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Case Report

Transcanal Endoscopic Ear Surgery for Pediatric Bilateral Congenital Cholesteatoma: A Report of Two Cases

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Bilateral congenital cholesteatoma (BCC) is rare among congenital cholesteatoma (CC) cases, and bilateral surgery poses a significant psychophysical burden on pediatric patients. Here, we present 2 pediatric cases of BCC that were successfully managed using simultaneous bilateral transcanal endoscopic ear surgery (TEES). The surgical approach provided clear visualization of the middle ear structures, enabling successful cholesteatoma removal with minimal invasiveness and a short operative time. In both cases, follow-up computed tomography showed no recurrence, and pure-tone audiometry results demonstrated good outcomes. Simultaneous bilateral TEES can be a viable option for pediatric BCC, as it reduces patient burden due to its less invasive procedure and short operative time.

KEYWORDS: Pediatric, congenital cholesteatoma, endoscopic ear surgery

INTRODUCTION

Congenital cholesteatoma (CC) is characterized by a mass of keratinizing epithelium ectopically located in the middle ear behind an intact tympanic membrane, with no history of otologic procedures or prior perforation. Bilateral congenital cholesteatoma (BCC) is rare; Lee et al[1] reported that BCC accounts for 1.8% of all CC cases. The treatment for CC is surgical removal; however, managing residual recurrence is challenging when lesions extend into the ossicles and mastoid.[2] In pediatric BCC cases, sequential surgeries, particularly those involving a postauricular incision, impose a significant psychophysical burden on the patient and delay the treatment of other lesions. An ideal surgical approach should minimize the invasiveness of simultaneous treatment on both sides. Additionally, it should provide an excellent field of view to prevent residual cholesteatomas from being overlooked.

Transcanal endoscopic ear surgery (TEES) is a minimally invasive approach that provides a clear view of the middle ear with few blind spots. Transcanal endoscopic ear surgery has been increasingly used to treat middle ear lesions and is comparable to postauricular surgery in terms of postoperative outcomes and recurrence rates.[3,4] Here, we present 2 cases of pediatric BCC in which TEES was successfully performed and their long-term postoperative follow-up.

CASE PRESENTATIONS

Case 1

A 5-year-old boy who presented with a whitish mass visible through the right tympanic membrane was referred to our hospital (Figure 1A and B). He had no complaints of aural symptoms and no history of otologic procedures. Computed tomography (CT) revealed a small, low-density mass in the middle ear on the contralateral side (Figure 2A and B), suggesting BCC.

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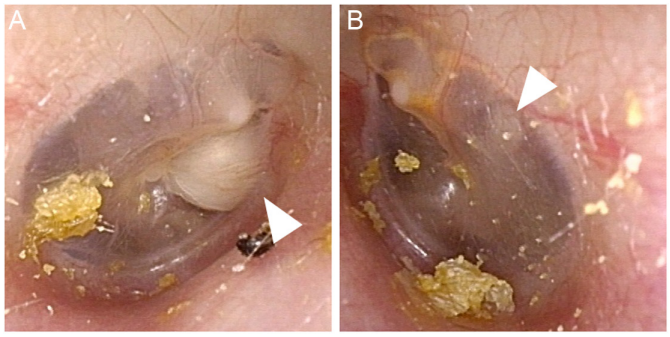

Figure 1. A, B. Preoperative endoscopic findings (case 1). (A) A white, round mass is observed behind the anterosuperior quadrant (ASQ) of the intact right tympanic membrane. (B) A white, round mass is observed behind the posterosuperior quadrant (PSQ) of the intact left tympanic membrane.

