CASE REPORT
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Symmetric Bilateral Congenital Middle Ear Cholesteatoma: A Case Report

Joon Pyo Hong and Min-Beom Kim

Department of Otorhinolaryngology-Head and Neck Surgery, Kangbuk Samsung Hospital, Sungkyunkwan University School of Medicine, Seoul, Korea

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Address for correspondence
Min-Beom Kim, MD, PhD
Department of Otorhinolaryngology-Head and Neck Surgery,
Kangbuk Samsung Hospital,
Sungkyunkwan University
School of Medicine,
29 Saemunan-ro, Jongno-gu,
Seoul 03181, Korea
Tel +82-2-2001-2269
Fax +82-2-2001-2273
E-mail minbeom.kim@samsung.com

Congenital middle ear cholesteatoma (CMEC) accounts for 2%−5% of all cases of middle ear cholesteatoma. CMEC is rare and diagnostically challenging; therefore, only a few cases are reported in the literature, and the pathophysiology of this condition remains largely unclear. Currently, epidermoid formation, amniotic fluid contamination, lack of the tympanic ring, and squamous metaplasia are among the mechanisms implicated in the pathogenesis of CMEC; however, no single theory satisfactorily explains the pathophysiology of this disorder. We report a case of CMEC in a young male patient, who showed a symmetric, binaural whitish mass posterior to the anterosuperior quadrant of the tympanic membrane. A few reports have described bilateral CMEC; however, no study has reported symmetrical CMEC as observed in this case. Bilateral tympanoplasty concomitant with cholesteatoma removal was performed, and histopathological evaluation of the resected specimen showed closed-type cholesteatoma. In this report, we describe a rare case of symmetric CMEC, which supports the epidermoid formation theory associated with CMEC, together with a literature review.

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Keywords: Bilateral; Congenital; Cholesteatoma; Symmetrical.

Introduction

Congenital cholesteatoma of the middle ear is a relatively rare disease that accounts for about 2% of all cholesteatomas, and is found as a white mass inside the normal tympanic membrane [1]. Although the frequency of detection of congenital cholesteatoma is increasing due to the recent development of diagnostic technology, the mechanism of occurrence of congenital cholesteatoma has not yet been clearly elucidated. There are several known hypothesis including epidermoid formation theory, amniotic fluid contamination theory, lack of tympanic ring theory, and the epithelial metaplasia theory, but none of them are fully satisfactory [2-5].

The definition presented by Levenson, et al. in 1986 is currently the most widely used to diagnose congenital cholesteatoma. A whitish mass should be observed medial to the tympanic membrane and there should be no previous history of otorrhea, perforation, or otologic surgery. However, there is a limitation in that it is difficult to distinguish from acquired cholesteatoma when the lesion progresses and affects surrounding structures such as perforation or adhesion of tympanic membrane.

The authors experienced a unique case of bilateral congenital cholesteatoma, which occurred completely symmetrically in the middle ear cavity. Even though there have been few reports of bilateral congenital cholesteatoma [6-8], totally symmetric case like this has never be reported. The lesions were successfully removed through bilateral exploratory tympanotomy, and authors report this case as a supporting evidence of epidermoid formation theory.

Case Report

A young 3-year-old male visited our hospital with an incidentally founded whitish mass inside the tympanic membrane on both sides. He had no previous medical history and

experiences of otologic symptoms. On the endoscopic examination, a whitish mass was observed in the anterior superior quadrant of the tympanic membrane on both ear (Fig. 1), and the hearing threshold confirmed by play audiometry showed 21 dB in the right (Rt) side and 23 dB in the left (Lt) side (Fig. 2). Temporal bone computed tomography (CT) of the Rt ear showed a 2.5×3 mm sized round mass in the anterior portion of the malleus, and the Lt ear showed 3.5×3 mm sized round mass in the symmetrical location (Fig. 3).

