Acquired cholesteatoma in children: Clinical features and surgical results

Olfa Ben Gamra, Wafa Abid, Ines Nacef*, Nadia Romdhane, Ines Hariga, Chiraz Mbarek

Department of Otorhinolaryngology, Head and Neck Surgery, Habib Thameur Hospital, Tunisia

Received 8 July 2015; accepted 27 September 2015

Abstract Acquired cholesteatoma in children is an aggressive disease due to its rapid growth and high recurrence rate.

Objective: To assess clinical features of cholesteatoma in children and evaluate our experience in the surgical management of this disease.

Methods: Forty children aged 5–16 years operated on for acquired middle ear cholesteatoma (2002–2011), were included in our study. Surgery was bilateral for 3 patients, which makes a total number of 43 operated ears. Functional and anatomical results were evaluated after a minimum follow-up of 3 years.

Results: Mean age at the first operation was 11.7 years. Canal wall-up tympanoplasty (CWUT) was performed as first-line procedure in 74% of cases. Canal wall-down tympanoplasty (CWDT) was performed in 26% of cases when cholesteatoma was hardly controllable with a CWUT. Cartilage graft was used in all cases for eardrum reinforcement. After a mean interval of 12.2 months, second look procedure aimed to verify a residual cholesteatoma in 8 cases and perform an ossiculoplasty in one case. Total cholesteatoma recurrence rate was 18% with CWUT and 9% with CWDT ($p = 0.4$). Predictive factors for recurrence were: children < 10 years ($p = 0.02$) and ossicular chain resorption ($p = 0.04$). Post-operative ABG $\leq 25$ dB was seen in 53% of CWUT against 18% of CWDT ($p = 0.04$).

Conclusion: CWUT is the optimal management technique offering better hearing results, however, prevention of recurrences are considered to be better with CWDT.

CrossMark

1. Introduction

Acquired cholesteatoma in children is an aggressive disease due to its rapid growth and high recurrence rate. Surgical management aims to eradicate the disease process, prevent...
reurrences and preserve auditory function thus preventing learning difficulties. The choice of the adequate surgical technique, either canal wall down tympanoplasty (CWDT) or canal wall up tympanoplasty (CWUT) must be determined for each individual case according to the extent of the cholesteatoma. CWUT technique has gained favor among most surgeons for treating cholesteatomas in the pediatric population. It is currently considered to be the reference procedure when it is technically feasible.

1.1. Objective

To assess clinical features of cholesteatoma in children and evaluate our experience in the surgical management of this disease.

2. Methods

We present a retrospective descriptive study (January 2002–December 2011) reviewing a total of 40 children aged 5–16 years, operated on for acquired middle ear cholesteatoma in our ENT Department.

Preoperatively, all patients had temporal bone tomodensitometry and tonal audiometry. Selection of the applied surgical technique (CWUT or CWDT) was made according to pre- and intraoperative ascertainment. CWUT was indicated as a first-line procedure for patients with a large well pneumatized mastoid and a well-controlled disease. CWDT was performed, depending on anatomical conditions, when cholesteatoma was hardly fully controllable with a CWUT technique. A planned second look surgery was indicated after a mean interval of 12 months to rule out a residual cholesteatoma or perform an ossiculoplasty if needed. Patients were operated on by four different surgeons.

The results were evaluated in terms of hearing improvement and recurrence rate. Preoperative audiometry and the last available audiometry were analyzed with calculation of air conduction (AC), bone conduction (BC) thresholds and air-bone gap (ABG) by averaging thresholds at 500 Hz, 1 kHz and 2 kHz.

Statistical analysis was performed using SPSS-19. We conducted a descriptive study of quantitative and qualitative variables, as well as univariate Chi-square study of qualitative variables. Statistical significance was assigned to a p-value of 0.05.

3. Results

Middle ear surgery was bilateral for three patients which makes a total number of 43 operated ears. For all cases, primary surgery was done in our institution with a minimum follow-up period of 3 years.

Mean age at the first operation was 11.7 years ranging from 5 to 16 years with a male predominance (sex-ratio of 2.3). A history of serous otitis media was observed in 2 cases. Among the cases of bilateral cholesteatoma, two had a cleft palate. There was no history of eardrum trauma.

