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CASE REPORT Open Access

Bilateral congenital cholesteatoma: a case report

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Abstract

Background Cholesteatoma is regarded as a critical health issue in the world. Nevertheless, congenital cholesteatoma (CC) is an unusual problem, and bilateral CC is extremely rare.

Case presentation The present work reports a young boy with conductive hearing loss because of CC. The left side was operated using a canal-wall-down mastoidectomy method for immense destruction of the middle ear structures and mastoid air cells, and endoscopic ear surgery was performed on the second side.

Conclusion Although CC is very rare, it should be considered in the differential diagnosis of hearing loss in children. Moreover, it should be carefully followed up because it can probably occur on the second side.

Keywords Middle ear, Congenital cholesteatoma, Ear surgery

Background

Congenital cholesteatoma of the middle ear is defined as a keratinizing epithelial remnant found medial to a healthy tympanic membrane without a previous history of infection or trauma. Although its cause is not completely clear, several theories have been proposed to explain its origin [1, 2]. Generally, only one ear is involved in this disease, and bilateral congenital cholesteatoma is extremely rare. However, there are rare reports on congenital bilateral cases, including a few cases in the past decades [3–6].

Bilateral CC can cause difficulties in surgical and clinical management. Since it differs from acquired cholesteatoma, the parents of the patient child would not know about the cholesteatoma presence by malodorous otorrhoea, and this disease is generally diagnosed by otoscopy as an incidental finding. Maintaining a standard hearing threshold is challenging not only due to the difficulty in easy detection but also because of the lesion's location. A cholesteatoma often is seen in the super anterior mesotympanum and may spread to the medial epitympanum. In this case, its removal by surgery without ossicular damage will be difficult [7].

With progress in otologic surgery during the past decade, new horizons have been opened for optimizing care for children suffering from bilateral CC. The use of an endoscope can reduce the need for secondary surgery. Herein, we present the case of a young male with bilateral CC as an educational example and a reminder to consider the intense follow-up of children with CC.

Case presentation

The pediatrician referred a 4-year-old boy to an Ear Nose and Throat Clinic to be evaluated for hearing loss in his left ear. The left ear canal showed a polypoid mass, and it was impossible to observe the tympanic membrane via otoscopy. The right ear examination showed normal middle ear. Moderate hearing loss was noted in the left ear through the auditory brainstem response testing, and the right ear was normal. The right and left ear tympanometry presented type A and type B, respectively.

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It was planned to remove suspected CC, and thus, he was sent to the surgery room. Mesotympanum, hypotympanum, and epitympanum showed extensive cholesteatoma. Erosion of the malleus was observed, and it was removed. Incus and stapes superstructure were destructed due to cholesteatoma. Because of massive spread of cholesteatoma, we have to perform canal-wall-down mastoidectomy for better visualization and eradicate cholesteatoma from the middle ear. No erosion was found in the facial nerve and lateral semicircular canal. After the first surgery during 24 months' follow-up, the left open cavity was stable, and the graft was normal. But a white shadow was found behind the right eardrum (Fig. 1).

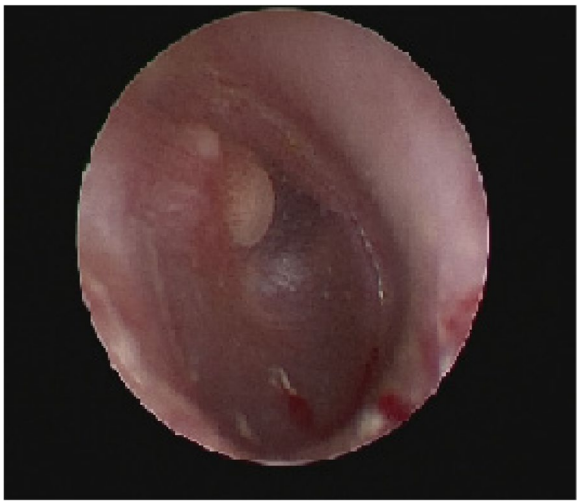

Fig. 1 Otoscopic presentation of the right ear

After performing a temporal computed tomography scan, we saw a little hyper dense mass in the right middle ear cavity. We also observed the findings of the previous surgery in the left ear (Fig. 2).

