Original Article

Congenital cholesteatoma of the middle ear – a report of 10 cases

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Abstract
Objectives To study clinical, imaging features and treatment outcomes of congenital cholesteatoma of middle ear (CCME). Methods This is a retrospective review of 10 CCME cases selected from 952 cholesteatoma cases treated between January 1995 and December 2005 at the Department of Otolaryngology-Head and Neck Surgery, Chinese PLA General Hospital. The main outcome measures were the site of origin, clinical features, surgical findings, imaging characteristics and hearing results. Results The mean age of the 10 patients was 16 years (ranged from 10 to 24 years), with 6 being older than 18 years. There were 7 males and 3 females. The average delay to diagnosis was longer than 2 years. The mean preoperative PTA was 55 dB HL, with a mean ABG of 45 dB. Typical cholesteatomas were seen behind the tympanic membrane in the superoposterior quadrant on otoscopy only in 2 patients. High resolution CT was completed in all patients. Most of the patients (8/10) were diagnosed with otosclerosis or ossicular abnormality before operation. All patients underwent a one-stage tympanoplasty following transmeatal explorative tympanotomy and complete cholesteatoma removal, except one, who underwent a CWU mastoidectomy due to extensive cholesteatoma involvement. The cholesteatoma lesion was confined to the superoposterior mesotympanum in all patients. The mean postoperative PTA was 20 dB HL. All patients were followed-up for at least 1.5 years postoperatively. Revision procedures were performed in 2 patients for hearing deterioration. No residual or recurrence of cholesteatoma was found. Conclusion CCME is a rare disease that often gets delayed diagnosis. Residual lesions and the prognosis mainly depend on the extent of the lesion.

Key words congenital cholesteatoma, middle ear, surgery

Introduction

Congenital cholesteatoma of middle ear (CCME) is a rare pathology that can go undiagnosed for years. This article reports 10 cases of CCME focusing on lesion location, clinical presentation, peri-operative evaluations, surgical techniques, and outcomes.

Patients and methods

The 10 CCME cases were identified among 952 cholesteatoma cases treated between January 1995 and December 2005 at the Department of Otolaryngology-Head and Neck Surgery, Chinese PLA General Hospital. The cases were identified based upon modified criteria of Levenson et al., i.e. 1) no history of otorrhea, tympanic membrane perforation, or otologic procedures; 2) normal tympanic membrane on otoscopy; and/or 3) a pearly white mass medial to the tympanic membrane.

Each patient underwent thorough examination of the ear, nose and throat. Hearing was assessed using a pure tone audiometry (GSI61, Madsen, Denmark) before and after the operation. Hearing outcomes were determined using mean residual air-bone gap, based on the difference between air and the bone conduction thresholds at 250, 500, 1000, 2000, 4000 and 8000 Hz. Other assessments included impedance tympanometry (GSI33 version 2 Middle ear analyzer, Granson-Stadler Inc., USA), ABR(Nicolet-Bravo, Nicolet Biomedical, USA) and High-resolution computed tomography (HRCT) scanning.

Surgical approaches were selected based upon CT findings, hearing test results, location of the cholesteatoma and integrity of the ossicular chain. Surgical procedures started with transmeatal exploratory tympanotomy in all 10 patients. Following removal of the lesion, the integrity of the ossicular chain was examined...
and ossiculoplasty was performed with PORT(type II), TORP(type III) or allogenic/autogenous ossicles grafts, as appropriate.

Results

The mean patient age was 16 years (ranging from 10 to 24 years), with 6 being older than 18 years. There were 7 males and 3 females(2.3:1). Average disease duration before diagnosis was 2 year or longer in more than half of the cases. The right ear was involved in 4 cases and left ear in 5 cases. Bilateral involvement was seen in 1 patient.

Most patients had no ear complaints other than hearing loss(10/10) and tinnitus(2/10) at the time of presentation. There were no intracranial complications, vertigo or facial paralysis. The tympanic membrane appeared normal in 8 patients and signs indicative of cholesteatoma were visible on otoscopy in 2 patients (Fig.1).

The mean pre-operative PTA was 55 dB HL, with a mean air-bone gap of 45 dB(Fig. 2). Type “A” tympanograms were seen in 7 cases and type “Ad” in 2 cases. Absolute wave I latencies on ABR were prolonged in all cases, consistent with conductive hearing loss.

HRCT scans were completed in all 10 patients. Lesions were characterized by a homogeneous mass of soft-tissue density in the middle ear space, similar to that of an acquired cholesteatoma except that the lesion was mainly limited to posterior mesotympanum(Fig. 3). Eight of these lesions were predominantly confined in the middle ear space. The rest two showed infiltration in the antrum and epitympanum. The mastoid remained pneumatic in most of these cases.

Otosclerosis or ossicular malformation was the working diagnosis in 8 cases before surgery. A definite diagnosis of cholesteatoma was established following explorative tympanotomy in all cases.