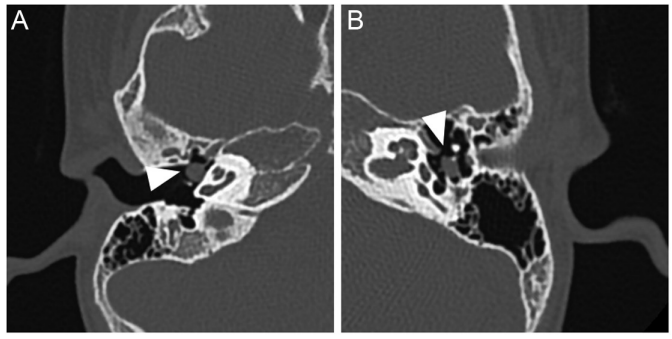

Figure 2. A, B. Preoperative CT findings (case 1). (A) A soft tissue mass is observed anterior to the malleus handle on the right side. (B) Soft tissue mass extending from the long process of the incus to the superstructure of the stapes on the left side.

A 2.7-mm-diameter endoscope with a working length of 11 cm and a 0° angle (Karl Storz Endoscopy Japan Co. Ltd., Tokyo) was used for surgery. The endoscope was connected to a high-definition camera and a monitor. A cholesteatoma measuring 3 mm in diameter was located anterior to the malleus and was removed from the surrounding tissue on the right side (Figure 3A). The tympanomeatal flap was then repositioned and covered with fibrin glue (Beriplast P Combi-Set; CSL Behring K.K., Tokyo, Japan). The left side was operated on consecutively. A cholesteatoma measuring 3 mm in diameter was identified between the stapes and the chorda tympani with an erosion of the incus. A portion of the horizontal segment of the facial nerve was exposed. The posterior portion of the tympanic sulcus was widened to visualize the ossicular chain and the cholesteatoma (Figure 3B). We extracted the incus and removed the cholesteatoma from the malleus, stapedius muscle tendon, and surrounding tissue. The remaining matrix was carefully dissected from the facial canal prominence to the stapes footplate. The remolded incus was placed on the head of the stapes as the columella. The operative time for the right and left side was 71 and 78 minutes, respectively, totaling 149 minutes. Computed tomography at 2.5 years postoperatively showed no recurrence. Pure-tone audiometry results for both ears demonstrated good outcomes at 3 years after surgery (Figure 4A). No abnormal findings were observed in the tympanic membrane at 3.5 years postoperatively.

Case 2

A 1-year, 8-month-old boy who presented with a whitish mass visible through the bilateral tympanic membranes was referred to our hospital. Computed tomography revealed small, low-density masses anterior to the malleus bilaterally. A cholesteatoma measuring 2.5 mm in diameter was observed anterior to the malleus without adhesion to the surrounding area and was easily removed. A cholesteatoma measuring 2.5 mm in diameter was also found anterior to the malleus and was similarly removed on the left side. The operative time for the left and right sides was 29 and 49 minutes, respectively, totaling 78 minutes. Computed tomography showed no recurrence 5 years postoperatively. Pure-tone audiometry results for both ears demonstrated good outcomes at 4.5 years after the surgery (Figure 4B). No abnormal findings were observed in the tympanic membrane at 5 years postoperatively.

The patients' legal guardians provided consent for the publication of this case report.

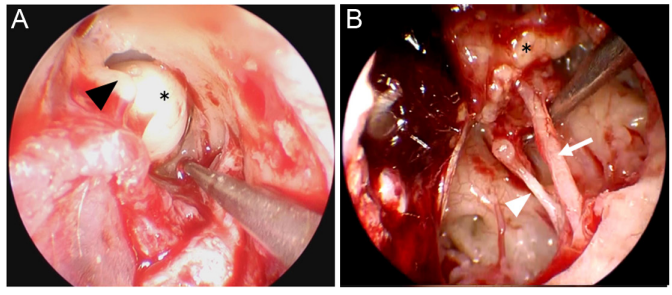

Figure 3. A, B. Intraoperative findings (case 1). (A) Endoscopic view shows a cholesteatoma anterior to the malleus on the right side. (B) The posterior portion of the tympanic sulcus is widened to visualize the ossicular chain and cholesteatoma on the left side. Asterisk: cholesteatoma, black arrowhead: malleus, white arrowhead: stapedius muscle tendon, white arrow: chorda tympani.