A hearing examination revealed the presence of moderate conductive hearing impairment in the left ear, while the right ear exhibited normal auditory function. Consequently, the second ear surgery was performed to remove the suspected CC observed in the right ear by the endoscopic transcanal approach.

A pearl of cholesteatoma was detected during surgery in the medial ear anteromedial to the malleus. An endoscopic transcanal method was used for its removal (Figs. 3 and 4). During the postoperative follow-up, the patient had no problem but a moderate hearing loss on the left side.

Discussion

Bilateral congenital cholesteatoma (BCC) presents unique diagnostic challenges and requires the expertise of a pediatrician. The pathophysiology of BCC is still not fully understood, with various theories proposed to explain its development. These include penetration of squamous epithelium through microscopic tympanic injury, metaplastic transformation of middle ear mucosa, and development from embryonic tissues.

The clinical presentation of BCC often includes routine otoscopic examination, recurrent otitis media, and hearing loss. Diagnosing BCC involves otoscopic or microscopic examination, which reveals a white mass deep to an intact tympanic membrane. However, effusion can sometimes obscure this finding. Further evaluation through imaging techniques such as temporal

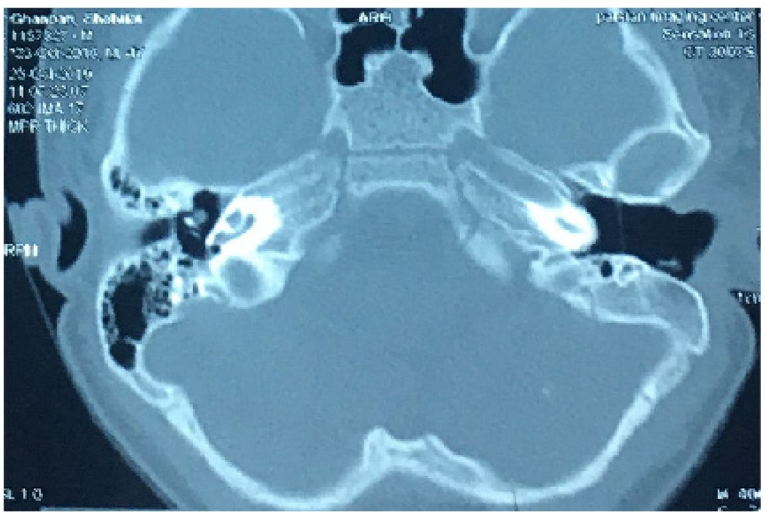

Fig. 2 Axial temporal computed tomography scan indicating a density of soft tissue medial to the malleus