The main symptoms reported by our patients were chronic otorrhea (93%) and hearing loss (72%), evolving for a mean duration of 38 months (Fig. 1). Four children presented a complication revealing the disease: acute mastoiditis (2 cases), facial palsy (1 case) and dizziness due to a labyrinthine fistula (1 case).

Preoperative otoscopy revealed cholesteatoma in 69%, attic perforation in 51%, pars flaccida retraction in 25% and an attic polyp in 25% (Table 1).

Abnormalities in the contralateral ear were observed in 11 ears (25%): attic cholesteatoma (3 cases), attic retraction (4 cases), adhesive otitis (2 cases) and tympanic perforation (2 cases). Nasal endoscopy showed a non-obstructive septal deviation in 3 cases and adenoid vegetations in 7 patients.

Preoperative audiometry revealed a mean air conductive hearing loss of 40 dB in 32 cases. A mixed hearing loss was found in 5 cases, with a mean threshold of 50 dB and a mean ABG of 40 dB. Auditory function was normal in 6 patients.

On CT scan, cholesteatoma extended to the epitympanum and the antrum in most cases (79%) (Table 2). Ossicular chain was complete in 25% (n = 11) and eroded in 75% (n = 32). The incus was the ossicle most frequently lysed (84%), followed by the malleus (55%) and the stapes (32%). CT also revealed complications including a facial canal lysis in 2 cases, a labyrinthine fistula in one case and a protruding jugular bulb in another.

Intra-operatively, sites involved by cholesteatoma are detailed in Table 3. Ossicular chain was disturbed in 38 cases (88%). The ossicle most commonly destroyed was the incus (86%), followed by the malleus (53%) and the stapes (46%).

Among our patients, 35 underwent a single operation, 4 were operated twice and one had a third operation. The mean interval between the first two procedures was 12.2 months.

CWUT is the preferred operative technique in our center, performed in 74% of cases (n = 32). It was indicated for patients with a large well pneumatized mastoid and a well-controlled disease. It was combined with a mastoidectomy in 24 cases (75%) and a posterior tympanotomy in 2 cases (6%).

CWDT was performed in 26% of cases. It was advocated to the following findings: lateral sinus protrusion (6 cases), meningeal protrusion (3 cases) and extension of cholesteatoma to the retrotymanum (5 cases). Considering these anatomical conditions, cholesteatoma was hardly controllable with a CWUT technique.

One child had a large mesotympanic cholesteatoma extending to the protymanum, responsible for a labyrinthine fistula of the lateral semicircular canal (Fig. 3). A CWDT helped to control the disease, remove the matrix completely and seal the fistula.

Figure 1 Main symptoms reported by patients.
Second look procedure was planned for 9 patients operated on by CWUT after a mean delay of 12.2 months [11–18 months]. It aimed to verify a residual cholesteatoma in 8 cases and to perform an ossiculoplasty in one case. Ossicular reconstruction was possible in 37% (n = 16) during the first-line procedure and was deferred until the second procedure in one case. Ossicles were removed during the first procedure in 46% of cases (incus in 32% and head of malleus in 14%). Reconstruction was carried out by means of columelization with shaped autologous ossicles or cartilage. It consisted of incus transposition in 9 cases, cartilage graft onto stapes in 6 cases, and titanium total ossicular replacement prosthesis in 1 case.

For eardrum and canal wall reconstruction, we used total cartilage in all cases: conchal cartilage in 81% and tragal cartilage in 19%.

After a mean follow up period of 44 months [3–10 years], the total cholesteatoma recurrence rate (residual and recurrent) was 16% (n = 7). This rate was not statistically influenced by the surgical technique (Table 4).

Residual disease was detected in 2 cases (CWUT and CWDT) after a mean delay of 12 months. All recurrences (n = 5) occurred with CWUT after a mean delay of 22 months requiring subsequent CWDT in 3 cases. These recurrences concerned an attic cholesteatoma extended to the atrium and the mastoid. Intraoperatively, we found a cholesteatoma invading the window region (2 cases), a lateral sinus protrusion (2 cases) and a meningeal protrusion (1 case). For the latter, a second recurrence occurred after a mean period of 3 years treated by CWDT.

