Indian J Otolaryngol Head Neck Surg
(Oct–Dec 2018) 70(4):578–582; https://doi.org/10.1007/s12070-018-1362-z

ORIGINAL ARTICLE

Congenital Cholesteatoma in Adults-Interesting Presentations and Management

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Received: 19 November 2017 / Accepted: 10 April 2018 / Published online: 13 April 2018

Abstract To report a series of adult patients diagnosed with congenital cholesteatoma (CC) with respect to symptoms, different varieties of presentation, surgical findings and approach used, complications and the post-operative results. A retrospective chart review of adult cases of CC who were treated in the period from January 2014–2017 was carried out in a tertiary care center. Levenson's criteria were used for diagnosis. Diagnosis was confirmed by imaging and intraoperatively. Postoperative results and complications were also analyzed. Six adult cases of CC were studied with a mean follow up of 10 months. Interesting presentations included otitis media with effusion, non-resolving facial nerve palsy, post aural discharge and meningitis. It included 3 cases of petrous apex cholesteatoma, 2 patients with cholesteatoma involving both the middle ear and mastoid and 1 patient with mastoid cholesteatoma. The operative procedures included canal wall up mastoidectomy (1 patient), atticotomy (1 patient), canal wall down mastoidectomy (1 patient), translabyrinthine and transotic excision of mass with blind sac closure (2 patients) and partial labyrinthectomy (1 patient). Complications encountered during surgery were cerebrospinal fluid leak and worsening of hearing in 2 patients and 1 patient respectively. CC can have variety of interesting presentations in adult population and they may or may not have the classical white mass behind the tympanic membrane. Appropriate individualized surgical planning and intervention gives good results.

Keywords Congenital cholesteatoma · Adults · Mastoidectomy · Labyrinthectomy

Introduction

Congenital cholesteatoma (CC) is relatively rare and Tos has reported an annual incidence of CC as 0.12 per 100,000 individuals [1]. CC may be diagnosed based on the criterion given by Levenson et al. [2]: (A) a whitish mass medial to the normal tympanic membrane; (B) normal pars flaccida and pars tensa; (C) no prior history of otorrhea or perforation; (D) no prior otological procedures; (E) exclusion of canal atresia, intramembranous or giant cholesteatoma. In this criterion prior bouts of otitis media were not grounds for exclusion. Peron and Schuknecht classified five sites from where CC can arise in the temporal bone: petrous apex, mastoid, middle ear, middle ear and mastoid, and external auditory canal [3].

In literature various theories have been proposed for evolution of CC like metaplasia [4], aberrant differentiation of pluripotent cells [5], misplaced epithelial tissue [6], persistence of 'epidermoid formation' [7], etc. For petrous apex cholesteatoma, Gacek [8] proposed that these arise from epithelial remnants of Seesel's pocket. Mastoid CC is said to arise from implantation of epithelium in the sutures surrounding the mastoid [9].

Many reports of pediatric cholesteatomas exist but studies in adult population are relatively less. Here we present a case series of six adults describing their interesting presenting features and also outline the management undertaken in each case.

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Methods

A retrospective chart review of all adult cases diagnosed with CC was carried out at a tertiary referral center between the period of January 2014 to January 2017. This study was approved by the institutional review board and ethics committee dated 21.06.2017, IRB min no: 10726(retro).

Levenson's criterion was used for diagnosis. All patients underwent high resolution computerized tomography (HRCT) of temporal bones and magnetic resonance imaging (MRI) with contrast was additionally undertaken in 4 patients. The patients were followed for a mean period of 10 months ranging from minimum of 5 months follow up to a maximum of 18 months. The presenting features, ear microscopic findings, imaging findings, type of surgical approach used for every case, surgical findings, preoperative and postoperative hearing results, complications associated during and post-surgery and follow up findings were noted.