Bilateral endoscopically assisted exploratory tympanotomy was performed. Operational findings of both ears were the same, 3 mm-sized well capsulated cholesteatoma originating from cochleariform process were observed in both the Rt (Fig. 4) and Lt (Fig. 5) side (Fig. 6). Ossicular invasion of the cholesteatomal stroma was not observed, and the mobility of the ossicles was intact. Both lesions were removed as a single mass,

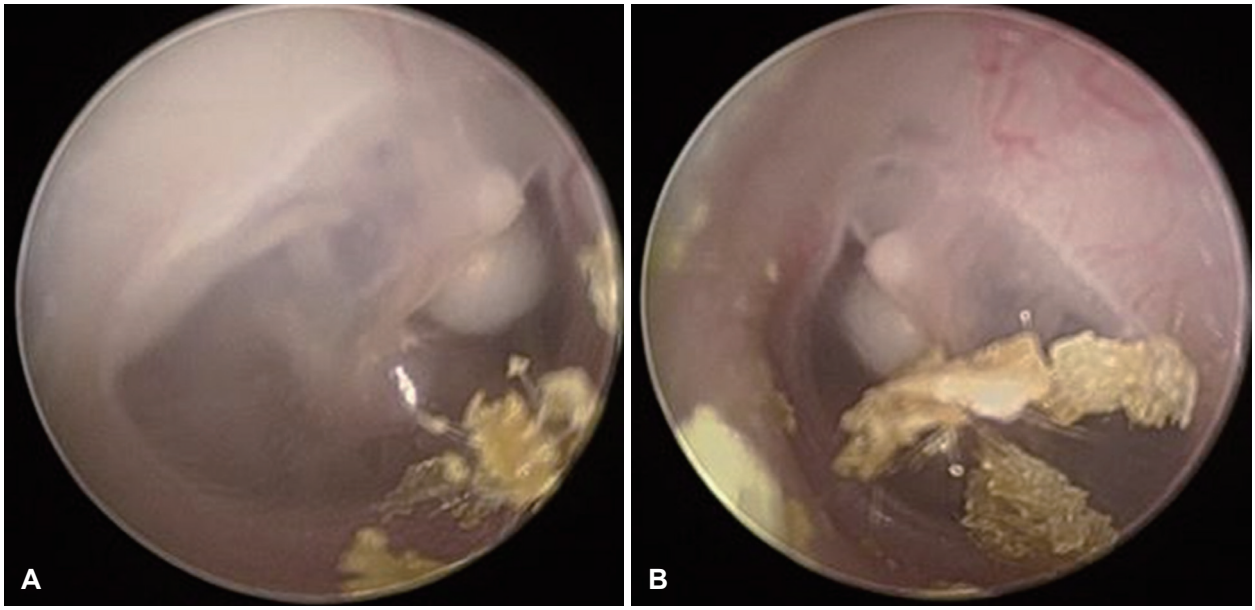

Fig. 1. Preoperative endoscopic finding. There were whitish round mass in right (A) and left (B) anterior superior tympanic membrane. Otherwise, there was no abnormality in both middle ear cavity.

