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A Case of Intratympanic Membrane Congenital Cholesteatoma

Yee Hyuk Kim

Department of Otorhinolaryngology-Head & Neck Surgery, School of Medicine, Catholic University of Daegu, Daegu, Korea

선천성 고막내 진주종 1예

김 이 혁

대구가톨릭대학교 의과대학 이비인후과학교실

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Address for correspondence
Yee Hyuk Kim, MD
Department of Otorhinolaryngology-Head & Neck Surgery,
School of Medicine,
Catholic University of Daegu,
33 Duryugongwon-ro 17-gil,
Nam-gu, Daegu 705-718,
Korea
Tel +82-53-650-3189
Fax +82-53-650-4533
E-mail yhukim@cu.ac.kr

Intratympanic membrane congenital cholesteatoma (ITMCC) is extremely rare in patients without previous trauma or surgery of the ear. There are multiple theories to the pathogenesis of middle ear congenital cholesteatoma but none are applicable for ITMCC. When the size of ITMCC is small, there are usually no ear symptoms. However, surgery is needed early, as in any other type of cholesteatoma, because early treatment before middle ear involvement enables enucleation without the need for other procedures such as myringoplasty. We report an incidental finding of a small pearl at the umbo in a 2-year-old boy. The cholesteatoma involved only the outer epidermic layer of the tympanic membrane (TM). A transcanal approach was performed and the cholesteatoma was enucleated from TM.

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Key Words Tympanic membrane · Congenital · Cholesteatoma.

Introduction

Despite the many reported cases of epidermal pearls encountered between the layers of the tympanic membrane (TM) following otologic surgery, the observation of an intratympanic membrane congenital cholesteatoma (ITMCC) - in other words, primary tympanic membrane cholesteatoma in childhood without history of otorrhea, otologic surgery, and ear trauma - is extremely rare.[1-3] To our knowledge, congenital cholesteatoma within the TM in infancy or early childhood has been reported in only 15 cases in the English literature,[1,4] and has never been reported in the Korean literature. Criteria have been proposed for clinical diagnosis and multiple theories exist for the pathogenesis of congenital middle ear cholesteatoma but not for ITMCC.[1,4] Although the natural course of ITMCC is not known, early treatment is needed - as in the case of any other type of cholesteatoma - because this condition can cause progressive destruction.[1,5]

Case

A 2-year-old boy was referred to our department by his pediatrician who noted a pearly white mass on the left tympanic membrane when he presented with an upper respiratory tract infection. Otoscopic examination revealed a normal right TM and external auditory canal. However, the left TM showed a small white mass centered on the umbo, which was considered to be a cholesteatoma (Fig. 1). The patient did not have any ear symptoms or any history of previous otitis media, otorrhea, ear trauma, or otologic surgery. The auditory evoked brainstem response test revealed normal hearing in both ears. A computed tomography scan of the temporal bones showed a small soft round tissue mass located on the external surface of the left TM and did not reveal any middle ear extension (Fig. 2). Cho-

lesteatoma pearl could easily be enucleated from the TM by a transcanal approach and involved only the outer epidermic layer of the TM, without extension into the fibrous layer. After removal, no myringoplasty was necessary since the integrity of the fibrous layer of the TM had not been violated.

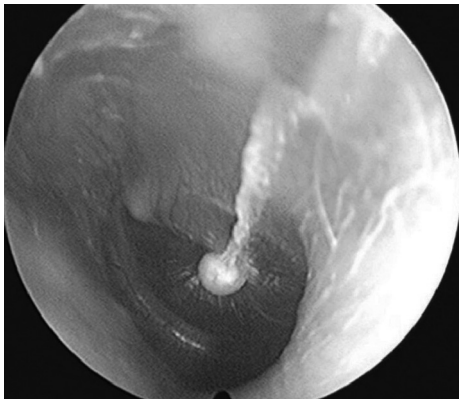

Fig. 1. Otoscopic view of the left tympanic membrane showing a small pearl-shaped mass centered on the umbo.

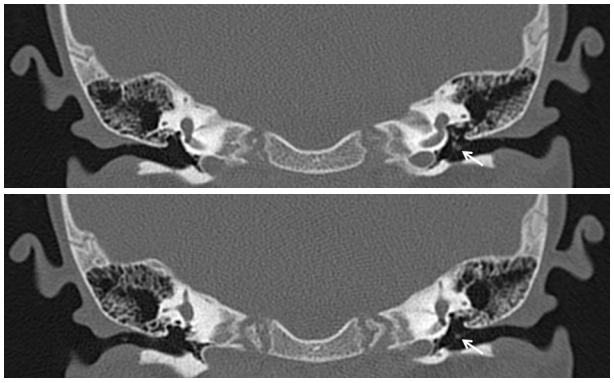

Fig. 2. Coronal CT scans of the temporal bones showing a small soft tissue mass (arrows) located on the umbo of the left tympanic membrane. CT: computed tomography.

