLEAR IMPLANTATION

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Introduction

In some cases, profound sensorineural hearing loss can be caused by chronic otitis media or cholesteatoma. Recently, even in such cases, cochlear implantation can be safely performed after the active middle-ear infection has settled down.[1] However, the rate of complications after cochlear implantation is higher in cases with middle-ear disease than without disease.[2,3] We report a case with cholesteatoma that occurred ten years after cochlear implantation.

Case history

A 69-years-old (in 2010), female suffered in 1996 from sudden deafness in the left ear. She was treated with steroids and batroxobin without effect.

In 1997, a tympanoplasty was performed in order to cure the adhesive otitis media of her right ear.

In 1998, a tympanoplasty with columella on the stapes was performed again because of adhesion of tympanic membrane and hearing loss. Insertion of the gromet tube was also performed. Cholesteatoma was seen. Just after surgery, she suffered from profound sensorineural hearing loss. She was treated with steroids without effect.

In 1999, a cochlear implantation was placed in the right ear. The mastoid was filled with granulation. There was a bone defect in the basal turn of cochlear. The electrode was inserted into this hole. Twenty-two active electrodes and five extra electrodes were inserted.

In 2009, cholesteatoma was identified (CT and tympanic membrane).

In 2010, a CT showed that the mass of mastoid had grown (Fig. 1). Surgery with the aim of removal of cholesteatoma was done.

We chose the canal-wall-down technique and removed the cholesteatoma from the matrix with the so-called Bondy technique. We could remove cholesteatoma without re-implantation. The electrodes were left as they were (Fig. 2).

Results

Three months after surgery, the electrodes are not exposed at either external ear canal or retro-auricle (Fig. 3). The cavity is almost free from maintenance. The post-operative hearing score is almost same as before (Fig. 4).

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Edited by Haruo Takahashi

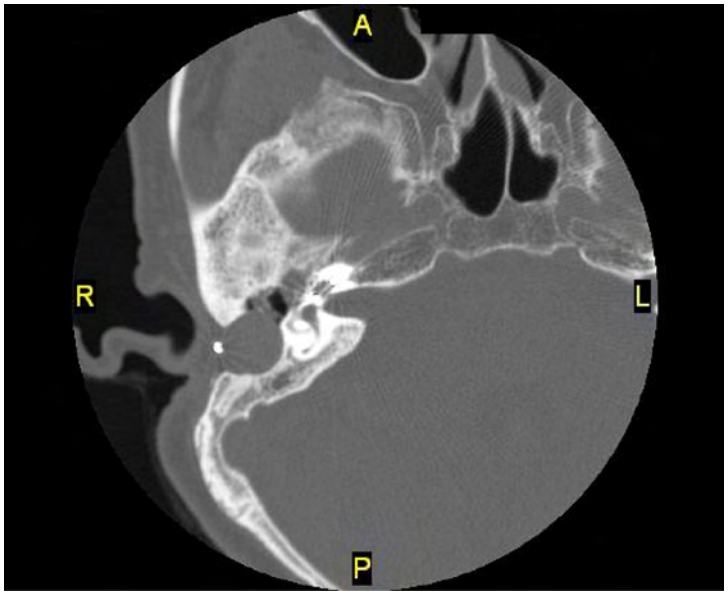

Fig. 1. CT (2010).

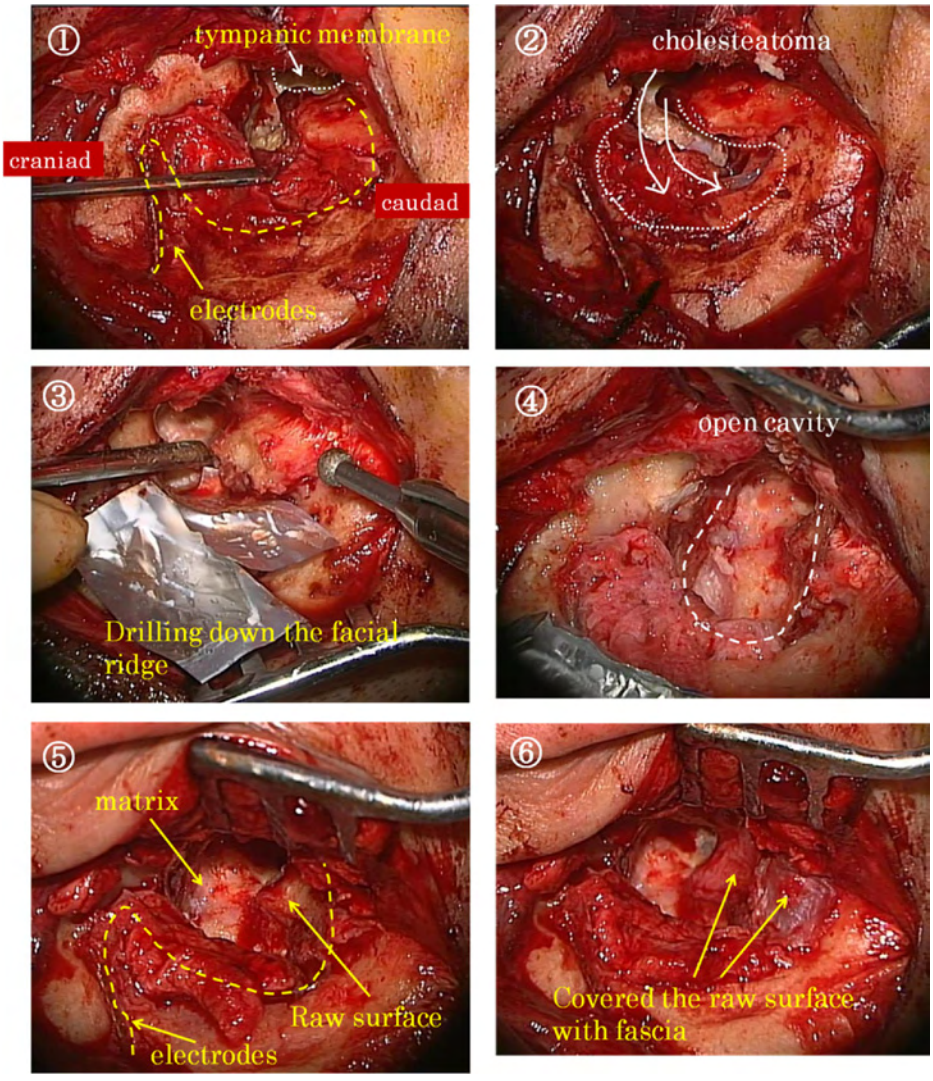

Fig. 2. Surgical findings (2010).

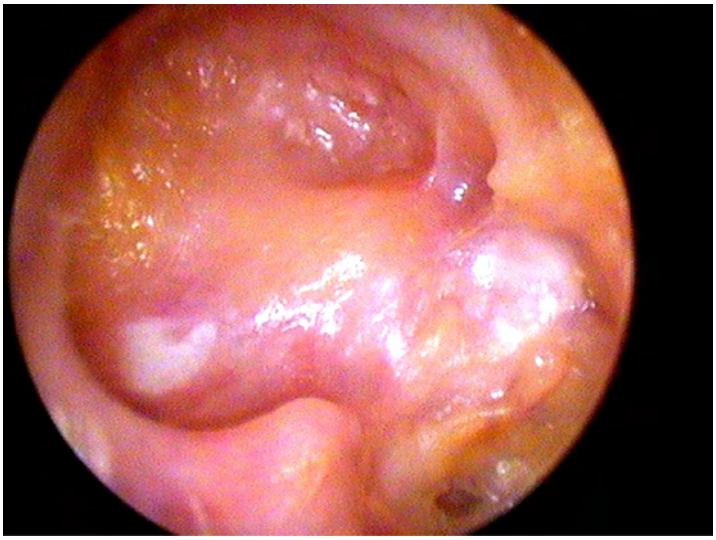

Fig. 3. External ear canal (three months after surgery).

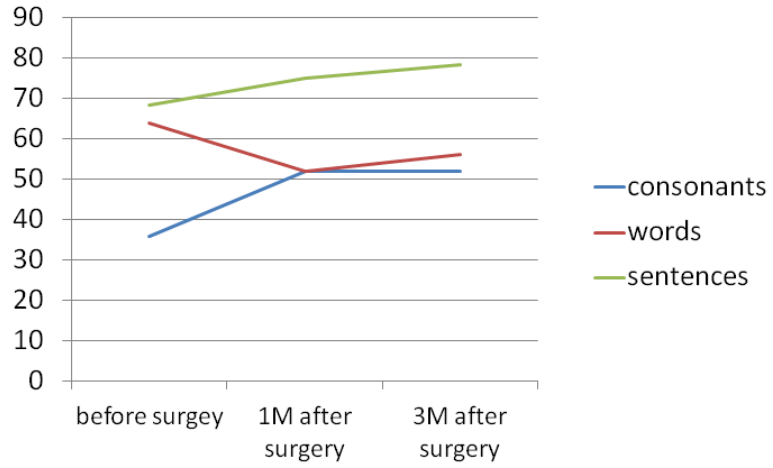

Fig. 4. Hearing (CI-2004). *CI-2004: Test battery for the patient with a cochlear implant to assess the audiological phase. The components are lists of consonants, syllables, words and sentences (Japanese).

Discussion

In this case, although the ear drum tended to be adherent, the cochlear implantation was performed in the ordinary method. Therefore, cholesteatoma recurred from the pars flaccida.

The rate of complication after a cochlear implantation for patients with middle-ear disease is higher than that of patients without middle-ear disease.[2,3] Especially the adhesive ear drum and poor aeration of tympanic cavity are factors of complications after cochlear implantation.

Therefore, when we plan cochlear implantation for the patients whose middle ear is difficult to aerate, some special considerations are required, such as, obliteration of mastoid and tympanic cavity (+ EAC closure),[1,4,5] canal-wall-down technique to leave no possibility of retraction, covering of the electrodes with cartilage.[6]

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THE CASE OF CONGENITAL CHOLESTEATOMA THAT SPONTANEOUSLY CURED DURING OBSERVATION

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Congenital cholesteatoma was first reported by Derlacki and Clemis.[1] Congenital cholesteatoma is characterized by a white pearly mass behind the intact tympanic membrane. The anterosuperior quadrant of the middle-ear cleft is known to be the most frequently involved site in over 80%.

We report the case of a child with congenital cholesteatoma spontaneously cured during the observation.

A three-year-old boy was examined by his local otolaryngologist for ear wax. In the clinic, a pearl was pointed out behind the anterior-superior quadrant of an intact tympanic membrane, and the boy was referred to our hospital. He was diagnosed as cogenital cholesteatoma, and a CT-scan of the temporal bone showed a soft-tissue density mass between the malleus and anterior wall in the tympanic cavity. An erosion of ossicles was not observed and hearing level was normal with visual reinforcement audiometry. Therefore, he was followed up in our hospital.

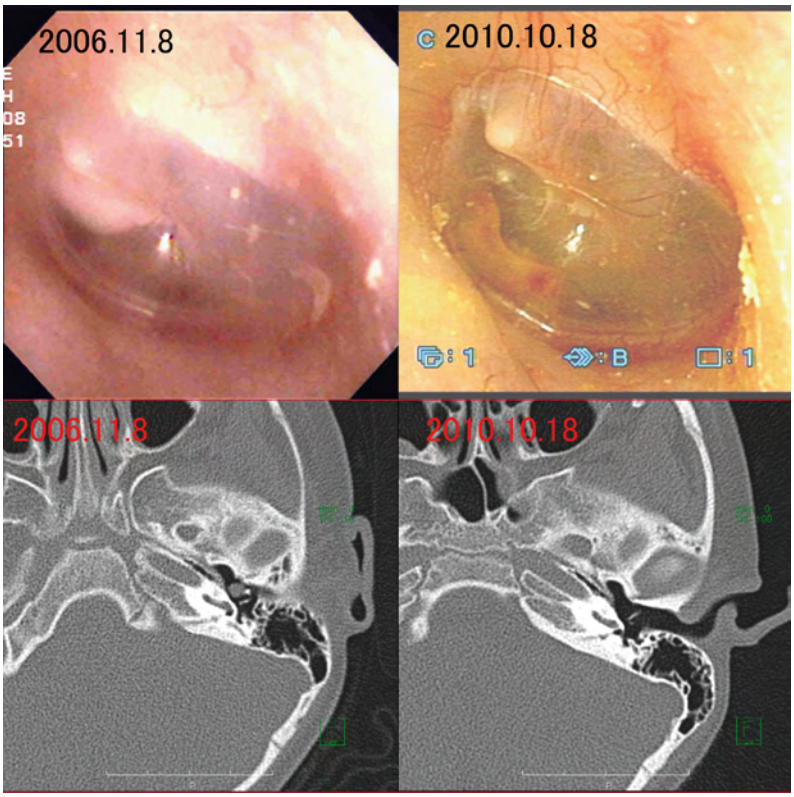

Fig. 1. Disappearance of the choleateatoma in the tympanic cavity

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Edited by Haruo Takahashi

During the follow-up period, the whitish mass behind the eardrum gradually decreased in size (Fig. 1). Three years later, at the first examination, there was no mass observed by oto-microscopy or temporal bone CT images of the middle ear. Kodama *et al*. reported two cases of spontaneous regression of congenital cholesteatoma in 2012.[2] There could be other cases of congenital cholesteatoma cured before diagnosis. The patients with cholesteatoma are usually encouraged to have a tympanoplasty at an early stage. On the basis of our case, careful observation can be one of the treatments for congenital cholesteatoma.

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CONGENITAL MIDDLE-EAR CHOLESTEATOMA IN CHILDREN: A RETROSPECTIVE REVIEW

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Background

Congenital cholesteatoma is defined as a middle-ear cholesteatoma located behind an intact tympanic membrane. Lesions typically occur in the anterior-superior quadrant (ASQ).[1] Lesions in the posterior-superior quadrant (PSQ) constitute a rare variant that is sometimes found among older individuals.[2] Posterior lesions have been observed among Asian populations[3]. The purpose of this retrospective study was to present the clinical features of congenital cholesteatoma in children treated at our institute.

Methods

We reviewed the clinical and surgical records of 15 patients who underwent primary surgery for congenital cholesteatomas at Kobe University Hospital between 1990 and 2011. The patients were followed up for at least one year after their last operation. The criteria for the diagnosis of congenital cholesteatoma included the presence of a white mass behind an intact tympanic membrane with no perforation and no history of otologic surgeries. The four-stage system proposed by Potsic *et al*.[4] was used (Table 1). Audiometric analysis was performed according to the guidelines recommended by the Japan Otological Society. The pure-tone average (PTA) was calculated from a three-tone average of 0.5, 1, and 2 kHz. Selected important characteristics for each event are summarized in Table 2.

Results

The median age at the primary surgery was 5.0 years (range 3-15 years). There were 13 male patients and two female patients. The left side was involved in four cases (27%); the right side was involved in 11 cases (73%). No case of bilateral congenital cholesteatoma was observed.

Hearing loss was the main presenting symptom and was observed in eight cases (53%). Among the 15 cases included in the study, five (33%) involved patients who complained of otalgia, one (7%) involved ear fullness, and one (7%) involved facial palsy.

The anatomic sites of cholesteatoma involvement at the time of the initial procedure are summarized in Table 3. The PSQ was affected most often, in nine cases (60%). The ASQ was affected in only six of 15 ears (40%).

Regarding the disease stage at the time of surgery, two cases were classified as surgical stage 1 (13%), two cases as stage 2 (13%), seven cases as stage 3 (47%), and four cases as stage 4 (27%). The surgical method used most frequently was canal-wall-up tympanomastoidectomy; this procedure was used in ten ears (67%)

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Edited by Haruo Takahashi

Table 1. Staging system for congenital cholesteatoma (Potsic et al., 2002).

Stage 1	Single quadrant: no ossicular involvement or mastoid extension
Stage 2	Multiple quadrant: no ossicular involvement or mastoid extension
Stage 3	Ossicular involvement: includes ossicle erosion and surgical removal to eradicate disease; no mastoid involvement
Stage 4	Mastoid extension (regardless of findings elsewhere)

Table 2. Characteristics of 15 ears in 15 patients.

Characteristic	*Value*
Age, median (range), y	5 (3–15)
Age Category, no. (%)	
< 4 y	3 (20)
4–5 y	6 (40)
≧ 6 y	6 (40)
Sex, no. (%)	
Male	13 (87)
Female	2 (13)
Ear, no. (%)	
Right	11 (73)
Left	4 (27)

Table 3. Anatomical involvement.

	No. (%)
Anterior-superior quadrant	6 (40)
Posterior-superior quadrant	9 (60)
Anterior-inferior quadrant	5 (33)
Posterior-inferior quadrant	4 (27)
Mastoid region	4 (27)

to eradicate the disease. A canal-wall-down procedure was required in five ears (33%). Ossiculoplasty with a columella on the stapes was performed in six (40%) patients, ossiculoplasty with a columella on the footplate in six (40%) patients, and the Wullstein type-I method in three (20%) patients. The operation procedures and ossiculoplasty protocol are summarized in Table 4.

Pre-operative hearing data were available for nine of the 15 ears. The initial mean (± SD) hearing loss was 31.5 ± 14.8 dB. Post-operative hearing data were available for nine ears, and the mean (± SD) hearing loss was 23.0 ± 11.4 dB. The residual rate was 0% in stage 1, 50% in stage 2, 57% in stage 3, and 50% in stage 4. The residual rate and the pre-operative and post-operative PTAs are shown in Table 5.

Table 4. Operation procedure and ossiculoplasty.

		Canal wall (n)		*Ossiculoplasty (n)*		
Stage	*No. of ears*	*Intact*	*Down*	*Type 1*	*Columella on the stapes*	*Columella on the foot plate*
1	2	2	0	2	0	0
2	2	2	0	1	1	0
3	7	6	1	0	4	3
4	4	0	4	0	1	3
Total	15	11	5	3	6	6

Table 5. Residual rate and hearing data.

Stage	*No. of ears*	*Residual Rate (%)*	*Pre-operative PTA (n)*	*Post-operative PTA (n)*
1	2	0/2 (0)	19.2 ± 2.5 (2)	14.2 ± 5.8 (2)
2	2	1/2 (50)	5 (1)	16.7 (1)
3	7	4/7 (57)	35.4 ± 8.1 (4)	20.0 ± 5.4 (4)
4	4	2/4 (50)	49.2 ± 2.5 (2)	40.8 ± 7.5 (2)
Total	15	7/15 (47)	31.5 ± 14.8 (9)	23.0 ± 11.4 (9)

Discussion

Congenital cholesteatoma is thought to start most often in the ASQ, grow gradually into the posterior quadrant and the attic, and then finally invade the mastoid.[2] On the other hand, Inokuchi *et al*. reported that posterior-type congenital cholesteatoma was seen more often in Asian patients.[3] In this study, posterior-type cholesteatoma was dominant in Japanese patients, which is consistent with previous reports.[3,5,6]

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A CASE OF CONGENITAL CHOLESTEATOMA WITH ANTERIOR SEMICIRCULAR CANAL FISTULA

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Introduction

Labyrinthine fistula is a severe complication of cholesteatoma. The incidence of labyrinthine fistula is 7% in reported series of mastoidectomies for chronic ear disease.[1] It is reported that the prevalence of the anterior semicircular canal fistula is respectively 6% of the labyrinthine fistulae.[1,2] We experienced a case of congenital cholesteatoma and the malformation of auditory ossicles with anterior semicircular canal fistula.

Case report

A 15-year-old male had suffered from right hearing loss in childhood. He was diagnosed with right hearing loss at a health-care examination in May 2011. He visited our hospital for further examination.

The right tympanic membrane was intact. Pure-tone audiogram revealed a 38 dB conductive-hearing loss in the right ear (Fig. 1).

Auditory ossicles were detected (Fig. 2A). The attic was filled with soft-tissue density (Fig. 2B). Soft-tissue density existed in the aditus ad antrum and the mastoid antrum. Part of the bone surrounding the anterior semicircular canal was unclear. The anterior semicircular canal fistula was suspected (Fig. 2C and D).

A part of the long process of the incus was incomplete and the incudo-stapedial joint showed fibrous chain (Fig. 3A). The superstructure of the stapes was intact. The cholesteatoma existed widespread from attic to mastoid antrum (Fig. 3B). Then, the cholesteatoma matrix penetrated into the small space of mastoid cells and behind the lateral semicircular canal (Fig. 3C). An anterior semicircular canal fistula was confirmed (Fig. 3D). Based on the CT scan and intra-operative findings, a labyrinthine fistula was classified in six stages (Table 1). The anterior semicircular canal fistula of this case was stage III. The removal of the cholesteatoma matrix might be incomplete behind the lateral semicircular canal and the vicinity of the anterior semicircular canal fistula. Therefore, a planned staged tympanoplasty without ossiculoplasty was performed.

Post-operative vertigo and nystagmus was observed until one month after surgery. The post-operative pure-tone audiogram is shown in Figure 4. Bone-conduction threshold showed a 60 dB hearing loss at 4,000 Hz in the right ear. The patient has been under observation as an outpatient for nine months and has presented no signs of recurrence of cholesteatoma. A second-look operation was planned.

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Edited by Haruo Takahashi

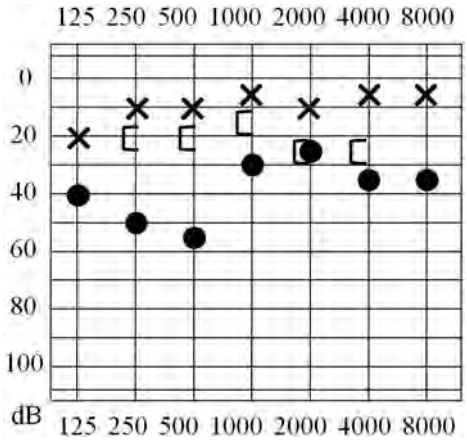

Fig. 1. Pure-tone audiogram revealed a 38dB conductive hearing loss in the right ear.

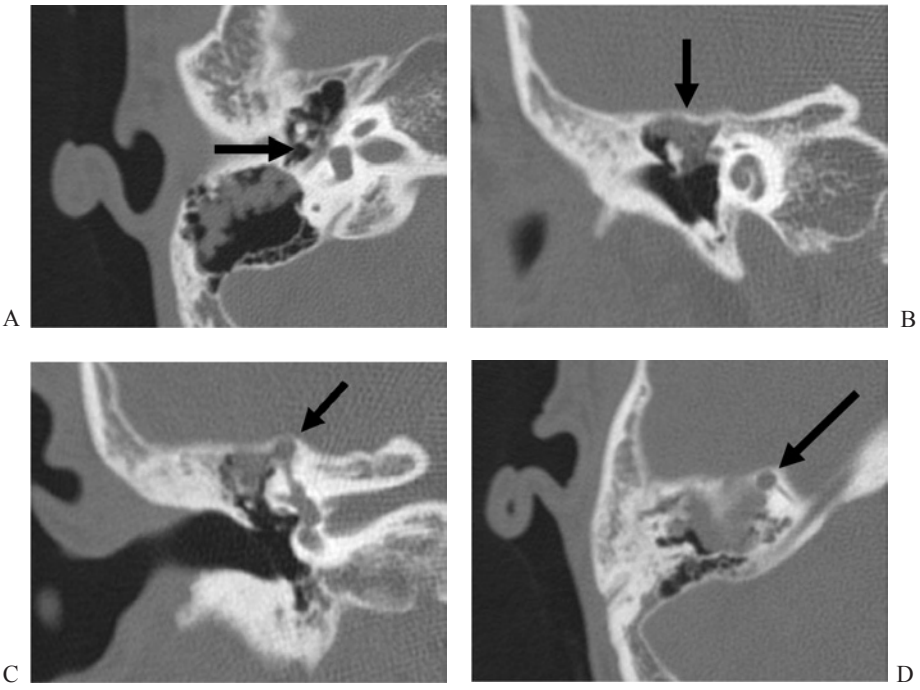

Fig. 2. Axial section CT. A. The superstructure of the stapes was detected (arrow); B. Attic was filled with soft tissue density (arrow). Coronal and axial section CT. C, D. Part of the bone surrounding the anterior semicircular canal was unclear (arrow). The anterior semicircular canal fistula was suspected.

Discussion

The incidence of a labyrinthine fistula is 7% in a reported series of mastoidectomies for chronic ear disease.[1] The location of the labyrinthine fistula was predominantly of the lateral semicircular canal (87%) followed by the promontry (8%), anterior semicircular canal (6%), and posterior semicircular canal (2%).[1] The incidence of post-operative sensorineural hearing loss with cholesteatoma matrix removal varies from 6% to 37%.[2] Table 2 shows the reported hearing in cases of labyrinthine fistulae since 2000. Fistula stages should theoretically correlate with the risk of sensorineural hearing loss. Much debate has arisen in the literature regarding the treatment of the fistula matrix. The first approach is to leave the matrix on the fistula, while the second approach advocates the total removal of the cholesteatoma from the fistula in one or two stages. Advantages of the matrix removal are the reduction of bone resorption, risk of suppurative labyrinthitis, and post-operative vertigo. The main disadvantage is the potential risk of post-operative sensorineural hearing loss.[3] Kobayashi *et al*. recommended a one-stage, open-method tympanoplasty and emphasized that careful manipulation of

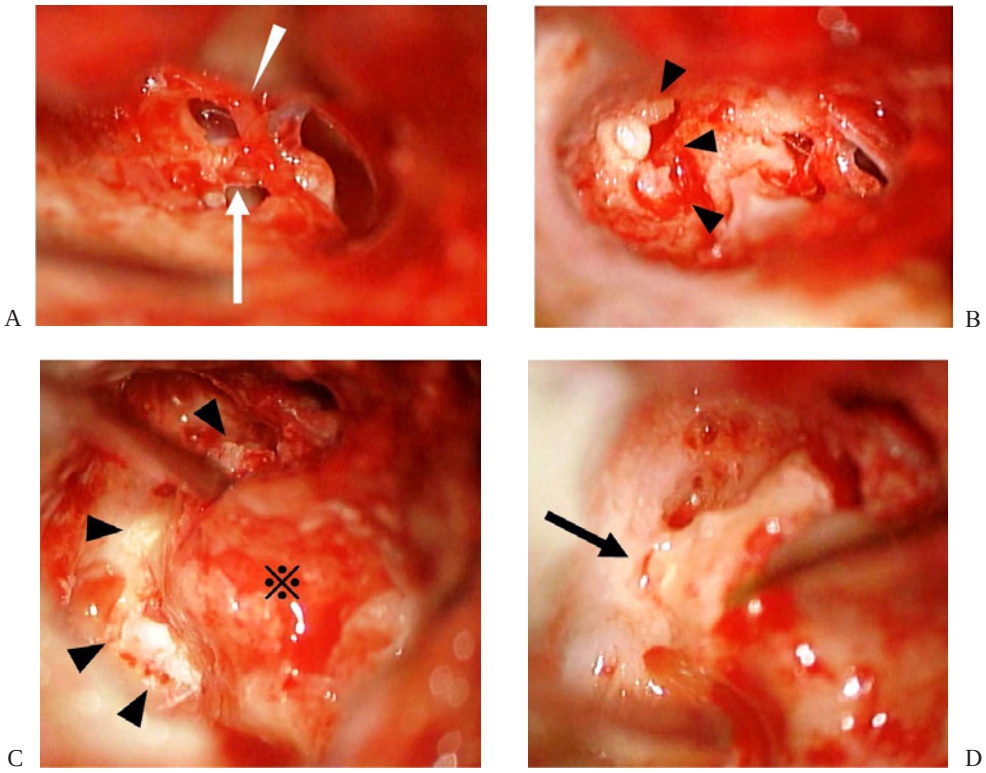

Fig. 3. A. A part of long process of incus was incomplete and incudo-stapedial joint was fibrous chain (arrow). Chorda tympani (arrowhead); B. The cholesteatoma (▲) was open type and existed widespread from attic to mastoid antrum; C. The cholesteatoma matrix (▲) penetrated into the small space of mastoid cells and behind the lateral semicircular canal (※). The removal of cholesteatoma matrix might be incomplete behind the lateral semicircular canal; D. Anterior semicircular canal fistula was confirmed (arrow). This case was stage III.

Table 1. Fistula staging.

Stage I: Pre-fistula (blue line)
Stage II: Small fistula ≤ two mm
Stage III: Fistula between two and four mm Stage IV: Invasion of one (a) or more (b) semicircular canal(s)
Stage V: (a) Invasion of vestibule
(b) Invasion of vestibule and cochlea
Stage VI: (a) Fistula limited to the stapes footplate
(b) Promontorial fistula

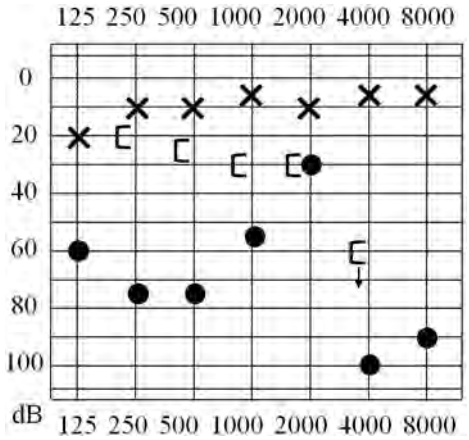

Fig. 4. Post-operative pure-tone audiogram. Bone-conduction threshold showed a 60dB hearing loss at 4,000 Hz in the right ear.

Table 2. Reported hearing in cases of labyrinthine fistulae.

Author	*Year*	*Pre-operative vertigo*	*Hearing decreased post-operatively*	*Dead ears post-operatively*
Gersdorff	2000	43/49	1/49	5/49
Soda Merthy	2000	20/23	3/23	0/23
Kvestad	2000	13/16	3/16	0/16
Habuka	2002	25/25	3/25	1/25
Ahmad	2002	7/22	2/22	2/22
Grewal	2003	25/50	0/50	0/50
Portier	2005		4/22	0/22
Ueda	2009	23/31	3/27	0/27
Quaranta	2009	43/36	5/46	0/46
Chen	2010	13/22	2/22	0/22
Yamamoto	2010		4/22	0/22
Ghiasi	2011	10/16	2/16	0/16
Gosea	2012	11/31	7/31	2/31

the semicircular canal can be conducted without damaging the cochlear function.[4] In the present case, the sac of cholesteatoma was not clear. As a result, total removal of cholesteatoma matrix vicinity the anterior semicircular canal fistula caused the sensorineural hearing loss. It is essential that treatment of the labyrinthine fistula is delicate in order to preserve hearing. The choice of surgical technique for a labyrinthine fistula is determined by the patient's general condition, the ipsi- and contralateral hearing thresholds, and the skill and experience of the surgeon.[5]

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AN UNUSUAL IATROGENIC CHOLESTEATOMA OF THE EXTERNAL AUDITORY CANAL: A CASE REPORT

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Case report

We present a patient with unusual, huge external auditory canal cholesteatoma (EACC) that developed during a three-year period after tympanoplasty. The patient suffered from otalgia and slight hearing loss and these symptoms started six months after the surgery. Ear discharge was not present. During the otoscopic examination, a sub-epithelial mass obliterating the left side of the external auditory canal was seen (Fig. 1). The patient's history revealed that otalgia and slight hearing loss began to develop six months after surgery. EACC excision and myringoplasty was performed (Fig. 2).

Discussion

External auditory canal cholesteatoma is an uncommon form of cholesteatoma originating from the external auditory canal extending into surrounding structures, including the mastoid and middle ear. It is first described by Toynbee.[1] There are no sufficient data about incidence of EACC; although there are authors who reported the incidence of EACC to be one in 1000 new otologic patients.[2,3] EACC is described as a late complication of temporal-bone trauma, tympanomastoid surgery, and radiation therapy. The potential for development of EACC in congenital and acquired aural stenosis is well recognized.[2] Chronic inflammatory disorders may also stimulate development. Depending on the etiology, EACC has been classified into six types: congenital, post-traumatic, iatrogenic, post-obstructive, post-inflammatory and spontaneous. There is little information on the incidence of ECC according to cause. Earlier literature on this topic suffers from an uncertain definition of the process and confusion with other disorders of the external auditory canal, mainly keratosis obturans.[3] An accurate description of each process by Piepergerdes and colleagues[4] was important in the resolution of this dilemma. They recognized that EACC develops secondary to a disorder of the canal bone, in many instances caused by a local osteitis. The diagnosis of ECC is based on history and physical examination. Radiographic assessment is useful to define the extent of the lesion and delineate its proximity to the middle ear and neurovascular structures. Presenting symptoms, physical findings, radiographic appearance, and management will vary according to the cause of the lesion.

Iatrogenic EACC develops as a late complication of tympanomastoid surgery. The lesion can present as invagination of skin through a defect in the posterior canal wall or as a subepithelial mass lesion.[5] Imaging reveals destruction of the canal wall lateral to the scutum and a normally aerated middle-ear space if the initial operation was successful. Hearing loss is variable, being dependent on the initial operation.

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Edited by Haruo Takahashi

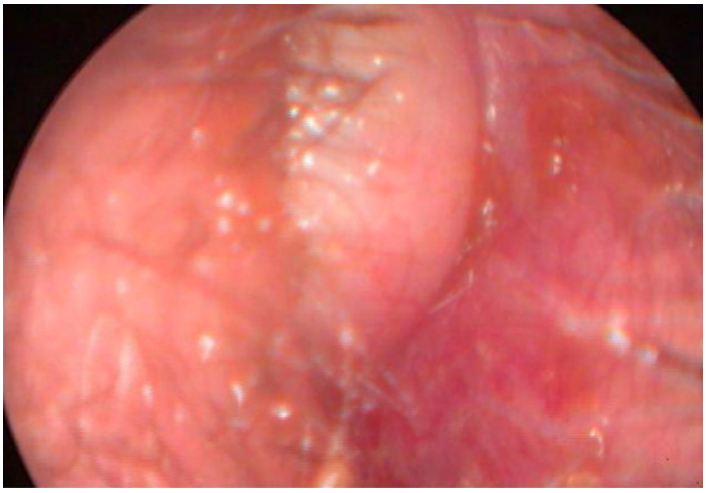

Fig. 1. Pre-operative view of subepithelial mass that is obliterating the left external auditory canal.