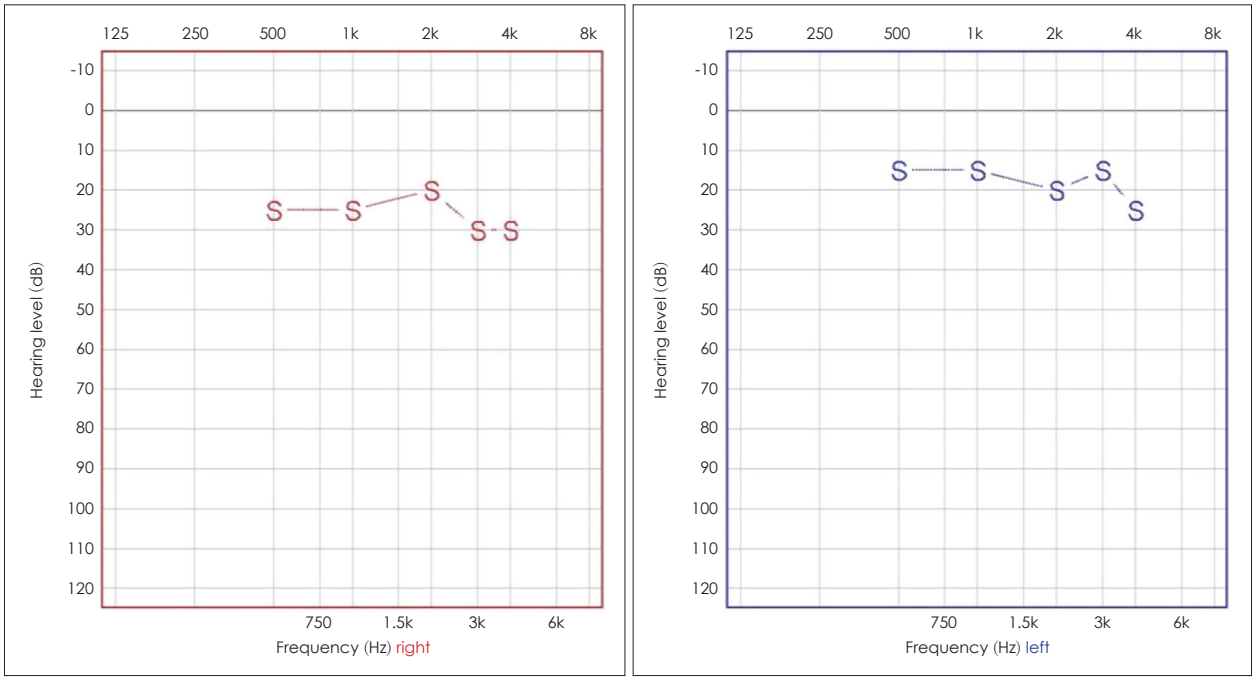

Fig. 2. Preoperative play audiometry. Auditory threshold was 24 dB (right), 18 dB (left). The test was performed under soundfield setting. S, soundfield.

and the operation was terminated after confirming no residual tissue in the middle ear cavity. Closed type cholesteatoma was confirmed by post-operative pathology report (Fig. 7). The endoscopic exam two months after the operation showed well adapted tympanic membrane without any evidence of recurrence (Fig. 8), and the audiometry confirmed the maintenance of hearing (Fig. 9). The patient is being followed up at our outpatient clinic without any postoperative complications for more than 1 year.

This report was approved by the Institutional Review Board of the Kangbuk Samsung Hospital (IRB File No. 2021-12-041), and the requirement of informed consent was waived.

Discussion

Embryologically, the middle ear is primarily formed when the

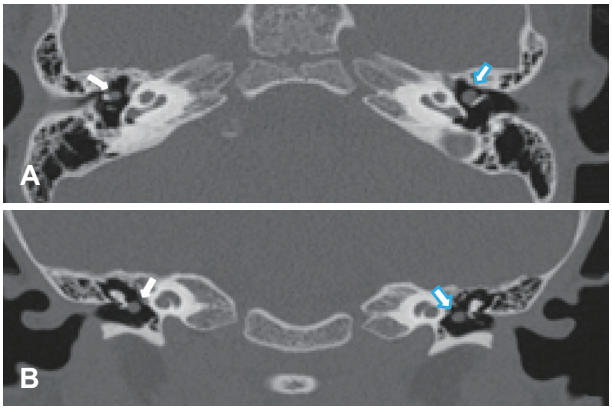

Fig. 3. Axial view (A) and coronal view (B) of CT scan shows bilateral round mass in both middle ear cavity. The mass were observed at same level in CT scan, and they were symmetrically located in anterior medial part of malleus. Right mass (white arrow) was 2.5×3 mm and left mass (blue arrow) was 3.5×3 mm sized. CT, computed tomography.

