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CASE REPORT

Simultaneous Open- and Closed-type Congenital Cholesteatomas in the Middle Ear Removed Using an Otoendoscope and Surgical Microscope

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ABSTRACT

Aim: Open- and closed-type congenital cholesteatoma in the middle ear can be found at the same time and an ear endoscope can be useful to identify and remove residual cholesteatoma during surgery.

Background: Congenital cholesteatoma of the middle ear usually manifests as a single growth. Open- and closed-type congenital cholesteatomas occurring simultaneously in the middle ear on one side are extremely rare. In recent years, the use of endoscopes for middle ear surgery has been gradually increasing.

Case description: This report describes the case of a 6-year-old boy in whom we observed both open-type and closed-type congenital cholesteatomas in the right middle ear. The closed-type congenital cholesteatoma extended from the malleus to the posterior wall of the tympanic cavity and had destroyed the long process of the incus and crura of the stapes. An open-type congenital cholesteatoma was observed on the medial side of the malleus, supratubal recess, and attic walls. The cholesteatoma was removed successfully via a transmeatal approach using an ear endoscope and surgical microscope. Using the ear endoscope, we could identify and completely remove the residues of cholesteatoma in the epitympanum, posterior tympanum, and supratubal recess.

Conclusion: It is rare for both open-type and closed-type cholesteatomas to exist simultaneously in the middle ear, as in this case. Although the cholesteatomas were more extensive than the single lesion that usually occurs, both could be removed successfully via a transmeatal approach using a surgical microscope and an ear endoscope.

Clinical significance: It is important to keep in mind the possibility of simultaneous open-type and closed-type congenital cholesteatomas in the middle ear, and the use of endoscopes can reduce the extent of the surgery and help identify residual cholesteatoma.

Keywords: Cholesteatoma, Congenital, Endoscope, Middle ear.

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BACKGROUND

Congenital cholesteatoma in the middle ear can be defined as open- or closed-type depending on histopathologic classification and is a relatively rare disease that occurs in 0.12 per 100,000 people annually.[1,2] Congenital cholesteatoma usually manifests as a single growth, so it is extremely rare for both the open and closed types to be observed in the middle ear simultaneously.[3] Congenital cholesteatoma in the middle ear can be removed via a transmeatal approach. However, this technique is difficult using a surgical microscope alone when the mass is present on the attic or posterior wall of the tympanic cavity.

For this reason, mastoidectomy is usually performed in such cases; more recently, the otoendoscope has increasingly been used.[4,5] Specifically, when the matrix of the cholesteatoma is difficult to observe using a surgical microscope, it can be viewed using an otoendoscope and removed, or its residues can be identified. In the present case, both open-type and closed-type congenital cholesteatomas were observed simultaneously in a child's right middle ear. The masses were removed successfully via a transmeatal approach under the guidance of both a surgical microscope and an otoendoscope.

CASE DESCRIPTION

The patient was a 6-year-old boy in whom a white mass was incidentally observed in the right tympanic membrane while he was brought to a private clinic with the flu-like symptom. He was immediately referred to the otolaryngology department at our hospital. According to his mother, the boy had never had otorrhea or undergone ear surgery. Otoscopy revealed a round, white mass with a smooth surface at the back of the tympanic membrane. A light-yellow mass with a jagged maple leaf-like appearance was also observed at the front of the tympanic membrane (Fig. 1). The left tympanic membrane was normal.