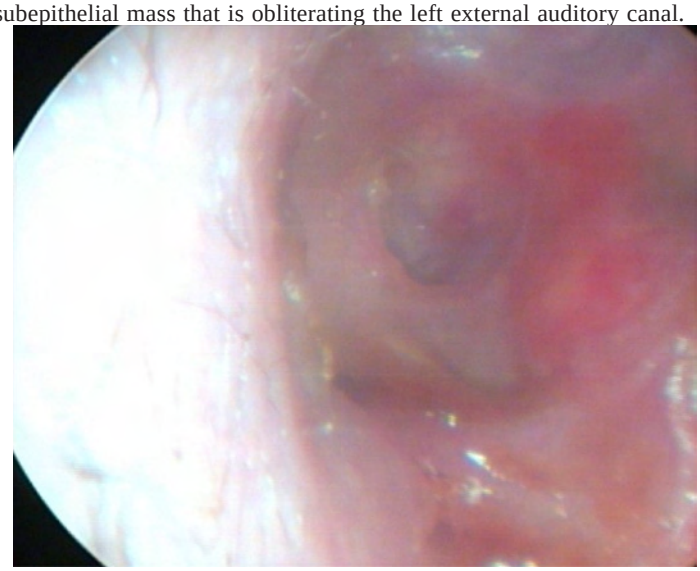

Fig. 2. Post-operative view of the left external auditory canal after surgery.

Conclusion

As seen in our case, we should be very aware not to implant squamous epithelium into deep tissue of mastoid cavity, tympanic cavity and external auditory canal during ear surgery.

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SEMICIRCULAR CANAL FISTULA WITH MIDDLE-EAR CHOLESTEATOMA – REPORT OF 24 CASES

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Introduction

Semicircular canal fistula occurs in approximately 5~10% of cases of middle-ear cholesteatoma and is reported to occur commonly in the lateral semicircular canal.

Our objective is to understand the incidence, location, pre- and post-operative symptoms, diagnosis and procedure in cases with semicircular canal fistula.

Materials and methods

Between January 2003 and December 2011, at the Department of Otolaryngology, Fukushima Medical University, 312 cases with middle-ear cholesteatoma underwent surgical treatment.

Pre-operative CT imaging was performed using MDCT (Aquilion®, TOSHIBA) and CBCT (3D-Accuitomo®, MORITA) systems. The depth of the labyrinthine fistula was classified using the Milewski & Dornhoffer classification (Table1).[1]

Table 1. Milewski & Dornhoffer classification

Type	
I	erosion of the bony labyrinth with an intact endosteum.
IIa	opened perilymphatic space with undisturbed perilymph
IIb	opened perilymphatic space with disturbed perilymph.
III	opened perilymphatic space with a disturbance of the underlying membranous labyrinth.

Results

In 24 of the 312 cases, a semicircular canal fistula was confirmed during the operation. The incidence of semicircular canal fistula was 7.7% in all patients.

Vertigo was present in 79.2% of all patients, fistula symptom was positive in 50% of all patients pre-operatively.

Diagnosis of semicircular canal fistula was made before surgery for 91.7% of patients on the basis of symptoms, signs and CT imaging.

The fistula was located in the lateral semicircular canal in 22 cases (91.7%), located in the anterior semicircular canal in one case (4.2%), and located in the lateral and anterior semicircular canals in remaining one case (4.2%)(Table2).

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Edited by Haruo Takahashi

Table 2. Location of the fistula (n = 24).

Location	*n*
Lateral semicircular canal	22 (91.7%)
Anterior semicircular canal	1 (4.2%)
Lateral and anterior semicircular canals	1 (4.2%)

Table 3. Classification of the semicircular canal fistula (n = 24)

Type	*n*
I	9 (91.7%)
IIa	12 (4.2%)
IIb	1 (4.2%)
III	2 (8.3%)

We classified the semicircular canal fistula according to the Milewski & Dornhoffer classification: nine fistulas were type I (37.5%), 12 fistulas were type IIa (50%), one fistula was type IIb (4.2%), two fistulas were type III (8.3%) (Table 3). Type I.

In 21 cases, semicircular canal fistulas were closed with temporalis fascia and bone paste (bone dust mixed with fibrin glue) or conchal cartilage, while in three cases the fistulas were closed with temporalis fascia only.

Vertigo improved in 17 out of the 19 cases with semicircular canal fistula accompanying vertigo.

Complications such as facial-nerve paralysis or meningitis did not occur in any cases.

After surgery, vertigo worsened in one case and bone-conduction hearing level deteriorated by more than 10 dB hearing level in six cases.

Case reports

Case1: A 9-year-old male with left middle ear cholesteatoma.
He had semicircular canal fistula diagnosed by preoperative CBCT scan (Fig1-3).
The bony labyrinth was destroyed and endosteum exposed (Fig.4).

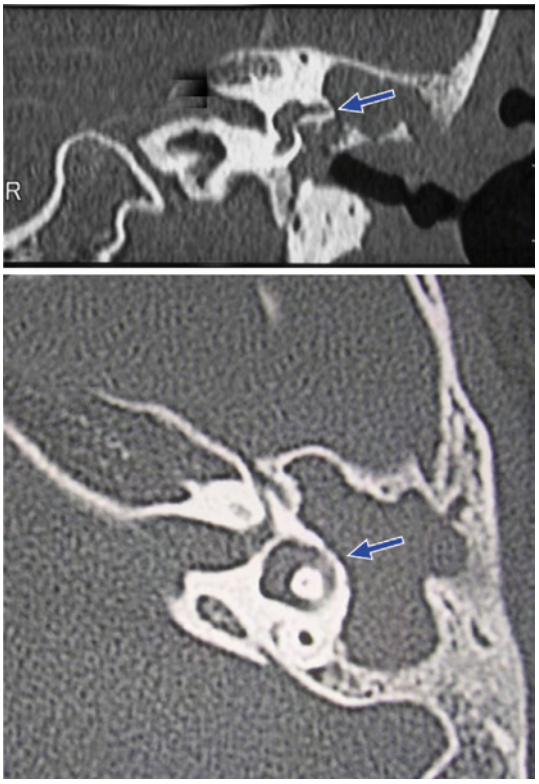

Fig. 1 and Fig. 2. Pre-operative MDCT scan of of left temporal bone in axial and coronal views, no fistula is apparent in the lateral semicircular canal.

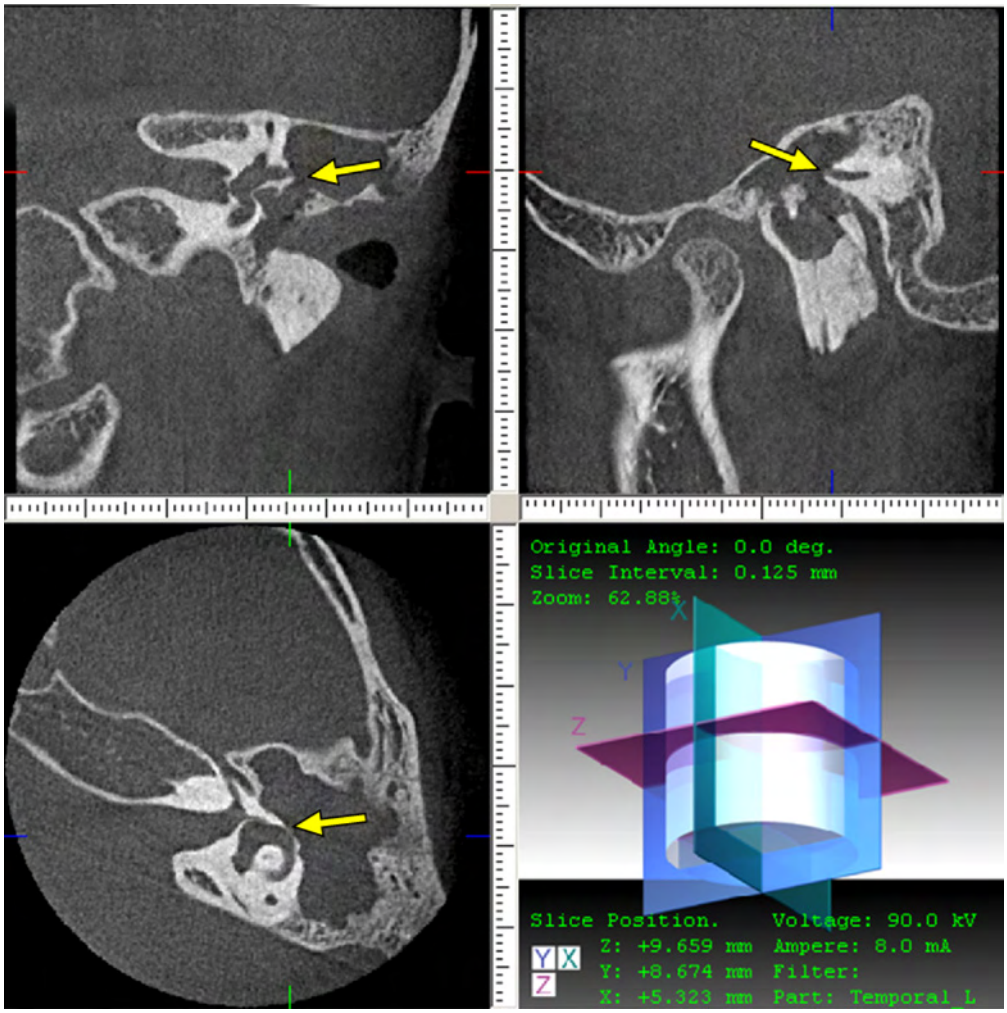

Fig. 3. Pre-operative CBCT scan, showing erosion of the otic capsule with lateral semicircular canal fistula.

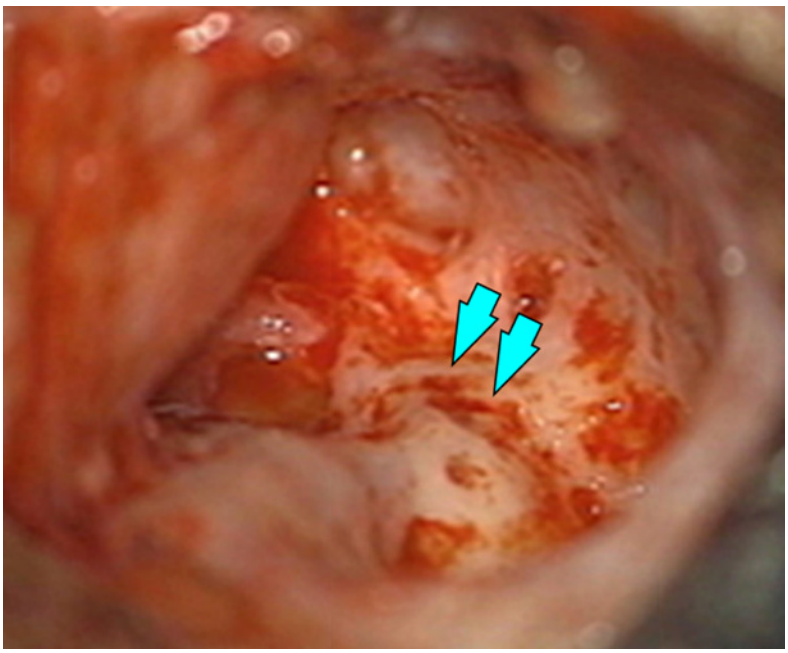

Fig. 4. The bony labyrinth was destroyed and the endosteum exposed.

Case2: A 37-year-old male with left middle ear cholesteatoma.
Destruction was found in lateral semicircular canal and anterior semicircular canal (Fig.5).
The bony and membranous labyrinth was destroyed by cholesteatoma (Fig.6).
Type III of Milewski & Dornhoffer classification was diagnosed in this patient.

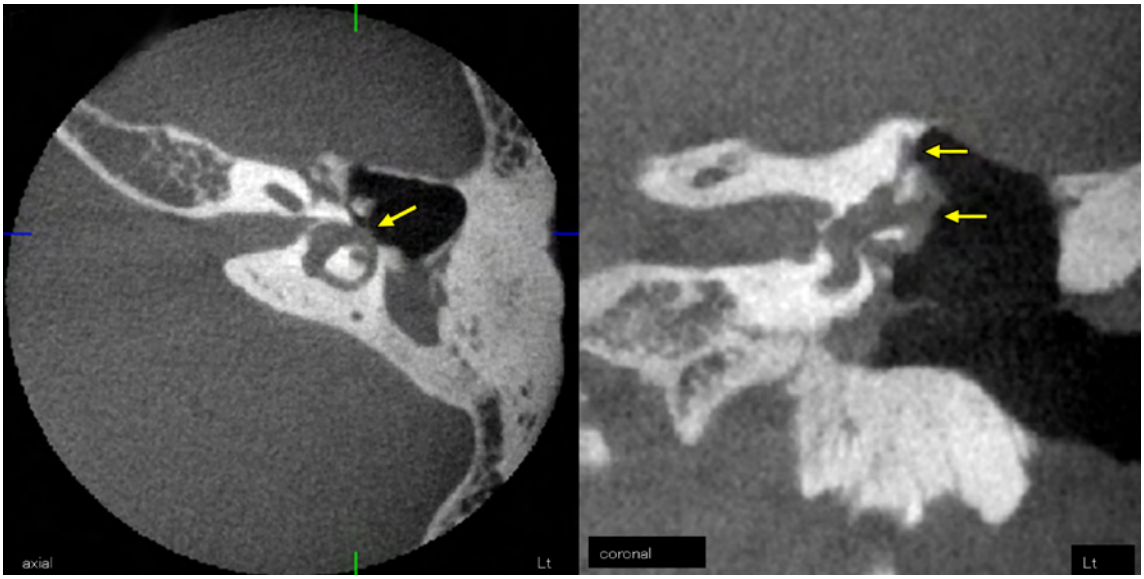

Fig. 5. Pre-operative CBCT scan, showing erosion of the otic capsule with lateral semicircular canal fistula.

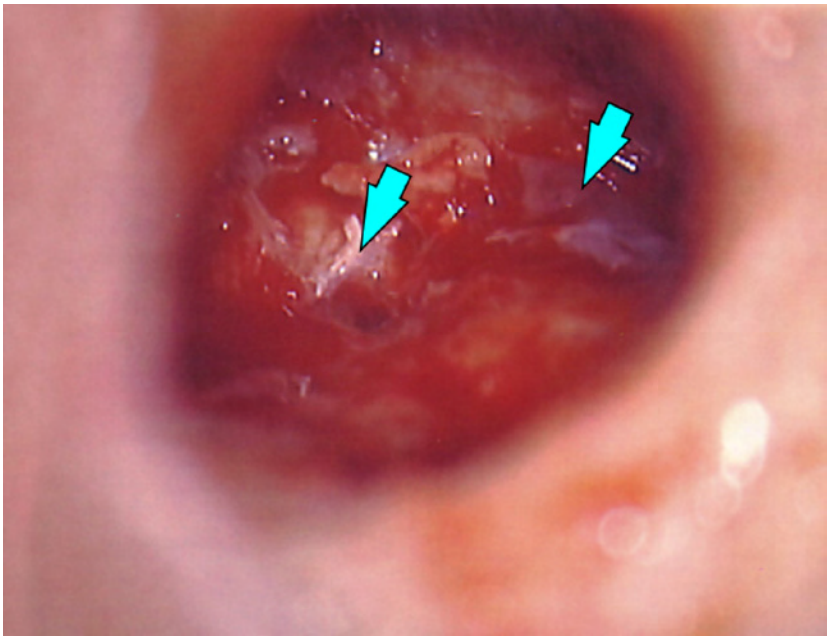

Fig. 6. The bony and membranous labyrinth were destroyed by cholesteatoma.

Discussion

In our study, only 50% of all patients were positive for fistulas and the diagnosis of semicircular canal fistula was made in 91.7% pre-operatively.

Since a perfect pre-operative diagnosis of LF is not possible, the surgeon needs to be prepared for unexpected fistulas. Peltonen[2] reported that CBCT was effective at the diagnosis of middle-and inner-ear areas. In our study, two ears misdiagnosed by MDCT were correctly diagnosed with labyrinthine fistula by pre-operative CBCT. For preservation of hearing and labyrinthine function, sealing the fistula with temporalis fascia and hard tissue are important.

Conclusion

Pre-operative diagnosis using CT imaging and surgical closure of the fistula with temporalis fascia and hard tissue are important in treatment of semicircular canal fistula with middle ear cholesteatoma.

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HEARING IMPAIRMENT IN NASOPHARYNGEAL CARCINOMA PATIENTS

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Background and purpose

As the prognosis of nasopharyngeal carcinoma (NPC) has improved with the introduction of concurrent high-dose chemoradiotherapy (CRT), the prevention of side effects of CRT has become increasingly emphasized. Ear symptoms such as otitis media with effusion (OME) and progressive sensorineural hearing loss (SNHL) are major complications and seriously erode the quality of life of NPC patients. We retrospectively analyzed the factors affecting progressive SNHL in NPC.

Objective and methods

Fifty-three NPC patients were treated in Kumamoto University Hospital between April 1996 and December 2010. Follow-up pure-tone audiometry was performed in 58 ears of 29 patients (23 males and six females), who were then enrolled in this study. The mean age at diagnosis was 59.3 (range 18-86) years. The standard treatment protocol for NPC in our hospital is concurrent CRT, based on cis-diamminedichloroplatinum (II) (CDDP) and 5-fluorouracil (5-FU). Three subjects received radiotherapy (RT) alone due to patient factors and the others received concurrent CRT. We assessed the pre-treatment and time-dependent bone conductive hearing threshold and endoscopic findings of the ear. The mean hearing level was calculated as the hearing at (500 Hz + 1 kHz×2 + 2 kHz)/4 (dB). The NPC patients were classified into two groups based on the follow-up audiometry: the SNHL group had a > 10 dB increase in the bone-conduction hearing threshold (29 ears), and the non-SNHL group had a < 10 dB increment in the bone-conduction hearing threshold (15 ears). Fourteen ears were excluded because their bone-conduction hearing threshold was beyond the measuring limits of the audiometer at the start of CRT. Age, tumor stage according to the International Union Against Cancer (UICC) T category (6^{th} edition, 2002), radiation dose to the cochlea, and total CDDP dose were compared between the two groups. Significance was analyzed using the Mann-Whitney *U*-test or chi-square test and *p*-values < 0.05 were considered to indicate significance.

Results

Age, local tumor stage, and pretreatment bone conductive hearing threshold differed significantly between the SNHL and non-SNHL groups. The mean age of the SNHL and non-SNHL groups was 66.0 (range 43-79) and 54.0 (range 18-72) years, respectively. The difference was significant ($p = 0.0042$, Mann-Whitney *U*-test). Figure 1 shows the patients' UICC T categories. The proportions of T3 and T4 were 55.2% (SNHL) and 13.3% (non-SNHL), respectively. The proportion of locally advanced tumors (T3 and T4) was significantly larger in the SNHL group ($p = 0.0075$, χ^2 test). The pretreatment bone-conduction hearing threshold also differed significantly between the two groups (Fig. 2). The median hearing level in the SNHL group was 25.0

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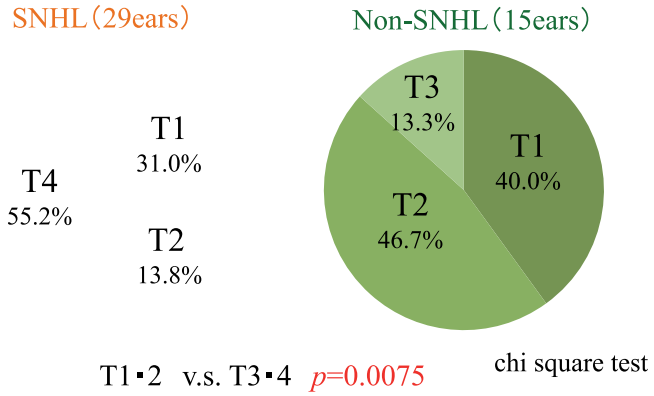

Fig. 1. T category according to the 6th UICC, 2006.

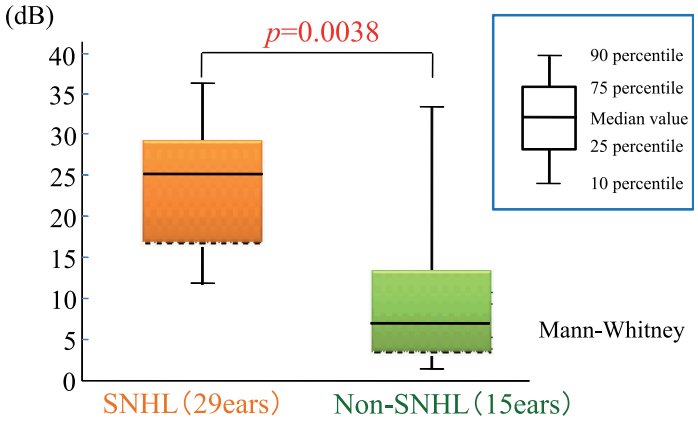

Fig. 2. Pretreatment BC threshold.

(range 6.7-41.7) dB *versus* 6.7 (range 0-35.0) dB in the non-SNHL group (p = 0.0038, Mann-Whitney U-test). The radiation dose to the cochlea and total dose of CDDP did not differ significantly (Figs. 3 and 4). Persistent otorrhea after RT was observed in 15 ears in the SNHL group, while no patients in the non-SNHL group suffered from post-irradiation otorrhea. Fifteen ears in the SNHL group showed progressive deterioration of hearing over two years after RT or CRT (Fig. 5).

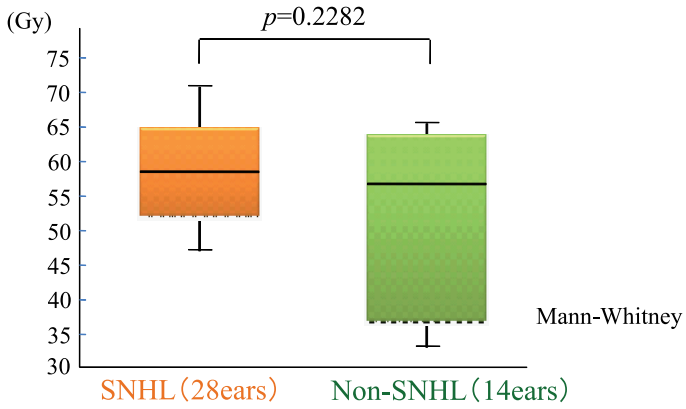

Fig. 3. Radiation dose to the cochlea.

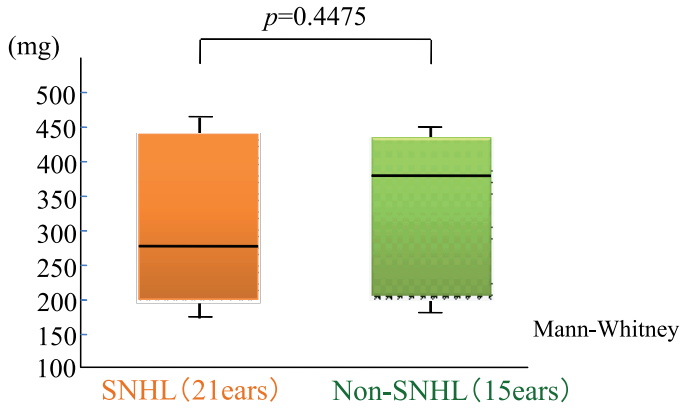

Fig. 4. Total CDDP dose.

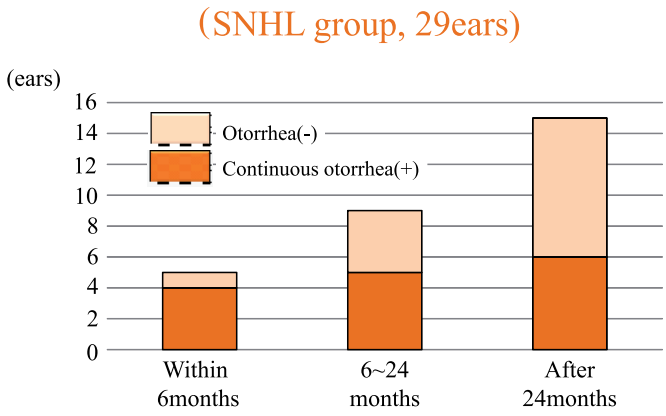

Fig. 5. Time from the end of therapy to the onset of SNHL.

Discussion

Nasopharyngeal carcinoma is a common disease in East and Southeast Asians. Standard therapy is concurrent CRT. The radiation field extends from the skull base to the lower neck, and high-dose cisplatin-based chemotherapy is given simultaneously. Consequently, most NPC patients suffer from side effects. SNHL is one of the most common late complications, and occurs more than 80% of NPC patients, including non-complaining patients.[1] It is reported that post-radiation SNHL is related to patient age.[1-3] Our data also suggest that old age is a risk factor for post-treatment SNHL. Moreover, the condition of patients who have pretreatment SNHL tends to worsen after the treatment. Inner-ear fragility might be related to the post-treatment SNHL. Periodic audiometric examinations are important in post-treatment NPC patients.

In this study, the radiation dose to the cochlea and total dose of CDDP did not differ significantly between the SNHL and non-SNHL groups. There are numerous reports that radiation might be related to the post-radiation SNHL, especially high-frequency SNHL.[1-4] In our study, the prescribed irradiation dose was completed in all patients, so the median dose of radiation to the cochlea was similar: 58.8 Gy (SNHL) and 54.9 Gy (non-SNHL). Chen *et al*. recommend a mean cochlea radiation dose of < 47 Gy to control the ototoxicity of CRT for NPC patients.[2] In our study, the radiation dose to 28 of 29 ears exceeded 47 Gy in the SNHL group. In comparison, five of 15 ears in the non-SNHL group had doses < 47 Gy. Such high-dose radiation to the cochlea might have led to the SNHL. To prevent late complications of RT, intensity-modulated radiation therapy (IMRT) is commonly used for NPC therapy. We started IMRT in 2008, but the mean radiation dose to the cochlea has not decreased (data not shown). A complete cure of their NPC is the most important issue for the patients, so there are practical limits to decreasing the radiation dose.

Continuous otorrhea has been suggested to be a risk factor for gradually progressive SNHL. RT causes immune or mucosal changes in the nasopharynx and middle ear,[5] so many NPC patients are tormented by stubborn otorrhea. However, the treatment of post-radiation OME remains controversial. Tympanoplasty or

grommet insertion was ineffective in some recalcitrant OME cases.[6] As the prognosis of NPC has improved, more patients suffer from continuous otorrhea and progressive SNHL. Further investigation is needed to control post-radiation OME.

Conclusions

Our data suggest that inner ear fragility and persistent otorrhea are risk factors for progressive post-treatment SNHL. In addition, the radiation dose to the cochlea is related to SNHL. Unfortunately, there is no effective method for preventing post-treatment SNHL. This remains a future challenge.

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CLINICAL FEATURES OF CHOLESTEATOMA WITH LABYRINTHINE FISTULAE

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Introduction

Chronic otitis media with cholesteatoma can cause various complications such as vestibular dysfunction, sensorineural hearing loss, facial palsy, and meningitis. Inner-ear disturbance is usually induced by labyrinthine fistulae, *e.g*., semicircular canal fistula or cochlear fistula. In this study, we evaluated the clinical features of cholesteatoma with labyrinthine fistulae.

Material and methods

We enrolled 220 patients with cholesteatoma; of these patients, 210 had acquired cholesteatoma and ten had congenital cholesteatoma. All patients underwent tympanoplasty and/or mastoidectomy surgical procedures. Pure-tone audiometry was performed, and computed tomography (CT) scans (axial and coronal views) were examined. Labyrinthine fistula was detected by CT scan pre-operatively and confirmed by observation during surgery.

Results

Fistula location

Cholesteatoma with labyrinthine fistula was diagnosed in 19 patients, including 18 patients with acquired cholesteatoma and one with congenital cholesteatoma. The fistulae were located in the following regions in the 19 patients: 13 in the lateral semicircular canal and six in the cochlea. The site of the cochlear fistulae was the oval window. Patients with lateral semicircular canal fistulae showed defects of incus by 73% and patients with cochlear fistula showed defects of stapes by 83%. The pre-operative CT scan detected 82% of the lateral semicircular canal fistulae, while 83% of the cochlear fistulae (oval window) were not revealed. Because a differential diagnosis between bony and membranous fistula in the lateral semicircular canal could not be confirmed by CT scan, this was confirmed by observation during surgery.

Fistula type

Labyrinthine fistula is classified into two types: bone fistula, which affects only the bony labyrinth, and membranous fistula, which erodes both the bony and membranous labyrinths. Bony and membranous fistulae were found in 11 and eight patients, respectively. Membranous fistulae were found in two of 13 patients

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Edited by Haruo Takahashi

with lateral semicircular canal fistulae, while the other 11 had bony fistulae. All six cochlear fistulae of the oval window were membranous fistulae. Labyrinthine fistulae often cause vertigo, dizziness, hearing loss, or tinnitus. Pre-operative vertigo was observed in two patients with membranous fistula of the lateral semicircular canal. Half of the patients with bony fistulae of the lateral semicircular canal experienced pre-operative vertigo, even though the perilymph did not leak out. Only 33% of patients with membranous fistulae of the cochlea complained of vertigo pre-operatively.

Hearing

The bone-conduction hearing thresholds did not deteriorate in the majority (89%) of patients who underwent surgical removal of the matrix covering the labyrinthine fistulae (Fig. 1). A patient with acute sensorineural hearing loss and vertigo secondary to labyrinthine fistula showed improvement in bone-conduction hearing thresholds after complete matrix resection and closure of the lateral semicircular canal fistula by surgery. Removal of the matrix covering the oval window did not affect bone-conductive hearing loss thresholds. However, one patient with a cochlear fistula who did not show sensorineural hearing loss in the first few months after surgery, became deaf six months after surgery.

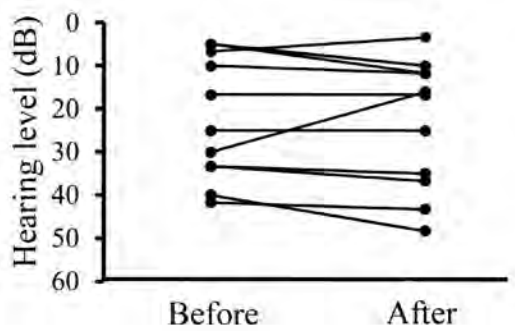

Fig. 1. Bone-conduction hearing thresholds before and after surgery. The bone-conduction hearing thresholds did not deteriorate in the majority of patients who underwent surgical removal of the matrix covering the labyrinthine fistulae.

Discussion

Although both acquired and congenital cholesteatoma can lead to several complications such as ossicular erosion, facial palsy, intracranial infection, and labyrinthine fistula, these complications tend to present less often in congenital cholesteatoma than in acquired cholesteatoma.[1] The reported frequency of labyrinthine fistula is lower in congenital cholesteatoma (0-5%)[1-4] than in acquired cholesteatoma (7-12.5%).[5-8] We postulate that the risk of labyrinthine fistula is related to various factors such as duration, location, and infection. Firstly, long-term exposure to the cholesteatoma matrix increases the probability of labyrinth destruction. Secondly, cholesteatoma usually occurs near the lateral semicircular canal and oval window, especially in acquired cholesteatoma. Thirdly, infection accelerates the destructive feature of the cholesteatoma matrix. These factors determine the destructiveness of the cholesteatoma.

Management of labyrinthine fistulae with total or incomplete matrix removal to prevent deterioration of cochlear or vestibular function is controversial.[9] Some surgeons recommend preservation of the matrix over the fistula with exteriorization through an open cavity, while others completely remove the matrix over the fistula to prevent further effects.[9] Careless removal of the matrix over the fistulae may result in deterioration of the bone-conduction hearing level. However, careful removal of matrix over the fistula could prevent sensorineural hearing loss. Our study suggests that complete resection of the matrix covering the labyrinthine fistula preserves bone-conduction hearing levels and is a safe and effective management method. This safety is supported by the report that cochlear function was preserved after interruption of lateral semicircular canals.[10] Furthermore, it may improve acute sensorineural hearing loss secondary to cholesteatoma fistula.[11] Improvement of sensorineural hearing loss after complete removal of the matrix over a lateral semicircular canal fistula was observed in one of our cases. Otherwise, preserving the cholesteatoma matrix over the fistula may induce delayed sensorineural hearing loss due to suppressive labyrinthitis by 10%.[12] Total removal

of the cholesteatoma matrix over the fistula is advantageous with respect to preventing both further possible osteoclastic bone resorption by the cholesteatoma matrix and possible erosion under the matrix.[9] However, the risk of complications increases for large fistulae. A procedure to remove the matrix may destroy the membranous labyrinth because the adherent membranous labyrinth is pulled. In this case, a second-look operation or preservation of the cholesteatoma matrix over the fistula through an open cavity may be appropriate.[12] Until the secondary operation, the matrix might dissolve spontaneously and the size of fistula may diminish if infection is not present.[13] In the absence of infection, the risk of adhesion of the matrix and the membranous labyrinth may decrease.

A CT scan may suggest the presence of lateral semicircular canal fistulae, but cochlear fistulae of the oval window are difficult to detect pre-operatively. Removal of the cholesteatoma is very risky in cases of covert labyrinthine fistulae. In cases with stapes defects, a cochlear fistula of the oval window should be considered.

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PRE-OPERATIVE EVALUATION OF PETROUS BONE CHOLESTEATOMA BY THREE-DIMENSIONAL (3D) RECONSTRUCTION IMAGES FROM CT

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Introduction

The approaches to petrous bone cholesteatomas (PBCs) are selected usually based on the information of pre-operative CT images, however, it is extremely difficult for surgeons to understand the three-dimensional (3D) relationship of the complicated temporal-bone structures. The aim of this study is to create the precise 3D images from CT easily, and to examine its usefulness in the surgical planning of PBCs.

Methods

First we manually colored pre-operative CT images using Photoshop CS Extended. The inner ear, auditory ossicles, facial nerve (+ internal auditory canal (IAC)) and PBC were shaded in blue, red, yellow and green, respectively (Fig. 1a). We then converted the colored CT images to 3D images (Fig. 1b) using 'Delta Viewer', a freeware for Macintosh available on the Internet (http://delta.math.sci.osaka-u.ac.jp/DeltaViewer/index.html). The 3D images can be rotated freely using the Delta Viewer application. Before surgery, we discussed any problems anticipated based on the 3D images, and planned for the surgery of a PBC within the surgery group.

