Case report

A rare case of bilateral congenital posterior mesotympanic cholesteatoma

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Abstract

Bilateral congenital mesotympanic cholesteatoma is a very rare disease. It can present differently from ordinary congenital cholesteatoma. We report a case of bilateral congenital cholesteatoma diagnosed at age of 22 years old. She presented with bilateral intermittent ear discharge since 10 years old that worsening two weeks prior to her presentation to our clinic and associated with bilateral reduced hearing. Clinically there was intact tympanic membrane with retraction of the mesotympanic area with present of mass medial to tympanic membrane. CT scan imaging showed there was soft tissue in the bilateral middle ear cavity with intact scutum and ossicles. Patient undergone canal wall down procedure and the diagnosis of congenital mesotympanic cholesteatoma was confirmed with present of cholesteatoma sac at the posterosuperior part, as opposed to anterosuperior quadrant, where the common site for congenital cholesteatoma.

Keyword: MeSH: Congenital cholesteatoma; middle ear cholesteatoma; cholesteatoma

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Introduction

Mesotympanic cholesteatoma is part of a congenital cholesteatoma, defined as cholesteatoma mass behind an intact tympanic membrane without any previous history of otologic surgery and tympanic membrane perforation[1]. Congenital cholesteatoma is commonly localized at the anterosuperior part of tympanic membrane and rarely localized at the posterosuperior area. Bilateral cholesteatoma are even rarely reported. Congenital cholesteatoma is commonly diagnosed incidentally when there is present of white mass in the middle ear with intact tympanic membrane and it is easily missed even by otorinolaryngologist. However there are still patient who presented with reduced hearing without any history of ear infection.

Congenital cholesteatoma usually diagnosed during chilhood mainly because of the screening programme and rarely presented late. However patient might present later in life because congenital cholesteatoma might not cause any troublesome symptoms except mild hearing loss that can be coped well by the patient.

Case report

22 years old lady presented to Otorhinolaryngology clinic for bilateral intermittent foul smelling ear discharge since 10 years ago that was self-limiting without seeking any treatment and it was worsening over the past tow months. It was associated with bilateral reduce hearing for 10 years and patient claimed it was not troubling her very much and she did not need any hearing assistance for that. There

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were no vertigo, tinnitus, ear bleeding and ear blockage. Patient denied any trauma any recurrent nasal symptoms such as nasal blockage, epistaxis and rhinorhea.

Otological examination of the bilateral ears revealed normal external auditory canal and intact tympanic membrane. Both tympanic membrane were inflammed with present of retracted pars tensa, granulation tissueat the posterior part of the tympanic membrane and whittish mass at posteroinferior part of tympanic membrane (figure 1).

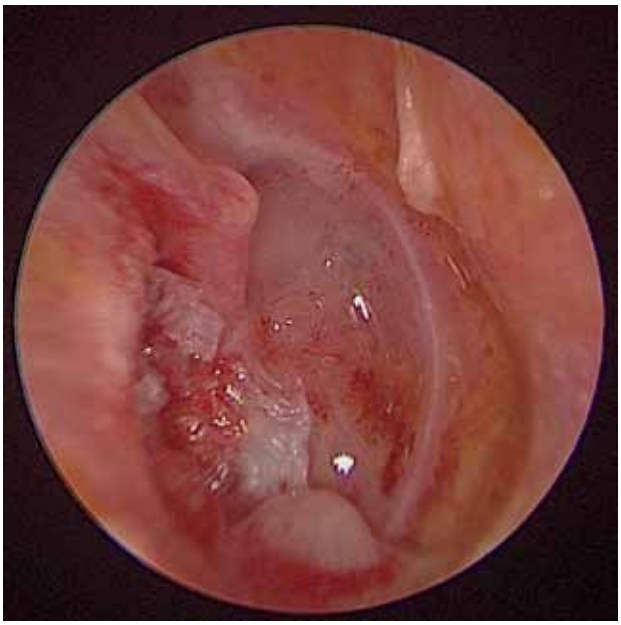

Figure 1: Retracted mesotympanum of right tympanic membrane. It is intact with present of granulation tissue at posterior inferior with mass in the middle ear cavity.

Audiological examination revealed Rinne's test were positive bilaterally and Weber test was central. Audiogram showed mild to moderate conductive hearing loss bilaterally (figure 2).

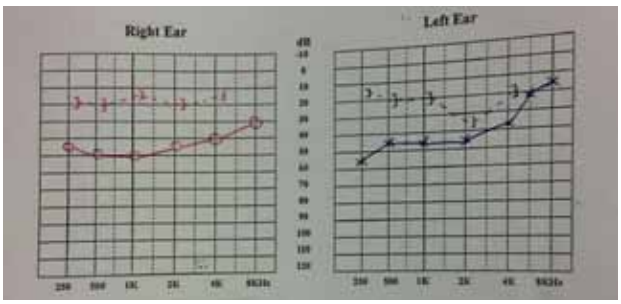

Figure 2: Right mild conductive hearing loss. Left ear has mild to moderate conductive hearing loss at low frequency and mild sensorineural hearing loss at high frequency

Computed tomography scan imaging was done and revealed there was mixed soft density in the bilateral middle ear cavity that extend into aditus ad antrum with bilateral sclerotic mastoid air cells. However, both scutum and bilateral osscicles were intact (figure 3).