Recurrence occurred more frequently in children < 10 years, where 80% had recurrence against 28% in older children (p = 0.02). Also, all patients with recurrent disease had ossicular chain resorption against 55% with no recidivism (p = 0.05).

Hearing failure was seen in 11% with a mean loss of 25 dB. Hearing improvement was observed in 56% with a mean gain of 15 dB. Auditory function was stable in 33%. Post-operative ABG ≤ 25 dB was seen in 41% of cases.

According to the surgical procedure, the mean final AC hearing loss was 8.59 dB in the CWUT group and 20 dB in the CWDT group (Fig. 2). A post-operative ABG ≤ 25 dB was observed in 53% of CWUT against 18% of CWDT (p = 0.04) concluding to better functional results with CWUT.

There were no cases of sensorineural hearing loss or facial nerve injury after surgery in our series. However, one patient developed post-operative infection and another presented a meatal stenosis due to meatoplasty infection. In both cases, infection resolved without sequelae after adequate antibiotherapy.

4. Discussion

Retraction pocket of the tympanic membrane, resulting from eustachian tube dysfunction is the commonest cause of acquired cholesteatoma in children. These pockets usually develop in the pars flaccida or the upper posterior part of the pars tensa. Most authors believe that cholesteatoma is more aggressive in children than in adults. Bujia analyzed the expression of MIB1 (a monoclonal antibody marker of cell proliferation) in child and adult cholesteatoma and found a higher proliferation rate in children. Mobeen also confirmed a significantly higher proliferative rate of the cholesteatoma matrix in this group. In addition, Dornelles showed that pediatric specimens expressed higher levels of matrix metalloproteinases and exhibited an exaggerated inflammatory profile.

Temporal bone CT assesses cholesteatoma extension, explores complications and detects anatomic variants before surgery. Nevertheless, cholesterol granuloma, frequently associated with cholesteatoma, cannot be differentiated on CT. MRI is indicated for initial extension assessment, in case of meningeal contact and during follow-up when CT scan is doubtful.

---

### Table 1: Otoscopy findings.

<table>
<thead>
<tr>
<th>Retraction pocket:</th>
<th>Number</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shrapnel membrane</td>
<td>11</td>
<td>25</td>
</tr>
<tr>
<td>Posterior</td>
<td>3</td>
<td>7</td>
</tr>
</tbody>
</table>

### Table 2: Cholesteatoma extension on CT scan.

<table>
<thead>
<tr>
<th>Tomodensitometry findings</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mesotympanum + epitympanum</td>
<td>34</td>
<td>79</td>
</tr>
<tr>
<td>Mesotympanum + mastoid</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Mesotympanum</td>
<td>4</td>
<td>9</td>
</tr>
<tr>
<td>Mesotympanum + mastoid + retrotympanum</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Mesotympanum + window region + retrotympanum</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td>100</td>
</tr>
</tbody>
</table>

### Table 3: Cholesteatoma extension: intraoperative findings.

<table>
<thead>
<tr>
<th>Cholesteatoma extension</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Attic</td>
<td>6</td>
</tr>
<tr>
<td>Attic + mesotympanum</td>
<td>9</td>
</tr>
<tr>
<td>Attic + hypotympanum</td>
<td>1</td>
</tr>
<tr>
<td>Retrotympanum/stapes/window region</td>
<td>12</td>
</tr>
<tr>
<td>Attic + mastoid</td>
<td>11</td>
</tr>
<tr>
<td>Attic + mesotympanum + mastoid</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
</tr>
</tbody>
</table>

### Table 4: Rates of recurrence and residual disease according to the surgical technique.

<table>
<thead>
<tr>
<th></th>
<th>Recurrence</th>
<th>Residual</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>CWUT (32)</td>
<td>5 (15%)</td>
<td>1 (3%)</td>
<td>0.4</td>
</tr>
<tr>
<td>CWDT (11)</td>
<td>0</td>
<td>1 (9%)</td>
<td></td>
</tr>
</tbody>
</table>
As pediatric cholesteatoma often involves the entire mastoid and mesotympanum, surgery is more difficult than in adults and results are considered to be poorer. It aims to eradicate the disease, preserve or improve hearing and prevent recurrence or residual disease. CWUT is the preferred operative technique in our center and for most authors. It is an adequate surgical option for treating most acquired cholesteatomas and is currently considered to be the reference procedure when it is technically feasible. This technique allows the use of post-operative hearing aids, rapid healing, water exposure and preserves the anatomy of the middle ear. This approach is widely performed in children also because of their difficulty to manage an open mastoid cavity. In our study, the canal wall was preserved in 74% of cases.