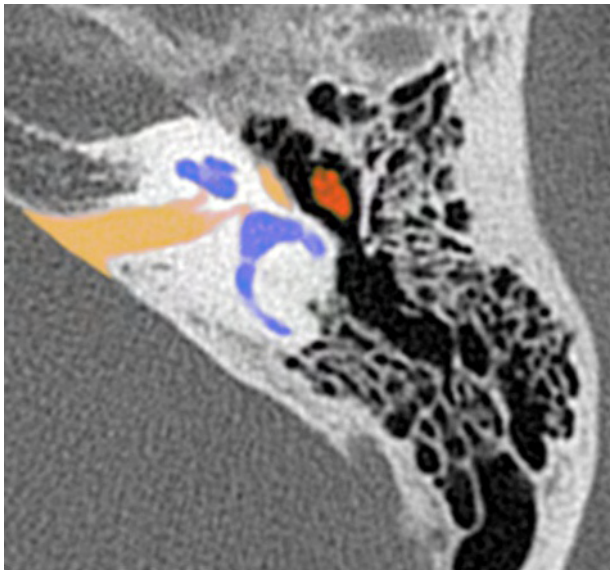

Fig. 1a. Colored CT image of normal left temporal-bone structures.

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Edited by Haruo Takahashi

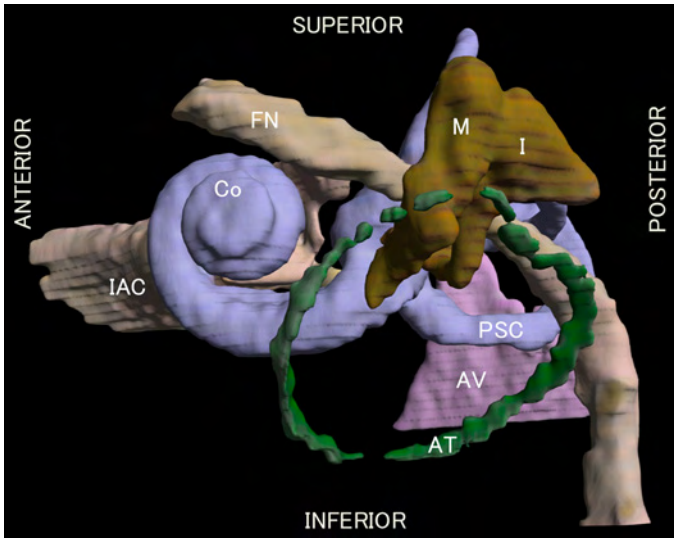

Fig. 1b. Lateral view of the 3D image of normal left temporal-bone structures, converted by Delta Viewer. Co: cochlea; PSC: posterior semicircular canal; IAC: internal auditory canal; FN: facial nerve; M: malleus; I: incus; S: stapes; AT: annulus tympanicus; AV: aqueduct of vestibule.

Case 1: a 17-year-old female

The CT findings of the left ear (Fig. 2) showed that the cholesteatoma had derived from epitympanum extending anteriorly up to anteromedial to the anterior semicircular canal (ASC). The 3D images (Figs. 3 and 4) indicated that a part of the PBC was hidden by the ASC when viewed from the direction of mastoid approach. Referring to the 3D images, we were able to find out the appropriate direction to approach the PBC from anterior to ASC, and successfully removed the PBC without any complication.

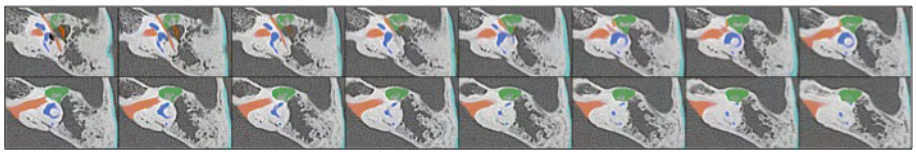

Fig. 2. Colored CT images of case 1; left ear. The cholesteatoma of petrous bone part is colored in green.

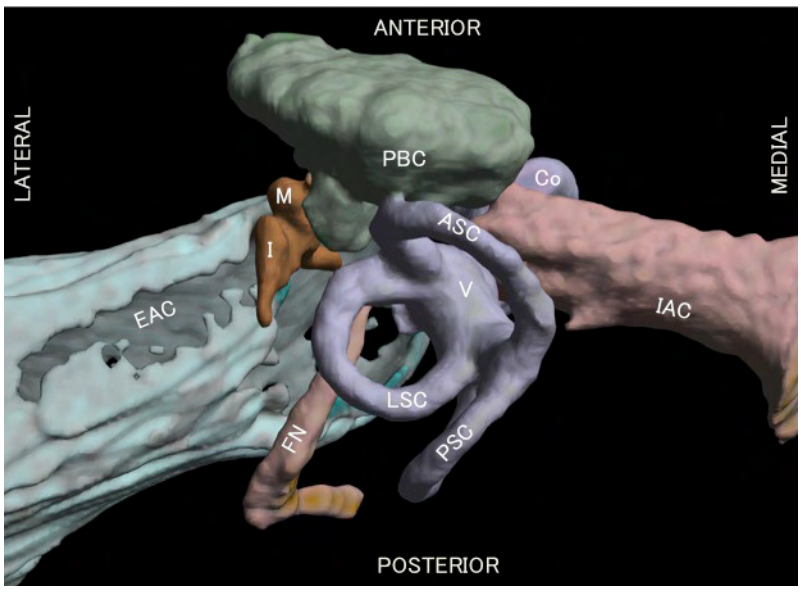

Fig. 3. Superior view of the 3D image of case 1. Co: cochlea; V: vestibule; FN: facial nerve; M: malleus; I: incus; IAC: internal auditory canal; EAC: external auditory canal; LSC: lateral semicircular canal; ASC: anterior semicircular canal; PSC: posterior semicircular canal.

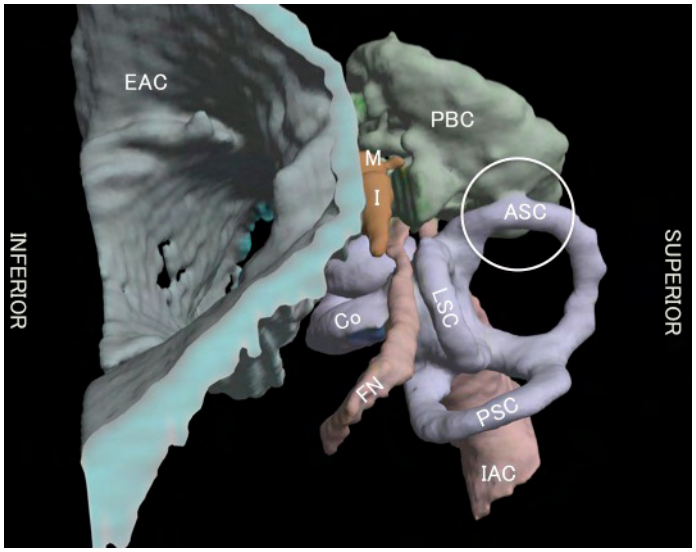

Fig. 4. 3D image viewed from the direction as in the surgery with the transmastoid approach. A part of the PBC is hidden by the ASC (white circle). Co: cochlea; FN: facial nerve; M: malleus; I: incus; IAC: internal auditory canal; EAC: external auditory canal; LSC: lateral semicircular canal; PSC: posterior semicircular canal.

Case 2: a 45-year-old female

The CT findings of the right temporal bone (Fig. 5) indicated a very complicated and odd shape of a cholesteatoma that had derived from the epitympanum and extensively destroyed the floor of the middle cranial fossa, and had extended around the IAC with fistula. The 3D images (Figs. 6 and 7) showed that most of the PBC was hidden by the semicircular canal and it was impossible to reach the part of the PBC around the IAC with the mastoid approach without destroying the ASC even partially. Then we decided to use the translabyrinthine approach and the middle-fossa approach for the part around the IAC. We were able to remove all of the PBC without any remnant of PBC and any complications except post-operative dizziness.

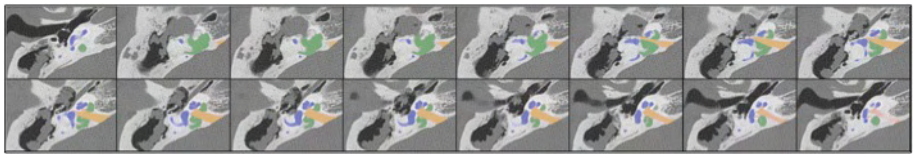

Fig.5. The colored CT images of case2; right ear. The PBC, shaded in green, was complicated and odd shape.

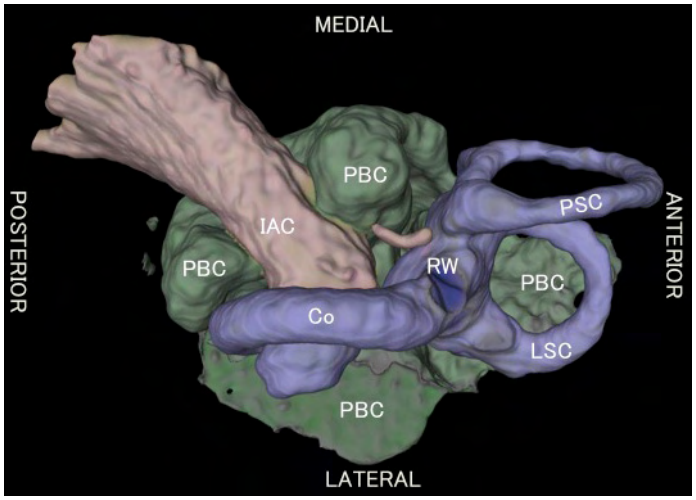

Fig.6. Inferior View of 3D image of case 2. Co, cochlea; RW, round window; IAC, internal auditory canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal.

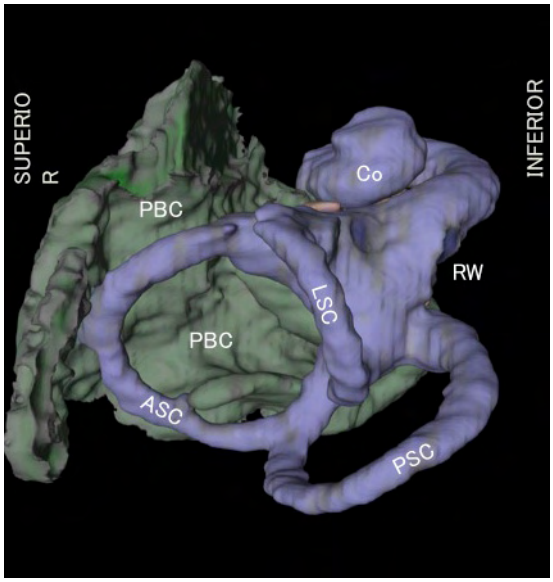

Fig.7. 3D image viewed from the direction as in the surgery with transmasitoid approach. Most of the PBC is hidden by the inner ear labyrinth. Co, cochlea; RW, round window; IAC, internal auditory canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal.

The other cases

The 3D images of 2 other cases are shown in Figures 8 and 9.

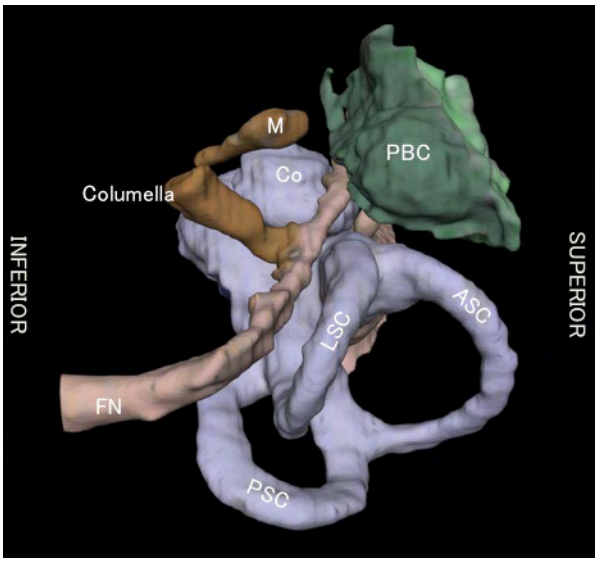

Fig.8. 3D image viewed from the direction as in the surgery with transmasitoid approach; 6-year-old boy, left ear. We performed normal mastoidectomy approach referring this 3D image. Co, cochlea; M, malleus; ASC, anterior semicircular canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal.

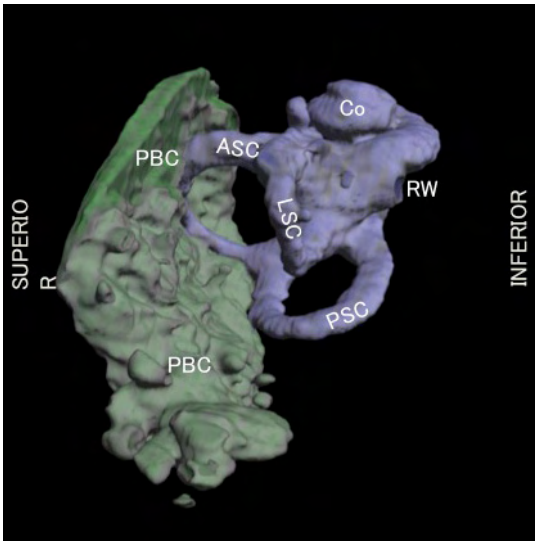

Fig.9a. 3D image viewed from the direction as in the surgery with transmasitoid approach; 67-year-old male, right ear. We performed translabyrinthine approach and sacrificed anterior semicircular canal. Co, cochlea; RW, round window; ASC, anterior semicircular canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal.

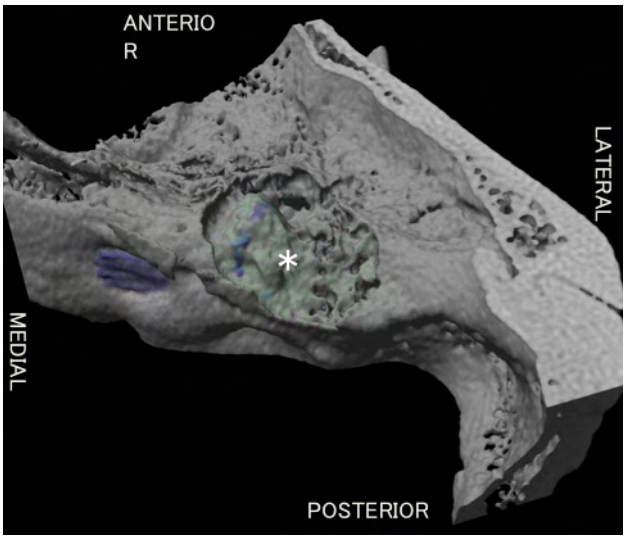

Fig.9b. 3D image of temporal bone viewed from posterosuperior direction; the same case as fig 9a. Asterisk (*) shows the bony defect due to the PBC.

Conclusions

We were able to successfully depict the temporal-bone structures and PBC as 3D images. They were extremely useful to build a strategy of approaching the PBC during the surgeries and we were able to complete the surgeries without any complications in all the cases. This method to make 3D images by Delta Viewer has been reported before.[1]

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RATE OF CHRONIC SUPPURATIVE OTITIS MEDIA IN AN OTOLARYNGOLOGY CLINIC OF A TURKISH MILITARY HOSPITAL

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Objective

Chronic suppurative otitis media (CSOM) is chronic inflammation of the middle ear and mastoid process and it is one of the common diseases of the ear causing mild to moderate hearing impairment.[1] Its incidence appears to depend to some extent on race and socio-economic factors. Poor living conditions, overcrowding, poor hygiene and nutrition have been suggested as a basis for the widespread prevalence of CSOM in developing countries.[2] If CSOM patients need surgical treatment, it may cause some changes in their hearing levels and life styles.[3] Because of that we aimed to find out the rate and course of CSOM in our patients that is representative for the Turkish population.

Material and methods

The present retrospective study is designed to assess the ratio of CSOM applicants and their profile of surgical treatment course in an otolaryngology clinic of a tertiary military hospital in Turkey during a period of two years. All the patients who applied to our clinic for an otolaryngological complaint from January 2010 to January 2012 were reviewed and CSOM patients were identified according to the diagnosis. Then, if surgery was performed, the surgery types were classified.

Results

In two years, a total of 64,792 patients admitted to our clinic for all kinds of otolaryngological diseases and their symptoms. Eight hundred forty-one (1.30%) of them were found to be CSOM patients and 58 (6,90%) of those 841 patients were operated. Operated patients were mostly male (n = 55, 94.83%) and their mean age was 21.8. Of the operated ears, 53.45% (n = 31) were right ears and the others were left ears (n = 27, 46.55%). Canal-wall-down mastoidectomy was applied to 45 patients (77,59%) and canal-wall-up mastoidectomy was applied to the remaining patients (n = 13, 22.41%). Of the canal-wall-down mastoidectomies 60% (n = 27) and of the canal-wall-up mastoidectomies 23,08% (n = 3) were combined with tympanoplasty.

Discussion

Most of the authors say that early diagnosis of the patients with CSOM may reduce the need for a major surgery. In a long-term study it has been shown that 72% of the patients with radical mastoidectomy had similar

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Edited by ww Takahashi

or better hearing levels after surgery compared to hearing levels before surgery.[4] We believe that an early diagnosis is important to prevent the patient from unwanted results and complications of surgery. However, surgical treatment in early stages of CSOM may be beneficial for preventing the residual hearing levels.

Conclusion

Although it seems that the rate of CSOM in our applicants is not high, it still is necessary to diagnose them during an early stage of CSOM development to reduce the need for major surgical treatments. It is also important for prevent hearing disability and other complications due to CSOM.

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MODIFIED BUROW'S SOLUTION IS EFFECTIVE ON REFRACTORY OTORRHEA

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Introduction

Burow's solution, which contains 13% aluminum acetate, has been shown to be effective against chronic otitis media.[1] Since the preparation of Burow's solution is time-consuming, its rapid preparation method has been recently developed.[2] In this study, we evaluated the therapeutic effects of the modified Burow's solution on refractory otorrhea in patients with chronic suppurative otitis and its anti-microbial activity *in vitro*.

Methods

Fourteen ears of 12 patients with chronic otitis media, granular myringitis, otitis externa and postoperative mastoid cavity problems were treated topically with cotton swab/ball soaked with modified Burow's solution or its four-fold diluted ear drops once a week. We then examined the antimicrobial spectrum of modified Burow's solution against clinical bacterial isolates from otorrhea and laboratory bacterial strains *in vitro*.

Results

In all ears, refractory otorrhea disappeared after one to17 weeks of treatment with modified Burow's solution with a mean of 5.4 weeks, without apparent side effects such as ototoxicity. Modified Burow's solution inactivated all Gram-positive bacteria within five minutes except *Enterococcus species*, all Gram-negative bacteria including *Pseudomonas aeruginosa* within 30 seconds and *Candida albicans* within two minutes (Table 1). In addition, modified Burow's solution inactivated MRSA completely within five minutes, while 80.6% of MRSA survived even a 20-minute contact with 0.3% ofloxacin.

Conclusion

These findings indicate that modified Burow's solution, in addition to bearing a broad antimicrobial activity, is as effective as the original Burow's solution in the treatment of chronic suppurative otitis.

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Edited by Haruo Takahashi

Table 1. Anti-microbial activity of modified Burow's solution against clinical bacterial isolates from otorrhoea and laboratory bacterial strains.

	Modified Burow's solution	*Survival rate (%)*						
	Species	Time(min)						
		0	0.5	2	5	10	20	40
Gram-positive species	MSSA	100	77.5	0				
	MRSA	100	97.9	58.0	0			
	Enterococcus sp	100	93.7	93.4	93.2	78.1	47.4	0
	Corynebacterium sp.	100	55.2	0				
	Bacillus sp.	100	0					
Gram-negative species	*Pseudomonas aeruginosa*	100	0					
	Escherichia coli	100	0					
	Aeromas sp	100	0					
	Providencia sp.	100	0					
	Pro. mirabilis	100	0					
	Burkholderia cepacia	100	0					
	K. pneumoniae	100	0					
Fungas	*Candida albicans*	100	55.3	0				

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A CASE OF SECONDARY ACQUIRED CHOLESTEATOMA WITH CARCINOID TUMOR OF THE MIDDLE EAR

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Abstract

Secondary acquired cholesteatoma may occur from trauma, surgical manipulation of the drum or acute otitis media. We presented a 73-year-old lady who had secondary acquired concholesteatoma with carcinoid tumor of the middle ear. While she underwent the myringotomy for right otitis media at another clinic, her hearing was not improved, she therefore visited our department. A perforation at the antero-inferior quadrant of the right tympanic membrane and a white mass both in the cavity of right middle ear and around of the edge of the perforation were observed. We diagnosed this case as secondary acquired cholesteatoma for these finding and performed the tympanoplasty. Most part of the tympanic membrane was adhered to the wall of the middle tympanum. A white mass was localized to the tympanic membrane and another yellowish mass was observed in the middle tympanum. The pathological diagnoses of the white and yellowish masses were cholesteatoma and carcinoid tumor, respectively. In this case, we thought that the blockade of the eustachian tube by carcinoid tumor caused otitis media with effusion, and secondary acquired cholesteatoma appeared at the edge of the perforation after the myringotomy. It is suggested that the careful evaluation would be needed when we see a case of secondary acquired cholesteatoma, since it might be accompanied by be other lesion such as carcinoid tumor.

Case

A 73 year-old lady visited the clinic located near her house, since she had suffed from right hearing loss. She was diagnosed as right otitis media with effusion and underwent the myringotomy twice. However, her hearing was not improved. She therefore visited our department again. On the otoscopic examination, a perforation was observed at the antero-inferior quadrant of the right tympanic membrane (Fig. 1). The white masses were also observed both in the cavity of right middle ear and around the edge of the perforation (Fig. 1). An audiogram revealed mixed hearing loss, a pure-tone average of 76.7 dB on right ear (Fig. 2). The temporal bone computed tomographic scan (CT) revealed the soft tissue density area extending from right middle tympanum to the eustachian tube without any destruction of ossicles (Fig. 3). We diagnosed this case as secondary acquired cholesteatoma for these finding and planned the tympanoplasty. Most part of the tympanic membrane that we thought the perforation was retraction and adhered to the wall of the middle tympanum (Fig. 4A). A white mass was localized just behind the tympanic membrane and another yellowish mass was observed in the middle tympanum (Fig. 4B). Since the rapid pathological diagnosis of yellowish mass was low-malignant potential tumor, these masses were completely removed with a part of the tympanic membrane and the repair of the eardrum was not performed. The pathogenic examination of yellowish tumor revealed the proliferation of cuboidal cells without any nuclear irregularity arranged in glandular pattern (Fig. 5). Immnohistochemical

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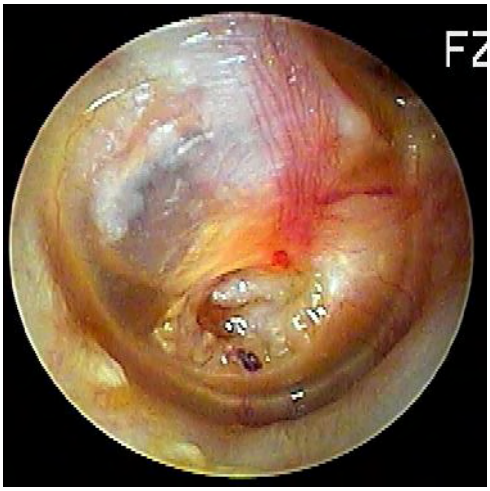

Fig. 1. Otoscopic examination. A perforation at the antero-inferior quadrant of the right tympanic membrane and the white masses both in the cavity of right middle ear and around the edge of the perforation were observed.

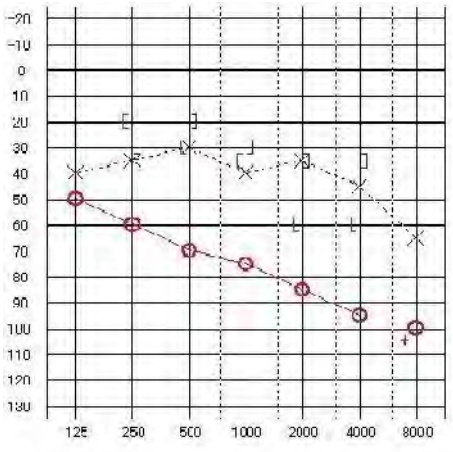

Fig. 2. Audiogram. The mixed hearing loss, a pure-tone average of 76.7 dB, on right ear was revealed.

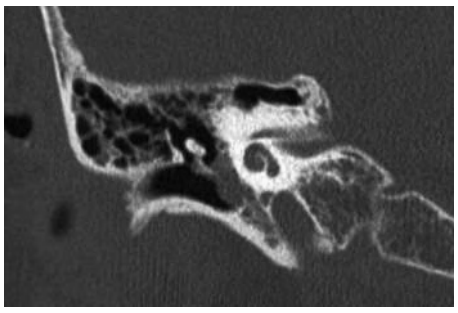

Fig. 3. CT finding. The soft tissue density area extending from right middle tympanum to the eustachian tube without any destruction of ossicles was revealed.

stains for chromogranin and symaptophysin were positive. S-100 protein, Ki-67 and p53 were negative (Fig. 6). The pathological diagnoses of the white and yellowish tumors were cholesteatoma and carcinoid tumor, respectively. The recurrence finding was not observed about for 3 years after the operation (Fig. 7).

Discussion

Cholesteatoma is classified into congenital and acquired. Furthermore, acquired cholesteatoma is divided into primary and secondary according to the cause. Secondary acquired cholesteatoma may occur from a trauma,

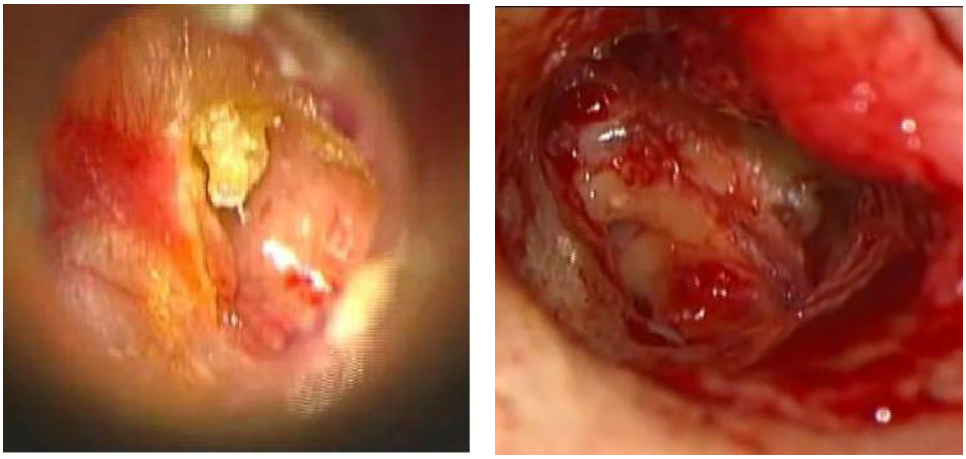

Fig. 4. Otoscopic finding in the operation. A: The retraction at the antero-inferior quadrant of the right tympanic membrane was observed. B : The yellowish mass in the middle tympanum was observed.

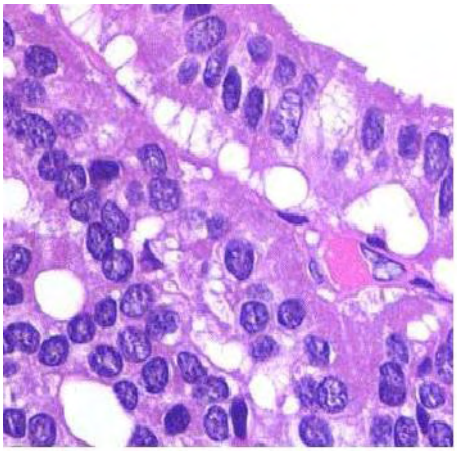

Fig. 5. Pathological finding of yellowish tumor, hematoxylin-eosin stain (×400). The proliferation of cuboidal cells without any nuclear irregularity arranged in glandular pattern was observed.

surgical manipulation of the drum or acute otitis media. Several pathogenic theories to induce secondary acquired cholesteatoma have been reported follow as 1) implant theory[1] 2) metaplasia theory,[2] 3) epithelial invasion theory.[2] It has been reported that the incidence of secondary acquired cholesteatoma following the myringotomy and the ventilation tube insertion in children was ranged from 0.48 to 1.1%.[1,3,4] Carcinoid tumor was one of neuroendocrine tumors and occurs rarely in the middle ear.[5] Carcinoid tumor was observed mostly around ossicles and sometimes extending to the eustachian tube such as this case.[6] Any case of secondary acquired cholesteatoma with carcinoid tumor has been reported ever. In the present case, we thought the blockade of the eustachian tube by carcinoid tumor caused otitis media with effusion first and secondary acquired cholesteatoma then appeared at the edge of the perforation after the myringotomy.

Conclusion

A case of secondary acquired cholesteatoma with carcinoid tumor was presented. We thought the blockade of the eustachian tube by carcinoid tumor caused otitis media with effusion first, and secondary acquired cholesteatoma then appeared at the edge of the perforation after the myringotomy. It is suggested that the careful evaluation would be needed when we see a case of secondary acquired cholesteatoma, since it might be accompanied by be other lesion such as carcinoid tumor.

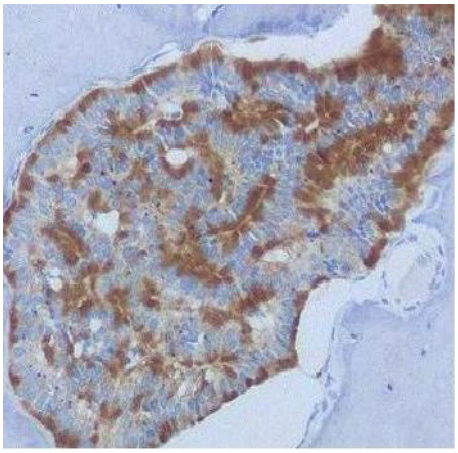

Fig. 6. Immnohischemical stain of yellowish tumor. Symaptophysin was positive.

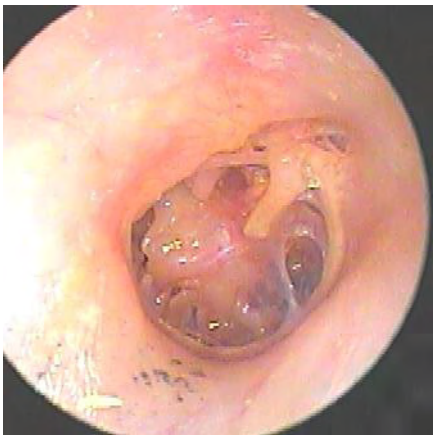

Fig. 7. Otoscopic examination after the operation

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EPIDURAL ABSCESS DUE TO FOREIGN BODY INSERTION INTO THE EXTERNAL AUDITORY CANAL IN AUTISM

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Introduction

Although the occurrence of intracranial complications due to otitis media has declined because of improved antibiotic therapy,[1] they remain to be major challenges for physicians once they occur. We report a case of epidural abscess caused by the insertion of foreign bodies into the external auditory canal in an autistic patient.

Case

A 38-year-old male patient with autism and epilepsy presented with a two-day history of left ear discharge, fever, and vomiting. Purulent otorrhea and inflammatory granulation tissue were seen in his left ear, and several cotton-like foreign bodies were seen in his right ear. Audiometry was not possible due to mental retardation. He was febrile (39.2 degrees), with impaired consciousness (Glasgow coma scale: E3V3M5). Initial laboratory tests showed severe inflammation: white blood cell count, 14,100 /μL and C-reactive protein level, 33.4 mg/dL. Cerebrospinal fluid examination showed bacterial meningitis: increased leukocyte count (3,436 /mm^3), increased protein level (292 mg/dL), and decreased glucose level (22 mg/dL).

CT and MRI findings

Head computed tomography (CT) scans of the axial view showed left tympanomastoiditis without aeration, and contained a hyperdense object lateral to ossicles (Fig. 1A), epidural abscess with gas formation in the posterior cranial fossa and sigmoid sinus occlusion (Fig. 1B). Head T1WI magnetic resonance (MR) with gadolinium contrast showed non-enhancing crescent in the lateral part of the posterior cranial fossa which was considered as abscess with sigmoid sinus occlusion (Fig. 2A). DWI MR illustrated hyperintense signal in the left posterior cranial fossa (Fig. 2B). Both CT scans and MR images revealed no midline shift of his brain. He was diagnosed as epidural abscess of the left posterior cranial fossa induced by tympanomastoiditis due to acute exacerbation of chronic otitis media.

Surgery

An emergent surgery was performed on the same day. We drained the posterior fossa abscess by drilling a burr hole in a retro-sigmoid region. Mastoidectomy was then performed and the foreign bodies of wood and cotton were removed. The bone covering the sigmoid sinus was removed to drain the posterior fossa abscess. The necrotic tissue around the sigmoid sinus was cleaned. A blind sac closure was performed to prevent the

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Edited by Haruo Takahashi

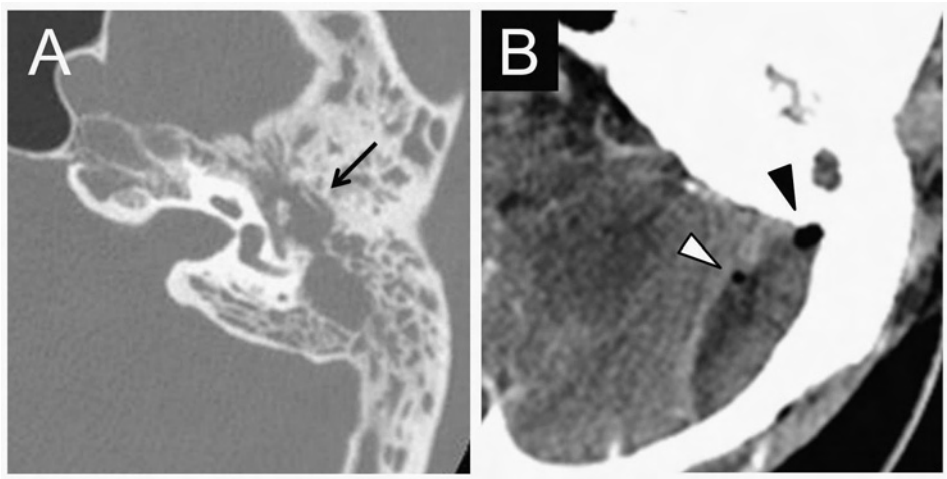

Fig. 1. Head CT scans of the axial view showed left tympanomastoiditis without aeration, and contained a hyperdense object lateral to ossicles (arrows) (A), epidural abscess with gas formation (arrowheads) in the posterior cranial fossa and sigmoid sinus occlusion (B).