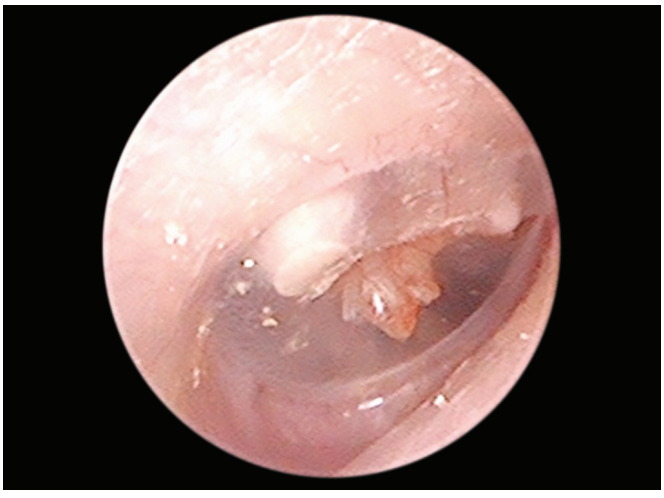


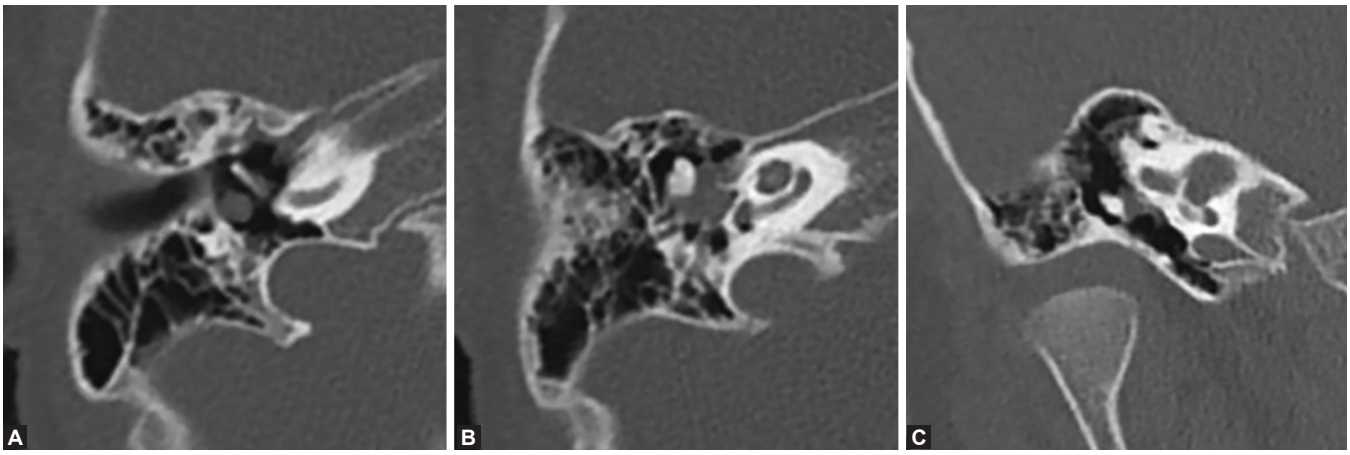
Fig. 1: Preoperative findings on otoscopy of the tympanic membrane. The otoscopic image shows two masses in the middle ear. One was a round and white mass at the back of the handle of the malleus, and the other was a light-yellow mass with a jagged maple leaf-like appearance at the front of the handle of the malleus