One-stage close typanoplasty was performed in 9 cases following explorative tympanotomy and complete cholesteatoma removal. CWU mastoidectomy was the choice in one case due to extensive cholesteatoma involvement.

Epitympanum and antrum involvement was uncommon in this series. Lesion extension into the attic below the ossicles was seen in only 2 cases and the origin could not be determined. In the rest cases, a well-encapsulated cystic lesion limited to the posterior mesotympanum was typically seen. These lesions seemed to arise from the middle ear, especially the

![Fig 1](image1.jpg)

**Fig 1.** Otoscopic view of the CMCC in case 1. The cholesteatoma is located in posterior superioer mesotympanum and seen behind tympanum.

![Fig 2](image2.png)

**Fig 2.** Before and after operation audiograms in case 10. The post-operative air-bone gap is reduced to 30 dB (red line) from 53 dB before operation (blue line)

![Fig 3](image3.png)

**Fig 3.** High resolution CT scan of CCME in a 24 years old man with conductive hearing deficit (case 10). (3a) Axial section showing abnormal soft-tissue debris in posterior mesotympanum (arrow). (3b) Axial section showing implanted Teflon artificial stapes and no residual or recurrence of disease at 1 year after surgery.
incostapedial joints. Middle ear mucosa was nearly normal in 7 cases, and the Eustachian tube was normal in all the cases. The incus (10/10) and stapes (8/10) were often eroded, especially at the incostapedial joints, whereas the handle of the malleus was involved in only 2 cases. Exposure of the facial nerve was seen in 2 cases. Lateral semicircular canal erosion was seen in 1 patient.

All patients underwent ossicular chain reconstruction. Materials used for ossicular reconstruction included PORT (type II, n = 1), TORP (type III, n = 6), autogenous incus (type III, n = 1), allogeneic malleus (type III, n = 1), and artificial stapes (n = 1) (Table 1).

All patients were followed-up postoperatively for at least 1.5 years with an interval of 12 ~ 24 months. The postoperative air-bone gap was 20 dB or less. The middle ear was revisited in 2 patients at 1 and 6 years in face of continuous hearing decline. No residual or recurrence of disease was found on high resolution CT scans or during second-look operation in these 2 cases. The causes of hearing loss were polyps and ossicular prosthesis dislocation.

### Discussion

Temporal bone histopathologic studies have demonstrated two distinct types of congenital cholesteatoma[3]. One type appears to be cystic and localized of anterior mesotympanum, in which ossicular lesions are uncommon. These cholesteatomas are most commonly documented and diagnosed in very young children (mean age, 4 years) at a systematic evaluation, and can be removed easily. Hearing loss is usually minimal (<20 dB HL)[5]. The other type is of a more diffuse, open form, in which cholesteatoma matrix forms part of the middle ear mucosa. It most often invades the superoposterior tympanum but progressively concerns the epitympanum and the posterior tympanic cavity. The ossicular chain is often eroded, mainly at incostapedial joints, inducing a consistent conductive deafness that leads to diagnosis in older children (over age 5) and adults[3]. Hearing loss is often more severe (>30 dB HL)[4].

CCME is thought to be a rare condition in the clinic, but the incidence (4% to 24%) is probably underesti-

### Table 1. Patients Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age(y)</th>
<th>Gender</th>
<th>Ear</th>
<th>Type</th>
<th>Site</th>
<th>Preoperative PTA(dB HL)</th>
<th>CT Finding</th>
<th>Procedure Performed</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10</td>
<td>M</td>
<td>R</td>
<td>Closed</td>
<td>Post/superior</td>
<td>53</td>
<td>Normal</td>
<td>Ossiculoplasty Type III (TORP)</td>
</tr>
<tr>
<td>2</td>
<td>14</td>
<td>F</td>
<td>B</td>
<td>Open</td>
<td>Unclear</td>
<td>43</td>
<td>Normal</td>
<td>Ossiculoplasty Type III (autogenous incus)</td>
</tr>
<tr>
<td>3</td>
<td>15</td>
<td>M</td>
<td>L</td>
<td>Closed</td>
<td>Post/superior</td>
<td>55</td>
<td>incus erosion, mass inside ossicular</td>
<td>Ossiculoplasty Type III (TORP)</td>
</tr>
<tr>
<td>4</td>
<td>18</td>
<td>F</td>
<td>L</td>
<td>Closed</td>
<td>Post/superior</td>
<td>40</td>
<td>Small density at lateral canal</td>
<td>Ossiculoplasty Type III(TORP)</td>
</tr>
<tr>
<td>5</td>
<td>19</td>
<td>M</td>
<td>R</td>
<td>Closed</td>
<td>Post/superior</td>
<td>70</td>
<td>Mass in posterior and superior tympanum</td>
<td>Ossiculoplasty Type III (TORP)</td>
</tr>
<tr>
<td>6</td>
<td>19</td>
<td>M</td>
<td>L</td>
<td>Open</td>
<td>Unclear</td>
<td>60</td>
<td>Normal mastoid, small density at IS joint</td>
<td>Ossiculoplasty Type III (PORP)</td>
</tr>
<tr>
<td>7</td>
<td>20</td>
<td>M</td>
<td>R</td>
<td>Closed</td>
<td>Post/superior</td>
<td>48</td>
<td>Mass in mesotympanum and atic</td>
<td>Ossiculoplasty Type III (TORP)</td>
</tr>
<tr>
<td>8</td>
<td>20</td>
<td>F</td>
<td>L</td>
<td>Closed</td>
<td>Post/superior</td>
<td>60</td>
<td>Soft-tissue in posterior mesotympanum</td>
<td>CWU with Ossiculoplasty Type III (allogeneic malleus)</td>
</tr>
<tr>
<td>9</td>
<td>22</td>
<td>M</td>
<td>L</td>
<td>Closed</td>
<td>Post/superior</td>
<td>50</td>
<td>Ossicular erosion,</td>
<td>Ossiculoplasty Type III (TORP)</td>
</tr>
<tr>
<td>10</td>
<td>24</td>
<td>M</td>
<td>R</td>
<td>Closed</td>
<td>Post/superior</td>
<td>70</td>
<td>Soft-tissue in posterior mesotympanum</td>
<td>Ossiculoplasty (artificial stapes)</td>
</tr>
</tbody>
</table>