Currently, the main recent advances are represented by the generalized use of total cartilage graft for tympanoplasty in the pediatric population. This material should be used for eardrum reinforcement and also canal wall reconstruction, given the preponderance of recurrence and retraction pockets in this age group. The problem encountered with cartilage is failure to control retrotympanic liquid effusion. The use of otoendoscopy is of great value in pediatric middle ear surgery. It has advantages of visualization of hidden areas such as: sinus tympani, facial recess, anterior epitympanic space, attic, hypotympanum, medial epitympanum and retrotympanum. For some authors, if the disease is limited to the middle ear and entirely accessible through the ear canal, it can be removed via an endoscopic permeatal atticoantrostomy, thus avoiding the disadvantage to the child of an external incision. For Adrian, tragus cartilage is the optimum graft for endoscopic ear surgery because the incision for access can be hidden on the posterior surface.

On the other hand, CWDT is reserved for large cholesteatoma that cannot be adequately removed by CWUT, children with a small mastoid, limited air cells or meningeal prolapse and patients with poor follow up. It is associated with a lower recurrence rate, varying from 13% to 17%. However, the mastoid cavity is more prone to otorrhea, requires regular cleaning and causes difficulties in applying hearing aids.

Recurrences are more often seen in children than adults and are most likely to be found in the attic and mesotympanum. Predictive factors for recurrence retained by many authors are: children <8 years, ossicular lysis especially stapes superstructure, large cholesteatoma, eustachian tube dysfunction and round window or sinus tympani involvement. Furthermore, Nelson reported that the cholesteatoma involving the posterior superior quadrant or the mastoid recurred more frequently than the cholesteatoma in anterior mesotympanum. Although many authors have shown that the choice of the initial surgical technique does not significantly modify the control of the disease, Drahy reported all recurrences with CWUT (Table 5).

Second look surgery is still controversial. For Schraff, staging is systematically done at approximately 6–12 months after first surgery to assess for recurrent cholesteatoma and ossicular chain reconstruction if needed. For Soldati, second surgery was planned for poor hearing results either because the ossicular chain reconstruction had not been possible at first surgery or because hearing has worsened. Recently, intraoperative use of endoscopes to reduce residual disease rates, as well as the use of MRI has the potential to reduce the need for second-look surgery.

In our institution, second look operation was planned for 9 children operated on by CWUT after a mean delay of 12.2 months. This procedure was performed to rule out a residual cholesteatoma in 8 cases evaluated by a diffusion-weighted MRI, and to perform an ossiculoplasty in one case.

Recurrence prognosis seems to depend not only on the surgical technique but also on the disease extension, the condition of

<table>
<thead>
<tr>
<th>Table 5 Recurrence rate of cholesteatoma.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Authors</td>
</tr>
<tr>
<td>Drahy</td>
</tr>
<tr>
<td>Gocmen</td>
</tr>
<tr>
<td>Soldati</td>
</tr>
<tr>
<td>Schraff</td>
</tr>
<tr>
<td>Hajri</td>
</tr>
<tr>
<td>Our series</td>
</tr>
</tbody>
</table>
the middle ear mucosa and the stapes superstructure. Indeed, many authors found that patients with CWDT had less improvement post-operatively compared with children with the CWUT approach. This was the case in our series (Table 6).

5. Conclusion

The optimal management of pediatric cholesteatoma remains challenging because of the high recidivism compared with adults. According to this study, children aged < 10 years and erosion of the ossicular chain were at risk of recurrence. CWUT is the optimal management technique offering better hearing results, however, prevention of recurrences are considered to be better with CWDT.

Conflict of interest

None.

References