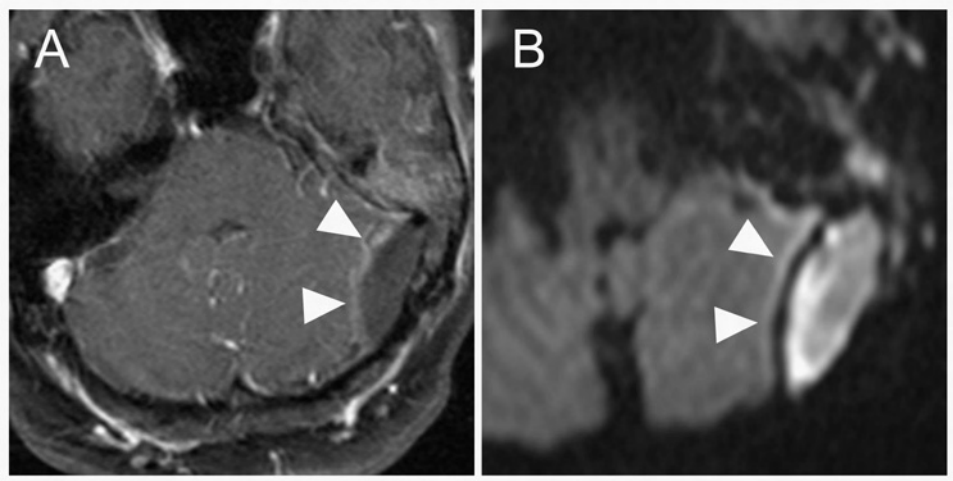

Fig. 2. Head T1WI MRI with gadolinium contrast showed non-enhancing crescent in the lateral part of the posterior cranial fossa which was considered as abscess with sigmoid sinus occlusion (arrowheads) (A). DWI MRI illustrated hyperintense signal in the left posterior cranial fossa (arrowheads) (B).

patient from touching his ear and inserting objects into it. Foreign bodies in his right auditory canal were then removed.

Post-operative course

The patient was treated with broad-spectrum antibiotics (MEPM 4 g/day, VCM 2 g/day). *Prevotella sp.* was isolated from the blood and the pus from the posterior cranial fossa. On post-operative day 7, his consciousness became clear and an enhanced head CT scan did not show the pus accumulation. On post-operative day 16, cerebrospinal fluid examination was normal. He was discharged on post-operative day 21. He had a habit of touching his ears and inserting things into them. After the surgery, we repeatedly showed him a picture saying ‘Don’t touch your ears’, since autistic patients tend to rely more on visual perception. He has never attempted to touch his ears and we have never detected foreign bodies in his right ear after the discharge. A head CT scan taken after three months the surgery revealed that the sigmoid sinus occlusion was cured. A follow-up CT scan one year after the surgery showed no iatrogenic cholesteatoma and showed an aeration cavity in the middle ear.

Discussion

Recently, the occurrence of intracranial complications due to otitis media has declined. The incidence of otogenic intracranial complications is between 0.13 % and 1.97 %.[1] In this case, however, epidural abscess occurred due to the following reasons. It was challenging to examine and treat the patient because of severe mental retardation. He habitually inserted foreign bodies to his external auditory canals. It was supposed that the abscess had been established secondary to inflammatory granulation of the external auditory canal and sustained infection derived from foreign bodies inside granulation.

Because epidural abscess caused meningitis and loss of consciousness, we performed emergent surgery to control the infection. Audiometry was not possible due to mental retardation, but his left hearing was presumed to be poor because of the erosion of ossicle chain and granulation of the tympanic cavity. Blind-sac closure was performed to avoid the secondary infection and to prevent him touching the ear canal.

In addition to blind-sac closure, psychotherapeutic approaches were effective. Autism is a neurodevelopmental disorder characterized by impaired communication and social skills and repetitive or stereotypical behaviors.[2] Because autistic patients tend to rely more on visual perception, it was reported effective to use pictures instead of words to help autistic patients to communicate.[3]

There was no trouble with his ears after the surgery. Both surgical and psychotherapeutic approaches were necessary to cope with such difficulties.

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A CASE OF BRAIN ABSCESS SECONDARY TO MIDDLE-EAR CHOLESTEATOMA

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Introduction

Despite the benefits of antibiotics and computed tomography (CT) examinations, chronic otitis media with cholesteatoma is sometimes accompanied by brain abscess. We report a case of brain abscess due to incomplete treatment for meningitis secondary to middle-ear cholesteatoma.

Case report

A 55-year-old man was referred to our hospital with high fever, nausea, persistent right otorrhea and right hearing loss. A white debris was observed (Fig. 1A). CT revealed bony destruction in the right tegmen (Fig. 1B). Magnetic Resonance Imaging (MRI) revealed the existence of epidural abscess (Fig. 1D). Audiogram showed right mixed hearing loss (Fig. 1C). The examination of spinal fluid obtained by lumbar puncture showed marked elevation of the white blood cells count. He was therefore diagnosed with otogenic bacterial meningitis.

After he was treated using intravenous antibiotics for about one month, the epidural abscess had disappeared. A surgery for middle-ear cholesteatoma was then planned, however, he refused the surgery and was discharged.

As a result of incomplete treatment to his middle-ear cholesteatoma, the patient was admitted to our hospital by ambulance three weeks later, suffering from high fever, headache, right otalgia and disordered consciousness. CT (Fig. 2A) and MRI (Fig. 2B) revealed a brain abscess in the right temporal lobe in addition to right middle-ear cholesteatoma.

Pseudomonas aeruginosa which was sensitive to cephem series and carbapenem series antibiotics was detected in the right otorrhea at second admission. Meropenem (MEPM) and vancomysin (VCM) were administered because anaerobic bacteria and methicillin-resistant *Staphylococcus aureus* could not be ruled out. *Pseudomonas aeruginosa* was detected again one month later at second admission, however, it was resistant to MEPM. MEPM was therefore replaced by ceftriaxone (CTRX).

After the brain abscess reduced and the patient's condition had improved, the surgery was then performed to remove the cholesteatoma, using canal-wall-down procedure. Since a dehiscence in the tegmen was observed (Fig. 3A), it was covered with bone tips and bone paste (Fig. 3B).

The brain abscess was cured conservatively with a course of antibiotics for as long as seven months (Fig. 4A). The patient has been under observation as an outpatient for seven months already and has presented no signs of recurrence of a cholesteatoma (Fig. 4B).

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Edited by Haruo Takahashi

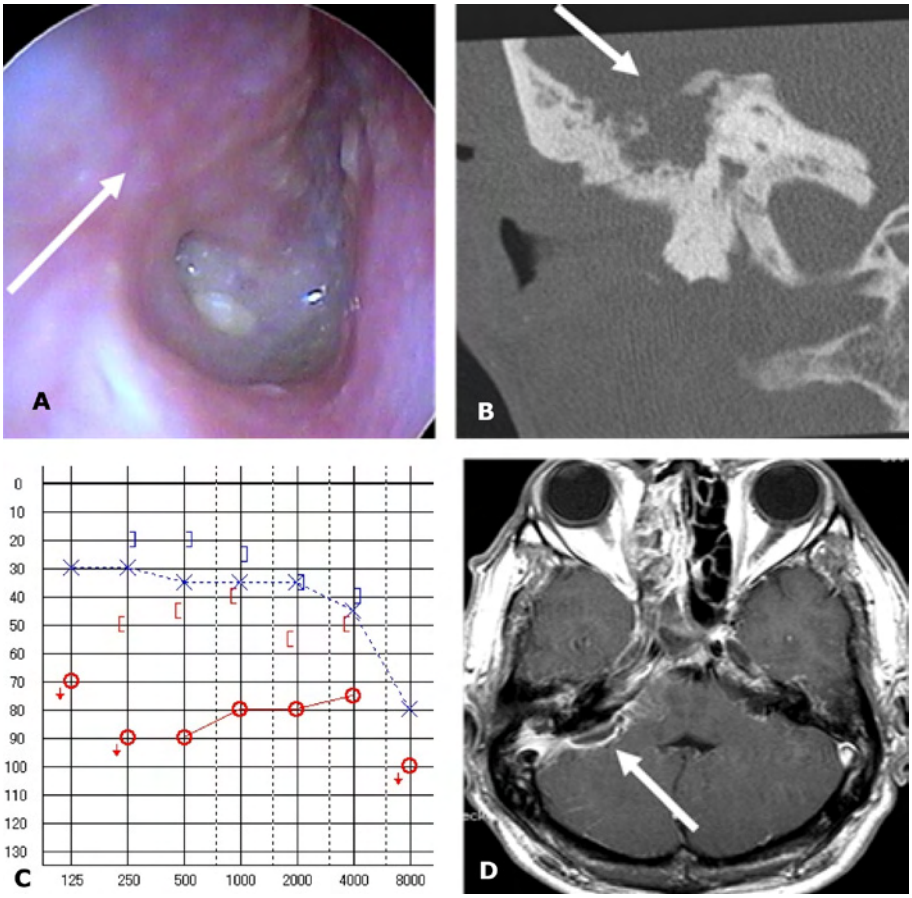

Fig. 1. A. A white debris was observed. The swelled posterior wall of the external auditory canal (arrow). *B.* Coronal section CT. Bony destruction in the right tegmen (arrow). *C.* Audiogram showed right mixed hearing loss. *D.* Axial section MRI. Epidural abscess in front of the right cerebellar hemisphere (arrow).

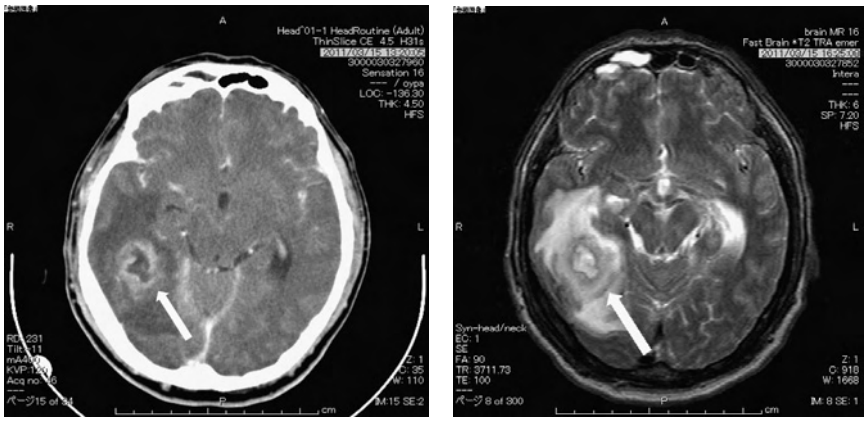

Fig. 2. A. Axial section CT at second admission. Brain abscess in the right temporal lobe (arrow). *B.* Axial section MRI at second admission. Brain abscess in the right temporal lobe (arrow).

Discussion

Nowadays, otogenic brain abscesses are rarely encountered in the ENT clinic in developed countries. Osama *et al*. has reported that the most frequent intracranial complication of chronic otitis media was meningitis (1.4%), followed by brain abscesses (0.35%).[1] In our institute, 251 patients with suppurative otitis media underwent tympanoplasty during an eight-year period (2003-2010). Two patients presented with meningitis (0.79%) and only one patient (reported in this article) presented with brain abscess (0.39%).

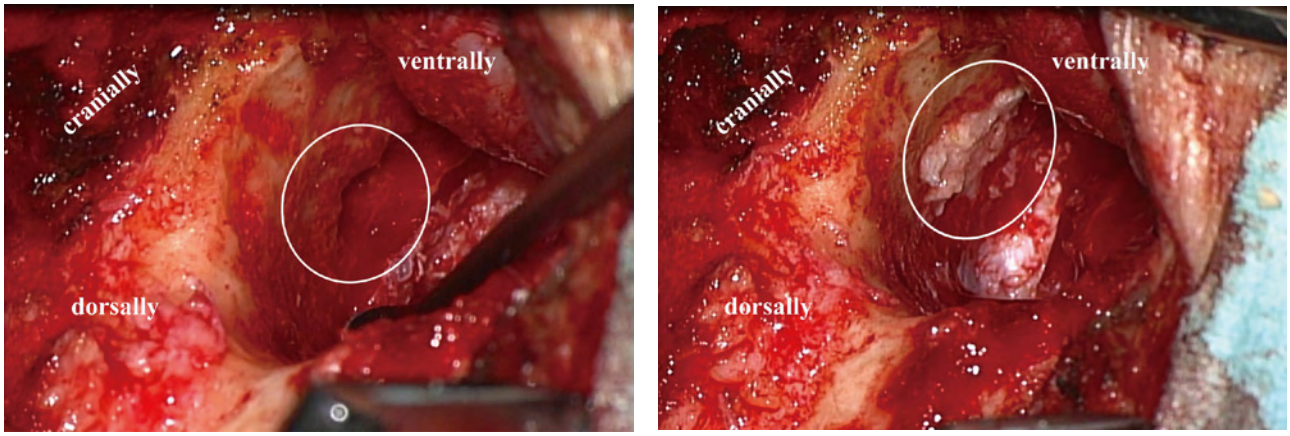

Fig. 3. A. A dehiscence in the tegmen (circle). *B.* A dehiscence was covered with bone tips and bone paste (circle).

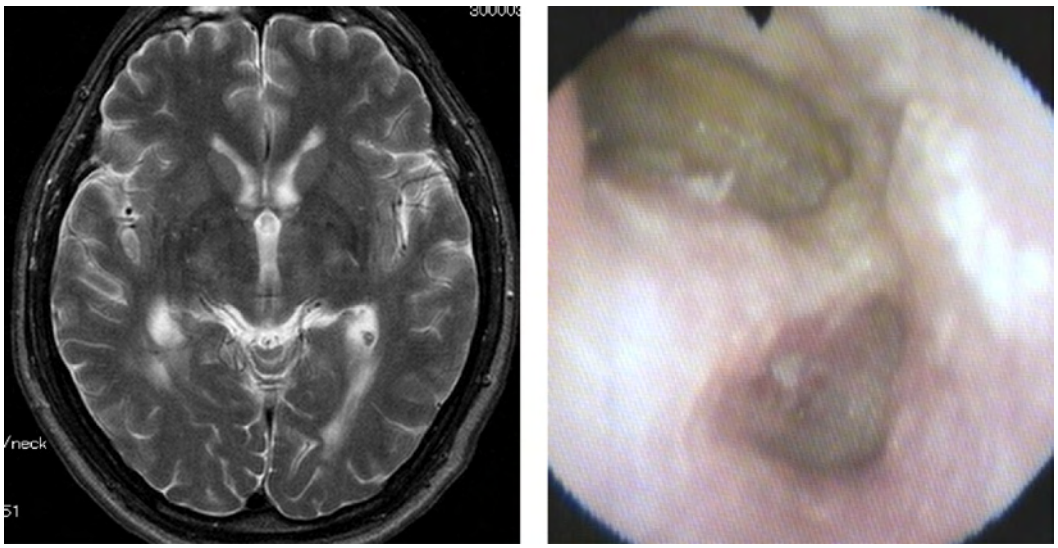

Fig. 4. A. Axial section MRI. Brain abscess has disappeared. *B.* No recurrence of a cholesteatoma.

It is controversial when to operate for a cholesteatoma with intracranial complications. In this case, the patient presented with brain abscess after he denied to undergo middle-ear surgery at first admission. Penido *et al*. have reported two patients who did not take surgery for a cholesteatoma and died from brain abscess. Therefore, the procedures concerning a cholesteatoma with intracranial complications should be performed sooner, preferably before or concomitantly with neurosurgical intervention.[2] Regarding treatment for brain abscess, Rosenblum *et al*. have proposed that a patient with otogenic brain abscess with severe disordered consciousness should be operated as soon as possible. However, if the patient is alert or lethargic, antibiotics would be administered for two weeks to maximize the chance for a well-formed abscess wall to develop.[3]

In this case, the brain abscess was cured conservatively with antibiotics for seven months. On the other hand, Morwani *et al*. have reported that transmastoid drainage of intracranial abscess and concurrent treatment of the otogenic pathology is an effective treatment for otogenic intracranial abscesses.[4]

It is important to have appropriate conservative therapy using antibiotics, appropriate neurosurgical or otological procedures with neurosurgeons and neurologists.

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REGENERATION OF MASTOID CAVITY USING POLYGLYCOLIC ACID SHEETS

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Objective

It is very important to recover mastoid aeration after ear surgery, especially in case of cholesteatoma sometimes recurs. Gas exchange via the middle-ear mucosa functions actively in the normal middle ear and plays an essential role in the regulation of middle-ear pressure.[1-8] Therefore, it is thought that regeneration of the middle-ear mucosa after removal of a nidus by ear surgery contributes to the aeration of the mastoid cavity after ear surgery. Polyglycolic acid (PGA), which is absorbed into the body, is often used in regenerative medicine as scaffold. It suppresses scar formation and regenerates the original tissue. The aim of this study was to regenerate the normal middle-ear mucosa and the pneumatic mastoid cavity after ear surgery in cases of cholesteatoma, using PGA sheets.

Materials and methods

Five patients with attic cholesteatoma underwent tympanoplasty with mastoidectomy. At the operation we stuck PGA sheets on attic and mastoid cavity, the mucosa of which was resected in order to remove cholesteatoma, and fixed them with fibrin glue (Fig. 1). Age at operation was from 23 to 67 years old (median 42 years). Four of the five patients underwent planned staged tympanoplasty. The remaining one had single-stage tympanoplasty. The observation period from the initial surgery was nine to 17 months (median 12 months). Four of the five patients had a poorly developed mastoid cavity. In these cases there was no aeration in the attic and mastoid cavity before operation. Degree of the pneumatization of attic and mastoid cavity was estimated by CT scan which was performed after initial operation from six months to one year.

Results

All patients had a well-pneumatized attic after the initial surgery. In two cases the mastoid cavity was fully pneumatized and also in two cases partially pneumatized (Table 1). In three cases that underwent a second-stage operation, PGA sheets used at the first-stage operation were absorbed and replaced by normal mucosa. The form of the tympanic membrane remained almost normal in all cases.

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Edited by Haruo Takahashi

Table 1. Results of all cases

case	age	sex	postoperative period (month)	operation	development of mastoid	attic aeration		mastoid aeration	
						preop.	postop.	preop.	postop.
1	42	f	17	staged	very poor	(+)	(++)	(+)	(++)
2	37	m	15	staged	poor	(-)	(++)	(-)	(+)
3	23	f	15	staged	well	(-)	(++)	(-)	(++)
4	46	f	10	staged	very poor	(-)	(++)	(-)	(+)
5	67	f	9	single- stage	very poor	(-)	(++)	(-)	(-)
						(-) no pneumatization			
						(+) partially pneumatized			
						(++) almost or fully pneumatized			

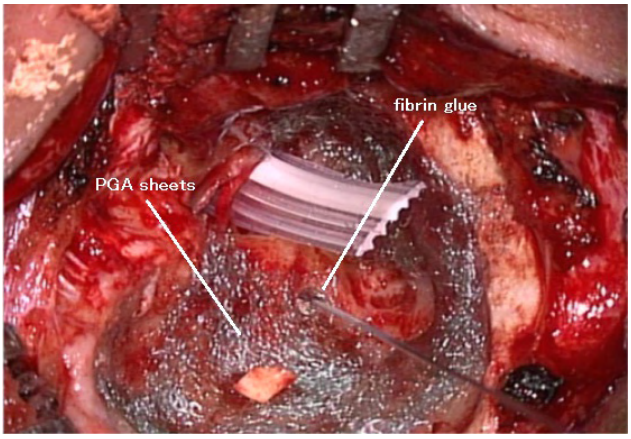

Fig. 1. PGA sheets are fixed with fibrin glue (case 3).

Case presentation (Case 3)

The case involves a 23-year-old female patient. She had been diagnosed with cholesteatoma in her left ear and underwent surgery a few years ago, but purulent otorrhea persisted even after the operation. Therefore she consulted Osaka University hospital. At the first examination, attic retraction pocket, in which purulent otorrhea existed, was seen in her left ear. She was diagnosed with recurrent cholesteatoma. A CT scan before the operation revealed that no aeration could be seen in her attic and mastoid cavity (Fig. 2). Therefore, a planned staged tympanomastoidectomy using PGA sheets was performed. Six months after the first-stage operation, a CT scan was made. It was found that her mastoid cavity was fully pneumatized after the first-stage operation (Fig. 3). At the second-stage operation, performed 12 months after the first operation, the PGA sheets were absorbed and replaced by a normal mucosa (Fig. 4).

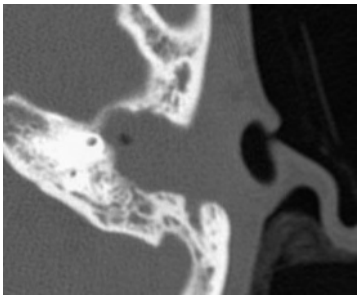

Fig. 2. CT scan image before the first-stage operation. No aeration can be seen in the attic and mastoid cavity.

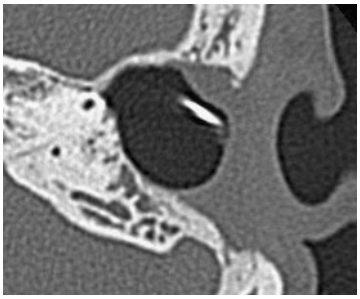

Fig. 3. CT scan image after the first-stage operation. The mastoid cavity is fully pneumatized.

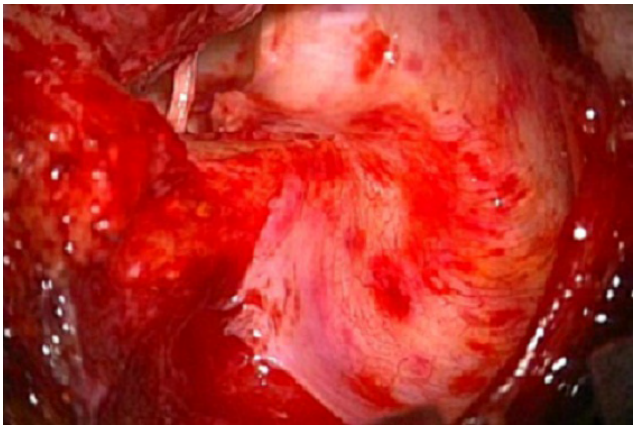

Fig. 4. PGA sheets used at the first-stage operation are absorbed and replaced by normal mucosa.

Discussion

The middle-ear mucosa plays an essential role in the regulation of middle-ear pressure. Gas exchange according to concentration or pressure gradients via the mono-layered mucosa supplies adequate pressure in the middle ear. A defect of the normal middle-ear mucosa after removal of a nidus by ear surgery causes a loss of pressure regulation in the middle-ear cavity. Therefore, it is very important to regenerate the middle-ear mucosa after ear surgery, especially in recurring cases of cholesteatoma because of the negative pressure in the middle ear.

Recently, it is believed that wound healing due to scar tissue formation is faster than the regeneration of an original organ when a wound occurs in mammals. Therefore, the mucosa of the middle ear that has been resected in order to remove cholesteatoma is replaced by granulation tissue after the operation.

PGA has the following characteristics: 1) It is absorbed into the body; 2) It is used in regenerative medicine as scaffold; 3) It suppresses scar formation and regenerates the original tissue.

So it is possible that PGA sheets help the mucosa of the middle ear to regenerate, and suppress attic retraction by scar contraction. As a result of this study, the improvement of pneumatization after operation was

observed in all cases. Also, the form of the tympanic membrane remained almost normal in all cases. This study demonstrated that PGA sheets were useful for regeneration of the mucosa in the middle ear cavity. This method may prevent the recurrence of cholesteatoma.

However, the problems of this study are: 1) The observation period is short; 2) This study does not include comparison with a control group; 3) It is still uncertain whether the regenerated mucosa has a gas exchange function or not. Therefore, further investigation is needed in future.

Conclusion

PGA sheets may be useful for regeneration of the mucosa in the middle-ear cavity.

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REDUCTION IN THE RATE OF RE-PERFORATION RATE AFTER SIMPLE UNDERLAY MYRINGOPLASTY BY USING FIBRIN GLUE

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Introduction

Myringoplasty with a post-auricular incision is routinely performed to repair tympanic membrane perforation. The transcanal approach has also been used in an attempt to simplify myringoplasty by using substances such as fat or micropore tape. However, these techniques did not produce satisfactory results comparable to those of conventional myringoplasty. Yuasa[1] developed a simple underlay myringoplasty (SUM) method using fibrin glue in 1989. Although SUM is a satisfactory technique for repairing tympanic membrane perforation, its success rate is lower than that of simple myringoplasty. Therefore, a re-closure operation is required in unsuccessful cases. A method that decreases the rate of re-perforation may improve the effectiveness of SUM. The objective of the present study was to evaluate the efficacy of the gelatin sponge in preventing postoperative re-perforation.

Materials and methods

SUM was performed in 25 ears for repairing tympanic perforation due to chronic otitis media. The subjects comprised 11 men and 14 women with a mean age of 60.9 years. Indications for SUM included a simple dry perforation, the absence of cholesteatoma, hearing gain in a paper patch, and the lack of a shadow in the attic on computed tomography. Typically, SUM was performed under local anesthesia, except in patients who preferred general anesthesia. The margin of the perforation was excised and removed by using a fine pick through an ear speculum. Subcutaneous tissue from the retro-auricular region was used for the graft. The graft was inserted under the eardrum, and the perforation was closed by using the graft. After ensuring contact between the graft and the margin of the perforation, several drops of fibrin glue were applied. Following this step, a gelatin sponge (Gelfoam®, Pfizer, USA), which was slightly larger than the perforation, was immediately placed on the eardrum. Closure of the eardrum perforation was examined during the postoperative course and finally at 6 months after surgery.

Results

We were able to perform this operation in 25 ears and observe the results for a follow-up period of at least 6 months. The gelatin sponges placed on the eardrum disappeared after a mean duration of 12.8 days. The postoperative re-perforation rate was 1/25 (4.0%). No serious complications were observed in this study.

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Cholesteatoma and Ear Surgery – An Update, pp. 433-434
Edited by Haruo Takahashi

Discussion

The success rate of closure by using conventional myringoplasty for chronic otitis media is often more than 90%. The advantages of SUM are that it is minimally invasive, easy to perform, and has no serious complications; however, postoperative re-perforation has been found in 20 to 30% of the patients undergoing this procedure.[2,3] Therefore, patients in whom closure is unsuccessful require a re-closure operation performed using an autologous frozen material. The high rate of postoperative re-perforation may be because the tampon was not placed in the ear. A tampon is required to attach the harvested graft to the eardrum in conventional myringoplasty. In SUM, fibrin glue plays the role of a tampon; however, the adhesive action of fibrin glue may be weak and may disappear in approximately 1 week. A gelatin sponge has a good affinity with human tissues and is routinely used as a stabilizing material in surgical procedures of the middle ear in many countries. In 1983, Hellstrom *et al.* reported increased fibrosis in rats after insertion of a gelatin sponge in the middle ear.[4] In the present study, the gelatin sponge may have increased fibrosis between the edge of the eardrum perforation and the graft. Thus, placing a gelatin sponge on the eardrum is recommended as the last step in SUM because this procedure is very easy and is useful for the prevention of re-perforation.

Conclusion

SUM is recommended for repairing tympanic membrane perforation. Although the postoperative re-perforation is generally 20 to 30%, the re-perforation rate decreased to 4% when a gelatin sponge was placed on the eardrum immediately after the operation.

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CHRONIC INFLAMMATORY LESION OF TEMPORAL BONE INVOLVING INNER EAR WITH MULTIPLE OCULAR MUSCLES PALSY

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Introduction

Inflammatory pseudotumor (IPT) is an uncommon fibro-inflammatory lesion, also referred to as inflammatory myofibroblastic tumor, occurring most frequently in the lung, abdominal cavity, retroperitoneum, and extremities. Its occurrence in the head-and-neck region is less common.[1] Temporal bone IPT is known to destruct adjacent bone structures and to invade the inner ear with or without cranial-nerve paralysis. We studied a case of temporal bone IPT extending to the petrous apex with multiple cranial-nerve paralysis.

Case report

A 52-year-old unmarried male was referred to our clinic with brain MR images, having complained of progressive diplopia for one month. On physical examination, a pinkish, pulsating middle-ear mass was found in his right ear. Left extra-ocular muscle (innervated by CN IV) and right extra-ocular muscle (CN VI) paralyses were confirmed by a specialized ophthalmologist (Fig. 1). A high-resolution temporal bone CT scan was

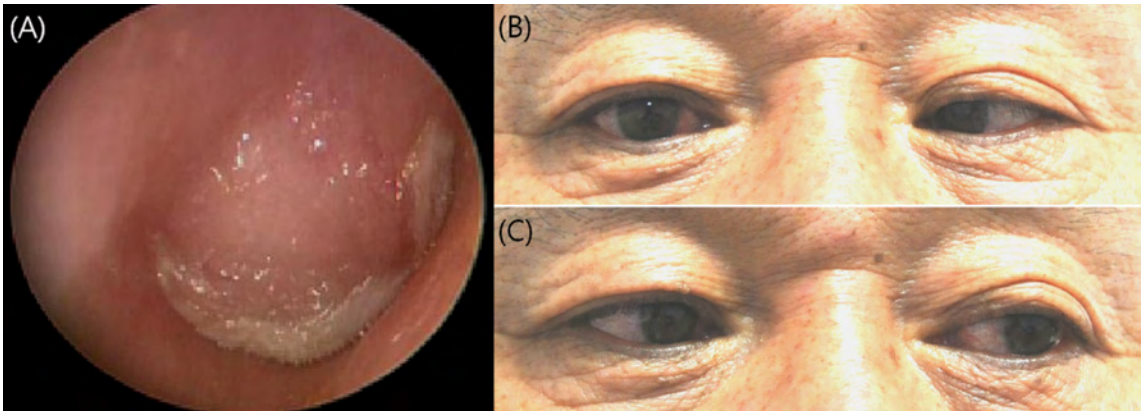

Fig. 1. Clinical features of a 52-year-old man with inflammatory pseudotumor of temporal bone. A: Oto-endoscopy of right eardrum showed pinkish pulsating mass. Abnormal eye movement of left IV and right VI extraocular muscles paralysis; B: Right gaze; C: Left gaze.

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Edited by Haruo Takahashi

made to obtain additional information on bone destruction and extent of the disease. Infiltrative soft-tissue densities of the mastoid, middle ear, and petrous apex were found (Fig. 2,3). Our patient experienced sudden hearing loss in his right ear three years ago. Pure-tone audiometry and a vestibular-function test showed total deafness and canal paresis of the lesion side (Fig. 4). Subtotal petrosectomy with partial otic capsule removal was planned for diagnostic and curative purposes (Fig. 5).

Histopathology demonstrated massive mononuclear cell infiltration in the fibrocollagenous tissue without nuclear pleomorphism or mitosis (Fig. 6). Plasma cells were dominant among the chronic inflammatory cells and the immunohistochemistry did not show monoclonality. After the negative bacterial, fungal, and tuberculosis cultural report, high-dose oral steroids (methyl-prednisolone, mPd 60 mg/day) were applied for ten days and tapered to the maintaining dose of 5~10 mg per day.

The patient returned to the clinic at one, three, six, and 12 months after surgery. Diplopia had slowly improved and subsided at three months after surgery. Eye movement was completely normalized at the postoperative six-month evaluation. He still used a low dose of steroids (8 mg/d mPd) at the last follow-up visit.

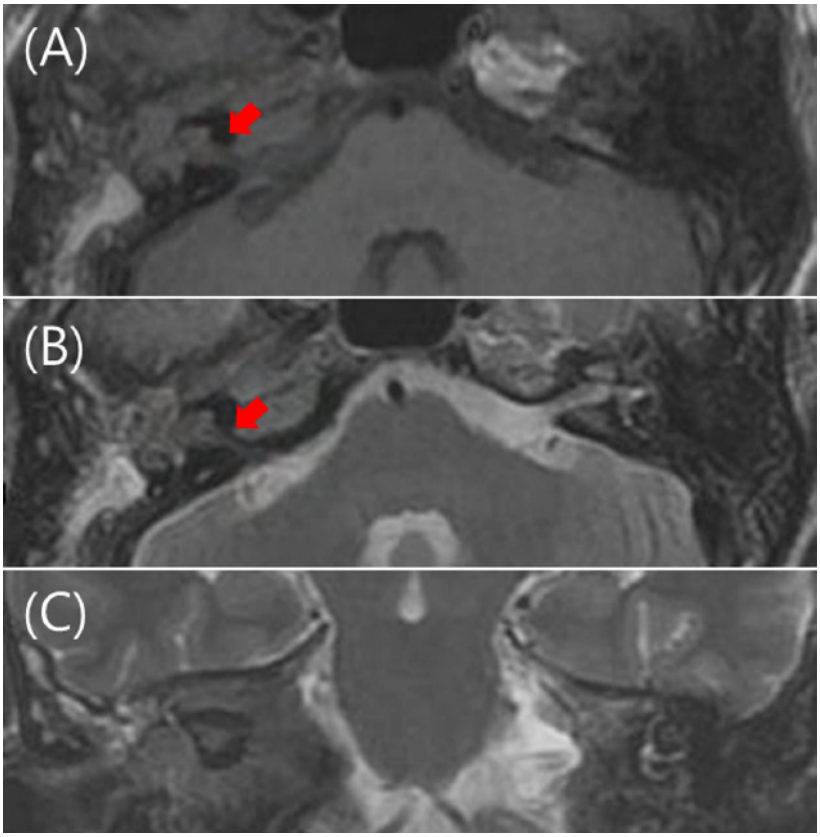

Fig. 2. Temporal bone images. Infiltrative tumorous lesion involving inner ear (red arrows) and inflammatory lesion of the temporal bone; A: Post-gadolinium T1-weighted MR axial image; B: T2-weighted MR axial image; C: T2-weighted MR coronal image.