Results

A total of 6 adults, 5 males and 1 female with CC were included in the study. 5 patients had left sided and 1 patient had right sided cholesteatoma. The patient characteristics, their presenting features, ear microscopic findings and hearing results are summarized in Table 1. The cholesteatoma affected the middle ear and mastoid in 2 cases, mastoid alone in 1 case and petrous apex in 3 cases. There was no history of head injury, surgical intervention or otorrhoea in any of the cases. The average duration of symptoms was 3.8 years. The most common symptom was hearing loss (100%) followed by tinnitus in 4 patients, facial palsy and vertigo in 3 and 2 patients respectively. Subject 1, 5 and 6 had grade 4 facial palsy.

Some other presentations noted were of otitis media with effusion in 1 patient (subject 2), history of meningitis with incidental finding of mass in petrous apex (subject 6), post aural fistula (subject 3) and of facial palsy which failed to improve (subject 1). On ear microscopy, a whitish mass behind an intact tympanic membrane was found in 4 patients and in two patients no mass was appreciated. The most common site of visualization of mass was in antero and postero superior quadrant in 2 patients. Out of the 4 patients, 1 had single quadrant involvement and 3 had 2 or more quadrants involved. Preoperatively, 2 patients had profound hearing loss, 2 patients had moderate conductive hearing loss, 1 patient had severe to profound hearing loss, and 1 patient had severe-mixed hearing loss. Postoperatively the hearing in 1 patient (subject 6) worsened from severe-mixed to profound and in 1 patient (subject 2) the hearing improved from moderate conductive hearing loss to mild conductive hearing loss. 1 patient (subject 4) with moderate conductive hearing loss was lost to follow up.

Table 2 summarizes the type of surgical intervention carried out and the surgical findings. Most common ossicular erosion was that of incus (4 patients) followed by stapes (3 patients). Dehiscence of tegmen, sigmoid plate and posterior cranial fossa plate was noted in 3, 2 and 1 patients respectively. Petrous apex cholesteatoma was noted in 3 cases; in these cases the cochlea was eroded in 2 cases and the semicircular canals and the vestibule was involved to various degrees in 3 cases. Subject 1 (Fig. 1) had cholesteatoma medial to the cochlea and the 3 semicircular canals; carotid canal was also eroded. This patient had to undergo translabyrinthine transotic procedure with blind sac closure for total excision of the mass. Similarly, subject 5 had cholesteatoma eroding the middle and apical turns of cochlea, lateral and superior semicircular canals; the mass was also eroding the internal auditory meatus and hence also had to undergo translabyrinthine, transotic procedure with blind sac closure. Subject 6 had intact cochlea but had partial erosion of the ampullary end of superior semicircular canal along with extension of cholesteatoma into the internal auditory meatus (IAM); while clearing disease from the IAM, CSF leak was encountered; this patient underwent partial labyrinthectomy with sealing of the CSF leak and blind sac closure. Subject 3 (Fig. 2) who had mastoid cholesteatoma eroding the posterior canal wall, sigmoid sinus plate and the dural plate underwent canal wall down mastoidectomy. In this case the cholesteatoma was engulfing the mastoid segment of the facial nerve which could be peeled off. During surgery the sac was found to be adherent to the dura of the posterior canal fossa dura, which could not be cleared completely and thus immediate obliteration of the cavity was not undertaken. Postoperatively this patient maintained facial nerve function. CSF leak was noted in 2 patients who had cholesteatoma eroding the internal auditory meatus. 1 patient (Subject 4) was lost to follow up and we encountered no recurrent or residual disease in the remainder (5 patients).

Discussion

There are many articles published describing CC in pediatric age group; however studies in adult population are relatively less. CC's are seen more frequently in children than in adults. This study describes CC in adult population. A large series of CC studied by Doyle et al. [10] which included both adult and pediatric population showed males outnumbered females. Our study showed similar result where male patients were more in number.