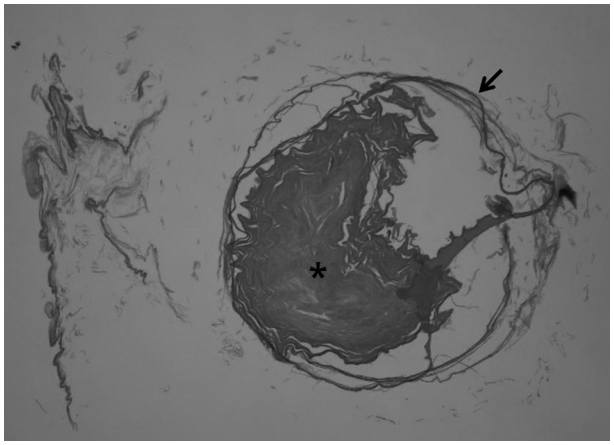

Fig. 3. Histopathologic findings showing a closed keratotic cyst (arrow: squamous epithelial lining, asterisk: keratinous materials, H&E stain, ×40).

Histopathology confirmed a cholesteatoma (Fig. 3).

Discussion

Primary cholesteatoma within the TM in infancy or early childhood has been reported in 15 cases in the English literature.[1,3,5-9] Table 1 shows the clinical features and treatment of these 15 cases of ITMCC.[1,3,5-9]

Several theories have been proposed to explain the formation of congenital middle ear cholesteatoma; for example, epidermoid formation, epithelial migration, and squamous metaplasia theory.[1,5,7,8] The pathogenesis of ITMCC is still unknown and theories are lacking regarding the formation of ITMCC. Some authors postulated that previous repeated bouts of middle ear inflammation stimulate active basal cell ingrowth and proliferation of the TM epithelium, which then leads to proliferated prickle cells that coalesce into an intramembranous keratoma.[6,10-12] However, the reported cases without previous otitis media do not support this theory. When cholesteatoma develops in subjects without any history of an inflammatory response of the external auditory canal and middle ear, an epithelial rest of embryonal origin is suspected.[8] This could be explained by the persistence of an epidermoid formation derived from the first branchial groove, which normally regresses after the 33rd week of gestation.[8,13,14] Aimi proposed that the

Table 1. Cases of primary cholesteatoma within the tympanic membrane in infancy or early childhood

No.	Age	Gender	OM	Site	ME	Tx.	Ref.
1	4	M	+	A	−	Ex	3
2	3	M	+	A	+	Ex	3
3	1	M	+	P	+	Ex	3
4	1	F	+	AI, PI	−	En	6
5	1	M	+	P	−	En	6
6	2	M	+	PI	−	En	6
7	2	F	+	PI	−	Ex+Mp	6
8	3	M	+	I	−	Ex+Mp	6
9	1	F	−	PI	+	Ex+Mp	7
10	3	F	−	P	+	Ex+Mp	7
11	4	F	−	PS	+	Ex+Mp	8
12	1	F	+	AI	−	En	9
13	1	F	−	U, AI	−	En	1
14	3	F	−	U	−	En	5
15	3	F	−	W	−	Ex	5

No.: patient serial number, M: male, F: female, OM: history suggestive of previous episodes of otitis media, A: anterior part of TM, P: posterior, I: inferior, U: umbo, AI: anteroinferior, PI: posteroinferior, PS: posterosuperior, W: whole of TM, ME: middle ear extension, Tx.: Treatment, Ex: excision, En: enucleation, Mp: myringoplasty, Ref.: reference, TM: tympanic membrane

ectodermal cells of the developing external auditory canal migrate to the endodermal side, and that these cells form the epithelial rest and may participate in the development of the tympanic membrane. The epithelial rest was postulated to disappear after the structure is fully developed, whereas persistence of this rest would result in the development of a congenital cholesteatoma within the tympanic membrane.[8,15)]

The natural course of the ITMCC is not known for certain, but ITMCC is postulated to increase in size and to expand into the middle ear space, as occurs in the case of cholesteatoma in other parts of the temporal bone.[1,7,8)] Therefore, surgical removal of ITMCC must be performed as soon as possible. Early diagnosis allows a complete removal by a transcanal or endaural approach and if the fibrous layer is not violated, neither myringoplasty nor tympanoplasty is necessary.[1,5)] Because it does not develop any symptoms in a small sized ITMCC, the lesion can go undetected for several years and can develop beyond the TM. Therefore, careful otoscopic examination is critical in the early diagnosis of ITMCC even in children with no ear symptoms. In the present case, cholesteatoma was centered on the umbo, but ITMCC was seen in all quadrants of the TM. ITMCC should be considered in the differential diagnosis of white TM lesions such as tympanosclerosis.[1,5)]

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