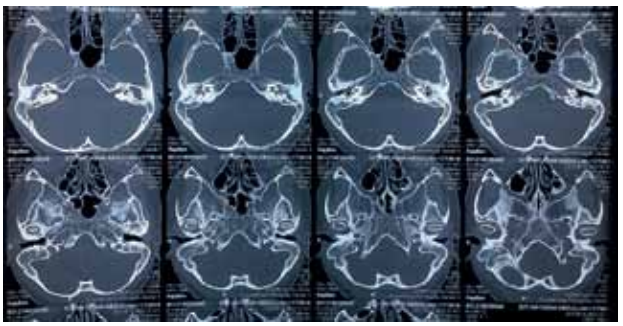

Figure 3 : CT scan image showing gsoft tissue density in the middle ear cavity with sclerotic mastoid air cells bilaterally. Both scutum were intact.

Patient underwent canal wall down procedure on the right ear.Intraoperative findings were sclerotic mastoid air cell with cholesteatoma originate from posteroinferior part of pars tensa extended to the fossa incudis and mastoid antrum preserving the anterior part of middle ear cavity. The incus and stapes suprastructure were eroded with intact malleus. Patient was discharged well two days post operation and subsequent follow up showed a very well epithelialized mastoid cavity. Patient was planned for another operation on the left side but patient was undecided yet even after a very well counselling given.

Discussion

Bilateral congenital colesteatoma is very rare. A series by Potsic[2] et al. showed that on 5 patients out of 167 patient had bilateral congenital cholesteatoma. Another series by Nelson[3] et al., 2 patients out of 119 patients has bilateral congenital cholesteatoma. Overall even congenital cholesteatoma only accounted for 0.12 per 100000 people-approximately 1 in 1 million per year[4].

The age of diagnosis congenital cholesteatoma ranging from 2 years old to 14 years old with mean of 5-7 years old[2,3,5]. In our case the age of diagnosis was made at 22 years old. The reason for this late diagnosis could be due to the patient's previous ear discharge was self-limiting and and patients activity of daily living was not much affected. It is also supported by the audiogram result in which that patient can still hear and can live her own life well with mild to moderate conductive hearing loss bilaterally. However this current presentation to

our centre is due to worsening ear discharge which caused by acute suppurative otitis media. Otoscopic examination support our clinical diagnosis as the tympanic membrane look inflammed with retracted pars tensa and whitish mass present.

Location of congenital cholesteatoma is mainly at the anterosuperior quandrant, classified in relation to four quadrants of the ear drum. The other group is posterosuperior mesotympanic cholesteatoma and its also less common than anterosuperior quadrant[3]. All these patient can only be classified if patient presented early and the cholesteatoma is relatively small thus, fit in the quadrant. These classification were supported by others such as in Friedberg's[6] series, only 5% involving posterosuperior and another 31% involving anterosuperior quandrants. In our case, cholesteatoma sac was mainly located at posterior mesotympanum area. This findings contradict with the theory that congenital cholesteatoma arise from an embryonic cell rest in the anterior superior quadrant[8]. The posterior mesotympanum cholesteatoma may have multifactorial origin possible due to Sade's[9] theory of metaplasia of the middle ear and Aimi's[10] epithelial migration.

This patient underwent canal wall down and meatoplasty procedure two months after the diagnosis was made. Intraoperatively, cholesteatoma was quite extensive. It fits the stage IV, as ossicles and mastoid involved. It is a system introduced by Potsic[2] et al. Stage I includes cholesteatoma involving one quadrant with no ossicular and mastoid involvement. Stage II includes case when multiple quadrants involved but not the ossicles and mastoid. When ossicles are eroded but mastoid is not, it is stage III, whereas involvement of ossicles and mastoid is stage IV. Late presentation to otorhinolaryngologist might contribute to patient's extensive disease. Canal wall down procedure in congenital cholesteatoma have raised some controversies. Nelson[2] et al. stated, canal wall down procedure should be considered when there is destruction of posterior canal wall, labyrinthine involvement, petrous apex extension, or concern about reliability follow up. A few literatures were against canal wall down procedure because it will result in undesirably large mastoid cavity as congenital cholesteatoma occur in well pneumatized mastoid, thus in children it will requiring ear care for life[3]. In our case considering the patient was diagnosed with congenital cholesteatoma at the age of 22 years old, it seems the canal wall down is the best procedure for her.

Conclusion

Congenital cholesteatoma is a very rare disease. To find bilateral mesotympanum cholesteatoma at 22 years old of age is even rarer entity. Because of untraditional presentation of this rare disease, the managing clinician has to have high index of suspicion to look at and appreciate this condition.

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