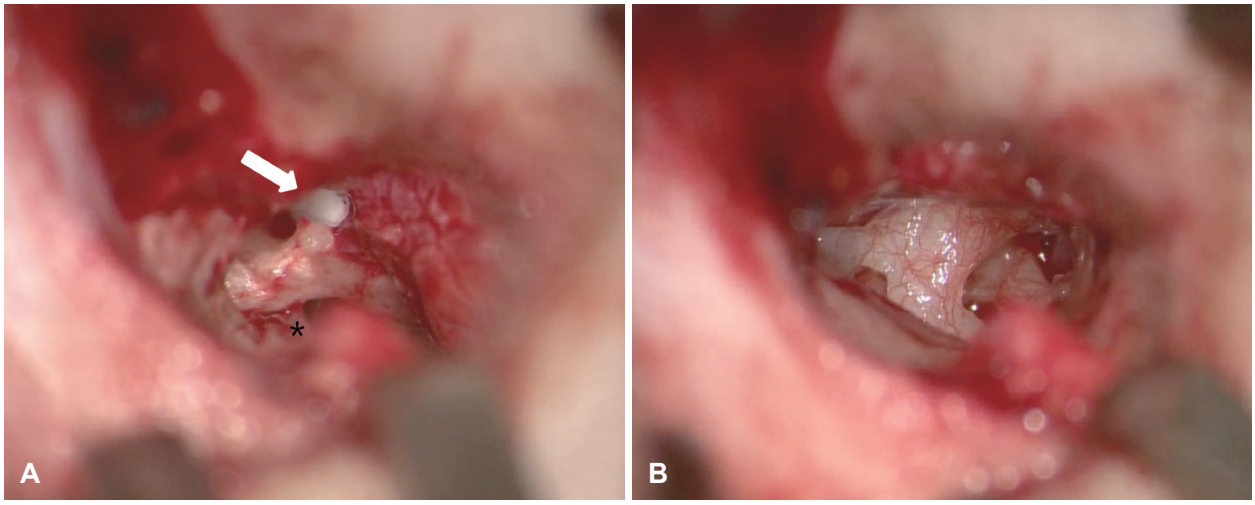

Fig. 4. Operative finding of right ear. (A) The mass (arrow) was located under the malleus handle. The origin of mass was cochleariform process (asterisk). (B) The mass was clearly removed without remnant tissue.

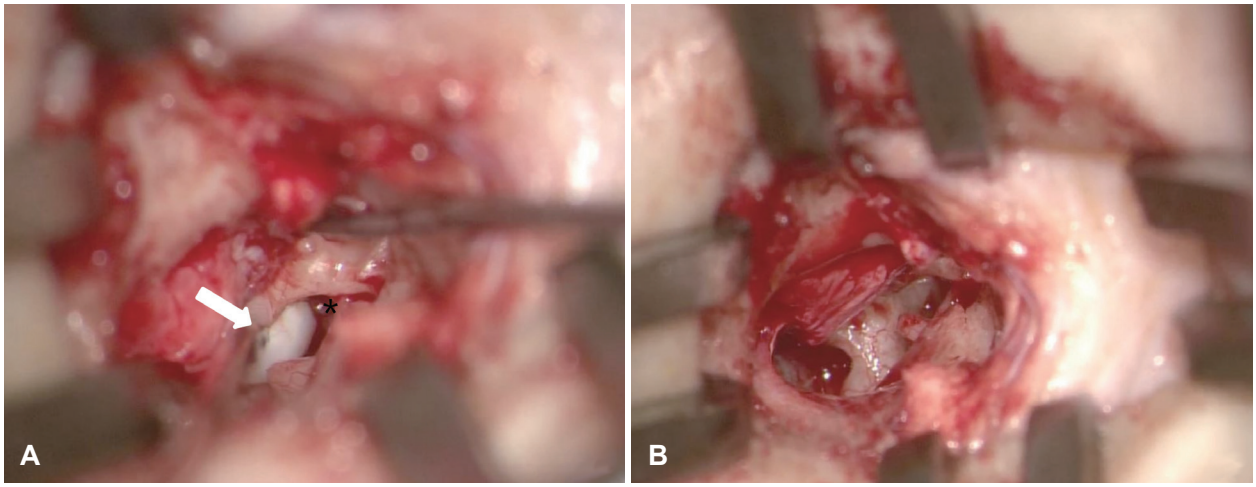

Fig. 5. Operative finding of left ear. (A) The mass (arrow) was located under the malleus handle and originated from cochleariform process (asterisk). which showed symmetric occurrence with right ear. (B) The mass was clearly removed without remnant tissue.