Discussion

There were not many reports of IPT involving the ear. Almost all reported cases involved temporal bone IPT with or without skull-base extension. Although two cases were confined to the middle ear, most cases involved mastoid and middle-ear cavity with local aggressiveness of neural and inner-ear destruction.[2-4] There are controversies concerning the optimal treatment modality of IPT including surgery, radiotherapy and corticosteroids.[5] Surgery is primarily considered for many cases depending on tumor location and behavior. A high dose of oral steroid can be used solely or combined with surgical resection. Some lesions, such as orbital IPTs, showed a good response to primary radiotherapy, with up to 75% showing a reduction of mass.[6] Tumors

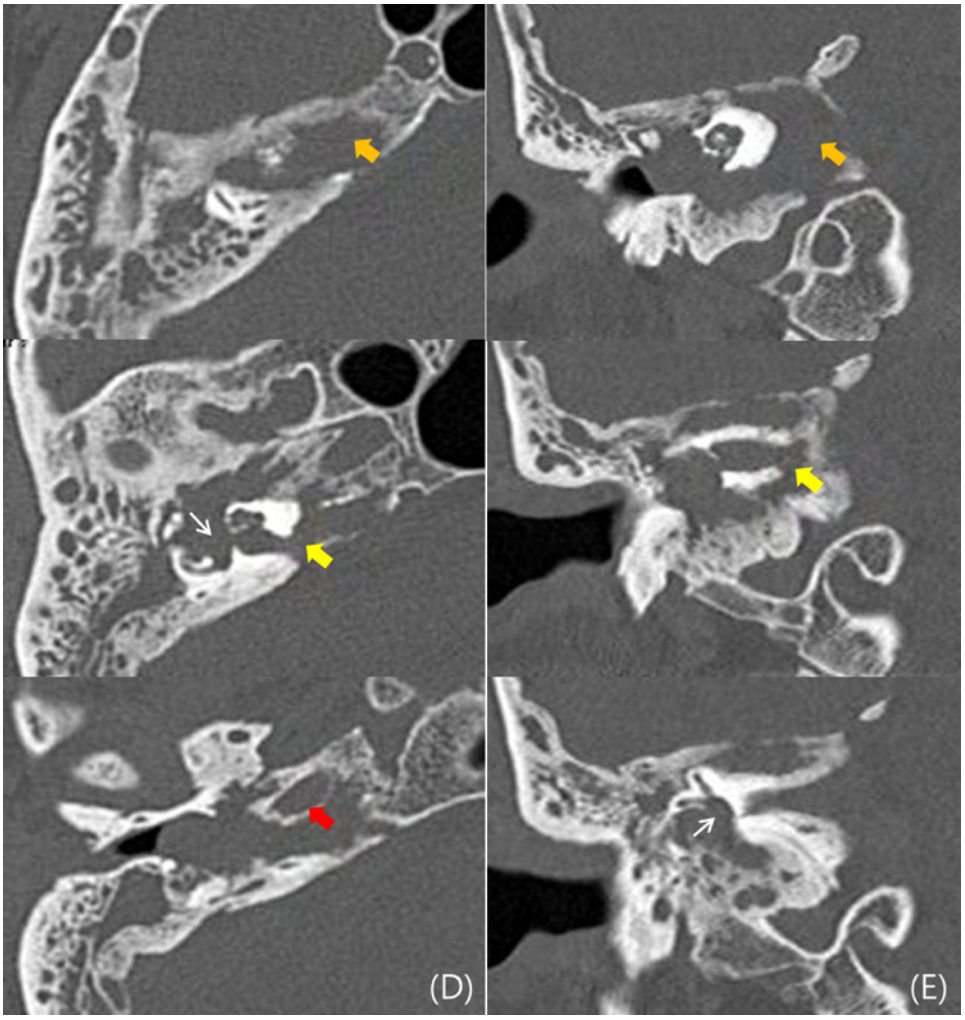

Fig. 3. Temporal bone images. D: Axial; E: Coronal CT scan images. D, E: Computed tomography (CT) images of infiltrative soft tissue filled the middle-ear cavity and extended to petrous apex (orange arrows), erosion in medial wall of the internal auditory canal (yellow arrows), stapes and oval window destruction (white arrows), and internal carotid canal surrounded by the lesion (red arrow)

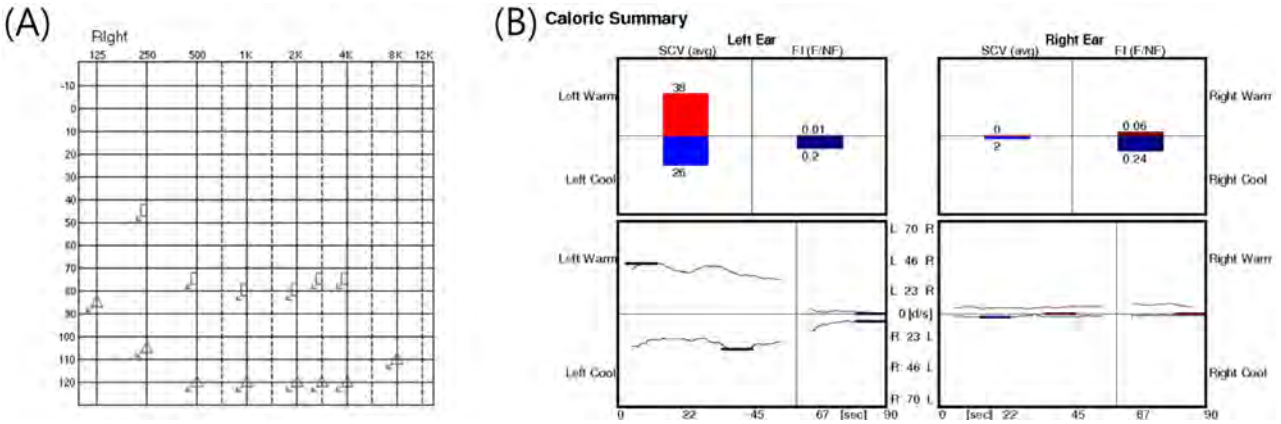

Fig. 4. Pure-tone audiometry and vestibular function test. A: Pure-tone audiometry showed total deafness of right ear; B: Bithermal caloric test revealed 94% weakness of the lesion side.

with a good response to corticosteroids are thought to be the most appropriate for radiotherapy. However, there are documented reports of local recurrence up to 15%, an incidence that can be reflecting inadequate resection of the lesion or tumors with behavior resembling inflammatory fibrosarcoma.[7] Coffin *et al*. also showed that IPT can be a cause of death because of uncontrolled local growth.[1] Temporal bone IPT in our case was clinically aggressive but was treated optimally with surgical excision with steroid maintenance therapy.

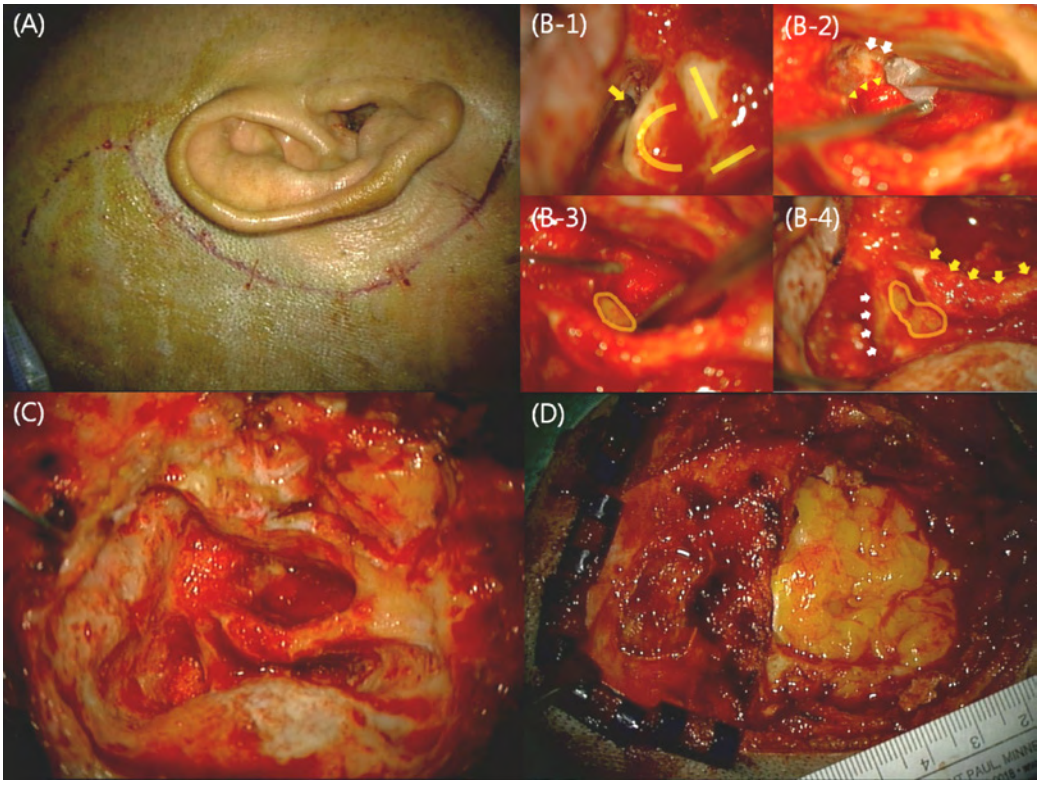

Fig. 5. Subtotal petrosectomy with partial otic capsule removal. A: Design for post-auricular incision line; B-1: Supralabyrinthine air cell tract exenteration was not sufficient for total removal of petrous apex lesion (orange arrow), translabyrinthine approach was conducted to remove the deeply seated petrous apex lesion (yellow lines); B-2: After removal of the middle-ear mass, the Eustachian tube orifice was obliterated with bone wax. Pseudocanal of tensor tympani m.(white arrow), internal carotid canal (yellow arrowhead); B-3: Destructed oval window was sealed with fat graft (orange loop); B-4: Fat graft obliteration of the opened vestibule (orange loop). All air cells and the mass around the vertical portion of the facial nerve (orange arrows) and the internal auditory canal (white arrows) were removed; C: Gross feature after subtotal removal of the mass; D: Obliteration of opened cavity with abdominal fat.

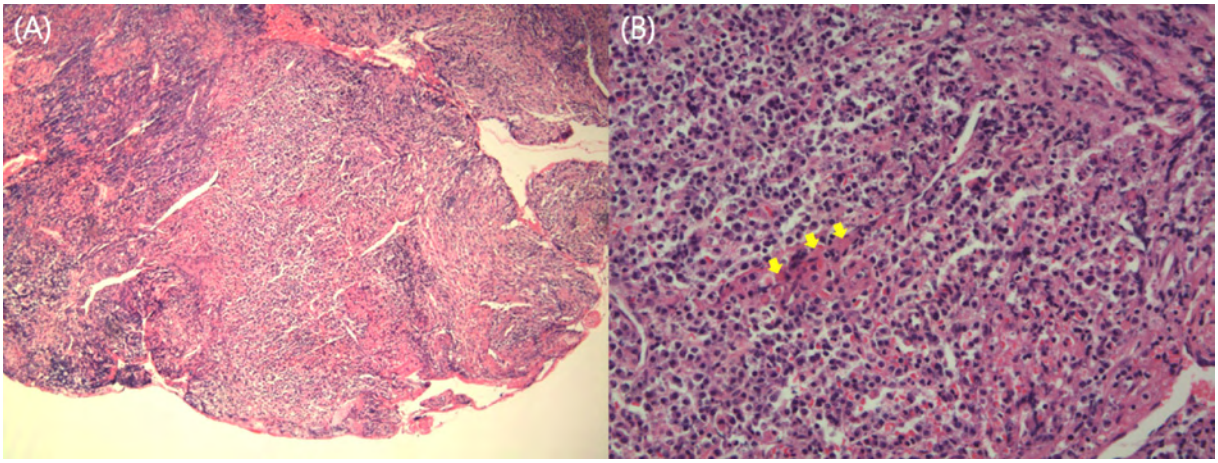

Fig. 6. Pathology of the middle-ear mass. Massive mononuclear inflammatory cell infiltration in the fibrocollagenous tissue without cellular atypism (A, H&E, ×100). Chronic inflammatory cells, mainly plasma cells (arrows, Russell bodies) are seen (B, H&E, ×200).

Conclusion

Temporal bone IPT often shows aggressive features with central neurological symptoms. Complete surgical excision should be considered first and steroids could be helpful for the suppression of the remnant disease progression.

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POST-OPERATIVE CONDITION FOLLOWING CANAL-WALL-UP TYMPANOPLASTY FOR PARS FLACCIDA CHOLESTEATOMA

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Introduction

Our principle strategy for surgery in middle-ear cholesteatoma has been the canal-wall-up tympanoplasty (CWU). Although a well-pneumatized cavity after surgery is the goal of CWU, the extent of re-pneumatization varies widely between tube and mastoidectomy cavity. This study assessed the relationship between 1) the extent of cholesteatoma and re-pneumatized spaces after CWU; 2) the extent of cholesteatoma and hearing improvement after CWU; and 3) the extent of cholesteatoma and recurrence rate after CWU.

Materials and methods

One hundred and nine ears with pars-flaccida type cholesteatoma underwent CWU with anterior tympanotomy[1] and scutum plasty at the University of Miyazaki Hospital from 1998 to 2004. All cases were followed up for more than five years.

Extent of cholesteatoma (staging)

Each ear was assigned to one of three groups according to the extent of the cholesteatoma demonstrated at surgery based on the staging criteria for cholesteatoma 2010 Japan: stage I: cholesteatoma localized within the attic (21 ears, 19%); stage II: cholesteatoma extends into the mastoid cavity (64 ears, 59%); stage III: large defect of the scutum and external auditory canal due to cholesteatoma extension (24 ears, 22%).

Distribution of pneumatization (pre-and post-operative CT)

Within three months before surgery and one year after surgery, a CT was performed in all patients. Each ear was assigned to one of four groups according to the degree of pneumatization: 1) no aeration; no aerated space within the middle ear; 2) mesotympanum; partially or totally aerated bony Eustachian tube, meso-, hypo-, and retrotympanum; 3) epitympanum; aerated tube, mesotympanum, and epitympanum, but no air space in the mastoid; and 4) mastoid; aerated from tube to mastoid antrum.[2]

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Edited by Haruo Takahashi

Results

The distribution of pneumatization (Fig. 1)

Figure 1 shows the distribution of pneumatization in 109 ears pre- and post-operatively. The tube/mesotympanum group accounted for up to 74% on pre-operative CT. Ninety-four percent of the ears showed improved pneumatization after operation. The mastoid (47%) and epitympanum (16%) groups in particular showed significant improvement of post-operative pneumatization ($p < 0.01$, χ^2 test).

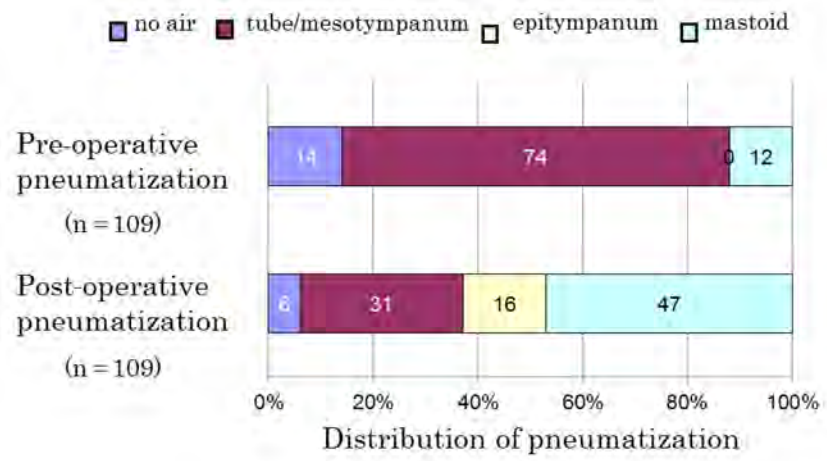

Fig. 1. Distribution of pneumatization.

Re-pneumatization according to preoperative pneumatization (Fig. 2)

The extent of re-pneumatization varied widely, even within the same pre-operative pneumatization group. There were 13 cases (12%) that had good pneumatization to the mastoid on pre-operative CT. In this group, the mastoidectomy cavity showed good re-pneumatization in 84% (ten of 13 ears). Even in the poorly pneumatized group, re-pneumatization to the mastoid was achieved in up to 41~47 percent. In contrast, no aeration in the pre-operative period indicated that the Eustachian tube had provided insufficient ventilation, which prevented even slight improvement (27%, four of 15 cases).

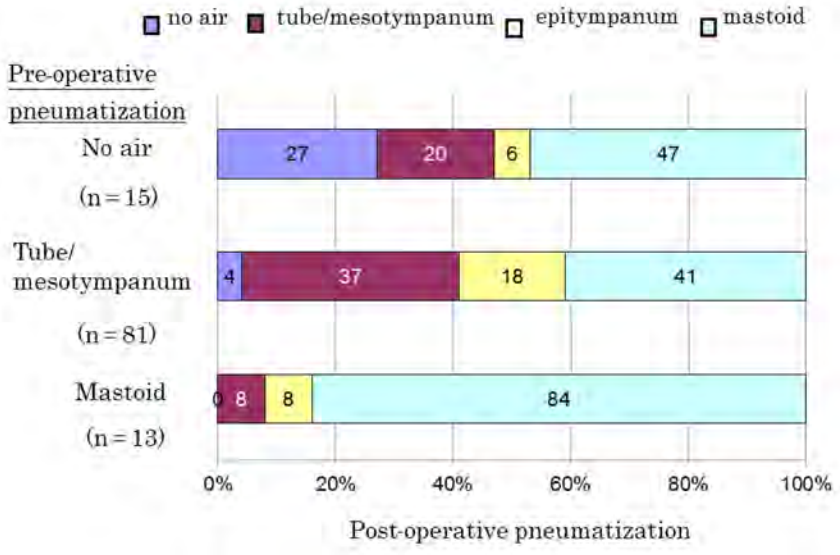

Fig. 2. Re-pneumatization according to preoperative pneumatization.

Extent of cholesteatoma and re-pneumatization (Fig. 3)

The extent of re-pneumatization varied widely, even within the same staging group. Stage II showed great improvement, however, there was no statistical difference among three groups. More extensive cholesteatoma did not always suggest poorer post-operative pneumatization.

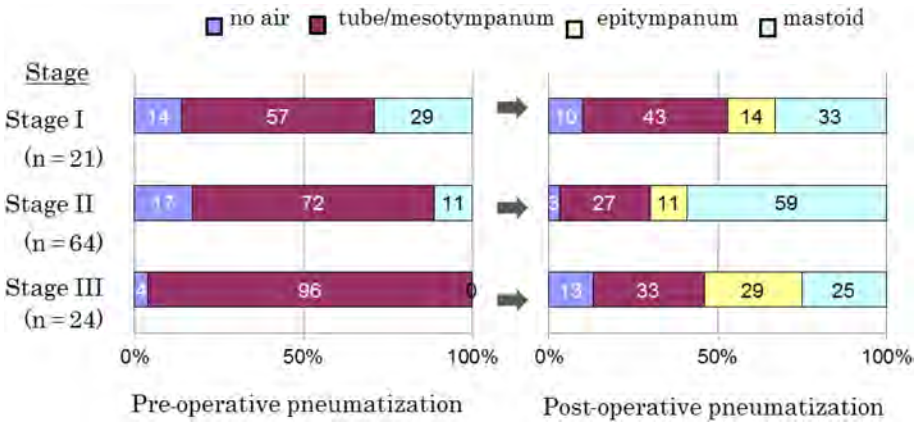

Fig. 3. Extent of cholesteatoma and re-pneumatization.

Extent of cholesteatoma and hearing, recurrence rate (Table 1)

We evaluated the hearing improvement, residual, and recurrence rate according to the stage. Hearing improvement of each stage was obtained in around 70% of the cases. Residual cholesteatoma occurred in about 10% of ears in every stage. The distribution of recurrence rate was: stage I: 4.7%; stage II: 10.9%; stage III: 20.8%. Although the incidence increased at higher stages, there was no statistical difference among those groups.

Table 1. Extent of cholesteatoma and hearing, recurrence rate.

	Hearing improvement	Residual	Recurrence
Stage I	14 (67%)	2 (9.5%)	1 (4.7%)
Stage II	47 (73%)	8 (12.5%)	7 (10.9%)
Stage III	16 (67%)	3 (12/5%)	5 (20.8%)

Conclusions

The extent of cholesteatoma with or without mastoid involvement did not affect the range of re-pneumatization after CWU. A large defect of scutum and posterior canal wall might make it more difficult to achieve mastoid re-pneumatization. A recurrent cholesteatoma resulting from a deep retraction pocket might increase in the same group. On pre-operative CT images, the extent of pneumatization and development of air cells can also be helpful in estimating re- pneumatization after CWU.

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MASTOID CAVITY OBLITERATION WITH PALVA FLAP AND BONE PATÉ

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Background

Chronic otitis media with cholesteatoma is a common disease in the field of otology. Canal-wall-down (CWD) tympanomastoidectomy is a well-established method in surgery to complete removal of cholesteatoma. It provides a good intra-operative exposure and an easy post-operative monitoring. A large cavity of mastoid in CWD tympanomastoidectomy technique could be troublesome. Problems are cosmetic, cleaning, draining ear, bathing, swimming, dizziness, and difficult ossicular reconstruction. The size of the surgical cavity can be diminished with obliteration to create a small cavity that is self-cleaning and easily maintained. Both autologous and synthetic materials have been used for obliteration, such as fat, cartilage, bone, periostomascular flaps and hydroxtapatitis. In this study we obliterated the mastoid cavity with bone paté and a Palva flap. Objective of this study is evaluation of outcome of mastoid cavity obliteration where bone paté and a Palva flap were used.

Material and methods

Between 2008 and 2011 in a prospective longitudinal study with a minimum follow up of 20 months, a series of 42 ears in 40 patients with chronic otitis media, due to a cholesteatoma, underwent surgery. Of these, 13 were revision surgery, the others were primary surgery. All patients underwent open-cavity tympanomastodectomy in which their mastoid cavity was obliterated with bone paté and a meatally-based musculoperiosteal flap (Palva flap).

Surgical technique

In general anesthesia and by post-auricular approach, a meatally-based musculoperiosteal flap (Palva flap) was prepared (Fig. 1), CWD tympanomastoidectomy (Fig. 2) was performed and bone dust was collected by means of a bone dust collector, from healthy cortical mastoid bone (Figs. 3 and 4). Then all of the pathologic material, such as cholesteatoma, was removed. After meatoplasty and placing of an underlay of fresh temporal fascia graft, the mastoid cavity was filled up by bone paté and a Palva flap (Fig. 5). Then a panrose was put in the cavity and the post-auricular incision was closed using Vicryl 3-0 subcutaneously. An antibiotic was given intravenously for one week.

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Edited by Haruo Takahashi

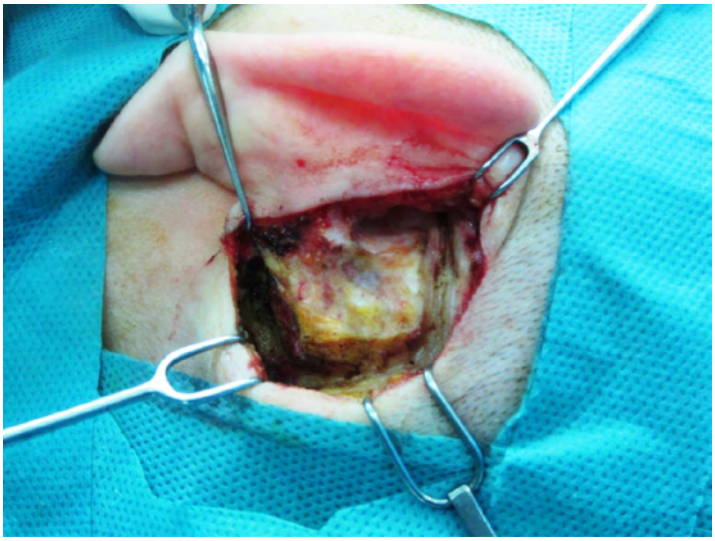

Fig. 1. Palva flap.

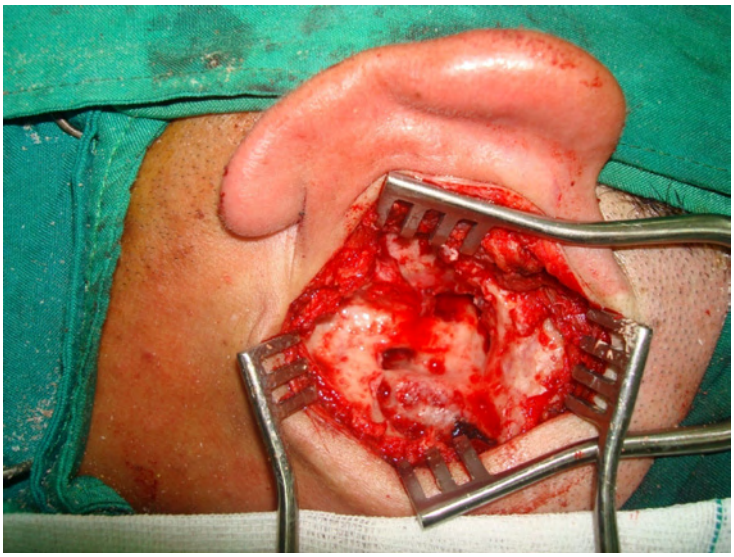

Fig. 2. CWD tympanomastoidectomy.

Fig. 3. Bone dust collector.

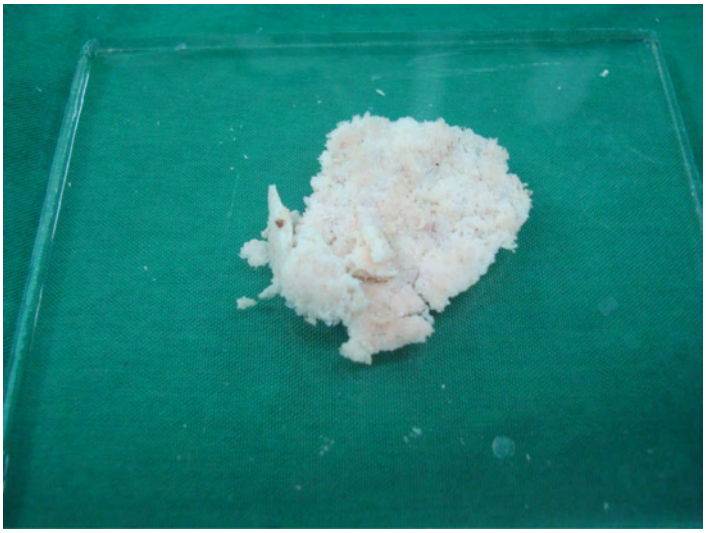

Fig. 4. Collected bone paté.

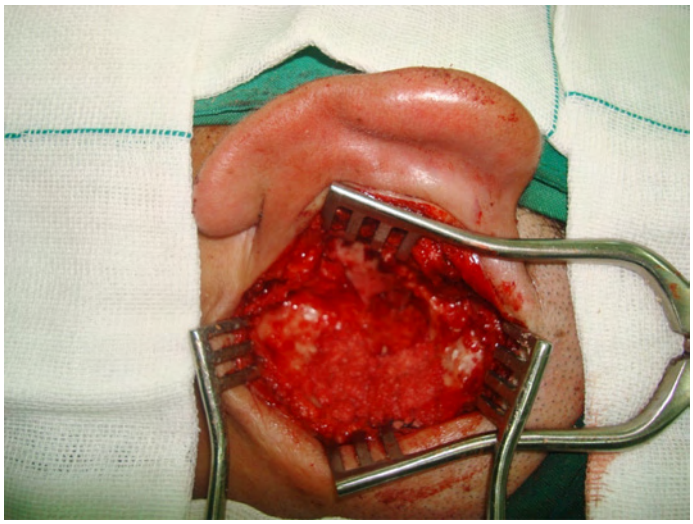

Fig. 5. Obliterated mastoid cavity with bone paté.

Results

In our study there was a female preponderance, 23 females versus 17 males participated. The mean age of the patients at the operation was 31 (± 11 SD) years (range from 9 to 58 years).

A total of 42 consecutive ears underwent open-cavity tympanomastoidectomy with mastoid obliteration; 35 (83%) had a very small dry healthy mastoid cavity, five ears (12%) had occasional otorrhea that was relatively easily managed by topical therapy, two ears (5%) had small granulation tissue that was treated with silver nitrate. In all patients ossicular reconstruction was needed. The most common ossicular lesion was incus lenticular erosion. All patients had second-stage surgery for detection of cholesteatoma recurrence and ossicular reconstruction.

Conclusion

In modern ear surgery, all mastoid cavities due to CWD tympanomastoidectomy were obliterated by various techniques and materials. In our experience, mastoid cavity obliteration using a Palva flap and bone paté is an effective method to manage patients with pre-existing mastoid cavities and also those not previously operated upon.

THE EFFECTIVENESS OF TYMPANIC ATTIC OBLITERATION TO PREVENT A POST-OPERATIVE RETRACTION POCKET

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Introduction

The basic surgical procedure at our department is canal-wall-down tympanoplasty with canal reconstruction using hard tissue such as auricular cartilage. In the observation of post-operative developments, there has been a growing number of cases in which obliteration with hard tissue of the tympanic attic is executed simultaneously. In order to study the effectiveness of this surgical procedure, a study was conducted on the post-operative retraction pocket in cases of chronic otitis media with cholesteatoma that have been examined and treated on an outpatient basis during the six-months period ending on April 25, 2011.

Subjects and results

The subjects were 112 cases of cholesteatoma outpatients of our department during the six-months period from October 25, 2010, to April 25, 2011. Of these, 38 underwent tympanic attic obliteration and 74 did not undergo the surgical procedure. Of the cases of tympanic attic obliteration procedure, eight (21.1%) developed a retraction pocket, and 30 (78.9%) did not. Of those who did not undergo tympanic attic obliteration, 30 (40.5%) developed a retraction pocket, while 44 (59.5%) did not. Of the total of 112, 38 (33.9%) developed a retraction pocket. The analysis of the 38 cases of retraction pocket by area of retraction revealed that the postero-superior quadrant (PSQ) was the area for the largest number (three or 37.5%) in the obliteration group, and the tympanic attic was the area for the largest number (15 or 50.0%) in the non-obliteration group. Of the total of 112, the number of cases of re-operation due to post-operative recurrence was 27. In these cases, the retraction pocket was examined post-operatively whether obliteration was done or not. In the obliteration group, a retraction pocket was found in three out of 14 (21.4%). In the non-obliteration group, the ratio was eight out of 13 (61.5%)

Discussion and conclusion

Tympanic attic obliteration was found to reduce a post-operative retraction pocket in the attic naturally. However, the retraction pocket increased for PSQ. It is believed that it is necessary to reinforce the PSQ area with thinly-sliced cartilage in the future. For the re-operation cases due to recurrence, it was found that obliteration of the tympanic attic clearly reduces a post-operative retraction pocket.

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Edited by Haruo Takahashi

RESULTS OF CANAL WALL DOWN TYMPANOPLASTY WITH SOFT-WALL RECONSTRUCTION FOR CHOLESTEATOMA

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Introduction

In cholesteatoma surgery, we use canal-wall-down tympanoplasty with soft-wall reconstruction for almost all cases. This method has the advantages of both canal-wall-up procedures, such as early wound healing, and canal-wall-down procedures, such as good radicality. There is another advantage, which is the relative ease with which this procedure can be performed. Furthermore, an even greater advantage of this procedure is that the soft posterior ear auditory canal skin changes its shape and position according to the residual capacity of the mastoid function of each individual patient after mastoid surgery, and this results in long-term safety and stability of the post-operative condition.

In the present study, we assessed incidence of the residual and recurrent cholesteatomas, post-operative conditions of the soft posterior meatal wall, post-operative aeration of middle ear cavity and hearing outcome.

Subject

The study subjects comprised 104 ears in 99 patients (56 males and 43 females) with acquired middle-ear cholesteatoma who had undergone canal-wall-down tympanoplasty with soft-wall reconstruction from 2004 to 2010.

The patients ranged in age from 15 to 84 years (mean 53 years). All the patients were followed for more than one year after surgery (ranging one year to seven years, mean three years and eight months). In 90 ears, post-operative aeration could be evaluated on CT scans. In 98 ears, post-operative hearing was assessed.

The type of cholesteatoma was attic cholesteatoma in 60 ears, sinus cholesteatoma in 24 ears, recurrent cholesteatoma in 17 ears and residual cholesteatoma in three ears.

Surgical technique

In all patients, canal-wall down and mastoidectomy were carried out, preserving the intact parts of the eardrum and external auditory canal (EAC) wall skin as much as possible. The defect on the eardrum or the posterior EAC wall skin after removal of cholesteatoma was covered (reconstructed) by a piece of temporalis fascia (soft-wall reconstruction). This method results in post-operative separation of the mastoid cavity from the EAC, making it different from the conventional CWD technique which opens the mastoid cavity to the EAC. No hard tissues or materials (*e.g.*, cortical bone or cartilage) were used to reinforce or reconstruct the posterior EAC wall in any of the ears in this study.

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Edited by Haruo Takahashi

At the same time, tympanoplasty was done in all of these ears except six. The ossicular chain was preserved in six ears (type I). In 92 ears, various kinds of columellas (incus, auricular cartilage, or artificial columella) were used for the reconstruction of the conductive system; on the stapes head in 64 ears (modified type III) and on the stapes footplate in 28 ears (modified type IV).

