Tympanic membrane cholesteatoma: a rare finding

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We report a rare finding of tympanic membrane cholesteatoma in a two-year-old girl.

Tympanic membrane cholesteatoma without trauma or surgery to the ear is a rare entity, with few cases documented in the literature. The exact etiology of this lesion is still unclear. The presentation, clinical course and management are discussed.

A whitish spot on the tympanic membrane should raise suspicion for cholesteatoma. Early diagnosis and treatment are imperative to allow an easy removal and avoid middle ear involvement.

Key words: cholesteatoma, tympanic membrane, congenital.

Congenital cholesteatoma was first described by House[1] in 1953. Derlacki and Clemis[2] proposed the clinical criteria to establish the diagnosis of middle ear (ME) congenital cholesteatoma, which were later revised by Levenson et al.[3] (Table I). The incidence of congenital cholesteatoma of the ME comprises 2% to 5% of all cholesteatomas[4-6]. Congenital cholesteatomas arising from the tympanic membrane (TM) are even less common[4,6-11]. Although the number of studies regarding congenital cholesteatoma of the ME has increased over the years, there are few reports on TM cholesteatoma.[4,6-13] Cholesteatoma should be considered in the differential diagnosis of white TM lesion[4,6,7,10,12,14,15]. Early treatment is essential to avoid progressive destruction[4-6,8-10,12,15,16]. We present a two-year-old girl with left TM cholesteatoma located in the posterior-inferior quadrant.

Table I. Criteria for Definition of Congenital Cholesteatomas by Levenson et al.[3]

White mass medial to a normal intact tympanic membrane
Normal pars flaccida and pars tensa
No history of otorrhea or perforation
No prior otologic procedures
Exclusion of canal atresia and intramembranous and giant cholesteatoma
Prior bouts of otitis media are not grounds for exclusion

Case Report

A two-year-old girl was referred to our outpatient department by her pediatrician for further investigation of the whitish spot on the left TM. Otomicroscopic examination revealed the presence of a white pearl, measuring 2 mm in diameter, located at the posterior-inferior quadrant of the TM (Fig. 1). The parents denied history of otitis media, ear trauma, otorrhea, or prior otologic surgery. Tympanograms were A type with intact acoustic reflexes bilaterally, and the parents believed she was responding appropriately to sound stimuli. A high resolution computed tomography of the temporal bones showed a round-shaped tissue located inside the TM without involvement of the ossicles or the ME space. Surgery was performed through a transcanal approach, and the cholesteatoma pearl was peeled off from the TM (Fig. 2). After removal, the fibrous layer of the TM was intact and no further action was taken. Histopathology confirmed a cholesteatoma. At the three-month follow-up, otoscopic examination was normal with no signs of recurrence, and the tympanogram was type A as well.

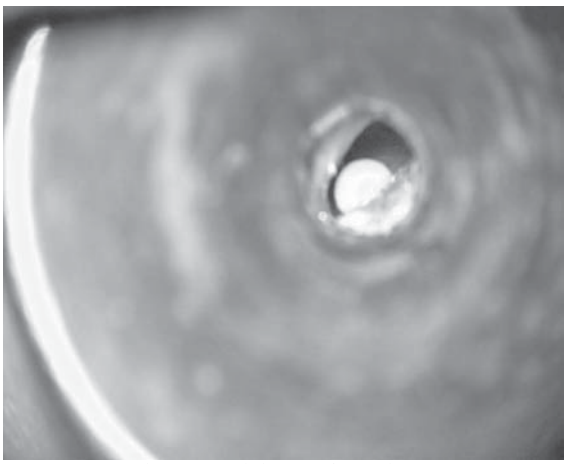

Fig 1. White pearl located at the posterior-inferior quadrant of the left tympanic membrane.

Discussion

Congenital cholesteatoma is now a relatively common diagnosis due to the recognition of the disease by otolaryngologists and pediatricians, but congenital cholesteatoma arising from the TM is very uncommon[4-11]. There are several theories as to the pathophysiology of congenital cholesteatomas. The "epithelial rest" theory postulates that persistence of epidermoid formations, derived from the first branchial groove, at the junction of the Eustachian tube and ME, results in congenital cholesteatomas[5-7,9-12,16,17]. The "metaplasia" theory supports transformation of inflamed ME mucosa into stratified squamous epithelium[4-6,9,17,18]. These two theories may explain the formation of congenital cholesteatomas of the ME, but the origin of the TM cholesteatomas may be best explained by Ruedi's[19] theory. He postulated that small inflammatory injuries to the TM *in utero* produce small perforations in the epithelium, through which the squamous epithelial basal layer proliferates into protruding cones[19]. These cones could then form TM cholesteatomas if retained within an intact TM. Our case also supports Ruedi's theory with the involvement of the epithelial layer of the TM with an intact fibrous layer. Tympanoplasty with an overlay or a combined overlay-underlay technique can be an important source of acquired TM cholesteatoma[11,13]. A whitish spot on the TM should raise suspicion for cholesteatoma[4,6,7,10,12,14,15]. Otomicroscopy is the gold standard for differentiating TM cholesteatoma from tympanosclerosis[6,10]. Surgery is mandatory because these lesions may potentially involve the ossicles and ME[4-6,8-10,12,15,16]. In our case, early diagnosis and treatment before violation of the fibrous layer provided easy removal of the disease and avoided a myringoplasty with grafting.

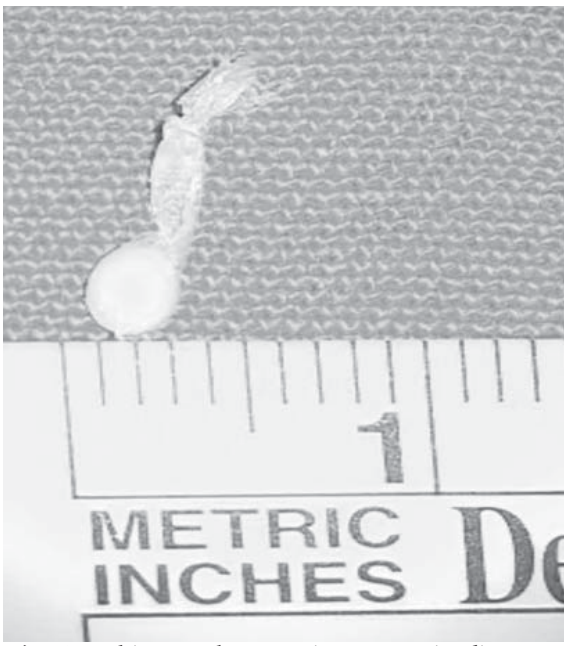

Fig. 2. White pearl measuring 2 mm in diameter after removal.

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