first pharyngeal pouch meet the first branchial cleft in 4 weeks, and after the continuous expansion, the formation is completed around 30−35 weeks of age [9]. In a study of 68 middle ear structures observed from fetuses between 10 and 33 weeks of age, Michaels reported that epidermoid formation occurred in the anterior superior middle ear mucosa near the tympanic membrane and disappeared around 33 weeks of age [4]. The epidermoid formation theory was first proposed that, in some cases, the fetus epidermoid can develop into congenital cholesteatoma if it remains and proliferates without degeneration.

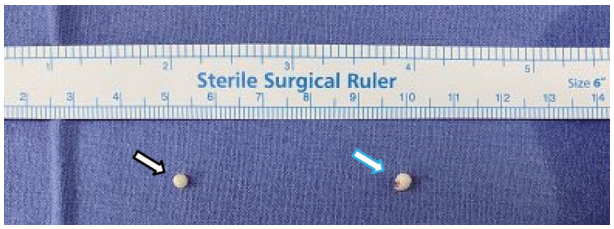

Fig. 6. Specimen photo. Specimen from right (black arrow) and left (blue arrow) middle ear cavity. There were symmetric 3 mm sized round whitish mass without adhesion to middle ear mucosa.

Initially, there was an opinion that the epidermoid formation theory was insufficient to explain cholesteatomas occurring in locations other than the anterior superior part of the tympanic membrane. However, despite vast majority of epidermoid formation truly does occurs in anterior superior quadrant, Liang, et al. [10] discovered that the formation can be seen in anywhere in temporal bone, and also small amount of an immune marker which related to epidermoid formation (CK 14) can be expressed in various locations in tympanic membrane, including posterior superior, anterior inferior, and posterior inferior quadrant [11]. Therefore, epidermoid formation theory overcame its limitation, and has now become the most widely accepted hypothesis among the mechanisms of development of congenital cholesteatoma. Nevertheless, other developmental hypothesis are also valid to explain various location of congenital middle ear cholesteatomas (CMECs), so more supplementary reports were needed for epidermoid formation theory.

In addition to the epidermal formation theory, there are several hypotheses that explain the development of congenital