R: Right, L: Left, B: Bilateral
mated, because the diagnosis is usually made only when the tympanic membrane has been perforated by the progressively growing cholesteatoma. Of the 952 cholesteatoma cases diagnosed in the authors’ practice, only 10 are congenital. Although the presentation of some other cases may suggest a congenital origin, they do not meet the Levenson et al criteria.

In this series, the most common site of cholesteatoma was in the posterior mesotympanum around the incostapedial joint and no involvement of the anterior tympanum was found. This is different from those reported by Levenson et al and Cohen, but consistent with findings by Karmarkar et al.

Regardless of the embryological origin, which may differ in individual cases, the so-called congenital cholesteatoma has to meet the very strict criteria defined by Derlacki and Clemis in 1965 and revised by Levenson et al. Many believe that these cholesteatomas behave in a more aggressive manner than does acquired cholesteatoma. This opinion is in general based on the frequent finding of more extensive invasion in a normally pneumatized temporal bone, ossicular erosion, and labyrinthine lesions at initial surgery.

In the clinic, the diagnosis of CMCC is often delayed for years. This is especially true for those originating from the posterior mesotympanum, whose progression is gradual and silent, with hearing loss being the only symptom, which is often compensated by the normal ear. All cholesteatomas in the present study arose from the posterior mesotympanum with no symptoms during the early stage of mesotympanum involvement. The patients did not seek medical attention until the interruption of the ossicular chain that resulted in notable hearing loss. In this group, the average delay in diagnosis was more than 2 year, longer than 4 year in half of the patients with the longest being 13 years.

In contrast to CCME in the posterior tympanum, those originating from the supraanterior quadrant of the middle ear tend to affect function of the Eustachian tube, leading to pseudo – otitis media with effusion. Early diagnosis is made when myringotomy is performed in these cases.

Clinical presentation of CMCC includes impaired hearing, tinnitus, facial palsy, ear discharge and vertigo. Some cases are incidentally diagnosed. The main complaints in our series were deafness (10/10) and tinnitus (2/10), with no facial palsy. Hearing loss was conductive in all the cases. Hearing loss was progressive in 3 cases, stable in 7 cases and incidental findings in 5 cases. In 2 cases, hearing loss was noticed after head shaking or trauma.

The association of congenital cholesteatomas with other malformations is well documented, occurring in almost 15% of patients. In this study, congenital cholesteatomas associated with malformation of the auricle, external auditory canal or middle ear are not included. The incidence of ossicular malformations is higher in some series (25% to 33%) than others, and seems higher than that of the general population, suggesting a genetic factor in congenital cholesteatoma.

In treating CCME, a planned second look operation is not advocated. Most prefer to follow patients clinically and radiologically with HRCT scans, performing a second surgical procedure only in cases of functional failure or suspicion of relapse. A 12 to 18 month period is the commonly reported interval for follow-up, but some have recommended a shorter interval (8 to 12-month) because of the particular aggressiveness of this disease and the need for functional rehabilitation. We performed second look procedures in 2 of our cases at 1 and 6 years after surgery, because of continuing hearing decline. No cholesteatoma recurrence was found and the causes of hearing loss in these 2 cases were polyps and prosthesis dislocation.

The rate of residual lesions and the prognosis in CCME are mainly dependent on the initial extension of the lesion. Rosenfeld et al suggest that residual lesions in CCME are not less common than in acquired cholesteatoma, and they do not consider congenital cholesteatomas to have a better prognosis than acquired cholesteatomas. When a congenital cholesteatoma is cystic and limited, the risk of residual lesion is low, but the risk can range from 20% to 52% for those with extensive invasion.

References


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