Evaluation

All data obtained from these patients were analyzed retrospectively:

- Incidence of residual and recurrent cholesteatomas, crust accumulation.
- Post-operative conditions of the soft posterior meatal wall. For analysis, the post-operative conditions of the soft posterior meatal wall were divided into four types according to a retracted extent: normal, slightly retracted, moderately retracted and like a radical mastoid cavity.
- Post-operative aeration in the middle ear. Computed tomographic scans (CT) performed more than one year after surgery were used to assess aeration. For analysis, the middle-ear space was divided into five segments: Eustachian tube (ET), hypo-mesotympanum, epitympanum, antrum and mastoid cavity.
- Hearing outcomes. Audiometric data were obtained at least 1 year after surgery by calculating the pure-tone average of 0.5, 1, 2kHz. Postoperative air-bone (A-B) gaps were calculated in each case.

Results

Incidence of residual and recurrent cholesteatomas, crust accumulation

Of the 104 ears operated on, seven ears (6.7%) had residual cholesteatomas and three ears (2.9%) had recurrent cholesteatomas, while crust accumulation was observed in five ears (4.8%).

Four residual cholesteatomas and all three recurrent cholesteatomas could be removed at the outpatient clinic, while three out of seven residual cholesteatomas were re-operated. Crust accumulation in five ears need to be removed at the outpatient clinic once every several months.

Post-operative conditions of the soft posterior meatal wall

The soft posterior EAC wall apparently retracted like a radical mastoid cavity in 45 of 104 ears (43.2%), moderately retracted in 48 ears (46.2%), slightly retracted in eight ears (7.7%), but showed no retraction in the remaining three ears (2.9%) (Fig. 1). In none of the ears, a defect or perforation occurred in the soft posterior EAC wall because of infection or necrosis of the graft after surgery.

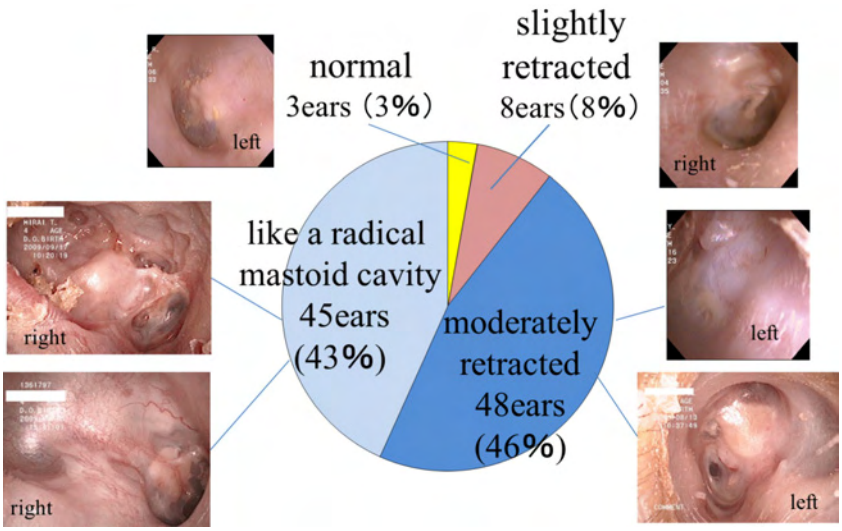

Fig. 1. Post-operative conditions of the soft posterior meatal wall (n = 104).

Post-operative aeration in the middle ear

In 90 ears, post-operative aeration could be evaluated on CT scans. Re-aeration was noted in the mastoid cavity in 30%. The rate of re-aeration was 53% in the epitympanum and 89% in the tympanic cavity. Post-operative conditions of the soft posterior meatal wall were not always correlated with these post-operative aerations in the middle ear.

Hearing outcomes

In 98 ears, post-operative hearing was assessed. At the latest hearing test, the mean air-bone gap was < 15 dB in 63 of 98 ears (64.3%) and < 20 dB in 72 of 98 ears (73.5%). As for the mode of tympanoplasty, numbers of ears showing less than 15 dB of ABG were 48 out of 64 ears (75.0%) with type-III or modified type-III tympanoplasty (interposition of an ossicle or an autogenous auricular cartilage columella between the stapes head and the eardrum or manubrium of the malleus), nine out of 28 ears (32.1%) with modified type-IV tympanoplasty (interposition of an ossicle or an artificial columella between the stapes footplate and the eardrum), and five out of six ears (83.3%) with type I.

Discussion

As advantages of the soft-wall reconstruction method, Smith *et al*.[1] stated early post-operative cure of the wound and its technical ease and little addition to operating time.

Hosoi *et al*.[2,3] have pointed out the low incidence of retraction pocket and recurrent cholesteatoma among ears after surgery by this method. In this study, the incidence of post-operative recurrent cholesteatoma was as low as 2.9%. This incidence is considerably lower than those after surgery by ICWT reported previously.[4,5] In three ears, a retraction pocket occurred in the soft posterior EAC wall to be a cholesteatoma in this study. The reason why this retraction occurred in such an extremely narrow portion of the soft wall may be partial vulnerability of the posterior EAC wall or ear drum. This suggests that even the soft-wall reconstruction method is not always perfect for the prevention of post-operative recurrent cholesteatoma.

The incidence of residual cholesteatoma was 6.7 %. This results indicate that the soft-wall reconstruction method may have at least an advantage similar to the canal-wall-down and open method in terms of the low incidence of the residual cholesteatoma.

In the present study, moderate retraction or large retraction like a radical mastoid cavity of post-operative soft posterior meatal wall was observed in more ears than those of previous studies.[2,3,6] Takahashi *et al*.[6] reported that the soft-wall reconstruction method let the mastoid cavity decide by itself which way to go (retract or not) after surgery according to its residual ventilatory function (gas exchange function). The reason why we had more retracted soft posterior EAC wall in most cases may be that we could not preserve enough intact mastoid mucosa during the surgery.

In this study, higher incidences of postoperative aeration were observed in the ET and hypo-mesotympanum followed by the epitympanum. This result suggests that post-operative recovery of aeration may begin in the ET and advance to the mastoid cavity through the hypo-mesotympanum, epitympanum, and antrum. The ventilation function of the ET is considered crucial for the post-operative recovery of aeration in the middle ear. Further studies of post-operative middle-ear aeration in relation to pre-operative ET ventilation function are needed.

Some studies have discussed which procedure, CWD and open (no aerated mastoid) or CWU, is beneficial with regard to hearing outcome.[7-13] Tos and Lau[7] and Stankovic[8] reported better outcomes with CWU tympanoplasty, whereas several studies indicated no differences in terms of post-operative hearing outcome between these two procedures.[9-13] There are no differences between these two methods and CWD tympanoplasty with SWR, as presented here, in terms of post-operative hearing outcomes.

Conclusion

Canal-wall-down tympanoplasty with the soft-wall reconstruction method is found to be a versatile method for cholesteatoma surgery compared with the canal-wall-down method because of earlier wound healing, whilst it preserves the advantages of the latter method, such as low incidence of residual and recurrent cholesteatomas.

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LONG-TERM RESULTS AND PROGNOSTIC FACTOR IN SINGLE-STAGE TYMPANOPLASTY

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Introduction

Various operative procedures have been developed for the surgical treatment of chronic otitis media with or without cholesteatoma. The principal objective in chronic otitis media with cholesteatoma surgery is the complete eradication of the disease to produce a safe ear and the improvement of hearing. Over the past years, one goal of the complete eradication of the disease has been consistently achieved using the canal-wall-down procedure.[1] However, this technique has several problems, such as difficulty with fitting a hearing aid and a higher rate of infection. For this reason, the canal-wall-up procedure is more commonly used.[2,3] In reality, the incidence of cholesteatoma recurrence is higher with canal-wall-up procedure than with canal-wall-down procedure. Therefore, staged operation is in heavy usage, and then patients with cholesteatoma must go through operations a number of times.[4,5]

We use one-stage tympanoplasty with mastoid obliteration, a modified canal-down procedure, for the treatment of otitis media with aeration trouble in the mastoid, while we use a tympanoplasty by transcanal approach for chronic otitis media without aeration trouble in the mastoid. The operation is performed as day surgery.

Otologic surgeons have used a variety of materials for reconstruction of the ossicular chain.[6-8] In Japan, autograft replacement is still the most commonly used; biocompatible synthetic materials are very rarely used. However, we have used ceramic prostheses (Type P and Type T) and reported excellent results, including good hearing improvement and low extrusion rates.[9]

Here we describe the effect of one-staged tympanoplasty with mastoid obliteration and tympanoplasty by transcanal approach using a ceramic prosthesis.

Materials and methods

A retrospective chart review was performed on 516 patients undergoing ossicular chain reconstruction between June 2001 and December 2005. The procedure included one-stage tympanoplasty with mastoid obliteration and tympanoplasty by transcanal approach. Demographic information about these patients is summarized in Table 1.

Table 1. Demografic information

	Modified CWD	TC approach
Patients	100	416
Mean age±SD	44.3±15.3	55.5±14.5
Sex (M:F)	42:58	187:229
Affected side (R:L)	54:46	203:213

CWD; Canal Wall Down, TC; Transcanal

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Cholesteatoma and Ear Surgery – An Update, pp. 455-458
Edited by Haruo Takahashi

Air-bone gap (ABG) data were obtained by comparing the most recent bone- and air-conduction results. ABG was calculated by the use of four-frequency pure tone averages (500, 1000, 2000, and 3000 Hz) of air and bone conduction from the same test intervals. 'Successful' reconstruction was defined as a post-operative PTA-ABG $\leq$ 20 dB.

Surgical procedure 1

We use one-stage tympanoplasty with mastoid obliteration, a modified canal-wall-down procedure for chronic otitis media with aeration trouble in the mastoid.[9] In this procedure, the mastoid cavity is opened by resecting bone of the posterior and superior walls of the external ear canal, after which the antrum and epitympanum are opened. However, the intact canal skin is maintained. After cleaning of the lesions, changes in the ossicular chain, particularly at the long process of the incus and the superstructure of the stapes, are carefully observed. If cholesteatoma includes the whole incus, the incus is removed by separating it from the lesions around the stapes. The tendon of the tensor tympani muscle is also cut. After the opening of the Eustachian tube to the tympanic cavity is confirmed, the eardrum is reconstructed by closing the perforation underlying it with the fascia.

As shown Figure 1, the ossicular chain is reconstructed using a ceramic ossicular prosthesis (P-type or T-type). The ceramic ossicular prosthesis is used after trimming the shaft to the appropriate size. We perform a partial ossicular chain reconstruction using the P-type prosthesis when the superstructure of the stapes can be utilized, while we perform a total chain reconstruction using the T-type prosthesis when the superstructure of the stapes can not be used.

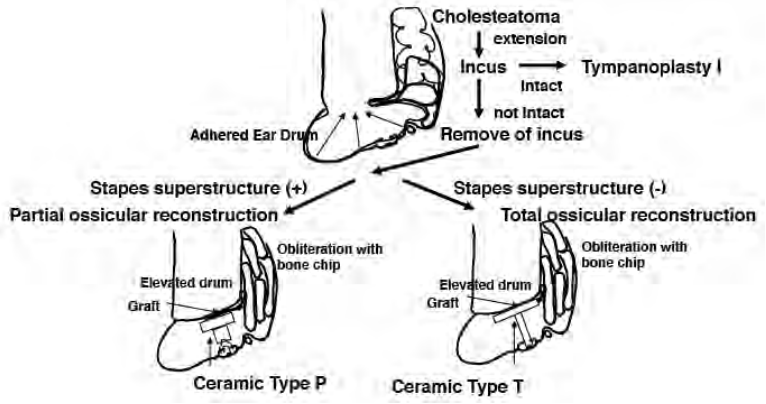

Fig. 1. One-stage tympanoplasty with mastoid obliteration, a modified canal-wall-down procedure. After the ossicular chain was reconstructed using the ceramic ossicular prosthesis (P-type/T-type), the reconstruction of posterior and superior walls of the external ear canal and the obliteration of the mastoid cavity was performed with the bone plate from the mastoid cortical bone.

After reconstruction of the ossicular chain, reconstruction of posterior and superior walls of the external ear canal and obliteration of the mastoid cavity was performed with the bone plate from the mastoid cortical bone. In this surgery, we consider preserving the skin of the external canal and keeping the position of tympanic membrane intact except for the perforation area of the tympanic membrane very important.

Surgical procedure 2

For this procedure we perform a tympanoplasty by transcanal approach for chronic otitis media without aeration trouble in mastoid. An incision is made with a lancet at 12 and 7 o'clock, and the meatal skin is elevated to the level of the fibrous annulus. The bone edge of the posterior canal wall is removed little by little with a chisel until the pyramidal eminence and the stapes tendon are clearly visible. After the annulus is raised, the middle ear is entered. We perform a partial ossicular chain reconstruction using the P-type ceramic prosthesis when the superstructure of the stapes can be used, while we perform a total chain reconstruction using the T-type ceramic prosthesis when the superstructure of the stapes can not be used.

Results

Ninety-three of 100 cases (93%) represented primary surgery in modified canal-wall-down, while 319 of 416 cases (77%) were performed as an unplanned revision surgery in tympanoplasty by transcanal approach (Table 2).

Table 2. Primary Surgery versus Revision Surgery

	Modified CWD		TC approach	
	Ceramic P	Ceramic T	Ceramic P	Ceramic T
Primary Surgery	68	25	89	8
Revision Surgery	4	3	208	111
Total	72	28	297	119

CWD; Canal Wall Down, TC; Transcanal

In modified canal-wall-down, mean ABG for partial ossicular replacement prostheses was 29.5 dB (SD ± 15.3) post-operatively. Mean ABG for total ossicular replacement prostheses was 38.1 dB (SD ± 12.8). Mean hearing gain in the P-type ceramic group was 10.3 dB (SD ± 7.41). Mean hearing gain for the group of patients following placement of T-type ceramic was 9.3 dB (SD ± 6.83). No patient had a significant acute worsening of bone conduction post-operatively. In tympanoplasty by transcanal approach, post-operative mean ABG for partial ossicular replacement prostheses was 40.4 dB (SD ± 11.9). Mean ABG for total ossicular replacement prostheses was 42.7 dB (SD ± 10.2). Mean hearing gain in the P-type ceramic was 7.8 dB (SD ± 12.8). Mean hearing gain for patients following placement of T-type ceramic was 6.2 dB (SD ± 9.54). No patient had a significant acute worsening of bone conduction post-operatively.

In the modified canal-wall-down group using P-type ceramic, 8.3% of the patients had a post-operative ABG of less than 10 dB. In the tympanoplasty by transcanal approach using ceramic P-type 25% of the patients had a post-operative ABG between 11-20 dB, no patients had a post-operative ABG of less than 10 dB. Ten percent of the patients had a post-operative ABG between 11-20 dB. In the modified canal-wall-down group using T-type ceramic, 17% of the patients had a post-operative ABG of less than 10 dB, 50% of the patients had a post-operative ABG between 11-20 dB, In tympanoplasty by transcanal approach, no patients had a post-operative ABG of less than 10 dB, 20% patients had a post-operative ABG of 11-20 dB. We defined success as post-operative air-bone gap of 20 dB or less. A modified canal-wall-down group using P-type and T-type ceramic prosthesis had a 33.3% and 67% success rate (Fig. 2). A tympanoplasty by transcanal approach using P-type and T-type ceramic prostheses had a 10% and 20% success rate.

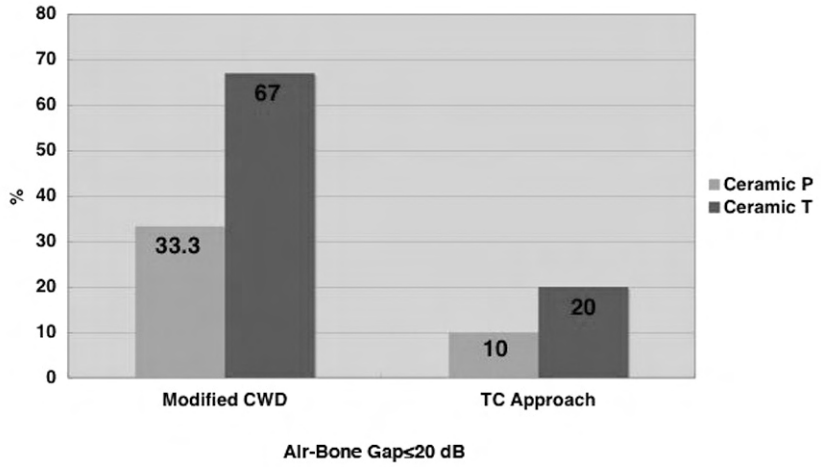

Fig. 2. Hearing results by operation performed. A modified canal-wall-down group with P-type ceramic prostheses and the group with T-type ceramic prostheses had a 33.3% and 67% success rate, resp. A tympanoplasty by transcanal approach using P-type and T-type ceramic prostheses had a 10% and 20% success rate, resp.

Discussion

We have used one-stage tympanoplasty with mastoid obliteration as a modified canal-wall-down procedure for chronic otitis media with aeration trouble in the mastoid.[9] The major advantage of the canal-wall-down procedure is that the surgical area is visible through a microscope. We maintain the intact external canal skin and fill the cavity formed by the removal of the bone of the external ear canal with the patient's own bone fragments obtained from the temporal bone, even though we remove the external ear canal bones and the mastoid cavity. After that, our procedure makes it easy to remove cholesteatoma and granulation, reduces the risk of infection, and prevents perforation of the tympanic membrane. Our results show that air-bone gaps could be reduced to 20 dB or less in 33.3% of the patients with P-type ceramic prosthesis and 67% of the patients with T-type ceramic prosthesis. In this study, we confirmed that the total ossicular reconstruction using a T-type ceramic prosthesis was satisfactory for one-stage tympanoplasty with mastoid obliteration. In fact, we broke down the disadvantage of the canal-wall-down procedure.

We also performed tympanoplasty by transcanal approach for chronic otitis media without aeration trouble in the mastoid. We reported a success rate of 10% for the P-type ceramic prosthesis and 20% for the T-type ceramic prosthesis in tympanoplasty by transcanal approach. In fact, the patients requiring tympanoplasty by transcanal approach had significantly worse results than those with one-stage tympanoplasty with mastoid obliteration. In this study, we considered that patients undergoing a tympanoplasty by transcanal approach had more severe disease including tympanosclerosis than those requiring one-stage tympanoplasty with mastoid obliteration. In addition, our results showed that the hearing results tended to be worse in unplanned revision surgery. Therefore, it is unfair to draw the conclusion that a modified canal-wall-down procedure has better hearing results than the transcanal approach. Our observation is that unsuccessful cases might not have a secure attachment to the stapes footplate because of not allowing tissue integrations. Therefore, we hope that a variety of materials for reconstruction of the ossicular chain will be used in the near future in Japan as well.

We confirmed there was a trend for a better air-bone gap when the middle ear was well aerated and the Eustachian tube functioned well. Conversely, insufficient middle-ear spaces due to post-operative Eustachian tube dysfunction was observed in several unsuccessful cases examined by post-operative Cone Beam CT scanning. We considered that the condition of the pre-operative aeration of the middle ear and the Eustachian-tube function would become an important indicator of post-operative results.

Conclusion

In conclusion, we confirmed the decade-long efficacy in the single-stage tympanoplasty. Especially the total ossicular reconstruction using ceramic T-type was satisfactory for one-stage tympanoplasty with mastoid obliteration. It is important to apply a single-stage tympanoplasty with mastoid obliteration and a tympanoplasty by a transcanal approach for the treatment of chronic otitis media selectively. We considered that the pre-operative important prognostic factors in the single-stage tympanoplasty would be the following two points: the condition of the pre-operative aeration of the middle ear and the Eustachian-tube function.

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EFFECTIVENESS OF CARTILAGE TYMPANOPLASTY AFTER EROSION OF THE INCUS IN CHILDREN WITH CHOLESTEATOMA

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Introduction

Cholesteatoma affects three in 100,000 children worldwide and can result in significant morbidity and mortality.[1] This disease can be locally destructive and is one of the most common causes of permanent conductive hearing loss in children secondary to incus erosion and/or removal in cholesteatoma surgery. Therefore, one of the goals of cholesteatoma surgery is the restoration of conductive hearing by reconstruction of the ossicular chain.

Hearing reconstruction is typically achieved with one or more of many reconstruction techniques which include myringostapediopexy (MS), cartilage myringostapediopexy (CMS), incus interposition (II), or synthetic material (titanium, ceramic, cement). Unfortunately, the literature offers little information regarding which choice of reconstruction is the best for children.

In this paper, we evaluate and compare hearing outcomes in children with erosion or absence of the incus from cholesteatoma and reconstructed with a myringostapediopexy, cartilage myringostapediopexy, or incus interposition.

Materials and methods

A prospective cohort analysis of all consecutive children undergoing cholesteatoma surgery between 2001 and 2012 at Sick Children's Hospital in Toronto, Ontario, Canada was performed. The procedures were performed by two paediatric otologic surgeons. The study was approved by the research ethics board at the University of Toronto. To be included in the study, an intact-canal-wall procedure was performed and an eroded incus with an intact stapes and handle of malleus was identified intra-operatively. All revision cases and patients with fixed stapes were excluded from the study.

Hearing reconstruction was achieved by a myringostapediopexy, cartilage myringostapediopexy, or incus interposition graft (Fig. 1). The choice of the reconstruction was made at the discretion of the surgeon. Tragal cartilage was used for cartilage myringostapediopexy and temporalis fascia was used for reconstruction of the tympanic membrane.

Four-frequency pure tone average (4PTA) air conduction audiograms (0.5, 1, 2, & 4 kHz) were completed on all patients pre-operatively and one year post-operatively. Serviceably normal hearing was defined as a 4PTA of less than 30dB HL.

Comparisons of means were done using unpaired Student's t-test for variables with Gaussian distribution, and Mann-Whitney for non-Gaussian variables. Comparison of binomial variables (normal versus abnormal hearing) was done using $\chi 2$ analysis. All statistical analysis was performed using SPSS (statistical package version 14.0; SPSS, Inc., Chicago, IL).

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Edited by Haruo Takahashi

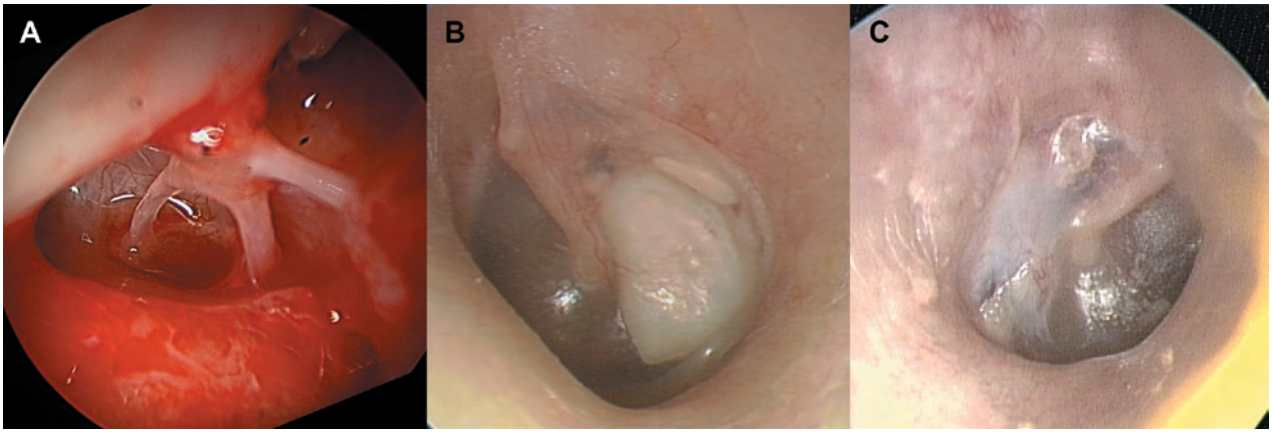

Fig. 1. A: Intra-operative image of a cartilage myringostapediopexy reconstruction. B: Post-operative image (18 months) of a cartilage myringostapediopexy reconstruction. C: Post-operative image of an incus interposition reconstruction.

Results

A total of 76 patients were included in the study: 14 patients with MS, 19 with CMS, and 43 with II (Table 1). No significant differences were found between groups with respect to mean age, sex, percentage of incus erosion, or Mills stage. Pre-operative 4PTA in the MS group (29.5dB) was significantly lower than the II group (36.0dB) ($p < 0.05$). No difference was found in the pre-op 4PTA between the CMS and II groups.

Table 1. Demographics

	MS	CMS	II	P
N	14	19	43	
Age (yrs)	11.3	12.0	11.0	>0.05
Sex (male)	50%	68%	62%	>0.05
Incus eroded	100%	95%	89%	>0.05
Incus removed	100%	100%	100%	>0.05
Mills stage	2.9	2.3	2.6	>0.05
Pre-op 4PTA	29.5 dB *	35.2 dB	36.0 dB *	0.04*

Demograpic data for study groups. The only significant difference was found in the pre-op 4PTA between the MS group and the II group.

Post-operatively, all groups had a significant change in their one-year 4PTA (Fig. 2). The post-operative 4PTA significantly improved in the CMS group (34dB to 23dB; $p < 0.05$) and II group (35dB to 26dB; $p < 0.05$). Interestingly, post-operative hearing significantly worsened in the MS group (26dB to 36dB; $p < 0.05$). The post-operative 4PTA in the MS group was significantly worse than both the CMS and II groups. No difference was found between the post-operative 4PTA in the CMS and II groups.

The percentage of patients who obtained post-operative normal serviceable hearing (4PTA of less than 30dB) was 21.4% with MS, 78.9% with CMS, and 60.5% with II (Fig. 3). The MS group was significantly worse than both the the CMS and II groups. There was no significant difference between the CMS and II groups ($p = 0.2$).

Discussion

The challenges related to performing ossiculoplasty in children have been attributed to Eustachian-tube dysfunction, higher rates of recurrent infection, and the associated difficulty controlling middle-ear disease. These factors potentially result in higher rates of extrusion or failure. Unfortunately, the literature offers the surgeon very little in helping choose the appropriate method of reconstructing hearing following cholesteatoma surgery in children. Most studies look at the adult population and few studies in children compare different ossicular reconstructive methods.

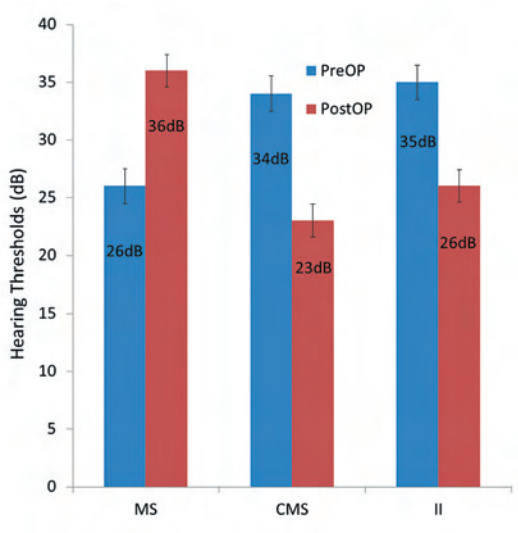

Fig. 2. Pre and post-operative 4PTA in MS, CMS, and II groups. All groups had significantly different hearing after surgery.

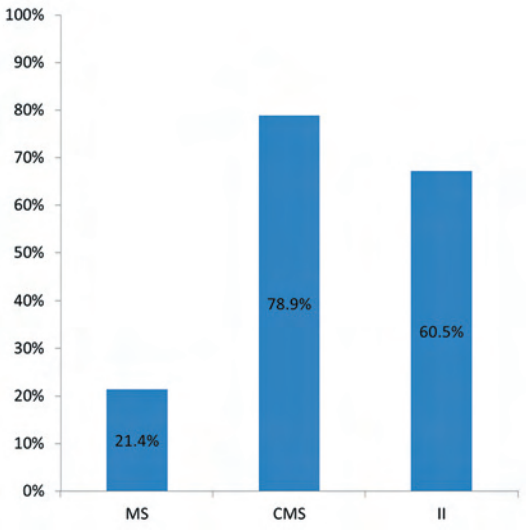

Fig. 3. Percentage of patients who achieved serviceably normal hearing (air conduction 4PTA < 30dB). He MS group percentage was significantly lower than both the CMS and II groups.

Silverstein *et al*.[2] evaluated 192 adults and 18 children looking at notched incus homografts and Plasti-Pore prostheses for reconstruction following surgery for chronic ear disease. Although they did find that both methods achieved good post-operative hearing results, the inadequate sample size in the children group did not allow significant conclusions to be drawn. It was noted that the Plasti-Pore extrusion rate was 17% in children and was advised against.

Iurato *et al*.[3] compared myringostapediopexy to incus/malleus interposition in adults and children and found that myringostapediopexy achieved better air-bone gap (ABG) closure within 10db (81%) when compared to ossiculoplasty (55%). The authors did not stratify the results based on adults and children and, therefore, it is difficult to extrapolate these results to children alone.

Nevoux *et al*.[4] looked at cartilage myringostapediopexy in 268 children following chronic ear surgery and found that 62.2% of patients achieved ABG closure within 20dB. Unfortunately, without a comparison group, it is unclear how this means of reconstruction compares to others.

One study by Daniels *et al*.[5] did compare ossiculoplasty techniques in children: porous polyethylene partial ossicular replacement prosthesis (POPs) and Schuring ossicle cup prosthesis (SOCs). They did show that ABG closure within 10dB was achieved in 77% of POPs and 61% of SOCs at one year. They were not significantly different. Only one extrusion occurred in these groups. Unfortunately, no autologous grafts were evaluated and/or compared in this study.

Although ossicular reconstruction in children remains a secondary goal after establishing a safe, dry, and stable ear, post-operative hearing remains a significant concern to patients, parents, and surgeons. Given the potential for extrusion and failure in children, allografts remain a viable option for hearing reconstruction following cholesteatoma surgery, particularly when the stapes is intact. Our study found that in children with cholesteatoma and incus erosion, cartilage myringostapediopexy and incus interposition did improve post-operative hearing. Myringostapediopexy actually worsened hearing post-operatively.

Conclusions

This evaluation of children with intact stapes following cholesteatoma surgery demonstrated that cartilage myringostapediopexy and incus interposition can improve post-operative hearing. Myringostapediopexy may worsen hearing so should be avoided. As cartilage myringostapediopexy may help prevent recurrent pars tensa retraction following surgery, this has become our preferred method of reconstruction for incus erosion in children with cholesteatoma.

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REPAIR OF SPONTANEOUS CEREBROSPINAL FLUID OTORRHEA FROM DEFECT OF MIDDLE CRANIAL FOSSA

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Introduction

Cerebrospinal fluid (CSF) otorrhea most commonly occurs as a result of trauma, surgery or congenital anomalies. Although spontaneous CSF otorrhea is a rare condition, it is a life-threatening disease because of the high incidence of subsequent meningitis. Immediate and definitive repair of CSF leak is important to prevent this potential complication. Surgical approaches include a transmastoid approach, middle fossa craniotomy, or a combined approach. We have recently experienced the case of a patient with spontaneous CSF otorrhea occurring through a bony defect in the anterior wall and roof of epitympanum. The defect was successfully repaired using bone chips with fascia via the transmastoid approach.

Case

A 49-year-old woman was referred to our hospital because of right pulsatile otorrhea after myringotomy. Initially, she had experienced right ear fullness and hearing disturbance for two weeks. When a local otolaryngologist examined her ear, there was visible fluid through the tympanic membrane. Thus, she was diagnosed as right serous otitis media and treated with antibiotics. However, after the two-week treatment, the fluid in the tympanic cavity was still present. Myringotomy was tried, but profuse pulsatile clear discharge from the myringotomy site was encountered.

Examination of her right ear revealed pulsatile clear discharge through the myringotomy incision site. The pure-tone audiogram showed right conductive hearing loss of 40dB (air-bone gap 32dB). She had no specific past medical history and family history. High-resolution computed tomography (HRCT) scan and magnetic resonance imaging (MRI) of the temporal bone were performed for further evaluation of the otorrhea. On the HRCT scan, anteromedially a large bony defect from the anterior wall to the roof of the right epitympanum was identified (Fig. 1). The right middle-ear cavity and mastoid cavity were filled with soft tissue density. A T2-weighted MRI image demonstrated high signal intensity in the right middle ear and mastoid cavity, which is consistent with CSF (Fig. 2).

Repair of CSF leak through a transmastoid approach was planned. First, after general anesthesia, lumbar drainage was done. Temporalis muscle fascia was harvested. Several pieces of bone chips were taken from the mastoid cortex. A cortical mastoidectomy was performed. On entering the antrum, a profuse clear discharge from the epitympanum was noted. To further evaluate the tegmen tympani, the incus and head of malleus were removed. There was a bony defect of approximately 8 mm from the anterior roof of the epitympanum extending to the anterior wall. Also, there was no identifiable healthy dura or brain tissue herniation at the CSF leak site.