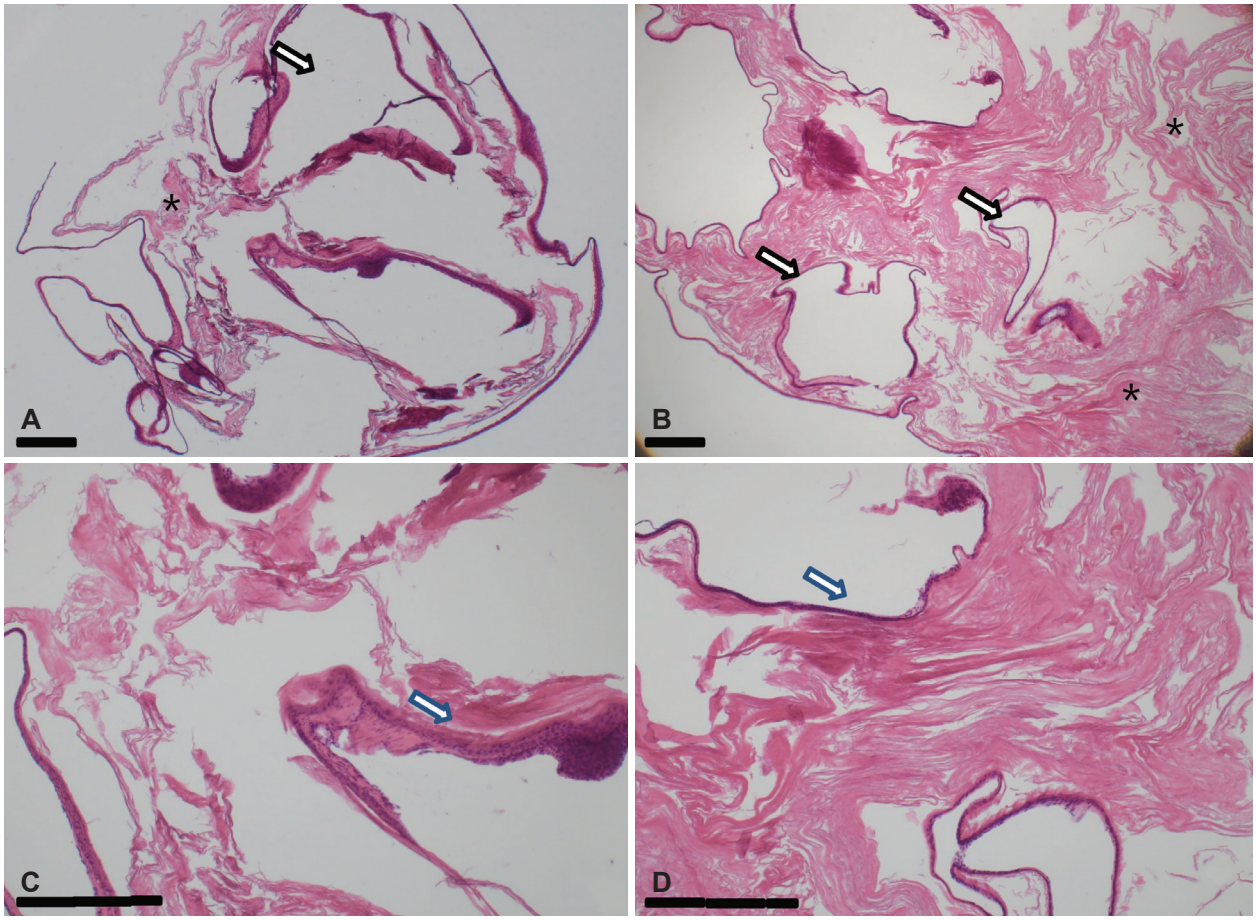

Fig. 7. Pathologic finding. Keratin tissues (asterisk) with cystic components (black arrow) were observed in both right (A) and left (B) specimen (H&E, ×40). Both right (C) and left (D) specimens showed intact epithelial lining (blue arrow) without evidence of foreign body reaction, suggesting closed type cholesteatoma (H&E, ×100). Scale bar=200 μm.

