Paediatric cholesteatoma: Experience of Universiti Kebangsaan Malaysia Medical Centre

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ABSTRACT

Introduction: Cholesteatoma is an aggressive disease and its management poses a greater challenge in children than in adults. This study reviews the experience of Universiti Kebangsaan Malaysia Medical Centre in the clinical presentation and management outcome of acquired cholesteatoma in paediatrics that required surgical interventions. Materials and Methods: A retrospective review of case records of patients below 18 years old who underwent surgery from 1999 to 2010. Results: A total of 46 patients presented with 53 cases of cholesteatoma in which seven patients had bilateral disease. The age of presentation ranged from four to 18 years old with a mean age of 12 years. Male and female patients were 65% and 35% respectively. Otorrhoea or previous history of otorrhoea on presentation was found in 94% and 96% of them had hearing impairment. Cerebellopontine angle abscess, sigmoid sinus thrombosis and mastoiditis were among the complications. Tympanic membrane was retracted in 64% while 47% having had attic retraction and 53% had total atelectasis. A majority (85%) underwent canal wall down surgery with or without tympanoplasty. Post-operatively, 71% had improvement or preserved hearing level. The duration of follow up ranged from one month to 13 years and a quarter had recurrent disease and underwent revision surgeries. Conclusion: Majority of the cholesteatoma patients suffered from hearing loss and otorrhoea. Tympanic membrane retraction remained the most common clinical finding. Hence, children with persistent otorrhoea after adequate treatment may represent cholesteatoma. Surgical options of canal wall up and canal wall down procedures have equal risk of recurrence.

Keywords: cholesteatoma, canal wall up, canal wall down, hearing loss, recurrence

INTRODUCTION

Cholesteatoma is a condition where keratin debris produced by keratinised squamous epithelial cells accumulate in the middle ear cavity or any pneumatised part of the temporal bone. The incidence in children ranges from...
five to 15 cases per 100,000 children. Cho- 

lesteatoma may be classified as acquired or congenital. The most common cholesteatoma is the acquired type that occurs as sequelae of middle ear disease or retraction pocket. Functional obstruction of the Eustachian tube is the basis for the pathogenesis of acquired cholesteatoma in children. Abnormal functioning of the tube results in an impaired ventilation of the middle ear or mastoid air cell system resulting in high negative middle ear pressure and formation of retraction pockets at the attic of tympanic membrane. For congenital cholesteatoma, it has been suggested that the underlying pathogenesis is from remnants of foetal epithelial cell rests with growth stimulated by otitis media.

Cholesteatoma in children is widely considered to be a more aggressive disease than in the adult. The disease is usually more extensive and have a higher rate of residual or recurrence after treatment, most probably secondary to anatomic and physiologic differences.

Well pneumatised mastoids in children can lead to more extensive disease compared with more sclerotic mastoid bones in adults. Sclerotic bones are more difficult to be eroded by cholesteatoma. Therefore the management poses a greater challenge in the paediatric patient. In paediatric acquired cholesteatoma, the most common defect of the tympanic membrane begins in the postero-superior quadrant of pars tensa (marginal) and in the pars flaccida (attic). In contrast, the congenital type usually develop behind an intact tympanic membrane most commonly at the anterosuperior quadrant.

Most cholesteatoma requires surgical intervention and various types of surgery can be done. The treatment depends on the size and location of the cholesteatoma. The treatment options include atticotomy, canal wall up (CWU) mastoidectomy and canal wall down (CWD) mastoidectomy. (Please refer to supplementary text for details of the various surgeries).

In the Otorhinolaryngology Department of Universiti Kebangsaan Malaysia Medical Centre, a tertiary referral centre, the majority of cases are operated using the CWD surgical techniques. This study reviews our experience with acquired cholesteatoma in paediatric patients who required surgical interventions.

MATERIALS AND METHODS

Patients below the age of 18 years were included in this retrospective study. All cases of cholesteatoma that were managed surgically from July 1999 to December 2010 were identified from operating records. Data on clinical presentations, pre- and postoperative hearing level using pure tone average at 500, 1000 and 2000 KHz, intraoperative disease extension and management outcome of the cases were reviewed systematically. We also included patients who had been previously operated on their ear in others hospitals to observe the overall clinical features and outcome. Cases with congenital cholesteatoma were excluded from the study.

RESULTS

There were altogether 46 patients with 53 cases of cholesteatoma treated during this period. Seven patients had bilateral disease. The age of presentation ranged from four to 18 years with mean age of 12 years. There was a male predominance, 30 (65%) com-
pared to 16 (35%) female. No ethnic predilection was noted.