Auditory acuity was measured by pure-tone audiometry that showed an air conduction hearing threshold of 27 dB and a bone conduction hearing threshold of 7 dB indicating a mild conductive hearing loss in the right ear. Computed tomography (CT) of the temporal bone showed good pneumatization of the mastoid on both sides, round soft tissue shades in the posterior wall of the right tympanic cavity (Fig. 2A), and irregular soft tissue shades with a rough surface across the mesotympanum, protympanum (Fig. 2B), and attic walls of the tympanic cavity (Fig. 2C). Congenital cholesteatoma was diagnosed based on the medical history, findings for the tympanic membrane, and the results of temporal bone CT. A tympanotomy was performed under general anesthesia via a transmeatal approach. After induction of general anesthesia, an incision was made in the skin of the ear canal in a 12 o'clock to 6 o'clock direction, passing through 9 o'clock. The ear canal skin flap was then elevated; to prevent lateralization of the tympanic membrane after the operation, the tympanic membrane was not separated from the handle of the malleus. The posterior wall of the ear canal was widened using a surgical drill to expose the posterior tympanum. A closed-type cholesteatoma extending from the malleus to the posterior wall of the tympanic cavity was then observed (Fig. 3A) and, an open-type cholesteatoma was also observed on the medial side of the malleus (Fig. 3B) and across the anterior tympanum and attic walls (Fig. 3C). The open-type and closed-type cholesteatomas were in contact with one another in the anterior tympanic isthmus but were easily separated. Consistent with the temporal bone CT findings, neither the long process of the incus nor the superstructure of the stapes were observed during surgery. The incus with an eroded long process was removed, after which the cholesteatoma was observed and removed using the surgical microscope. Next, the middle ear was observed using the otoendoscope, and the residues of the cholesteatoma in the posterior area of the stapes footplate (Figs 4A and B), medial side of the head and neck of the malleus, and supratubal recess were checked (Fig. 4C) and removed under the otoendoscope (Fig. 4D). The footplate of the stapes and tympanic membrane were connected by implantation of the patient's native incus, and the area between the tympanic membrane and the implanted incus, as well as the extended posterior wall of the ear canal, were reinforced using tragal cartilage.

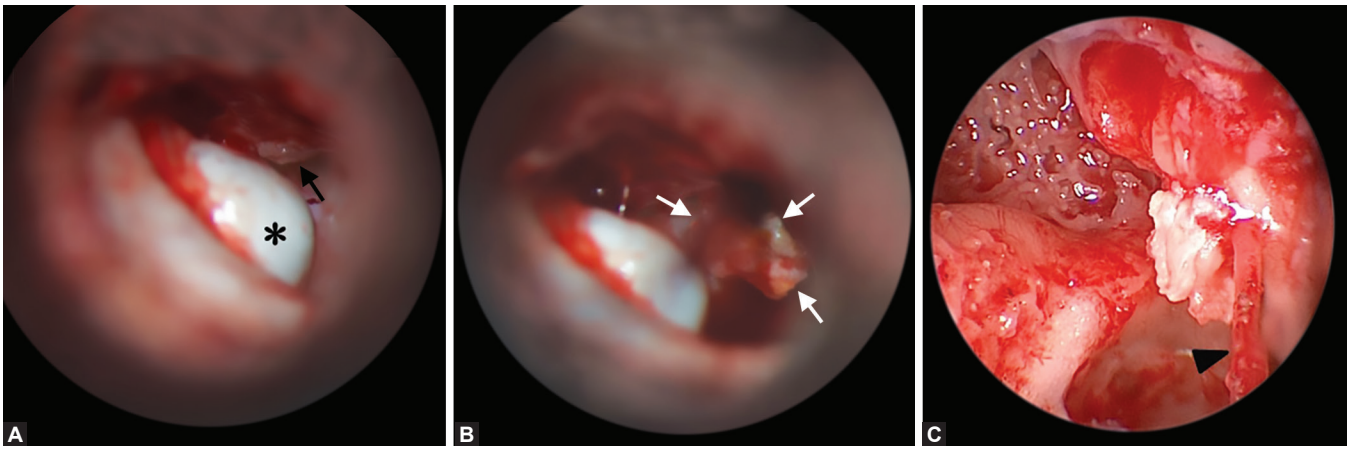
Tympanic membrane findings and CT images of the temporal bone obtained 2 years postoperatively showed that there had been no signs of recurrence of cholesteatoma (Fig. 5). At this time, pure-tone audiometry in the right ear was 18 dB, indicating normal auditory acuity.

DISCUSSION

It is not clear how an open-type and a closed-type congenital cholesteatoma could have developed simultaneously in the middle ear on one side in this patient.