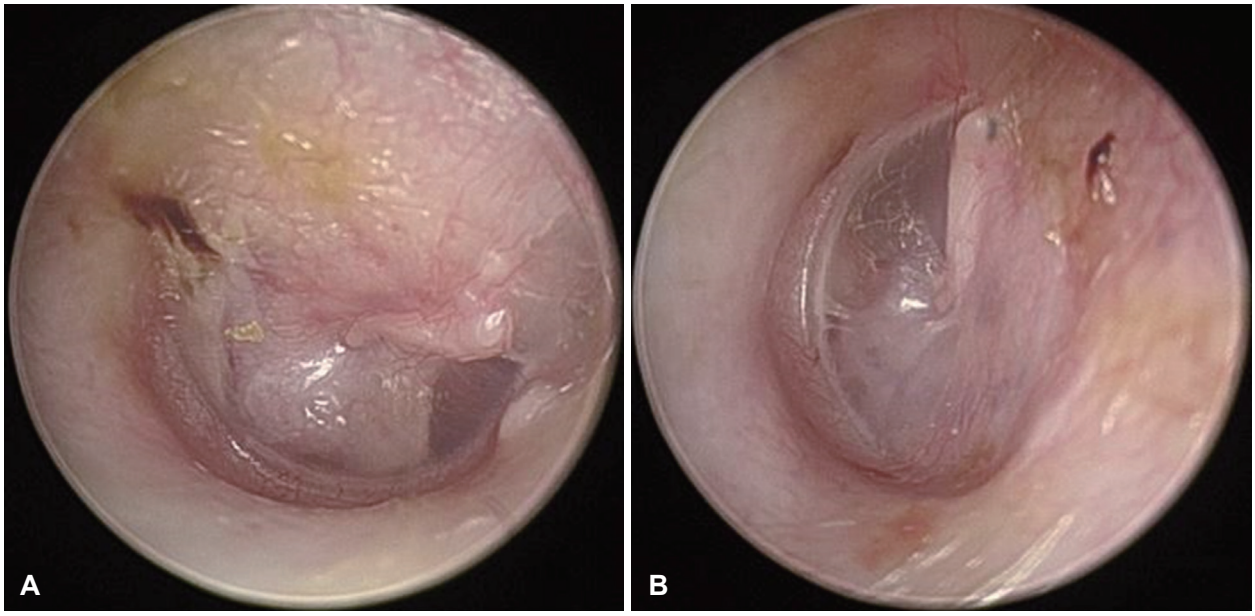

Fig. 8. Postoperative endoscopic finding after 2 month. Both right (A) and left (B) tympanic membrane were well formed after operation.

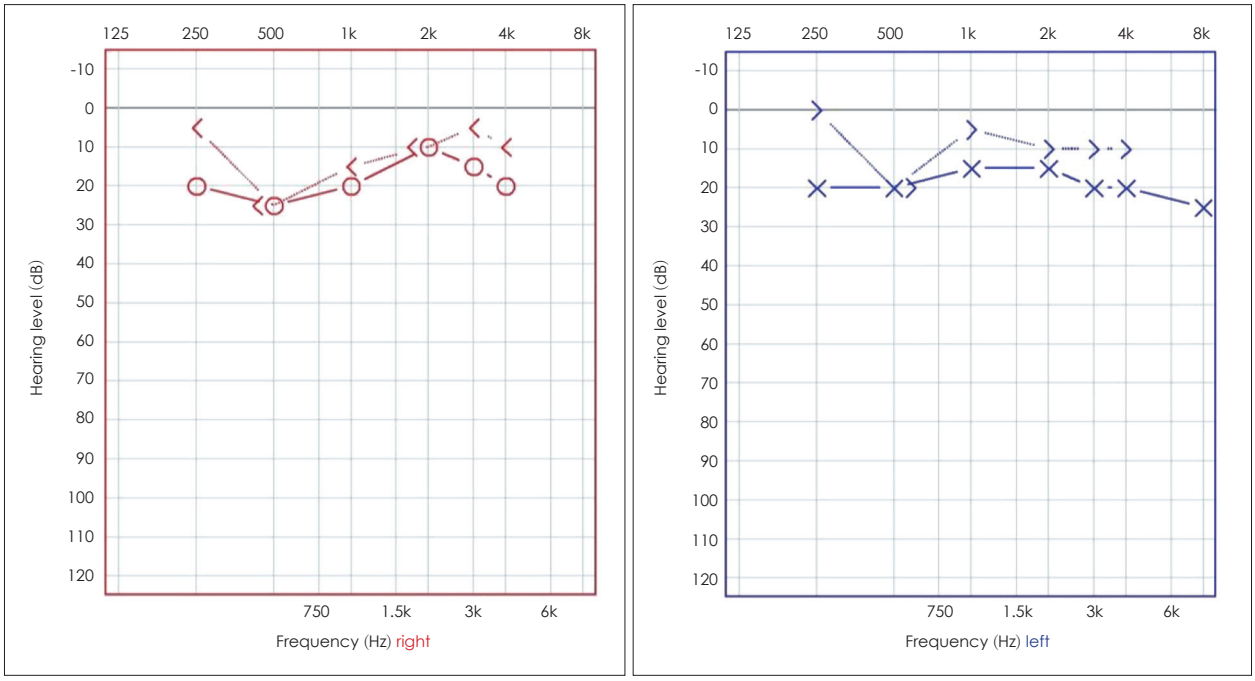

Fig. 9. Postoperative audiometry after 2 month. Auditory threshold was 18 dB (right), 17 dB (left). The thresholds were preserved in both ear after surgery. O, Air conduction of right ear. X, Air conduction of left ear. <, Bone conduction of right ear. >, Bone conduction of left ear.

cholesteatoma. Amniotic fluid contamination theory explains the formation of cholesteatoma by implantation of epidermal cells existing in the fetal amniotic fluid into the middle ear cavity through fetal eustachian tube, the tympanic ring defect theory explains that cholesteatoma occurs because the stop signal of the tympanic ring is not expressed in fetal period due to the absence of the tympanic ring, and the metaplasia theory explains the development of cholesteatoma as a metaplasia of mucosal cells in middle ear cavity to keratinized squamous epithelium.