Most patients presented with uncomplicated otorrhoea whilst three (6%) presented otorrhoea with complications (Table 1). Other symptoms included hearing loss, otalgia and dizziness. In 34 (64%) cases, tympanic membrane were retracted, 16 (47%) with attic retraction only (Figure 1) and 18 (53%) had total atelectasis. Seven (13%) had central perforation while only four (8%) had marginal perforation. There was no documentation on the tympanic membrane in the other eight cases.

The status of pre-operative hearing level was determined based on the pure tone average at 0.5 KHz, 1 KHz and 2 KHz (Table 2). Fifty-five percent had moderate hearing loss in which a majority of them were conductive in nature. Twenty-one percent had hearing level of over 60 dB and these consisted of mixed and sensori-neural type of hearing loss. Three patients did not have any pre-operative audiometry.

Intra-operatively, most cases had extensive disease in epitympanum, mesotympanum and mastoid cavity (Table 3). Twenty four (45%) had erosion of incus/maleus but intact stapes suprastructures, 20 (38%) had erosion of incus/maleus with loss of stapes suprastructures and eight (15%) had intact ossicular chain. The ossicular chain status was not described in one case.

The follow-up duration was between one month and 13 years with a mean of 30 months. Two patients were lost to follow-up after removal of their mastoid pack. Of the eight ears with a preoperative mild hearing loss, only two had post-operative audiometry which showed worsening hearing loss; moderate hearing loss (n=1) and profound hearing loss (n=1). Of the 29 ears with preoperative moderate hearing loss, only 12 had post-operative audiometry in which the hearing improved (Table 4).

Of the patients who were followed up, 13 (25%) had recurrent disease. This consisted of 12 ears which had CWD surgery and one ear had CWU surgery prior to their disease recurrence. All underwent revision surgeries and seven still had troublesome ear discharges that required frequent review and aural toileting.
DISCUSSION

Acquired cholesteatoma are common sequelae of middle ear disease despite the widespread use of antibiotics and tympanostomy tube. In children, the most common defect of the tympanic membrane begins in the posterior superior quadrant of pars tensa and in the pars flaccida or attic. A majority of the cases in our study had attic retraction pocket and total atelactasis.

The most flaccid part of the tympanic membrane (posterior superior quadrant of pars tensa and pars flaccida areas) lose elasticity and become atelactatic in the presence of chronic high negative pressure in the middle ear, usually secondary to Eustachian tube dysfunction. Another less common defect is central membrane perforation. Based on the invagination theory, migration of normal squamous epithelium from the external canal to the retracted attic or through a perforation into the middle ear can contribute to formation of cholesteatoma. Another factor in the development of an acquired cholesteatoma may be the blockage of the inter-fold space in the middle ear, especially at the postero-superior portion of the mesotympanum, Prussak space, and tympanic isthmus.

Our study showed that the majority presented with otorrhoea and a high proportion presenting with complications was associated with low socio-economic status group and inadequacies of primary care facilities. A majority had hearing loss greater than 40 dB with the presence of sensori-neural hearing loss. Only a small number had normal hearing at diagnosis. Therefore, hearing loss is a common feature and should alert clinicians to consider cholesteatoma. Pre-operative pure tone audiometry that averages more than 20dB and air bone gap above 20 are pre-operative factors that are associated with an increased risk of cholesteatoma recurrence.

In our study, another noticeable feature was the extent of disease found intra-operatively. Based on the retraction pocket theory that suggested a shorter period of disease development in young patients, a cholesteatoma should still be confined to the attic and anterosuperior region. However the disease in our patients were more aggressive with involvements of the attic, middle ear and mastoid cavity. A possible explanation is that more metalloproteinases factors are produced resulting in more inflammation resulting in a more aggressive form of the disease compared to adults. Other possible explanations include delayed presentations and delay in diagnosis and referral from primary healthcare.

The optimal treatment for paediatric cholesteatoma is still controversial. The goal of surgery is to achieve a dry and safe ear

Table 2: Pre-operative hearing levels.

<table>
<thead>
<tr>
<th>Severity</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>2 (4)</td>
</tr>
<tr>
<td>Mild (20-40dB)</td>
<td>8 (15)</td>
</tr>
<tr>
<td>Moderate (41-60dB)</td>
<td>29 (55)</td>
</tr>
<tr>
<td>Severe (61-90dB)</td>
<td>8 (15)</td>
</tr>
<tr>
<td>Profound (≥90dB)</td>
<td>3 (6)</td>
</tr>
</tbody>
</table>

Note: Three patients hearing level results were not available.