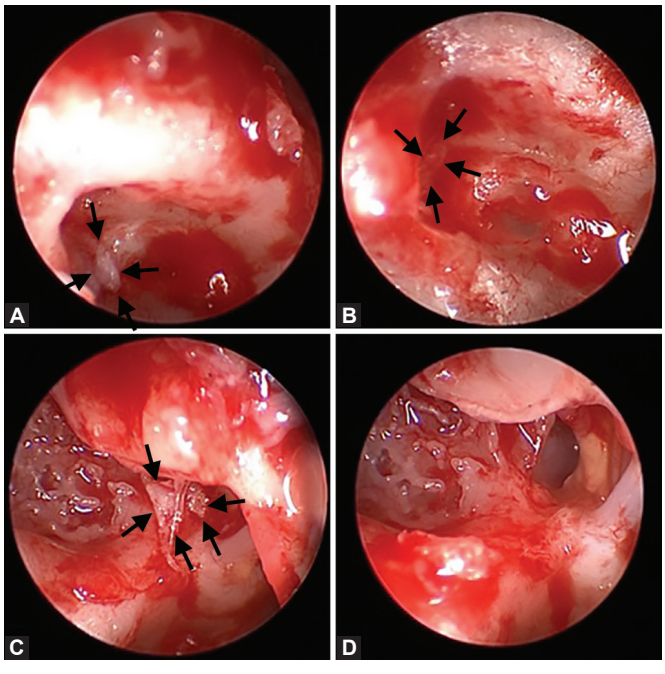
Figs 2A to C: Preoperative findings on computed tomography (CT) scans of the temporal bone. (A) Round soft tissue shades in the posterior wall of the right tympanic cavity; (B and C) Soft tissue shades with a rough surface on the medial side of the right malleus and incus. A and B are axial views and C is a coronal view



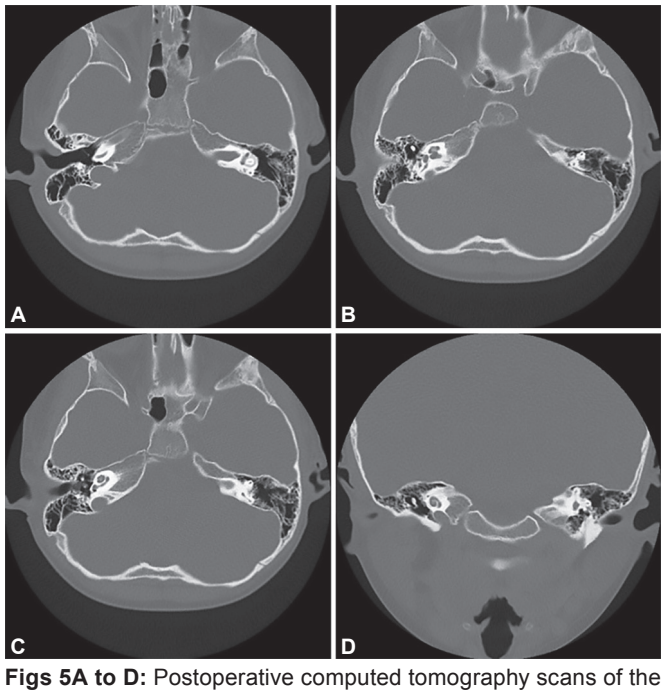
Figs 3A to C: Intraoperative findings using (A and B) the surgical microscope and (C) otoendoscope. (A) A surgical microscopic image acquired after the elevation of the tympanic membrane shows a closed-type congenital cholesteatoma (asterisk) and an open-type congenital cholesteatoma (arrow); (B) The open-type congenital cholesteatoma (arrows) was removed first, leaving the closed-type congenital cholesteatoma intact; (C) An otoendoscopic image shows part of the open-type congenital cholesteatoma on the medial side of the head and neck of the malleus. Black arrowhead indicates the chorda tympani nerve.