There was no single hypothesis that could completely explains the pathogenesis of congenital cholesteatoma. Clinically, since the formation process of cholesteatoma in the fetus cannot be continuously observed, it is thought that the proof of the hypothesis would be very challenging, and the continuous research with a verification through case reports will be required.

Bilateral congenital cholesteatoma is so rare that only about 60 cases have been reported worldwide, and there has been no report in which cholesteatoma was found in the completely symmetric location in bilateral middle ear cavities. There were few clinical reports of bilateral CMECs [6-8] and one of them explained the situation by accompanied syndromic disease [12], but the authors could not find any clinical report emphasizing on specific developmental hypothesis of CMEC. This is probably because the mere fact that the CMECs were bilateral can be explained by all of hypothesis known so far. However, if the premise that bilateral CMECs happened completely symmetrically with identical size like this is added, it is hard to explain with mere coincidence. To explain this complete symmetrical CMEC case by epidermoid formation theory, the epidermoid formation of the fetus has occurred in same embryologic position as in normal growth process, and if the previously formed bilateral fetal epidermoid did not degenerate, symmetrical occurrence can be explained. In order to explain the symmetrical cholesteatomas by mechanisms other than epidermal formation theory, implantation, stop, or metaplasia of cells must have occurred in the exact same location in the middle ear cavity by chance, but the possibility is extremely low.

McGill, et al. [13] classified congenital cholesteatoma into closed type and open type according to histological type. In the case of open type, the matrix in the cholesteatoma adheres to the middle ear mucosa after invasion of the capsule so the surgical removal is relatively difficult, and a foreign body reaction accompanied by multinucleated giant cells is observed microscopically.

In 2003, Chang, et al. [14] retrospectively analyzed 38 cases of CMEC surgery and compared the prognosis between the location of the lesion and the surgical method for each Potsic stage. Also in 2013, Kim [15] presented a new Anterior quadrants-Posterior quadrants-Mastoid involvement-Recurred (APMR) stage of congenital cholesteatoma by retrospectively analyzing 90 cases of CMEC surgery, and presented a new systematic treatment plan according to the APMR stage. Based on the preceding studies, the authors performed bilateral exploratory tympanotomy in this case, and was able to complete the operation with type 1 tympanoplasty after successful removal of cholesteatoma.

The epidermoid formation theory alone will not be able to explain the development of all CMECs. In addition, since bilateral CMECs can occur in various locations, it is difficult to explain with only single theory. However, in the case of this completely symmetric bilateral CMECs experienced by the authors, the epidermoid formation theory seems to be the most acceptable hypothesis than other theories, and we present this interesting case with some references.

Acknowledgments
None

Conflicts of interest
The authors have no financial conflicts of interest.

Author Contributions
Conceptualization: Min-Beom Kim. Data curation: Joon Pyo Hong. Formal analysis: Min-Beom Kim, Joon Pyo Hong. Funding acquisition: Min-Beom Kim, Joon Pyo Hong. Investigation: Joon Pyo Hong. Methodology: Min-Beom Kim, Joon Pyo Hong. Project administration: Min-Beom Kim. Resources: Min-Beom Kim, Joon Pyo Hong. Software: Min-Beom Kim. Supervision: Min-Beom Kim. Validation: Min-Beom Kim, Joon Pyo Hong. Visualization: Joon Pyo Hong. Writing—original draft: Joon Pyo Hong. Writing—review & editing: Min-Beom Kim. Approval of final manuscript: all authors.

ORCID iDs
Joon Pyo Hong https://orcid.org/0000-0002-9745-6164
Min-Beom Kim https://orcid.org/0000-0001-8849-5148

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