Two Cases of Congenital Cholesteatoma of the Tympanic Membrane

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Abstract Congenital cholesteatoma of the tympanic membrane is rare, and lesions without a history of otitis media or any other adverse events involving the tympanic membrane are extremely rare. We report two cases of this lesion; one was a 3-year-old girl who underwent removal of a cholesteatoma using retroauricular approach and a partial myringoplasty with an underlay technique, and the other was a 2-year-old girl in whom a cholesteatoma was enucleated without grafting. This disease is thought to be of embryonic origin.

Keywords Cholesteatoma · Tympanic membrane · Congenital · Excision · Enucleation

Introduction

Cholesteatoma within the tympanic membrane without previous ear surgery or trauma is rare [8]. Some cases of this disease have had episodes of otitis media [4, 6, 8–10], but cholesteatoma of the tympanic membrane without any other adverse events is extremely rare and this lesion is considered to be of congenital origin [7]. Reddy et al. [7] opined that primary care physicians and pediatricians should routinely perform otoscopy on every child because most cases are found only by chance. Cholesteatoma should be removed by excision of the involved part of the

tympanic membrane or enucleation before it begins to progress.

Case Report

Case No. 1

A 3-year-old girl was referred to our outpatient clinic by an otolaryngologist who found a white mass on the left tympanic membrane. Otomicroscopic examination revealed a pearly white mass on the posterior superior quadrant of the left tympanic membrane (Fig. 1). There was no history of otitis media, otorrhea, ear trauma, or ear surgery in the left ear, but she did have a history of acute otitis media in the right ear. When she visited an ear nose and throat clinic for right otalgia, an otolaryngologist noticed the white mass on the left tympanic membrane. A pure-tone audiogram revealed a 10–20 dB conductive hearing loss at the frequencies of 125, 250 and 500 Hz on the left side. Tympanograms were A type in both ears. A computed tomography (CT) scan showed a round low density mass, 3 mm in diameter, on the external surface of the left tympanic membrane, but showed normal findings in the middle tympanum, the ossicular chain, the mastoid cavity, and the inner ear.

The lesion was removed surgically using a retroauricular approach and then myringoplasty was performed. The mass was on the external surface of the left tympanic membrane, and behind the mass there was the mucosal layer of the tympanic membrane. The lesion did not touch the malleus, and the ossicular chain was intact. In the middle tympanum there was no lesion. After removal of the mass with a part of the surrounding tympanic membrane, a partial myringoplasty was performed with the temporal muscle fascia

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Fig. 1 Otomicroscopic finding of case 1. A pearly white mass is seen on the posterior superior quadrant of the left tympanic membrane

using an underlay technique. Pathological findings were compatible with cholesteatoma. There is no evidence of recurrence 31 months after surgery. Otomicroscopy shows a well-healed graft, and the audiogram is normal.

Case No. 2

A 2-year-old girl visited our outpatient clinic complaining of rhinorrhea, and a small round white mass was found on the anterior inferior quadrant of the left tympanic membrane (Fig. 2). She had had no adverse events, such as otitis media, trauma, or surgery, on the bilateral tympanic membrane. A CT scan revealed no abnormal findings in the middle ear.

Fig. 2 A small white mass is seen on the anterior inferior quadrant of the left tympanic membrane of case 2

The mass on the left tympanic membrane was enucleated by the transmeatal approach without grafting, because the fibrous layer of the tympanic membrane was intact. In pathological findings, there were stratified squamous epithelium and keratin debris without endodermal and mesodermal components. The patient is doing well 24 months after surgery with no evidence of recurrence.

Discussion

Cholesteatoma within the tympanic membrane is a potential complication of tympanoplasty [3], but cholesteatoma within the tympanic membrane without the previous otosurgery or ear trauma is rare [8]. Moreover, only a few reports of intratympanic membrane cholesteatoma without a history of otitis media or any other adverse events are found in the literature [2, 5, 7, 10].

In cases with previous otitis media, intratympanic membrane cholesteatoma may develop by proliferation of the basal cell layer of the tympanic membrane epithelium in response to an inflammatory process [5], as shown in the animal models [1]. Intratympanic membrane cholesteatoma without any adverse events, such as our two cases, can be considered of congenital origin [2, 5, 7, 10]. However, the true cause of this lesion is still unknown because the pathogenesis of congenital cholesteatoma in the middle ear is unclear.

Intratympanic membrane cholesteatoma without any adverse events in the middle ear or on the tympanic membrane has been detected as an incidental finding on routine examination of the ear [7]. Therefore, not only otolaryngologists but also pediatricians and primary care physicians should routinely perform otoscopy on every child in order to screen for this lesion.

Treatment of intratympanic membrane cholesteatoma is total removal as for cholesteatoma in the middle ear. In previous reports, some cases were treated by excision of the cholesteatoma together with the involved part of the tympanic membrane with or without myringoplasty, and some cases were treated by enucleation of the lesion alone. In case 1 we removed the cholesteatoma with a part of the tympanic membrane and performed partial myringoplasty using a retroauricular approach, because it was unclear whether the malleus was involved or not from the findings of CT scan and otoscopy. In case 2 we enucleated the cholesteatoma alone by the transmeatal approach, because the mass was small and separated from the malleus handle. Otosurgeons should select approach and removal area in according to the size and site of the cholesteatoma mass, and should perform myringoplasty if necessary.

Conclusion

Intratympanic membrane cholesteatoma without any previous events on the tympanic membrane is extremely rare. This lesion can be considered to be congenital and should be removed totally. Surgical techniques are varied according to the size and site of the lesion. In order to detect this disease at an early stage, pediatricians and primary care physicians should routinely perform otoscopy on every child because most patients do not complain of anything in the ear.

References

1. Chole RA, Tinling SP (1985) Basal lamina breaks in the histogenesis of cholesteatoma. *Laryngoscope* 95:270–275
2. Friedberg J (1994) Congenital cholesteatoma. *Laryngoscope* 104:1–24
3. Glasscock ME III (1973) Tympanic membrane grafting with fascia: overlay vs. undersurface technique. *Laryngoscope* 83:754–770
4. Kim SA, Haupt MS (2002) Congenital cholesteatoma of the tympanic membrane. *Otolaryngol Head Neck Surg* 127:359–360
5. Pasanisi E, Bacciu A, Vincenti V, Bacciu S (2001) Congenital cholesteatoma of the tympanic membrane. *Int J Pediatr Otorhinolaryngol* 61:167–171
6. Rappaport JM, Browning S, Davis NL (1999) Intratympanic cholesteatoma. *J Otolaryngol* 28:357–361
7. Reddy CEE, Goodyear P, Ghosh S, Lesser T (2006) Intratympanic membrane cholesteatoma: a rare incidental finding. *Eur Arch Otorhinolaryngol* 263:1061–1064
8. Smith R, Moran WB (1977) Tympanic membrane keratoma (cholesteatom) in children with no prior otologic surgery. *Laryngoscope* 87:237–245
9. Sobol SM, Reichert TJ, Faw KD, Stroud MH, Spector GJ, Ogura JH (1980) Intramembranous and mesotympanic cholesteatoma associated with an intact tympanic membrane in children. *Ann Otol Rhinol Laryngol* 89:312–317
10. Weber PC, Adkins WY Jr (1997) Congenital cholesteatomas in the tympanic membrane. *Laryngoscope* 107:1181–1184