However, four possibilities should be considered. First, two closed-type congenital cholesteatomas may have occurred and grown in size, and one could have ruptured subsequently and become an open-type mass. Second, a closed-type congenital cholesteatoma may have occurred, grown in size, and then ruptured, allowing the release of keratin from the cyst and formation of an open-type cholesteatoma. Subsequently, the site of rupture of the closed-type mass could have healed and become a cyst, i.e., a closed-type cholesteatoma that existed in contact with the open-type mass. Third, the closed-type and open-type congenital cholesteatoma may have occurred independently, grown in size, and come into contact with one another. Fourth, an open-type congenital cholesteatoma could have occurred and grown in size, with part of the cholesteatoma subsequently becoming a cyst (closed-type)and the rest of the cholesteatoma remaining of the open-type.

There have been reports of closed-type congenital cholesteatomas growing in size over time, rupturing, and then becoming open-type, consistent with the first and second mechanisms suggested above.[6,7] In other reports, the matrix of the cholesteatoma or desquamating keratin debris exposed outside the cyst in open-type congenital



Figs 4A to D: Intraoperative findings using otoendoscope. (A and B) After the closed-type congenital cholesteatoma had been removed using the surgical microscope, some cholesteatoma matrix (arrows) was observed in the posterior margin of the stapes footplate under the otoendoscope; (C) After the open-type congenital cholesteatoma had been removed using the surgical microscope, some cholesteatoma matrix (arrows) was observed in the upper part of the tensor tympani (arrowhead) under the otoendoscope; (D) It was confirmed using the otoendoscope that no cholesteatoma residues remained in the supratubal recess.



Figs 5A to D: Postoperative computed tomography scans of the temporal bone 2 years after surgery indicate no recurrence of cholesteatoma

cholesteatomas induced an inflammatory response in the mucos membrane of the middle ear. This led to granulation, which allowed the matrix of the cholesteatoma to grow such that an open-type cholesteatoma formed a cyst and became closed-type, as described in the fourth mechanism suggested above.[8,9]

According to the staging system introduced by Potsic et al.,[10,11] our patient had stage III cholesteatoma because of the erosion of the ossicles without invasion of the cholesteatoma into the mastoid. In such cases, even when the cholesteatoma does not invade the mastoid, mastoidectomy or posterior tympanotomy is often performed; alternatively, two-stage operations may be used to completely remove the cholesteatoma in the middle ear.[5,12,13] Combination of an otoendoscope and a transmeatal approach had several strengths over the use of a surgical microscope alone. For example, the otoendoscope passes through a narrow area in the outer ear canal, making it easier to observe lesions situated in the corner of the middle ear, such as in the attic or posterior wall of the tympanic cavity. Furthermore, it provides the surgeon with a better surgical field of view by magnifying the lesion at a closer distance.[4] Accordingly, although the child in the present case had a narrow ear canal, we were able to completely remove the cholesteatoma from the anterior wall, attic, and posterior wall of the tympanic cavity using a transmeatal approach under the guidance of both a surgical microscope and an otoendoscope.

CONCLUSION

A congenital cholesteatoma usually manifests as a single-growth, and the closed type occurs more frequently than the open type. It is rare for both open-type and closed-type cholesteatomas to exist simultaneously in the middle ear, as in this case. In our patient, the use of an otoendoscope helped the surgeons to remove the cholesteatoma from inside the middle ear, which has a narrow and complex structure, using a transmeatal approach alone. The otoendoscope also allowed the surgeons to identify residues.

CLINICAL SIGNIFICANCE

When performing physical examination and surgery, we should keep in mind that close type cholesteatoma and open type cholesteatoma can be rarely found at the same time. The closed type cholesteatoma was located in the posterior tympanum, although the margin of the lesion was clear. The open type cholesteatoma was located in the epitympanum and supratubal recesses without definite lesion boundaries. Therefore, it was difficult to confirm all the lesions with only the surgical microscope. The 6-year-old patient's ear canal was relatively narrow. However, by using an endoscope, the cholesteatoma could be completely removed by the transmeatal approach alone.

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