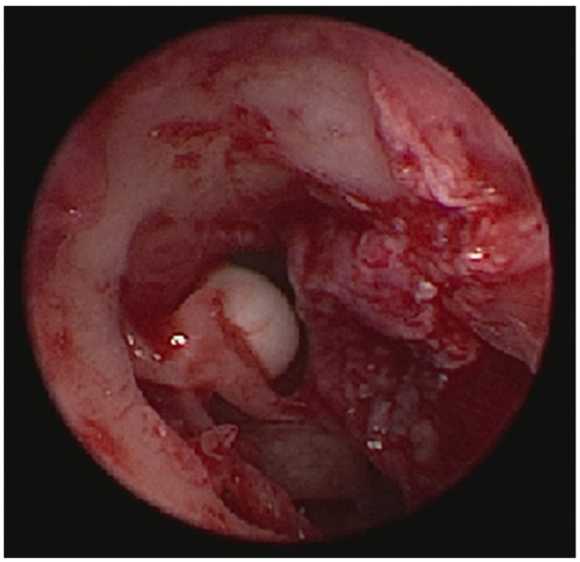

Fig. 3 Pearl of cholesteatoma in the right middle ear

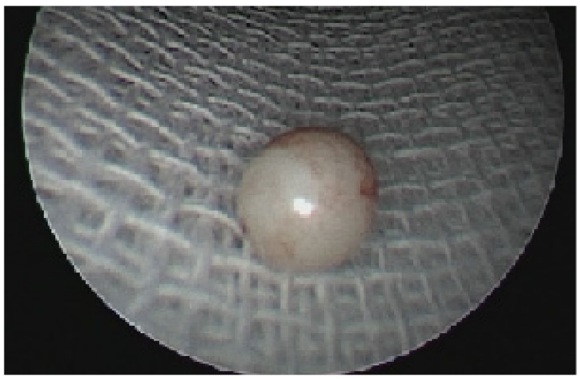

Fig. 4 Cholesteatoma view

bone CT or diffusion-weighted MRI can provide additional information, but definitive diagnosis is made through direct surgical visualization [8].

Bilateral congenital cholesteatoma is an extremely rare condition, with a global report of only approximately 60 documented cases. Some of which demonstrate ambiguous presentations, including otitis media with effusion [9]. The literature review revealed that the average (standard deviation) age at diagnosis for bilateral congenital cholesteatoma (BCC) was 7.77 years, with a male-to-female ratio of 8.3 to 1. Among cases where the chief complaint or reason for presentation was mentioned, routine otoscopic examination accounted for the highest proportion (29%), followed by recurrent otitis media (24%) and hearing loss (19%). Because of bilateral involvement, it is essential to preserve hearing, and its early detection is crucial. In our case, the second side CC was early detected due to the microscopic examination and close follow-up. As a result, an endoscopic operation could be conducted without postoperative hearing loss [10].

Hypothetically, it is possible to consider an ear affected by CC a normal ear physiologically and anatomically because the cholesteatoma has a congenital origin, and it is not because of dysfunction in the auditory tube or mastoid. The mastoid air cells in young pediatrics (age range: 3–6 years) with CC are pneumatized well with few large air cells. It structurally differs from adults' completely developed normal mastoids and nearly unpneumatized mastoids of children with chronic otitis media [9].

Ideally, it should be attempted to maintain normal physiology and anatomy of the middle ear, so the problem is eradicated from the latent parts (that is, protympanum, retrotympanum, supratubal recess) without disturbing ventilation routes or massive mucosa removal. It is possible to use a transmastoid posterior tympanotomy for a posteriorly located mass to have accessibility to the facial recess. However, in this way, the sinus subtympanicum and sinus tympani checkups are not facilitated [11]. Hence, it would be more effective to apply an endoscopic transcanal method to the retrotympanum.

With the introduction of completely transcanal endoscopic surgery, postoperative complications have been reduced, and it has brought about various advantages, including preventing post-ear incision pain, long-term bone work in the external auditory (EAC) canal and mastoid, soft tissue dissection, and rare complications of such interventions.

One of the benefits of an endoscope used in ear operation is that the probability of residual cholesteatoma is minimized [12]. Furthermore, the increased possibility of the complete elimination of the disease achieved by endoscopic visualization more clearly in hidden ear recess and fissures is significant. In addition, with endoscopic access to the medial attic, cholesteatoma clearance is increased without damage to ear ossicles, which leads to improved hearing results [13].

Conclusion

Despite the rare incidence of bilateral CC, there should be a precise management design because of the postoperative hearing complications. It is possible to add endoscopic operation to the management strategy, which results in control of hidden parts and successful surgery, while bone chains and mastoid cells are kept intact.

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Authors' contributions
NN manage the patient. MMR performed the radiologic examination of the patient, MA was a major contributor in writing the manuscript, and all authors read and approved the final manuscript.

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Declarations

Ethics approval and consent to participate
This study was approved by the Ethical Committee of Mashhad University of Medical Sciences.

Consent for publication
Written informed consent was obtained from the patient regarding participation and the anonymous publication of the results.

Competing interests
The authors declare that they have no competing interests.

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