Table 3: Intra-operative extend of disease.

<table>
<thead>
<tr>
<th>Site</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Attic/middle ear</td>
<td>5 (9)</td>
</tr>
<tr>
<td>Atticoantrum</td>
<td>11 (21)</td>
</tr>
<tr>
<td>Attic/middle ear/mastoid</td>
<td>37 (70)</td>
</tr>
</tbody>
</table>
and to improve hearing. The choice of surgery depends on the extent of the disease, surgical expertise and patient’s willingness for second look procedure and long term follow up. The treatment options include atticotomy, CWU mastoidectomy and CWD mastoidectomy. 8, 9

Management options that include either CWU or CWD continue to be debated. CWU operations result in an intact external ear canal and tympanic membrane making detection of residual disease more difficult. Recurrence rate is also higher in CWU operated group. Hence, a second look procedure is recommended in 12 to 18 months’ time. In CWD procedure, the mastoid antrum, middle ear, aditus ad antrum and attic are exteriorised to form a common cavity with external ear canal. This creates a self-cleansing cavity that may prevents recurrence. It also optimises subsequent cleaning and otologic monitoring for recurrence.

Generally, the type of surgery chosen for the management of paediatric cholesteatoma should be individualised. This should take into account anatomic, clinical and social factors. Patients who may be non-compliant with follow up and have extensive disease with large canal wall defect and/or erosion of semi-circular canal or have disease in their only hearing ear should undergo CWD procedure. 10 The type of mastoidectomy (CWU or CWD) usually do not affect the hearing outcome. 11

Besides eradicating the disease, cholesteatoma operation may improve hearing level in some patients. A study by Wetmore et al. showed that 29% had improvement in hearing levels after surgery. 12 In our series only 14 patients had post-operative audiometry; seven (50%) had improvement, three (21%) had preserved hearing level and only four (29%) had worsening of hearing level. Therefore, further study is required to confirm our findings.

Recurrence or residual disease usually takes several years to occur. Yung et al. recommended a spaced out duration of five years or longer after surgery before assessing the recidivism rate, postoperative otorrhoea and hearing outcome. 13 Regardless of techniques used, the recurrence rates for adults and children have been reported to be between seven and 57%. 14-18 The overall recurrence rate in our series was 18%, most occurring after CWD procedure. Few studies have shown that the types of procedure (i.e. CWU or CWD) was not associated with residual or recurring disease. The extent of ossicular erosions and disease in posterior mesotympanum including the oval and round windows and/or facial recess and sinus tympani have been reported to be associated with residual or recurring disease. 19, 20 In our study, 10 (77%) patients who developed disease

### Table 4: Comparison between pre-operative and post-operative hearing level in mild and hearing of moderate to profound hearing loss.

<table>
<thead>
<tr>
<th>Pre operative Hearing Level</th>
<th>Post operative Hearing Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild hearing loss</td>
<td><em>N = 2</em></td>
</tr>
<tr>
<td></td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Moderate to Profound Hearing Loss</td>
<td><em>N = 12</em></td>
</tr>
<tr>
<td></td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>7 (58%)</td>
</tr>
</tbody>
</table>
recurrence had ossicular erosion.

The limitation of this study includes small sample sizes, one centre study and that a majority of our patients presented with extensive disease. The retrospective nature of the study is also a limiting factor.

In conclusion, majority of paediatric patients with acquired cholesteatoma needing surgical interventions have hearing loss and non complicated otorrhoea. Tympanic membrane retraction remained the most common clinical finding. Patients with moderate to profound hearing loss will have a better chance of hearing improvement after surgery. Both CWU and CWD procedures have equal risk of recurrence. Long-term follow-up is important to monitor for disease recurrence. Patient should be educated regarding the nature of the disease and need for long term follow-up.

REFERENCES
17: Parisier SC, Hanson MB, Han JC, Cohen AJ, Selkin BA. Pediatric cholesteatoma: an individual-


This year’s World Kidney Day (18th March 2012) theme is ‘Donate Kidneys for Life Receive’ highlights the important role of kidney transplantation in reducing the burden of end-stage kidney disease (ESKD) such complications of ESKD, dependence and complications of renal replacement therapies, financial, social and psychological impact to patients and their families.

In Brunei Darussalam, the number of patients with ESKD is increasing but the number of patients undergoing kidney transplantation remains low at less than 7%, compared to other countries where this is approaching thirty percent. We have to do more to improve on this situation and help to improve the lives of people with ESKD and allow them to lead normal lives.