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Cholesteatoma and Ear Surgery – An Update, pp. 463-466
Edited by Haruo Takahashi

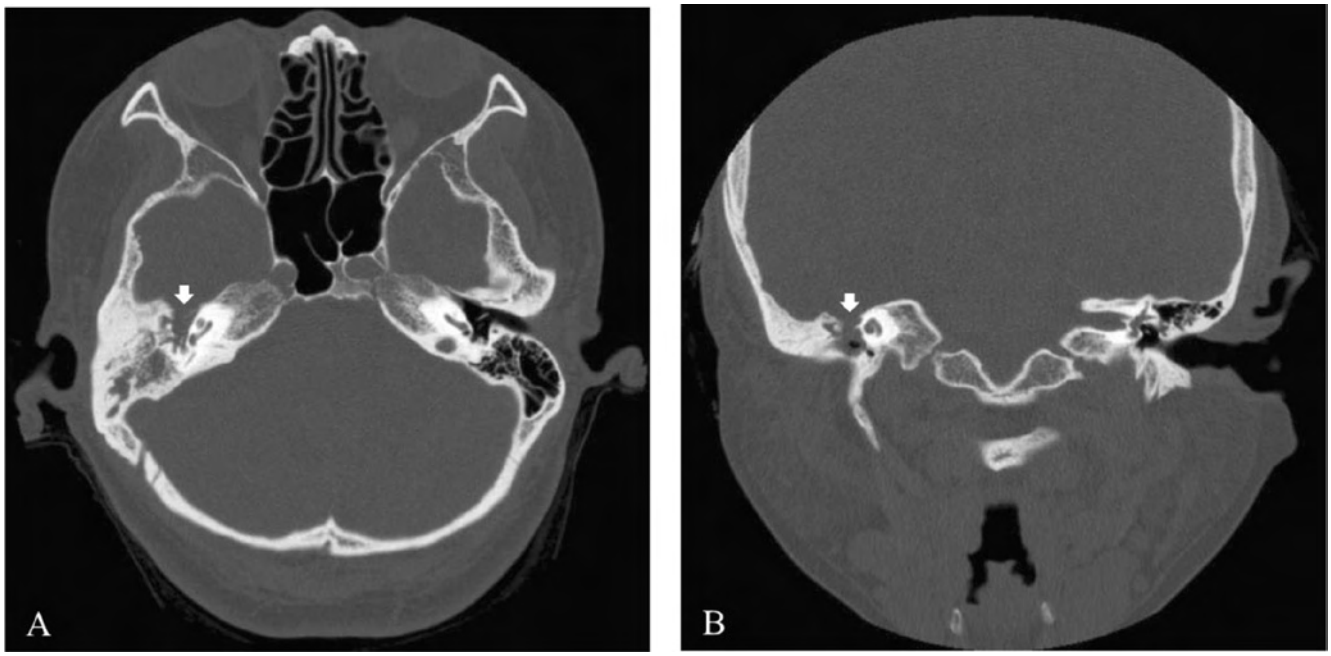

Fig. 1. Pre-operative HRCT scan of the temporal bone. The axial image (A) demonstrates a large bony defect of the right anterior wall of the epitympanum (arrow). The coronal image (B) shows a bony defect of the right middle cranial fossa. Also, the tympanic and mastoid cavities are filled with soft-tissue density.

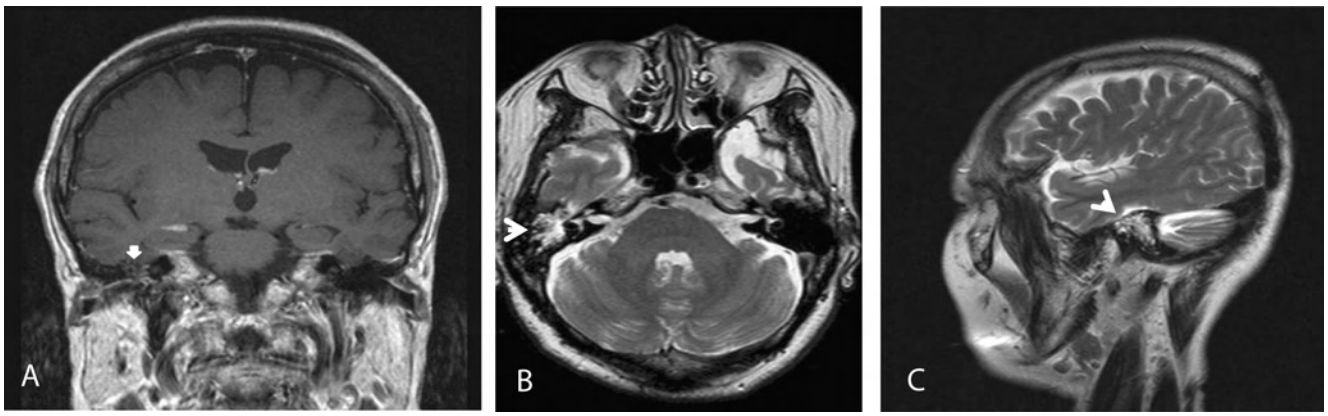

Fig. 2. Pre-operative brain MRI. Gd-enhanced T1-weighted coronal image (A) shows no evidence of brain herniation in the epitympanum and isosignal intensity (arrow). Unenhanced T2-weighted axial (B) and sagittal (C) images demonstrate high-signal intensity in the right middle ear and mastoid cavity (arrow head).

To repair the defect, fistly, prepared temporalis fascia was inserted in the bony defect. Then a large piece of bone chip was inserted into the defect to cover the fascia. The bony defect, however, was still visible because of its three-dimensional structure. More bone chips were put into the defect, until there was no evidence of the CSF leak left. Surgical glue was applied around the bone chips and gelfoam was inserted in the cavity.

Postoperatively, the CT scan showed a well-sealed defect, but also a pneumocephalus in the right temporal lobe (Fig. 3). But the patient did not complain of a headache or any neurologic deficits. Lumbar drainage was maintained for six days. Two weeks later, the pneumocephalus disappeared and the patient was discharged from hospital. One year later, the CT scan showed a well-healed defect (Fig. 4). There has been no evidence of recurrent CSF leak up to date.

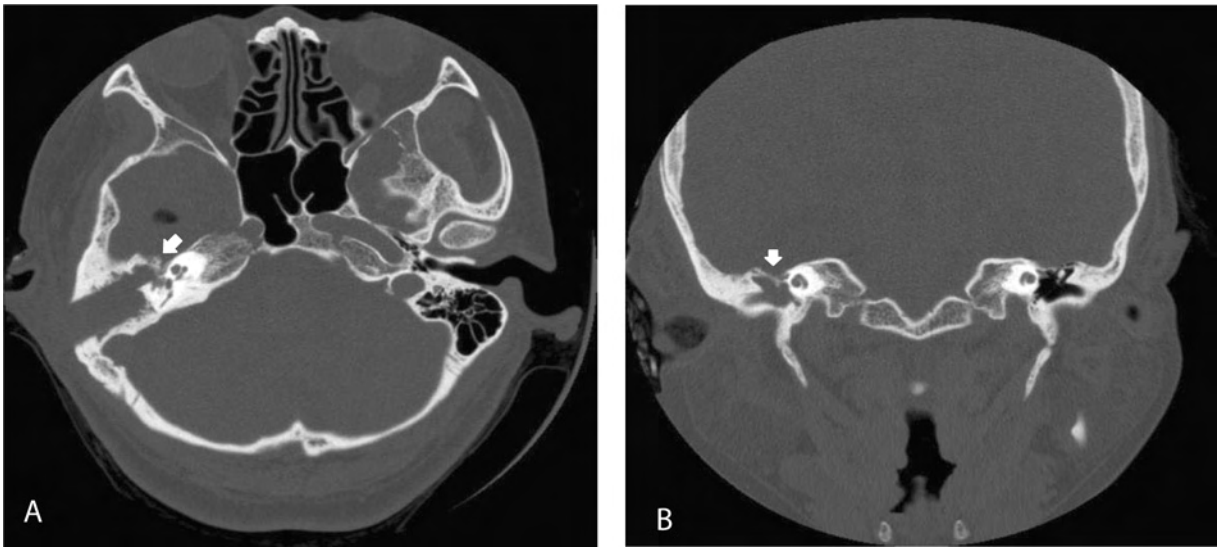

Fig. 3. HRCT of the temporal bone one week post-operative. Axial (A) and coronal (B) images show bone chip graft covering the defect of the right middle cranial fossa (white arrow). Pneumocephalus was noted in the right temporal lobe (black arrow).

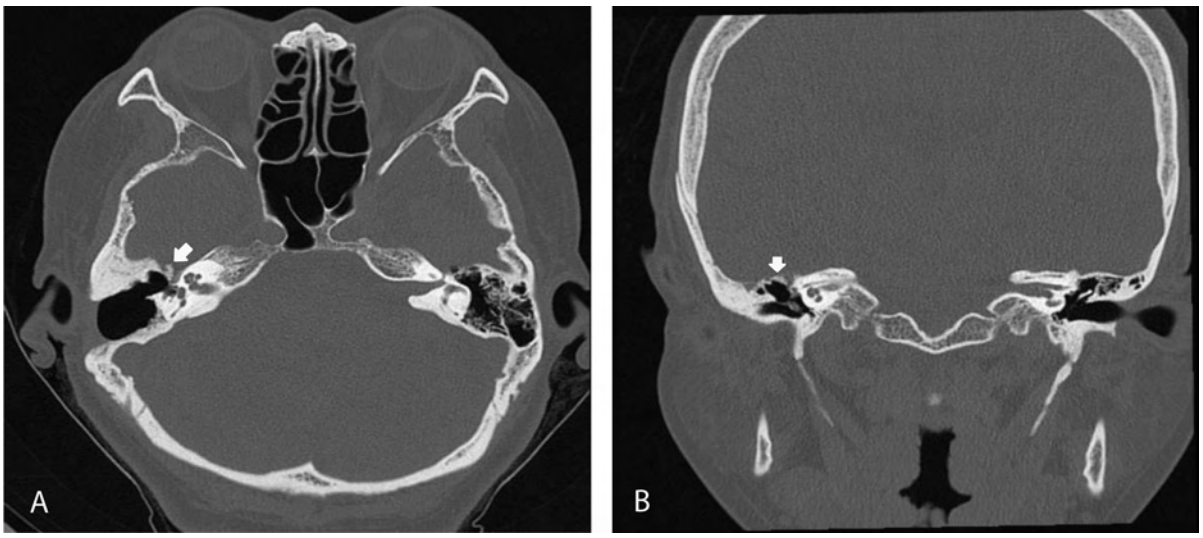

Fig. 4. HRCT scan of temporal bone one year post-operative. Axial (A) and coronal (B) images demonstrate a well-sustained anterior wall and roof of epitympanum (arrow). There is no evidence of CSF leak in the tympanic cavity.

Discussion

Spontaneous CSF otorrhea is defined when there are no identifiable causes such as past history of trauma, surgery, infection or tumor.[1] The origin of the CSF leak is most commonly a defect in the tegmen of the middle cranial fossa, less commonly in the posterior fossa.

The pathophysiology of spontaneous CSF otorrhea is not entirely clear but there are two main theories concerning the etiology of bony defect. The first is the congenital defect theory, where a tiny defect within the tegmen, caused by aberrant embryologic development, enlarges over time due to constant CSF pressure. This enlargement leads to eventual dural herniation and subsequent bony and dural thinning, resulting in CSF otorrhea.[2] The second theory, the arachnoid granulation theory postulated by Gacek, suggests that arachnoid granulations that do not find a venous termination during embryonic development come to lie in a blind end against the inner bony surface of the skull.[3] These granulations enlarge with age and may eventually erode bone. Aberrant arachnoid granulations located in the dural surface of the temporal bone are thought to be responsible for communication between the CSF space and the mastoid air-cell system. Gacek[3] identified an 8.5% incidence of arachnoid granulations in the posterior fossa of the temporal bone, while Ferguson[4] observed an incidence of 22% of pit holes created by arachnoid granulations in the middle fossa surface.

Bone defects of the cranial fossa can be closed using many autologous materials such as fascia, fat, muscle, cartilage and bone or synthetic materials such as silicone, silastic, Marlex, hydroxyapatite cement and titanium plate.[5] Each material must be strong enough to withstand intracranial pressure and be compliant enough to form a seal.

There are three surgical approaches for the repair of the CSF otorrhea: the transmastoid approach, middle fossa craniotomy, and a combined approach.[3,5,6] Surgeons can choose one of them by location and size of defect, their experience or preference, and better surgical view. The favored technique is a combined transmastoid-middle fossa approach, which gives a whole visualization of the tegmen and definite closure of the entire region. But this approach has a great potential for complications and should only be performed by experienced surgeons. Whereas the transmastoid approach is technically easier to perform, and includes fewer risks and complications, an anterior defect in the tegmen tymapni may require the removal of ossicles to ensure better exposure of the lesion. This causes hearing disturbance which can be restored with ossicular reconstruction in the same stage or the second stage. In our case, the lesion was located at the antero-medial side of the middle cranial fossa, so the combined approach could be suitable. However, we could also use the transmastoid approach with removal of ossicles only to obtain clear view of lesion and successfully manage the defect.

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RELATIONSHIPS BETWEEN THE LEVELS OF METYLMALONIC ACID, VITAMIN B12, HEARING LOSS AND TINNITUS IN SUBJECTS WITH ACOUSTIC TRAUMA

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Introduction

Acoustic trauma (AT) is a common cause of temporary or permanent sensorineural hearing loss that is caused by exposure to sudden excessive or long-term loud noise.[1]

Vitamin B12 deficiency has been based on low-serum vitamin B12 levels, usually less than 200 pg per ml. However, measurements of metabolites such as methylmalonic acid (MMA) and homocysteine have been shown to be more sensitive in the diagnosis of vitamin B12 deficiency than measurement of serum B12 levels alone.[1]

In the present study, we considered that the MMA level is more sensitive in measurement of serum B12 and possible diagnosis of vitamin B12 deficiency among patients with acute AT. We aimed to evaluate serum B12 and MMA levels, existence of tinnitus and hearing loss and to determine relationships between the levels of metylmalonic acid and Vitamin B12, hearing loss degree and tinnitus in subjects with AT.

Material and methods

This prospective, randomized study was conducted between January and September 2006. One hundred and thirty-five young healthy male subjects with acute acoustic trauma acquired during military service were studied prospectively. All subjects were exposed to loud noise during Habitable Place Battle Training. A G3-A3 automatic 7.62-mm caliber infantry rifle was used in training, and shootings were sequentially repeated for 21 times during a three-minutes period. All subjects were exposed to loud noise with an intensity of 134-142 dB Sound Pressure Level (average 138 dB). None of the participants used ear plugs during training (due to training standards). The volunteers were given an extensive explanation of the study before they accepted to participate and all signed an informed-consent form. Examination and laboratory study of participants were done in GATA Haydarpasa Training Hospital, Biochemistry and Audiology Laboratories. The study program was approved by the ethics committee of our hospital.

The following factors were examined: age, sound level of rifle-shot, degree of hearing loss, severity of tinnitus, serum levels of vitamin B12 and urine methylmalonic acid, increase in hearing loss, decrease of tinnitus severity. The severity of tinnitus was assessed by clinical grading on a 10-point scale. According to the severity of tinnitus, patients were divided into four groups. Tinnitus severity scale and pure-tone audiometry tests were performed on all subjects.

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Cholesteatoma and Ear Surgery – An Update, pp. 467-469
Edited by Haruo Takahashi

Results

All patients (n = 135) suffered from tinnitus after high-noise exposure. In 60 patients, the complaint had lasted more than 30 days. All subjects were divided into groups according to the tinnitus severity scale results (*1*-10). Subjects were subdivided into four groups according to the degree of tinnitus (Table 1).

Table 1. Tinnitus severity scale scores

	TINNITUS One day after		TINNITUS One month after
	Severity	n	n
Group 1	4	39	23
Group 2	3	24	10
Group 3	2	33	11
Group 4	1	39	16
TOTAL		135	60

Overall improvement rate of tinnitus after 30 days was 44.4%. The improvement rates after 30 days were 59%, 41.7%, 33.3%, and 41% in group 1 to group 4, respectively. Reduction in tinnitus severity was statistically significant (Spearman's rho test, $p < 0.01$). There was a significant difference between pre- and post-traumatic high-frequency audiometer thresholds ($p < 0.05$). There was a significant relationship between tinnitus and MMA levels measured in the first day after acoustic trauma (Spearman's rho test, $p < 0.05$, $r = 0.78$). But no significant relationship was found between tinnitus and MMA levels measured after 30 days (Spearman's rho test, $p > 0.05$). There was no significant relationship between hearing loss and tinnitus (Spearman's rho test, $p > 0.05$). There was no significant relationship between hearing loss, tinnitus and serum levels of vitamin B12 measured before and after acoustic trauma (Mann-Whitney U Test, $p > 0.05$) (Spearman's rho test, $p > 0.05$).

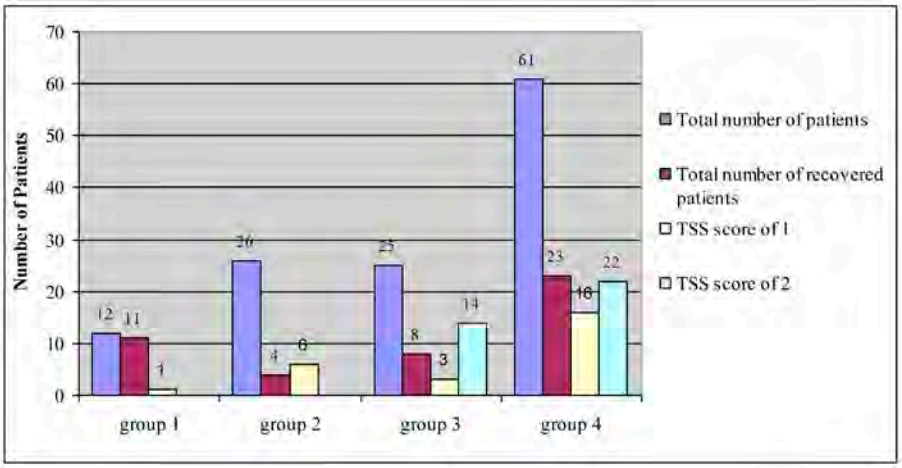

Fig. 1. Long-term changes in tinnitus severity

Discussion

Mrena *et al*. studied 119 patients of 163 AAT patients treated during the year 2000 and they found hearing loss in 46.7% and tinnitus in 94.2% of soldiers immediately after the high-noise exposure. Persistent hearing complaints were observed in 45% of the patients at the last control.[2] In our study, all patients (n = 135) had suffered from tinnitus and 83 subjects experienced hearing loss after high-noise exposure. Sixty patients had had a tinnitus complaint for more than 30 days.

Tinnitus is commonly associated with hearing loss.[3] In acoustic trauma, tinnitus may be present without detected hearing loss.[4] Dias and Cordeiro found the prevalence and severity of tinnitus to increase with increasing hearing loss. They concluded that tinnitus is less prevalent and less severe in milder hearing losses. On the other hand, in higher losses noise exposure results in greater discomfort.[5]

In our study we found no relationship between tinnitus severity and degree of hearing loss.

Shemesh *et al*. found a significant vitamin-B12 deficiency in patients with tinnitus and noise-induced hearing loss. After vitamin-B12 therapy, in some cases improvement in complaints were reported and routine vitamin-B12 level determination was recommended when evaluating patients for tinnitus.[6] Gok *et al*. studied the levels of homocysteine, folic acid, and Vitamin B12 in subjects with noise-induced hearing loss and they found homocysteine levels being significantly high and Vitamin B12 and folic acid levels significantly low compared to the controls.[7]

In our study, no relationship was found between urine MMA levels and hearing loss levels ($p > 0.05$). There was a relationship between urine MMA levels measured before noise exposure and tinnitus that occurred on the first day. However, there was no relationship between urine MMA levels measured one month after trauma and tinnitus severity ($p > 0.05$).

Table 2. Relationships bettween mean serum vitamin B12, methylmalonic acid levels and hearing levels

Levels	Hearing loss (n:83)			Normal hearing level (n:52)			Limits
	Before trauma	After trauma	P	Before trauma	After trauma	P	
Mean Serum B_{12} Levels (mg/dl)	(154-474) 257,3	(154-474) 269,6	p>0.05	(154-474) 189,7	(154-474) 242,7	p>0.05	220-914
Mean Urine MMA Levels (mg/dl)	(1,8-8,5) 4,24 (n:40)	(2,9-6,2) 4,21	p>0.05	(2,5-7,7) 4,5 (n:34)	(1,6-3,5) 4,1	p>0.05	0-6

Conclusion

There is a strong relationship between increased MMA levels and tinnitus. The MMA level is more sensitive in measurement of serum B12 and possible diagnosis of vitamin-B12 deficiency among patients with acute AT. We believe that low vitamin-B12 levels play a more active role in sensitivity of the ear to acoustic trauma. However, there is no relationship between persistent tinnitus and increased MMA levels. Therefore, MMA level is not a valuable test for prediction which patients will develop persistent tinnitus. The hearing system is highly complex and there are several unknown causes for damage to the inner ear. Further studies are needed to determine the possible effects of vitamin B12 on hearing protection and associations between acoustic trauma and tinnitus.

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PSYCHOLOGICAL STATUS OF PATIENTS WITH PATULOUS EUSTACHIAN TUBE

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Introduction

Patients with a patulous eustachian tube (PET) often complain of various symptoms, including autophonia, impaired hearing, ear fullness, tinnitus, dizziness, headache, and psychological symptoms. When treating PET patients, it is important to consider the patient's psychological state because they suffer from a variety of symptoms that may be long lasting and may affect, or be affected by, their mental state. This study focused on the psychological status of PET patients and compared these patients with healthy subjects.

We assessed the psychological status of PET patients using questionnaires that evaluated anxiety and depression, autonomic nerve imbalance, the degree of PET symptoms, and whether these symptoms interfered with daily life. We compared PET patients with healthy controls.

Materials and methods

We reviewed the medical records of 27 PET patients (ten males, 17 females; age 18-87 years, mean age 52.1) who visited our department between April 2010 and May 2012. Twenty healthy subjects were enrolled as controls (eight males, 12 females; age 24-67 years, mean age 48.0). To assess their psychological state and autonomic dystonia, all of the subjects completed the Hospital Anxiety and Depression Scale (HADS)[1] and Toho Medical Index (TMI), which evaluates autonomic nervous system symptoms and psychiatric symptoms.[2] The HADS consists of an anxiety score (HADS-A) and a depression score (HADS-D). Each item in the HADS is scored from 0-3, and the total score ranges from 0 to 21. A score ≥ 8 suggests the presence of a mood disorder such as an anxiety or depressive disorder.[3] The TMI score ranges from 0 to 33.[2] A score ≥ 8 indicates autonomic nerve imbalance. Additionally, in the 27 PET patients, we assessed the degree of each patient's symptoms and whether the symptoms interfered with daily life using a face scale ranging from 0 to 10 points (Fig. 1).

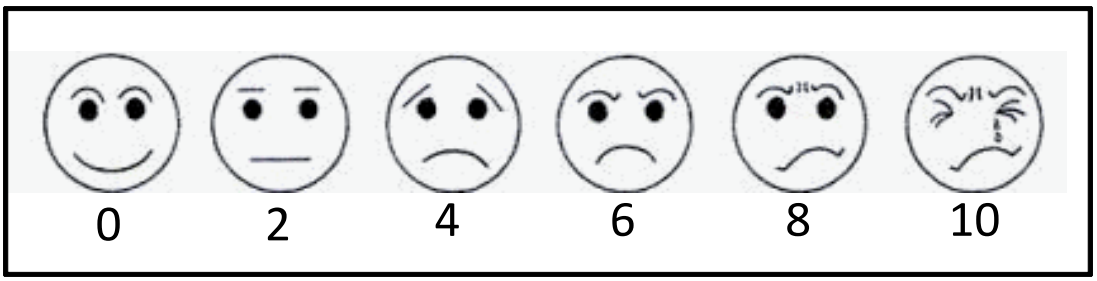

Fig. 1. Degree of the patients' symptoms and symptom interference with daily life.

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Edited by Haruo Takahashi

Correlations between the HADS scores and the degree of the patients' symptoms and whether the symptoms interfered with daily life were analyzed using Pearson's product-moment correlation coefficient. The PET patients and normal subjects were compared using a chi-square test.

Results

Comparisons of the HADS and TMI scores of PET patients and normal subjects

Seven of the 27 (25.9%) PET patients had positive HADS-A scores for anxiety (Fig. 2.1); significantly more PET patients than controls had high HADS-A scores (P = 0.04). Four of the 27 (14.8%) PET patients had positive HADS-D scores for depression (Fig. 2.2); there was no significant difference in the HADS-D scores between the PET patients and controls. Eight of the 27 (29.6%) patients had positive TMI tests (Fig. 2.3); there was no significant difference in the number of PET patients and controls with positive TMI scores.

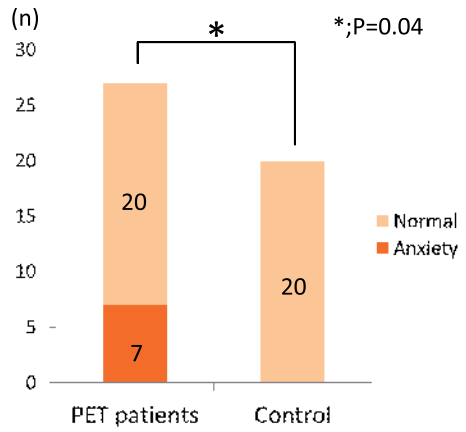

Fig. 2.1. Comparison of positive HADS-A scores in PET patients and controls.

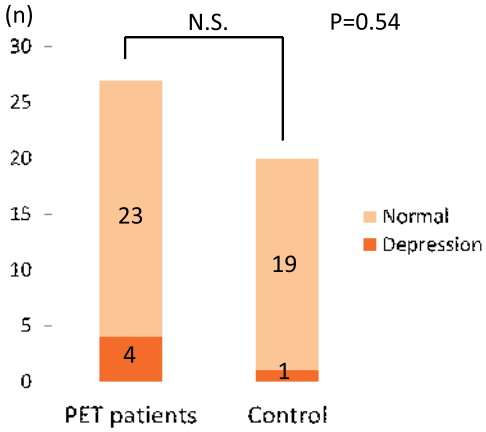

Fig. 2.2. Comparison of positive HADS-D scores in PET patients and controls.

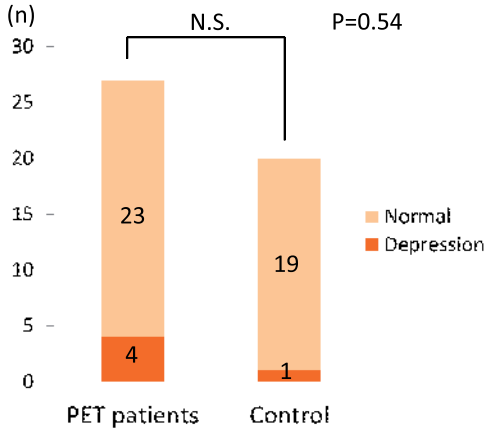

Fig. 2.3. Comparison of positive TMI scores in PET patients and controls.

Degree of symptoms and symptom interference with daily life in PET patients

The level of patients' symptoms was 6.1 ± 2.0 (mean ± SD). Of the 27 patients, 18 (66.7%) experienced severe symptoms, *i.e.*, higher than level 6. The mean score for symptom interference with daily life was 5.7 ± 2.1. Of the 27 patients, 18 (66.7%) experienced severe symptom interference with daily life, higher than level 6.

Correlations between HADS scores and degree of symptoms/symptom interference with daily life in PET patients

In PET patients, neither anxiety nor depressive disorder was significantly correlated with the degree of symptoms or symptom interference with daily life (Figs. 3.1 and 3.2).

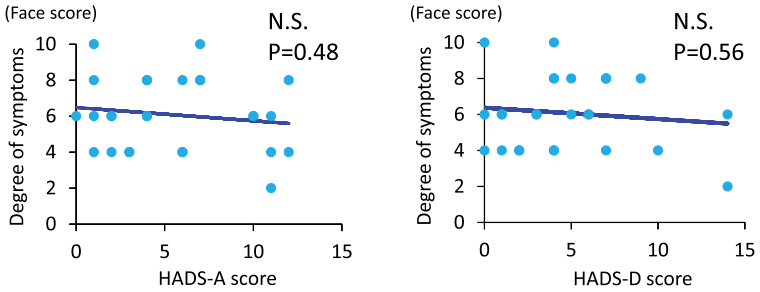

Fig. 3.1. Correlation between the HADS score and the degree of the PET patients' symptoms.

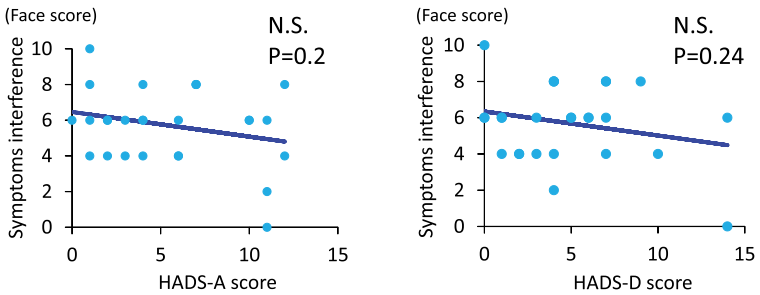

Fig. 3.2. Correlation between the HADS score and symptom interference with daily life in PET patients.

Discussion

A previous study found PET patients had tendency lacking in agreeableness than normal controls, and they scored lower on measures of though expression and neurotic tendencies, although these scores were not related to the duration of the disorder.[4]

In our series, around 70% of the PET patients experienced severe symptoms and symptom interference with daily life. Our PET patients were more prone to anxiety than the healthy subjects. No significant correlations were found between scores for anxiety and depression among the PET patients and the degree of symptoms or symptom interference with daily life. These results suggest that PET patients tend to develop anxiety, but the severity of their anxiety does not affect the severity of their symptoms or symptom interference with daily life. It is more likely that the anxiety preceded PET and is independent of their PET symptoms. Nevertheless, awareness of the mental state of PET patients is very helpful when treating these patients.

Conclusions

Patulous eustachian tube patients tend to be anxious, but the severity of their anxiety does not appear to affect the severity of their symptoms or symptom interference with daily life. Rather, it is their baseline personality character.

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AN ADULT CASE OF PERILYMPH GUSHER CAUSED BY HEAD CONTUSION

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Here we report a rare adult case of cerebrospinal otorrhea triggered by head contusion in a 46-year old male. Riding his bicycle, he fell to the ground and bruised his head. A month after the accident he had watery rhinorrhea and suffered from fever and headache. He consulted a neurosurgeon at a nearby hospital and was diagnosed with meningitis. He was admitted to the hospital and was treated with an intravenous drip infusion therapy of antibiotics. The patient consulted an otolaryngologist in our department and was diagnosed with CSF leak with temporal bone fracture. He is naturally completely deaf. The balance test revealed no particular remarks. The clear fluid outflow from the pharyngeal orifice of Eustachian tube was observed by means of fiberscopy. Brain MRI, temporal bone targeted CT and RI cisternography strongly suggested a CSF leak with Mondini's malformation of the inner ear and surgery was performed to explore the CSF leak and stop it. Mastoidectomy indicated no fracture. We found the fluid flow from the mesotympanum and finally found an extreme overflow of fluid from the footplate of the stapes. The stapes was removed and the vestibule was filled with fascia of temporal muscle and bone patty. About four years after the surgery, recurrence of CSF leak occurred and revision surgery was performed. The lesion of leakage was the same as the first surgery and the vestibule was filled with cartilage of the auricle and fascia. Although recurrent meningitis has sometimes been reported in patients with inner ear malformation, it is rare that head contusion is the trigger of a CSF leak from the inner auditory canal in an adult.

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Edited by Haruo Takahashi

CHORDA TYMPANI NERVE REPAIR WITH A POLYGLYCOLIC ACID-COLLAGEN TUBE IN CHOLESTEATOMA SURGERY

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Introduction

During cholesteatoma surgery in the middle ear, the chorda tympani nerve is frequently severed.[1] Resection of the chorda tympani nerve causes a defect of gustatory function in the affected side resulting in a taste disturbance. Although some patients experience gradual recovery of the symptoms, alteration or loss of taste forms a real problem for them. Restoration with sufficient functional recovery after nerve injury continues to be a clinical challenge. Various artificial nerve conduits have been tried as alternatives that regenerate severed peripheral nerves to autografts . There is no report of applying the device to the chorda tympani nerve reconstruction. The chorda tympani nerve seems hard to restore because this nerve runs through the aerial space without any floors as a scaffold for grafting. A bio-absorbable polyglycolic acid (PGA) tube filled with collagen sponge (PGA-collagen tube) (Fig. 1) has been developed recently. In experimental and clinical trials, it has proven to be effective for regeneration of peripheral nerve defect. Using a PGA-collagen tube, we attempted to reconstruct the chorda tympani nerve gaps in a patient with cholesteatoma who underwent a tympanoplasty, and examine whether the gustatory dysfunction restored.

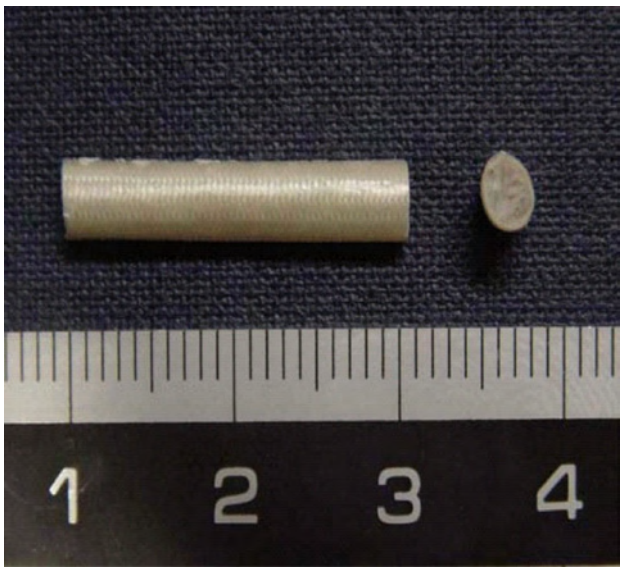

Fig. 1. Polyglycolic acid (PGA) tube and its transverse.

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Cholesteatoma and Ear Surgery – An Update, pp. 477-481
Edited by Haruo Takahashi

Case presentation

A 52-year-old male presented with a three-month history of taste disturbance and a two-month history of hearing loss in the right ear. Physical examination showed a cholesteatoma white mass with formation of inflammatory granulation tissue in the pars flaccida over the manubrium of the malleus. The pure-tone hearing testing revealed the moderate conductive hearing loss of 41.3 dB in the pure tone average (PTA). A CT scan with a bone window demonstrated the presence of soft-tissue mass in the epitympanum and antrum with ossicular erosion.

The patient underwent surgery for removal of cholesteatoma. During surgery, a cholesteatoma sac was found in the attic and antral cavity, that destroyed ossicles such as malleus and incus. It was found that the cholesteatoma sac was attached to the chorda tympani nerve and was likely to invade the nerve fibers on dissection of the sac. Since the chorda tympani nerve attenuated and was adhered to the lateral aspect of the sac, the decision was made to section the nerve in order to achieve the removal of the cholesteatoma sac. The chorda tympani nerve in continuity with the cholesteatoma sac was resected in the healthy portion of both proximal and distal nerve apart from the sac involvement. The resulting seven-mm nerve gap was reconstructed with a PGA-collagen tube (nine mm in length and three mm in diameter). Both the proximal and distal stumps of the severed nerve were inserted into the PGA-C tube to a depth of one mm. The tube was secured to the proximal and distal nerve ends with epineural 10-0 polypropylene monofilament sutures (Prolene, Ethicon, Somerville, NJ, USA) (Fig. 2). The tympanic membrane was reconstructed with the temporalis fascia.

