CASE REPORT

Congenital cholesteatoma: Different clinical presentation in two cases

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Abstract

Introduction: Congenital cholesteatoma (CC) of the middle ear is a rare entity that may be undiagnosed for years. The lesion can grow undetected until it produces symptoms such as reduced hearing or otalgia.
Method: Case report
Results: We report two cases of young ladies with CC who presented with different otological symptoms. The first case complained of recurrent unilateral ear pain while the second case presented with unilateral reduced hearing. Examination of both cases revealed a whitish mass seen behind an intact tympanic membrane. Both cases underwent surgery and histopathological examinations; findings were consistent with cholesteatoma.
Conclusion: There are varieties of clinical presentations of CC and the diagnosis is based on clinical findings.

Introduction

Congenital cholesteatomas (CCs), though uncommon, have been well documented and described in the literature. Congenital middle ear cholesteatoma is defined as a keratinising epithelial rest that occurs behind an intact tympanic membrane without a prior history of infection or trauma. Although its cause is unknown, several theories have been proposed to explain its origin. The theories include the induction of keratinising epithelium from the middle ear mucosa due to inflammation, squamous metaplasia and the 'migration' of epithelial debris into the middle ear through the eustachian tube.[1] We report two cases of CC with different presentations.

Case report

The first case is an 18-year-old lady who presented with a complaint of on and off right ear pain for 1-year duration. She denied any history of hearing loss but her family claimed otherwise. There was no history of vertigo, tinnitus or headache.

The examinations of the right ear revealed a whitish soft tissue mass behind an intact tympanic membrane at the anterosuperior quadrant (**Figure 1**), and the other ear showed no abnormality. Rinne test was negative on the affected side while Weber was lateralised towards the side. Pure tone audiometry showed mild conductive hearing loss of the right ear. Computed tomography (CT) scan of the temporal bone revealed a soft tissue density within the epitympanic recess extending into Prussak's space and mastoid air cells through the aditus and antrum. Atticotomy was done. Intraoperatively, a whitish mass was found in the attic but limited to the epitympanum space. The handle of malleus and incus were eroded. The mass was removed. The atticotomy area was covered with a tragal cartilage.

The second case was a 13-year-old girl who presented with progressive left ear reduced hearing for 1-year duration associated with ear discomfort and tinnitus. There was no history of vertigo, tinnitus or headache. The examination of the left ear revealed a bulging tympanic membrane with a whitish mass seen behind an intact tympanic membrane (**Figure 2)** while the contra lateral ear was normal. Rinne test was negative on the affected side with Weber lateralised towards the ear. Pure tone audiometry showed severe conductive hearing loss of the diseased ear. CT scan showed a soft tissue mass within the epitympanum extending into mastoid air cells with widening of the antrum. The ossicles were displaced laterally and the lateral semi-circular canal was eroded. Modified radical mastoidectomy with type 3 tympanoplasty was done.

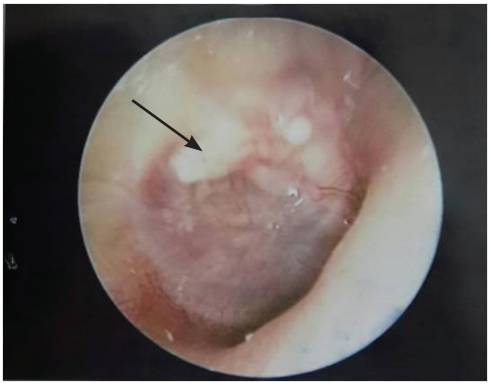

Figure 1. Otoscopic view of the right ear of the first case showing whitish mass behind an intact tympanic membrane

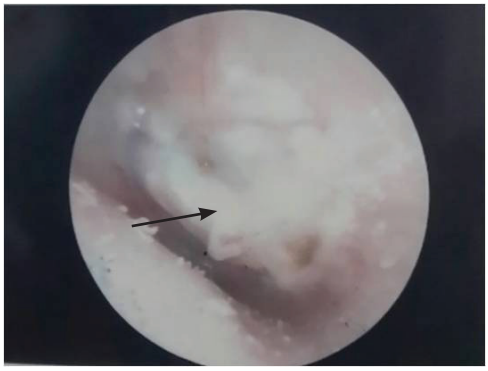

Figure 2. Otoscopic view of the left ear of the second case showing whitish mass behind an intact tympanic membrane

Discussion

CC is a relatively uncommon disease. It is diagnosed based on a finding of a white mass medial to a normal tympanic membrane with no prior history of otorrhea or tympanic membrane perforation or prior otologic procedures. The tympanic membrane can be visualised clearly by using a bright otoscope. The pinna is retracted backwards and upwards to straighten out the canal, allowing a more direct vision to the tympanic membrane. The pathogenesis of CC remains an area of controversy but it is thought to originate from rests of epidermoid cells incorporated into the temporal bone during embryonic development.

Though most of these patients complain of hearing loss, some are unaware of the condition until they are told that they have an abnormal tympanic membrane. Analysis by Kojima et al[2] in their study concluded 60.3% of their patients presented with hearing loss as a main complaint while 17.5% of the patients were initially examined for otorrhea/otalgia. According to Tagaki,[3] more than half of the patients with early-stage CC were diagnosed asymptomatically by a chance visit to a clinic while hearing loss is the main complaint in advanced stages. The patient in our case who complained of hearing loss was having a more advanced disease. However, the patient who presented with otalgia was actually having recurrent otitis externa but was noted to have CC as well.

It is known that CC undergoes progressive expansion and commonly presents at later age. Both of our patients are teenagers. Kojima[2] reported that the mean age at presentation for CC was 13.3 years. Over time, it grows with possible extension and erosion to the remainder of the tympanic cavity, antrum and mastoid. The complications of cholesteatoma include hearing loss and intracranial abscess.[4,5] The diagnosis of CC is usually made by clinical and otoscopic examination. CT scan of the temporal bone is performed to assess its extension, exploring for complications and detecting anatomic variants as preparation for surgery.

Treatment for CC remains surgical. The main objective is to eradicate the disease to restore a healthy aerated ear. Of no lesser importance, the second objective is to restore or improve the ossicular chain. The prognosis depends on its location, the age of the patient, degree of ossicular involvement and number of involved sites.

Conclusions

CC has a variety of clinical presentations. Hearing loss was the common presenting symptom but it can be diagnosed by chance at the early stage. The finding of a white mass medial to a normal tympanic membrane needs referral to an otorhinolaryngologist for early intervention and this can therefore can avoid further complications.

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