Table 1 Demographic data and clinical examination

Age range: 18–49 years (mean 27.5 years)
Gender
Males: 5 patients (83.3%)
Females: 1 patient (16.6%)
Ear affected
Left side: 5 patients 83.3 (%)
Right side: 1 patient (16.6%)
Duration of symptoms: range 1–7 years (mean 3.8 years)
Chief Complaints
Hearing loss: 6 patients (100%)
Facial palsy: 3 patients (50%)
Tinnitus: 4 patients (66.6%)
Pos taural fistula: 1 patient (16.6%)
Vertigo 2 patients (33.3%)
Past history of meningitis 1 patient (16.6%)
Tympanic membrane findings: whitish mass in the
Antero-superior and postero-superior quadrant: 2 patients (33.3%)
Postero-superior and postero-inferior quadrant: 1 patient (16.6%)
Postero-superior quadrant only: 1 patient (16.6%)
No opacity: 2 patient (33.3%)
Pre-operative hearing status:
Profound hearing loss: 2 patients (33.3%)
Severe-profound hearing loss: 1 patient (16.6%)
Severe-mixed hearing loss: 1 patient (16.6%)
Moderate conductive hearing loss: 2 patients (33.3%)
Post-operative hearing status
Profound hearing loss: 3 patients (50%)
Severe-profound hearing loss: 1 patient (16.6%)
Mild conductive hearing loss: 1 patient (16.6%)
Lost to follow up: 1 patient (14.3%)

Table 2 Surgical findings

Ossicular status: erosion of
Incus: 4 patients (66.6%)
Stapes: 3 patients (50%)
Dehiscence of
Tegmen: 2 patients (33.3%)
Sinus plate: 1 patient (16.6%)
Posterior cranial fossa plate: 1 patient (16.6%)
Sac involving/eroding the
Cochlea: 2 patients (33.3%)
Semicircular canals: 3 patients (50%)
Internal auditory meatus–2 patients (33.3%)
Others
Facial nerve status
Dehiscent tympanic segment: 2 patients (33.3%)
Surrounded 360° by cholesteatoma: 1 patient (16.6%)
Labyrinthine segment absent/eroded by sac: 1 patient (16.6%)
Cerebrospinal fluid leak: 2 patients (33.3%)
Surgical intervention
Transotic, translabyrinthine excision of cholesteatoma with blind sac closure: 2 patients (33.3%)
Transmastoid approach- partial labyrinthectomy: 1 patient (16.6%)
Canal wall down mastoidectomy: 2 patients (33.3%)
Atticotomy: 1 patient (16.6%)

CCs can present with a various symptoms. Subject 1 presented with history of unilateral facial palsy which was diagnosed elsewhere as Bell's palsy 3 years before visiting us. The palsy failed to improve and later he additionally developed symptoms of vertigo and progressive hearing loss for which he underwent imaging which revealed mass in the petrous apex. Subject 2 presented with allergic rhinitis and bilateral otitis media with effusion. Treatment with nasal steroid sprays and antihistamines improved the symptoms, however hearing loss persisted in one ear, the repeat ear microscopy revealed whitish mass behind tympanic membrane. House and Sheehy have also commented that CC should be considered in cases presenting with unilateral otitis media with effusion [11]. Subject 3 presented with post aural fistula and no other findings on ear microscopy. Imaging revealed mass limited to the mastoid region which was consistent with cholesteatoma. Very few cases of mastoid CC manifesting as mastoiditis have been mentioned in literature. Mastoid process is least frequently affected and often CC in this region have late presentation presumably because the disease may remain clinically silent for long periods and also because initially it does not