The gustatory function was assessed with electrogustometry (EGM) using an electrogustometer (Rion, TR-06, Rion Co, Tokyo, Japan) which is a more reliable and objective method to evaluate quantitative taste function.[1,2] A five-mm probe was touched to the lateral edge of the tongue at two cm from its tip to stimulate the CTN territory. The stimulation range of EGM was -8 to 34 dB (normal range ≤ 8 dB). Scale out, not detected with any EGM stimulation was taken as 36 dB.

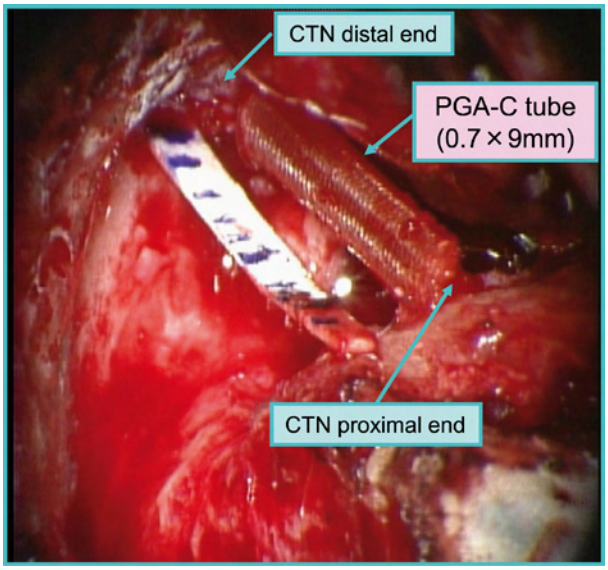

Fig. 2. Intraoperative view after bridge of a PGA-C tube. CTN: chorda tympani nerve.

Pre-operative EGM threshold scoring 25 dB showed objective gustatory mild disturbance. The threshold of EGM on the affected side elevated up to 32 dB in one week after the CTN reconstruction. However, the threshold started to decrease within ten days and returned to normal level (≤ 8dB) at two weeks, leading to complete recovery at 56 days, while the EGM threshold on the intact side showed little change all the time through the postoperative course (Fig. 3). Afterwards, the EGM threshold maintained the recovery level for 72 days.

This study was approved by the clinical research ethics board of the Nara Medical University Hospital.

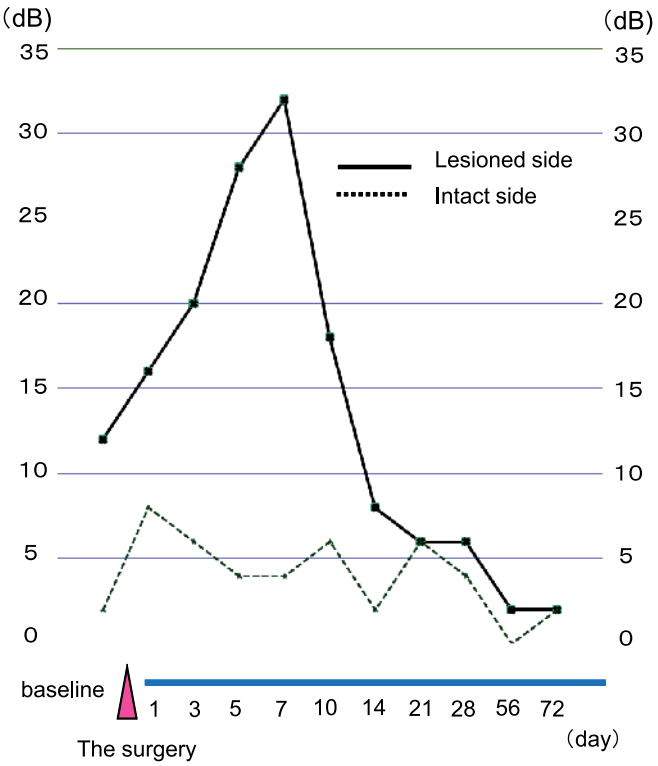

Fig. 3. The clinical course of the EGM threshold after reconstruction of CTN with a PGA-C tube.

Discussion

The artificial nerve conduit used in this study is composed of a biodegradable tube filled with biodegradable filaments. The tube framework is made of cylindrically woven PGA mesh of which outer and inner surfaces are coated with amorphous collagen layers. The inner space of this tube was filled with a 3D circumstance matrix spongiform collagen. Sponge-form collagen, which makes the surface area larger than fiber-form, seems to provide a favorable micro-environment for nerve progression (axonal sprouting cellular proliferation and tissue healing).[3,4] Although collagen has a tendency to dissolve easily in the body, it is likely able to remain the shape in the nerve conduit for periods long enough to complete the nerve regeneration in the body, since PGA was used as outer cover of collagen.[3,5] The PGA-C tube (30 mm in length, four mm in diameter) was trimmed in nine mm long pieves, in accordance with demand during surgery.

To take an advantage of this feature in the PGA-C tube, some experimental studies and clinical trials was extended to examine on the performance of PGA-C tube for nerve gap reconstruction. Histologic examination (four months) after implantation of the PGA-C tube showed regeneration of nerve tissue structure, including myelinated axons and Schwann's cells. Electrophysiological analysis demonstrated functional recovery of the regenerated nerves at a growth rate of 0.5-1.0 mm/day.[3,5]

PGA-C tubes have recently been applied for clinical cases with a defect of the peripheral nerve such as a proper digital nerve and superficial peroneal nerve with successful results regarding symptomatic and functional recovery.[6,7]

In this study, the nerve reconstruction for the severed chorda tympani nerve in the aerial space using a PGA-C tube, brought successful recovery of the taste function as assessed by EGM. There is little study reporting on the time course in the evaluation of EGM, a quantitative gustatory function test[2] used for each patient with a severed nerve. One study reported that the threshold of EGM, which was found to increase considerably two weeks after the section of CTN, did not get back to the baseline level in the follow-up cases. Thirty percent or less of the cases demonstrated partial recovery after two years.[8] Another report suggested that one to two years are needed until the EGM threshold is recovered and stabilized.[9]

According to these reports, the mechanism underlying the recovery of taste after unilateral CTN section seems to be associated with neuronal innervation to the lesioned side from contralateral intact side and /or from ipsilateral glossopharyngeal nerve.[10-14]

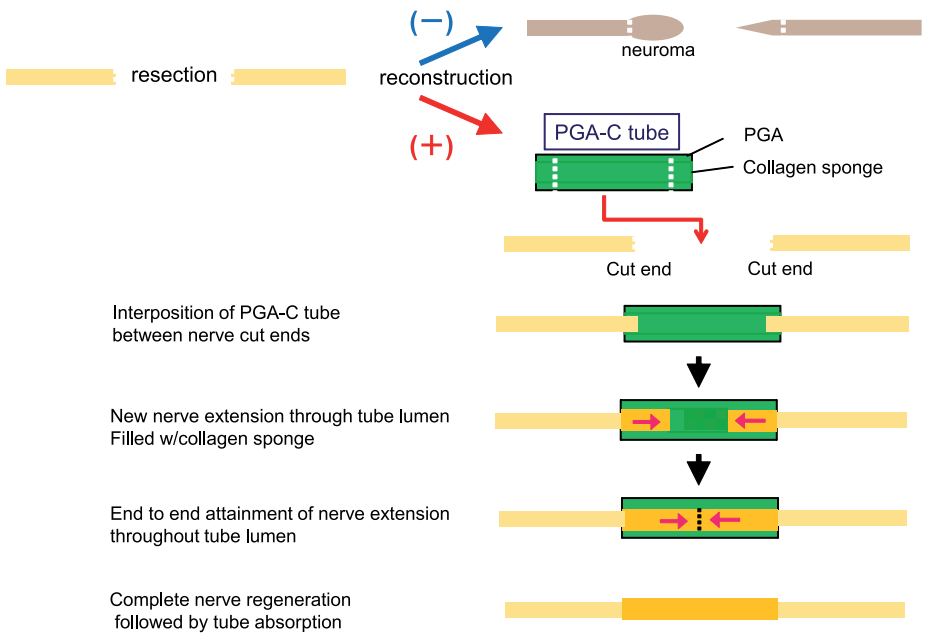

Fig. 4. Possible mechanism underlying a regeneration of severed nerve with a PGA-C tube.

In our study, the gustatory function as measured by EGM started improving at ten days, got back to normal range at 14-21 days and had recovered completely 56-72 days after the reconstruction of the severed chorda tympani nerve with a PGA-C tube. This fast improvement of the gustatory function appears to be caused not by the innervation from the other healthy regions such as contralateral chorda tympani or ipsilateral glossopharyngeal nerve, but by the regeneration of the nerve itself through the PGA-C tube.

Tube framework consisting of PGA likely plays a role in preventing nerve fibers from misdirection in a process of regeneration and guiding them correctly to each other nerve cut ends through tube lumen filled with collagen sponge, as nerve fibers extended and regenerated successfully toward a rectified way without any misleading even if it is in the air without any scaffold (Fig. 4).

In conclusion, these results suggest that reconstruction with an artificial nerve conduit, PGA-C tube involves functional and morphological regeneration of the chorda tympani nerve and is possible intervention to facilitate a recovery of the taste function.

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CLINICAL OBSERVATIONS IN 20 CASES OF POST-INFLAMMATORY MEDIAL MEATAL FIBROSIS

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Background and purpose

Medial meatal fibrosis (MMF) has numerous synonyms, including post-inflammatory acquired atresia, post-inflammatory medial meatal fibrosis, chronic stenosing external otitis, and recurrent acquired atresia, which reflect both its etiological and histopathology.

Katzke and Pohl (1982) proposed a classification of the disease categorizing the etiological factors (inflammatory and traumatic).[1] They reported cases of acquired atresia in which fibrous tissue in the medial portion of the external auditory canal developed as a consequence of chronic otitis externa, and called this condition post-inflammatory MMF (PIMMF), based on the morphology.

The purpose of this paper was to present our experiences with MMF.

Materials and methods

Over the past 20 years, The University of Miyazaki encountered a total of 20 (18 females and 2 males) cases of PIMMF. The patients' ages ranged from 54 to 80 years, with a mean age of 63 years. All patients complained of hearing loss, 13 cases also complained of chronic otorrhea, and seven cases habitually scratched their ears with ear picks or cotton buds because of an itching sensation in the ear. Six cases presented with bilateral disease. The pre-operative pure-tone average was 63.5 dB, and the average air-bone gap was 29.2 dB. Pure-tone audiometric examination revealed conductive or mixed hearing loss in all affected ears. There was no surgical history in any patient.

All patients underwent CT studies using a TOSHIBA TCT/900S scanner with 0.5-mm contiguous sections with a semi-axial projection (OM line 20° downward). Auditory status: Pure tone averages were calculated using 0.5, 1, and 2 kHz. Air and bone from the same test were used to calculate the air-bone gap.

Treatment: Seventeen patients underwent surgery. Type-1 tympanoplasty was performed in 13 cases, and the others were performed in four cases.

Results

Pre-operative CT findings

CT scans were useful for assessment of the soft tissue thickness within the bony canal and associated middle ear pathologies if present. High-resolution computed tomography showed a soft tissue density occupying the medial portion of the external auditory canal. Typical CT findings are shown in Figure 1. A partially remaining tympanic structure was observed in some cases. Severe MMF was observed in two cases (Figure 2). Extension of fibrosis to the tympanic cavity was observed in three cases (Figure 3).

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Edited by Haruo Takahashi

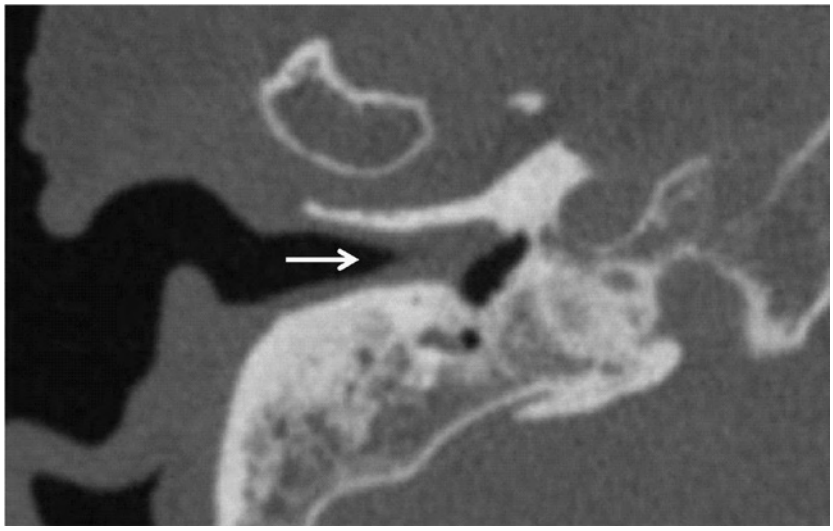

Fig. 1. CT image (horizontal plane, right ear). A soft-tissue density area (arrow) in the medial aspect of the external auditory canal.

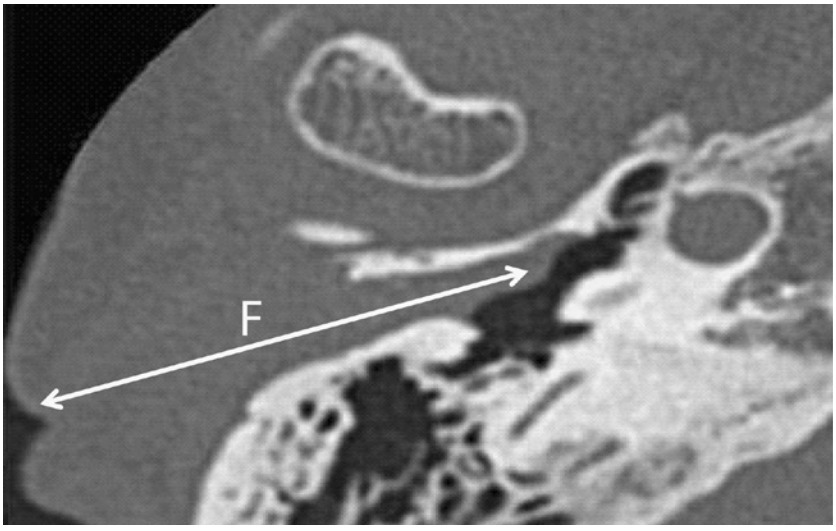

Fig. 2. CT image (horizontal plane, right ear). Atresia in the cartilaginous meatus.

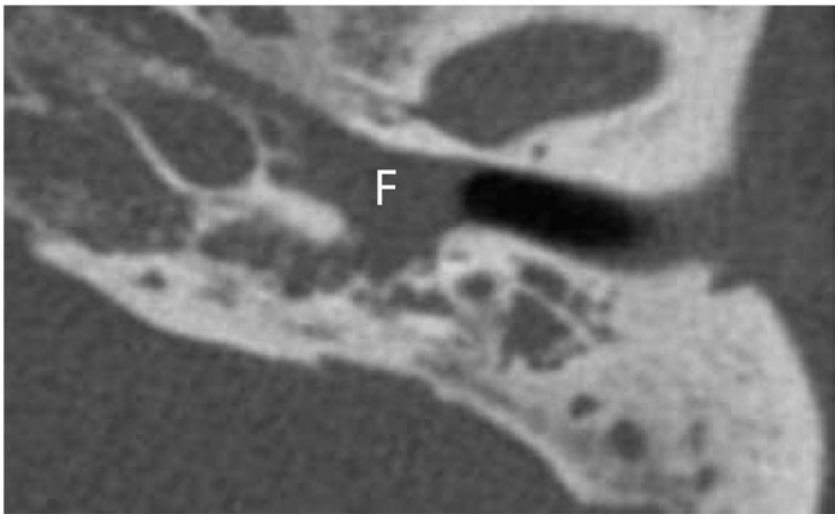

Fig. 3. CT image (horizontal plane, right ear). Extension of MMF to the tympanic cavity.

Pre-operative associated CT findings

Bony stenosis was concomitantly formed in 17 cases, and bony defects of the anterior wall were observed in six cases. An attic retraction associated with MMF was formed in one case.

Operative findings

Seventeen patients underwent surgery. Twelve patients had an intact lamina propria of the tympanic membrane, and five patients showed perforations. Ten patients had an intact ossicular chain, three patients showed caries, and two patients showed tympanosclerosis and malleus ankylosis.

Hearing results

Post-operative hearing results assessed at least after one year following surgery (ranging from one to five years, mean: 1.5 years). Hearing results have been favorable, from a mean of a 63.5-dB pure-tone average and 29.2-dB air-bone gap preoperatively to a 46.8-dB pure-tone average post-operatively.

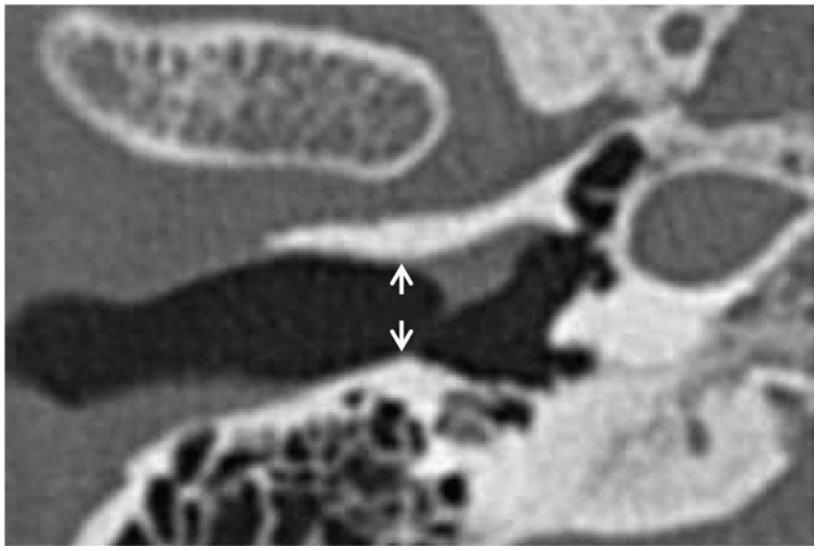

Fig. 4. CT image (horizontal plane, right ear). Bony stenosis of the external auditory canal (arrow).

0 ~ 10 dB	29%
11 ~ 20 dB	47%
21 ~ 30 dB	6%
> 30 dB	18%

Fig. 5. Evaluation of hearing results using the AAO-HNS guideline (n = 17).

Discussion

The term MMF was first described by Katzke and Pohl in 1982. In their article, the inflammatory or post-inflammatory medial meatal fibrosis was subdivided into an intact drum type and a perforated drum type. The traumatic type is subdivided into post-surgical, caustic, or external trauma. The PIMMF – intact drum type – is due to the fibrous healing of granulations resulting from either granular myringitis or granular otitis externa, and the perforated drum type is due to the organization of florid granulations resulting from chronic suppurative otitis media. In our study, all patients were diagnosed with PIMMF because of the existence of chronic inflammation in their affected ear for an extended amount of time. Twelve cases were the intact drum type and five cases were the perforated drum type. HRCT is useful not only to verify the presence of soft tissue thickening within the external auditory canal, but also to assess tympanic cavity involvement. The goal of MMF surgery was to remove the fibrous thickening of the subcutaneous tissue, creating a tympanic membrane as thin as possible, with preservation of the normal migration function of its skin layer. Additionally, the enlargement of the stenotic ear canal is important because 17 out of 20 ears with MMF had a concomitant bony stenosis. Hearing results of our series have been favorable, from a mean of a 63.5-dB pure-tone average and a 29.2-dB air-bone gap pre-operatively to a 46.8-dB pure-tone average post-operatively over an average follow-up period of 1.5 years. MMF is a good candidate for surgical treatment, but obviously a longer follow-up period is necessary for a stable canal skin condition.

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CLINICAL ANALYSIS OF CARCINOMA OF THE EXTERNAL AND MIDDLE EAR

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Introduction

Carcinoma of the external and middle ear is an uncommon neoplasm with annual incidence estimated at between one and six per million.[1] The incidence of the tumor is as rare as less than 0.2% of head and neck cancers.[2] Because of the rarity of the tumor, it has been difficult to formulate an evaluation and a treatment strategy. It is also sometimes difficult to diagnose the carcinoma at an early stage because it often resembles or coexists with chronic inflammatory diseases of the ear.

There is a consensus that an outcome of a treatment is poor for an advanced stage. Therefore it is necessary to find out a reasonable therapeutic strategy how the advanced stage of temporal bone carcinoma can be overcome. This study aims to analyze clinical characteristics and five-year survival of carcinomas of the external and middle ear in our department.

Subjects and methods

From February 2000 to April 2012, 28 patients with carcinomas of the ear were treated at the Department of Otolaryngology, Nagasaki University Hospital. Age and sex of these patients are listed in Table 1. In 25 patients, the carcinoma was mainly located in the external auditory canal and in three patients in the middle ear. Tumors were staged according to the Pittsburgh staging system[3] for the external auditory canal (Table 2).[3]

We have basically tried to select surgery whenever possible; mainly total resection of the tumor was performed. In T1 and T2 cases, we performed lateral temporal bone resection (LTB), preserving the facial nerve. Subtotal temporal bone resection (STB) was performed in T3 or anterolateral development cases of T4. The postoperative additional treatment was given to the patients who had positive margin (Fig. 1). Recently, super-selective arterial infusion (SAI) of cisplatin with concomitant radiotherapy is also chosen as treatment for patients who are considered contra-indicated for surgery. Intra-arterial infusion of cisplatin was delivered via the posterior auricular artery and/or superficial temporal artery.[4] The data of treatment and survival rate are summarized in Table 3.

Results

Eight patients had a history of otitis externa, and three had a history of chronic otitis media, respectively. Two had a history of ear surgery and five had a habit of ear-picking. All patients for whom more than one year was needed before the diagnosis could be established, had an advanced cancer. Histopathologically, there were 24 squamous cell carcinomas, two adenoid cystic carcinomas, one adenocarcinoma, and one undifferentiated

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Edited by Haruo Takahashi

Table 1.

Patient no.	Sex	Age (y. o.)	Stage (T N M)	Surgery Type	Positive Margin	Histopathologic Diagnosis	Radiotherapy (Gy)	Chemotherapy (CHE)	Follow-up (month)	Outcome	Chief Complaint	Duration before Diagnosis
1	M	62	I (1 0 0)	LTB,s ND	-	SCC	-	-	18	NED	Otorrhea	10m
2	M	39	I (1 0 0)	LTB,s ND	-	SCC	-	-	47	NED	Otalgia	1w
3	F	47	I (1 0 0)	LTB,s ND	-	SCC	-	-	105	NED	Bloody otorrhea	6m
4	F	90	I (1 0 0)	–		SCC	66	-	47	NED	Hearing loss	5m
5	M	67	II (2 0 0)	LTB,s ND	-	SCC	50	-	80	NED	Otorrhea	6m
6	M	69	II (2 0 0)	LTB	-	SCC	-	-	103	NED	Otorrhea	4m
7	F	47	II (2 0 0)	LTB,s ND	-	SCC	-	-	105	NED	Ear fullness	4m
8	F	67	III (3 0 0)	LTB,s ND	-	SCC	54	-	26	NED	Bloody otorrhea	3m
9	F	64	III (3 0 0)	LTB	-	Ceruminous adenocarcinoma	-	-	46	NED	Otorrhea	1y
10	F	62	III (3 0 0)	LTB,s ND	-	SCC	60	-	63	NED	Bloody otorrhea	1w
11	F	42	III (3 0 0)	STB,s ND	-	SCC	60	FP,TPF	64	NED	Otorrhea	7m
12	F	37	III (3 0 0)	LTB,s ND	-	ACC	-	-	71	NED	Otalgia	2y
13	M	69	III (3 0 0)	LTB,s ND	-	SCC	-	-	74	NED	Ear fullness	2m
14	M	62	IV (4 1 0)	LTB,s ND	-	SCC	63	FP×2	14	NED	Otalgia	2w
15	F	72	IV (4 1 0)	STB,s ND	-	SCC	63	CBDCA×3	15	DOD	Swelling	4w
16	F	34	IV (4 0 0)	LTB,s ND	-	SCC	pre30, post40, post SRT25	FP×2,TPF	23	DOD	Otorrhea	2.5m
17	F	46	IV (4 0 0)	Mastoidectomy	-	SCC	66	FP×3	42	NED	Otorrhea	1y
18	M	68	IV (4 0 0)	LTB,s ND	-	SCC	-	-	73	NED	Swelling	2m
19	M	61	IV (4 0 0)	LTB,s ND	-	ACC	-	-	86	NED	Otalgia	1y
20	M	59	IV (4 0 0)	LTB,s ND	+	SCC	59.4	FP×2	8	recurrence	Bloody otorrhea	4m
21	M	69	IV (4 0 0)	STB,s ND	+	SCC	60	-	9	DOD	Otorrhea	1w
22	F	64	IV (4 0 0)	LTB	+	SCC	60	TPF	61	NED	Otorrhea	3m
23	M	67	IV (4 0 0)	LTB	+	SCC	-	TS-1	9	DOD	Otorrhea	3m
24	F	85	IV (4 0 0)	–		SCC	63	SAI×3	5	NED	Bloody otorrhea	2w
25	M	75	IV (4 0 0)	–		SCC	60	FP×3	17	DOD	Otorrhea (facial palsy)	1y
26	F	89	IV (4 0 0)	–		Undifferentiated carcinoma	50.4	-	17	DOD	Swelling	6m
27	M	57	IV (4 1 0)	–		SCC	70	SAI×5	26	NED	Swelling	10m
28	F	66	IV (4 1 0)	–		SCC	70.2	SAI×2	27	NED	Swelling	1y

s ND: selective neck dissection, NED: no evidence of disease, DOD: died of disease, P: cisplatin, F: fluorouracil, T: docetaxel hydrate, TS-1: tegafur, SAI: superselective arterial infusion of cisplatin, m: month, w: week, y: year, SCC: squamous cell carcinoma, ACC: adenoid cystic carcinoma

Table 2. University of Pittsburgh TNM staging system proposed for the external auditory canal[1]

N status

Involvement of lymph node metastases is a poor prognostic finding ; Any node involvement should automatically be considered as advanced disease, ie, T1N1=stage III and T2, T3, T4, N1=stage IV

M status

Distant metastases indicate a very poor prognosis and should be considered as stage IV disease.

In the absence of metastastic lymph nodes or distant metastases, T status of the tumor defines the clinical stage.

T status

T1– Tumor limited to the external auditory canal without bony erosion or evidence if soft-tissue extension

T2– Tumor with limited external auditory canal bony erosion(not full thickness) or radiographic finding consistent with limited (<0.5cm) soft-tissue involvement

T3– Tumor eroding the osseous external auditory canal (full thickness) with limited (<0.5cm) soft-tissue involvement, or tumor involving middle ear and/or mastoid, or patients presenting with facial paralysis

T4– Tumor eroding cochlea, petrous apex, medial wall of middle ear, carotid canal, jugular foramen or dura, or with extensive (>0.5cm) soft-tissue involvement

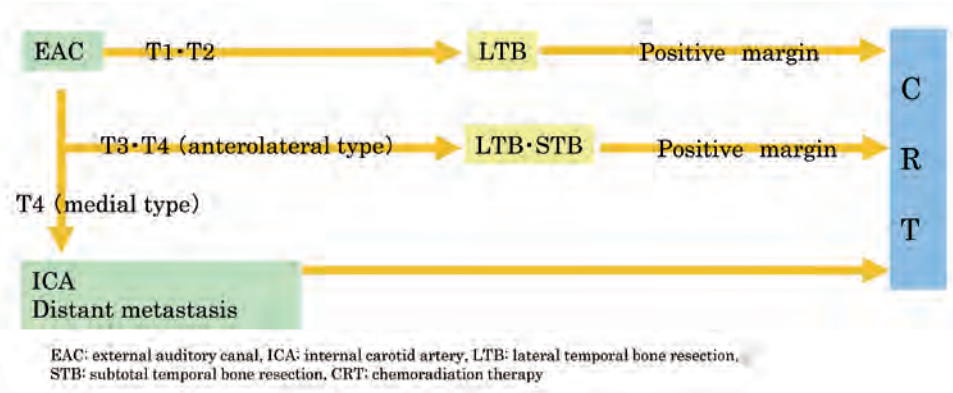

Fig. 1. Scheme of temporal bone resection.

Table 3. Treatment and survival

		Stage				Survival rate (%)	
	n	I	II	III	IV	Total	Stage III, IV
OPE+RT+CHE	7			1	6	57	57
OPE+RT	4		1	2	1	75	75
OPE+CHE	1				1	0	0
OPE	10	3	2	3	2	100	100
RT	2	1			1	50	0
RT+CHE	1				1	0	0
RT+SAI	3				3	100	100

RT: radiotherapy, CHE: chemotherapy, SAI: superselective arterial infusion of cisplatin

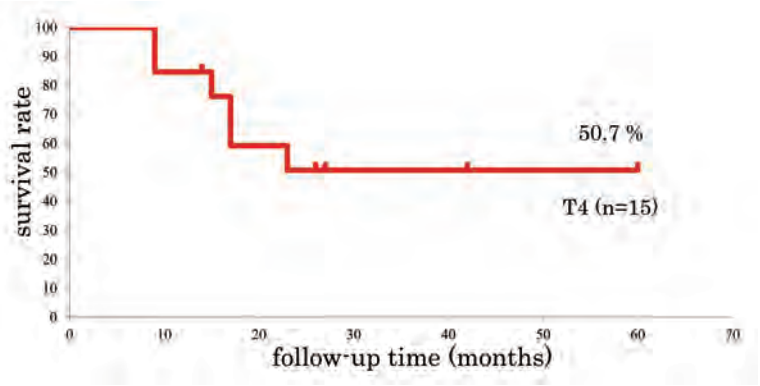

Fig. 2. Five-year survival rate of T4 patients.

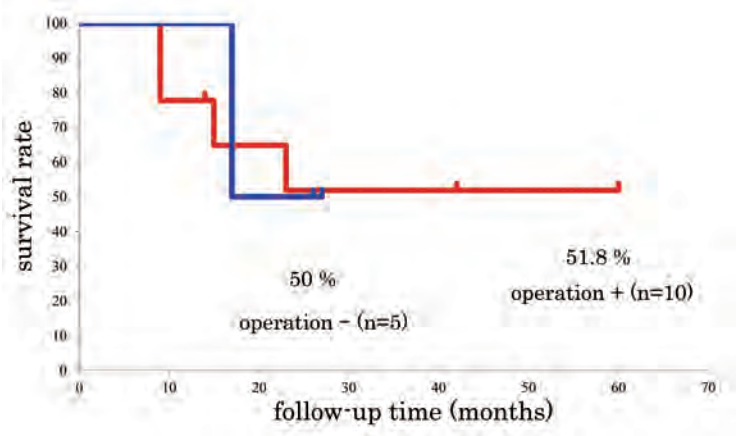

Fig. 3. Comparison of estimated survival in T4 patients treated with operation versus without operation.

carcinoma. We gave RT to T2 patients (no. 6) on whom we could not perform en-bloc resection of the tumor. The other T1 and T2 patients had no evidence of disease.

The five-year survival rate revealed by Kaplan Meier survival analysis was 100 % in T1, T2 and T3 cases, and 50.7 % in T4 (Fig. 2). In T4 cases, the survival rate of the patients who underwent surgery was 51.8 %, whereas that of the patients who had chemoradiation or SAI with radiotherapy was 50% (Fig. 3). Three patients who received SAI had a good outcome. Post-operative histopathological examination showed that four of 23 patients who underwent surgery had a positive margin, and all of them were T4. There was a considerable difference between positive and negative margin cases in the five-year survival rate. That is, regardless of the post-operative adjuvant treatment, the rates were 87% and 33%, respectively, as shown in Figure 4.

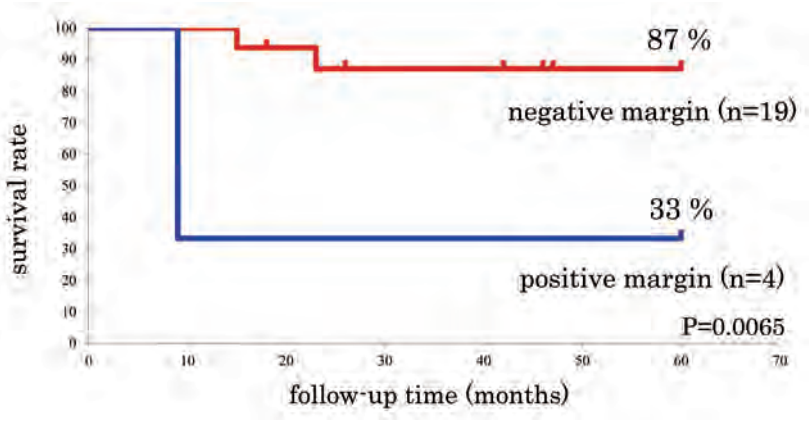

Fig. 4. Influence of histopathological margin on survival rate.

Conclusion

All the cases of the early-stage T1-3 showed good prognoses, while cases in the advanced stage, especially with T4 disease, showed poor five-year survival. These results suggest that prompt diagnosis and treatment are essential. When we examine a patient with repeated otorrhea for a long period, we should consider further examinations with CT scan and/or biopsy to exclude a neoplasm.

We reached the conclusion that current treatment including surgery with or without radiotherapy can control up to T3 cases. Even in advanced cases, surgical approach or SAI improved the survival rate and prognosis. In our view, this reaffirmed the importance of complete resection. On the other hand, one of four patients with a positive margin after surgery treated post-operatively with chemo-radiation survived more than five years with NED. This suggests that post-operative therapy for positive margin may be nevertheless worth trying. SAI could be an effective treatment for advanced cases.

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ISBN 978-90-6299-237-9