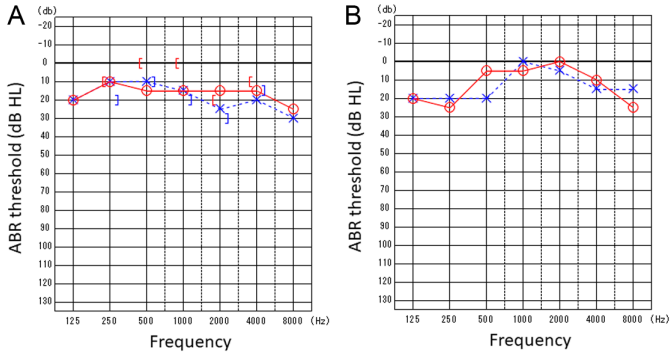

Figure 4. A, B. Postoperative pure-tone audiometry. Postoperative hearing results are good in both cases. (A) Three years after surgery (case 1: 8 years old). (B) Four and a half years after surgery (case 2: 6 years old).

MAIN POINTS

- Transcanal endoscopic ear surgery (TEES) is a minimally invasive approach that provides a clear view of the middle ear with few blind spots where congenital cholesteatoma can easily be left behind.
- TEES is a useful surgical technique in the selection of simultaneous bilateral surgery for the treatment of pediatric bilateral congenital cholesteatoma (BCC).
- We present 2 pediatric cases of BCC that were successfully managed with TEES with long-term outcomes.

DISCUSSION

Congenital cholesteatoma frequently occurs in the anterosuperior and posterosuperior quadrants, where it easily comes into contact with the ossicular chain or tensor tympani tendon.[5] We diagnosed the patient with CC based on typical tympanic membrane and CT findings; however, if the endoscopic finding is atypical and the preoperative diagnosis relies solely on imaging, combining CT and MRI is useful for an accurate diagnosis.[6] In pediatric cases, preservation of the tympanic membrane and external auditory canal morphology is desired; however, cholesteatomas often remain in blind spots and require revision surgeries. The burden of surgery in patients with bilateral CC has also increased.

The endoscopic approach provides a wide field of view and better visualization of the mesotympanic structures, such as the anterior part of the tensor tympani tendon, tympanic sinus, facial recess, and areas behind the ossicles, which may become blind spots with the microscopic approach. Transcanal endoscopic ear surgery is less invasive than postauricular approaches, making it easier to identify residual cholesteatoma and avoid damage to important structures by combining forward and oblique views. Dixon et al[4] reported that TEES was an effective alternative to postauricular surgery for pediatric cholesteatomas limited to the middle ear and attic region, with no significant difference in residual cholesteatoma rates. In a cohort study of patients with BCC, Lee et al[1] reported a shorter decline in postoperative hearing loss after simultaneous bilateral surgery than after sequential surgery. Therefore, performing simultaneous bilateral surgery using the less invasive TEES is considered beneficial. Transcanal endoscopic ear surgery has limitations when the cholesteatoma matrix extends beyond the attic into the antrum or other mastoid air cells.[4,5] Postauricular surgery is a more suitable approach for such cases. A narrow external auditory canal (EAC) diameter is another limitation of TEES.[7] However, Ito et al reported the mean smallest minimum Feret diameter of the EAC is 5.1 mm in children and 5.4 mm in adults, with no significant difference. Therefore, in the absence of EAC malformations, we consider that TEES can be performed using the 2.7 mm endoscope currently in use.[8] To determine the indications for TEES in pediatric BCC cases, it is essential to fully evaluate the extent of the cholesteatoma and the morphology of the EAC preoperatively using CT. Additionally, it is important to have a microscope on standby to switch the surgical approach to postauricular surgery, if necessary.

CONCLUSION

In this report, we presented 2 pediatric cases of BCC that were successfully managed with TEES, resulting in favorable long-term outcomes. Transcanal endoscopic ear surgery reduces the patient burden as it is a less invasive procedure and has a short operative time. Although BCCs are rare, simultaneous bilateral surgery using TEES is a viable option.

Informed Consent: Written informed consent was obtained from the patients' legal guardians who agreed to take part in the study.

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Declaration of Interests: The authors have no conflicts of interest to declare.

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