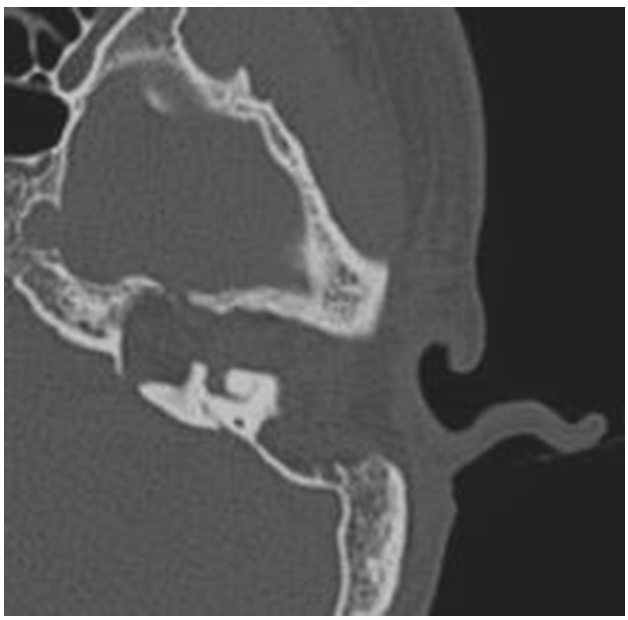

Fig. 1 HRCT showing cholesteatoma involving the petrous apex (subject 1)

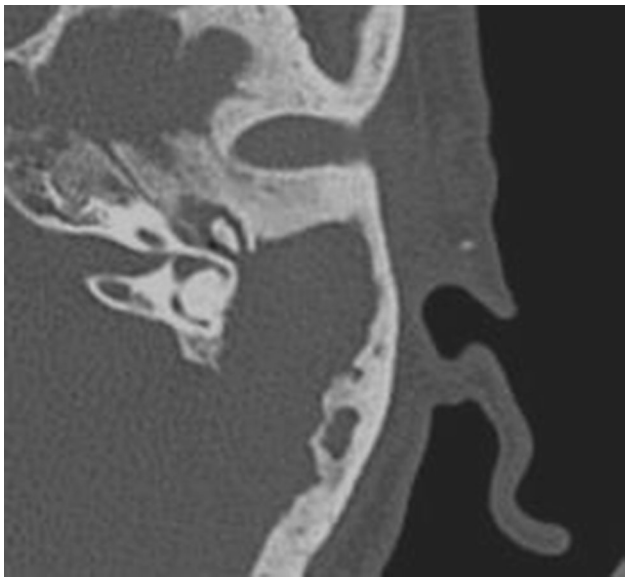

Fig. 2 HRCT showing cholesteatoma eroding the posterior boundary of the mastoid cavity (subject 3)

impinge upon any critical neurovascular structure [12, 13]. Subject 3 was a 49 year male with mastoid CC who had no other complaints except for sudden onset of post aural discharge following a mild swelling in the postaural region. Subject 6 was admitted in another medical center for meningitis and underwent imaging of the brain which revealed a mass in petrous apex following which he was referred to our hospital for further management. Rupture of the contents of CC into subarachnoid space with release of keratin, cholesterol and lipids cause chemical irritation of meninges and there are isolated case reports mentioned in literature which suggest petrous apex CC as a cause of meningitis [14, 15].

Imaging plays an important role in diagnosis especially when it is imperative to distinguish other lesions affecting the mastoid and the petrous apex. Computerized tomography (CT) reveals expansile well circumscribed mass and on magnetic resonance imaging (MRI), CC have low signal on T1 and high signal on T2, they don't enhance with contrast and show restricted diffusion on diffusion weighted imaging (DWI) sequences [12]. Additional role of DWI is in its role for identifying residual/recurrent disease especially in cases where the canal wall is not taken down during surgery, cases where cartilage is used for reconstruction of tympanic membrane and thus direct otoscopic visualization is insufficient to know the status; thus DWI may help to avoid unnecessary second look surgeries [16]. Its use is also extended in cases of petrous apex cholesteatoma to check for residual/recurrent disease and avoids unnecessary surgical reexploration. Pre-operatively, we used high resolution CT scanning in all our cases (100%) to study the extent of CC and MRI in 66.6% cases to differentiate CC with other lesions like cholesterol granuloma, arachnoid cyst, mucocoele [15] etc.

