Acquired Cholesteatoma in Pediatric Population: Clinical Features and Surgical Results

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Abstract

Introduction: Pediatric cholesteatoma is known to be more aggressive than in adults due to its rapid growth and the frequency of recurrences. Surgical treatment in children and adolescent has been a challenge to many surgeons. The effects of deafness can have social, educational and behavioral consequences, thereby necessitating the need for hearing preservation. Materials and methods: This is a retrospective study of 74 patients who underwent surgery for cholesteatoma in the ENT and CCF departments of the specialty hospital in Rabat, over a period of eight years between 2011 and 2018. Results: In eight years, 74 patients were treated for cholesteatoma, with a sex ratio of 1.46. The average age ranged from 6 to 16 years. 12 patients showed signs of complications. Otorrhoea and perforation of the tympanic membrane in 29 patients and lateral sinus thrombosis in four patients, peripheral facial paralysis in two patients and mastoiditis in five patients, brain abscess and meningitis in four patients, peripheral facial paralysis in two patients and lateral sinus thrombosis. Discussion and Conclusion: Eradication of cholesteatoma and restoration of hearing function present unique surgical challenges. An individualised approach is needed, and the choice of surgical technique should be based on anatomical, biological, radiological and social factors.

Keywords: Cholesteatoma, otoscopic examination, surgery, recurrence, hearing outcomes.

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Introduction

Cholesteatoma is defined as the presence of keratinising squamous epithelium within the middle ear, or in other pneumatised areas of the temporal bone. This keratinising epithelium exhibits independent growth, leading to expansion and to resorption of underlying bone. Its potential for causing central nervous system complications makes it a potentially fatal lesion.

Pediatric cholesteatoma is known to be more aggressive than in adults due to its rapid growth and the frequency of recurrences. Surgical treatment in children and adolescent has been a challenge to many surgeons. Various surgical procedures have been used to eradicate cholesteatoma: the open and closed methods. In children, the effects of deafness can have social, educational and behavioral consequences, thereby necessitating the need for hearing preservation.

Materials and Methods

This is a retrospective study of 74 patients who underwent surgery for cholesteatoma in the ENT and CCF departments of the specialty hospital in Rabat, over a period of eight years between 2011 and 2018.

Results

In eight years, 74 patients were treated for cholesteatoma, with a sex ratio of 1.46 (30/44). The average age ranged from 6 to 16 years. All 74 cases had a long-term history of otitis media such as otorrhoea and hearing loss. 12 patients showed signs of complications: mastoiditis in five patients, brain abscess and meningitis in four patients, peripheral facial paralysis in two patients and lateral sinus thrombosis. Otorrhoea examination revealed retraction pocket in 36 patients, perforation of the tympanic membrane in 29 patients, perforation of the tympanic membrane in 29 patients, and lateral sinus thrombosis in four patients.
and aural polyp in 9 patients. Pure tone audiometry showed a moderate conductive hearing loss ranging from 40 to 65 decibels in 41 patients. Computed tomography with thin cuts of the temporal bone detected soft tissue mass-like opacity in the middle ear cavity and mastoid antrum associated with erosion of the ossicles and pressure erosion of adjacent structures.

<table>
<thead>
<tr>
<th>CT scan</th>
<th>N</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Soft tissue density in the middle ear</td>
<td>74</td>
<td>100%</td>
</tr>
<tr>
<td>Expansion of the aditus and mastoid antrum</td>
<td>74</td>
<td>100%</td>
</tr>
<tr>
<td>Osseous chain erosion</td>
<td>61</td>
<td>82.4%</td>
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<tr>
<td>Eroded scutum</td>
<td>74</td>
<td>100%</td>
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<tr>
<td>Eroded Tegmen</td>
<td>14</td>
<td>18.9%</td>
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<tr>
<td>Eroded facial nerve canal</td>
<td>5</td>
<td>6.75%</td>
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<tr>
<td>Eroded lateral semi circular canal</td>
<td>2</td>
<td>2.7%</td>
</tr>
<tr>
<td>Brain abscess</td>
<td>2</td>
<td>2.7%</td>
</tr>
<tr>
<td>Lateral sinus thrombosis</td>
<td>1</td>
<td>1.35%</td>
</tr>
</tbody>
</table>

58 patients (78.37%) underwent canal wall up mastoidectomy (CWU). Canal wall down mastoidectomy (CWD) were performed in 16 patients (21.62%). The cholesteatoma was found to be diffuse in 57 cases (77%) and encapsulated in 17 cases (23%). Ossicles were found to be eroded in 68 patients (92%). No labyrinthine fistulae were detected in 31 patients with recurrences (41.89%). Pre- and postoperative audiograms were compared. Postoperatively, The mean air conduction improved significantly from 46.73 dB to 38.53 Db. No patients developed a dead ear after surgery.

**DISCUSSION**

There are two types of cholesteatoma, congenital and acquired, based on the pathogenesis of the lesion. Congenital or primary cholesteatoma refers to squamous epithelium medial to an intact tympanic membrane without a history of significant otitis media or Eustachian tube dysfunction. The more common type of cholesteatoma results as complication of chronic otitis media, that is, acquired or secondary cholesteatoma [1].

Incidence is hard to determine, especially for congenital cholesteatoma. Recent figures testify to a fall in the incidence of both adult and childhood acquired cholesteatoma: in 1925, the rate of cholesteatoma in the under-16's was one in three; it is now much less frequent, but incidence in the early 2000s was still three per 100,000 (compared to nine per 100,000 in adults) [2].

The main objectives of management of cholesteatoma are eradication of the disease and hearing rehabilitation [3]. Temporal bone CT assesses cholesteatoma extension, exploring for complications and detecting anatomic variants so as to prepare for any difficulties in surgery. Diagnosis is normally clinical and otoscopic. One essential issue in CT is radioprotection, especially in children presenting with chronic otitis media, requiring numerous scans over their lifetime. This mainly concerns the crystalline lens, and the risk of radio-induced cancer [4].

Middle-ear surgery is hampered in children by obstruction of the Eustachian tube and chronic inflammatory hyperplastic mucosa, increasing the risk of incomplete resection and residual lesion [5]. Cartilage should be used for eardrum reinforcement in children, whereas the temporal aponeurosis is preferred in adults. Posterior reinforcement is essential, given the preponderance of recurrence and retraction in this area of the tympanum. The problems encountered with cartilage are failure to control retrotypanic liquid effusion, and possible residual lesion [6].

Cholesteatoma in children is reputed to be more aggressive than in adults. Bujaia 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 [7]. For example, De Corso et al. reported a recurrence rate of 26.6% in children versus 6.8% in adults and more frequent ossicular lesions in children [8]. Charachon et al. reported residual disease in 31% of children after CWUT versus 16% in adults [9].

Hearing prognosis seems to depend not only on the surgical technique but also on the disease extension, the condition of the middle ear mucosa and the stalks superstructure. Indeed, many authors found that patients with CWDT had less improvement postoperatively compared with children with the CWUT approach [10].

**CONCLUSION**

The ideal surgical method for treatment of cholesteatoma in children remains a controversial subject. Eradication of cholesteatoma and restoration of hearing function present unique surgical challenges. An individualised approach is needed, and the choice of surgical technique should be based on anatomical, biological, radiological and social factors.
REFERENCES