Some studies [10] have also found that cases of CC may have normal looking tympanic membrane, thus suggesting that presence of a whitish mass behind an intact tympanic membrane may depend on factors like location of disease and extent of growth. In our study a whitish mass behind intact tympanic membrane was observed in only 4 out of 6 patients. Subject 3 had mass that was involving the mastoid region exclusively and hence no whitish mass was observed on ear microscopy. Subject 6 (with petrous apex cholesteatoma) also had no involvement of the mesotympanum and hence no whitish mass was appreciated. In the remaining cases multiple quadrant involvement was more common than single quadrant involvement, most commonly being antero-superior and postero-superior quadrant (33.3%). Schuknecht and Peron commented that CC occurs most commonly behind the antero-superior or postero-superior quadrant of the tympanic membrane and that it rarely arises in the mastoid cavity [3]. We did not find any such predilection and most of our cases had multiple quadrant involvement which can be explained based on the long duration of symptoms and the advanced lesion at presentation.

Postoperatively the hearing result worsened in subject 6 who initially had severe mixed hearing loss but after surgery had profound hearing loss. This can be explained based on 2 possible mechanisms. First, possible trauma to the inner ear while elevating the sac off the ampullary end of superior semicircular canal and secondly due to the fact that he had to undergo blind sac procedure as we observed CSF leak while dissecting the matrix off the internal auditory meatus. Canalis et al. [17] classified inner ear cholesteatoma into 3 groups and described the expected hearing outcome for every case depending upon the involvement of the inner ear. Group C of Canalis classification which includes involvement of cochlea and labyrinth has worst prognosis with respect to hearing. 2 of our patients (subjects 1 and 5) fell into this group and had profound hearing loss both pre and postoperatively. 1 patient (subject 3) with severe-profound hearing loss preoperatively continued having the same postoperatively. Subject 2 (pre-operative pure tone average of 48. 3 dB) who underwent canal wall up mastoidectomy with ossiculoplasty had improvement in hearing (pure tone average of 30 decibel postoperatively). Subject 5 who had moderate hearing loss preoperatively was lost to follow up.

Surgery is the modality of treatment and various procedures need to be carried out based on the extent and location of the disease. Canal wall up mastoidectomy with ossiculoplasty, atticotomy with ossiculoplasty and canal wall down mastoidectomy was carried out in 1 patient (16.6%) each. Translabyrinthine transotic excision of mass

with blind sac procedure was carried out in 2 patients (33.3%) as the mass was extending medial to the semicircular canals and cochlea and also eroding them. Subject 6 underwent partial labyrinthectomy with blind sac closure; in this case the ampullary end of semicircular canal was involved and thus the matrix was peeled off. Here there was no erosion of cochlea, however we encountered a CSF leak while clearing disease near the internal auditory meatus and thus the patient had to undergo blind sac closure for the same. In the blind sac procedure for all the 3 cases, the cavity was obliterated with layers of fat from thigh, fascia lata and tissue glue.

Of the 6 patients' only subject 4 who underwent atticotomy was lost to follow up. One patient who had disease limited to the mastoid and there was extensive erosion of the posterior cranial fossa plate, tegmen and sigmoid sinus plate underwent canal wall down mastoidectomy and was advised revision surgery at the end of 1 year for obliteration, however he came to us for only one follow up at the end of 5 months which showed well healed mastoid cavity and since then he is lost to follow up. 3 patients with grade 4 facial palsy showed no improvement and have been advised Ophthalmology consult for gold implants for eye closure. 1 patient of petrous apex CC in whom there was no involvement of the cochlea was advised appropriate surgery for hearing rehabilitation, however he deferred further intervention. The patients who have had blind sac closure have been in follow up with us and at the end of 1 year, underwent MRI with diffusion weighted imaging (DWI) sequences to look for recurrence/residual disease and none showed any evidence of recurrence or residual disease.

In conclusion CC can have various presentations. Surgical treatment results in good outcome and eradication of the disease. However surgery has to be individualized to suit each patient's disease location and extent.

Acknowledgements This study was approved by the Institutional Review Board (IRB) at Christian Medical College, Vellore, India. Study dated 21.06.2017, IRB Min No. 10726(retro).

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