CASE REPORT
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Large Congenital Cholesteatoma of Mastoid Origin Presenting as a Stenosis of the External Auditory Canal

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ABSTRACT

Congenital cholesteatomas represent a small part of all cholesteatomas and only rarely originate in the mastoid process. As such lesions grow slowly, they often remain asymptomatic for a long period. We report a case of a large congenital cholesteatoma of the mastoid process with invasion of the posterior semicircular canal, presenting as a stenosis of the external auditory canal. The aim of this case report is to describe the clinical and radiological features and to discuss its management.

Keywords: Cholesteatoma, congenital, mastoid, otologic surgical procedures

Introduction

A congenital cholesteatoma (CC) is a slow-growing lesion behind an intact tympanic membrane originating from remnants of keratinizing squamous epithelium in the temporal bone.[1,2] Congenital cholesteatoma can be found at various sites within the temporal bone. The most common location is the middle ear cleft. The mastoid process is a rare site of origin, with only a few reports worldwide.[3] Due to early conductive hearing loss by ossicular chain involvement, CC of the middle ear is mostly diagnosed during childhood.[1] Congenital cholesteatomas of the mastoid process on the other hand may be silent lesions for years.[3-5] As a result, they often have the time to cause substantial destruction of the temporal bone. Surgery may be challenging due to the involvement of the facial nerve, sigmoid sinus, dura, labyrinth, or external auditory canal.[3,5] Here, we present a case report of a large congenital cholesteatoma of mastoid origin causing stenosis of the external auditory canal.

Case Presentation

An 18-year-old woman presented at the ENT department with complaints of aural fullness and hearing loss on the right side. Except for ventilation tubes in early childhood, there was no history of otological interventions. On physical examination, otomicroscopy showed an extremely narrow external auditory canal, impeding the evaluation of the tympanic membrane. Pure-tone average (PTA), calculated for the 0.5, 1, 2, and 4 kHz hearing thresholds, was normal on both sides (left 10.0 and right 13.75 dB HL). Multislice computed tomography (MSCT) showed an expansile lesion in the mastoid process, with destruction of a large portion of the external auditory canal, the bony plate of the sigmoid sinus, and the mastoid segment of the facial nerve canal. Invasion of the otic capsule caused dehiscence of the posterior semicircular canal. The middle ear, attic, and antrum were free of pathology (Figure 1). Subsequent non-echo-planar diffusion-weighted imaging (non-EP DWI) confirmed the presence of a large cholesteatoma (Figure 2a). Based on the above findings the lesion was diagnosed as a CC of mastoid origin. A transmastoid procedure under general anesthesia with facial nerve monitoring was performed to remove the cholesteatoma and to rebuild the ear canal. As expected, a large portion of the mastoid cavity was filled with cholesteatoma. Only a small remnant of the posterior wall of the external auditory canal was left. By careful dissection, the matrix could be removed from the dura, sigmoid sinus, epineurium of the mastoid segment of the facial nerve, and eventually from the dehiscent posterior semicircular canal. The fistula of the canal was sealed with fascia and fibrin glue. Both the ear canal skin and tympanic membrane had a normal appearance.

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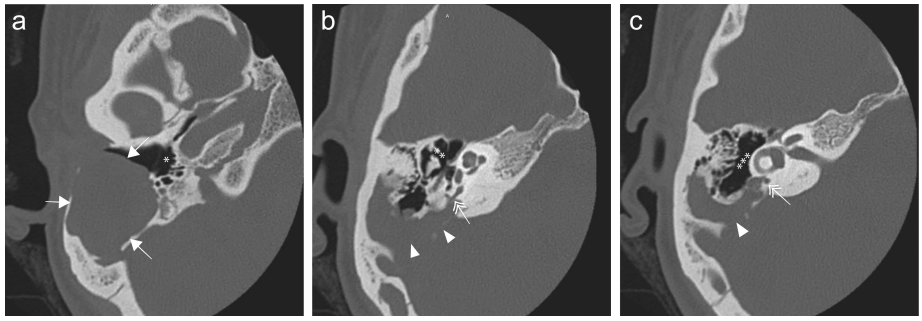

Figure 1. Preoperative axial MSCT images of the right temporal bone at different levels. (a) An extremely narrow external auditory canal can be visualized due to the bulging of a large soft tissue mass (arrows) through an eroded bony posterior canal wall. The hypotympanum is free of pathology (asterisk). (b) At a higher level dehiscence of the posterior semicircular canal can be seen (double arrow). The posterior fossa dura and sigmoid sinus have been exposed (arrowheads). The epitympanum is unaffected (double asterisk). (c) The aditus ad antrum and antrum have a normal appearance (triple asterisk). Note the dehiscence of the posterior semicircular canal (double arrows) and exposure of the posterior fossa dura (arrowheads). MSCT, multislice computed tomography.

Two large cortical bone chips and bone dust with fibrin glue were used to reconstruct the external auditory canal (Figure 3b). Dressing of the external auditory canal consisted of a silicon sheet, gelatin sponges, and a ribbon gauze wick with antibiotic ointment. In the immediate postoperative period, the patient experienced some dizziness, which slowly disappeared after a couple of days. Six months postoperatively PTA was 20.0 dB HL. The horizontal video head impulse test showed a normal gain on both sides, without corrective saccades. One year postoperatively non-EP DWI did not show residual cholesteatoma (Figure 2b). On CT imaging a wide external auditory canal could be observed (Figure 4). As palpation during otomicroscopic evaluation showed some space between the ear canal skin and the bony canal wall, the decision was made to perform a second-look intervention. A small additional bone chip was used to level the bony canal. No residual cholesteatoma was found. No problems were encountered during further follow-up. The patient gave oral informed consent for this case report.

Discussion

Congenital cholesteatomas may arise at different locations in the temporal bone. The exact pathophysiology is still unclear. The 4 main theories include development from an epithelial rest, invagination or implantation of squamous epithelium, and metaplasia of normal epithelium. The first theory, which states that a CC develops from an ectodermal remnant, is the most accepted.[1] The middle ear cleft and petrous apex are the most frequent sites of onset. In contrast, a CC originating from the mastoid process is rare.[3]

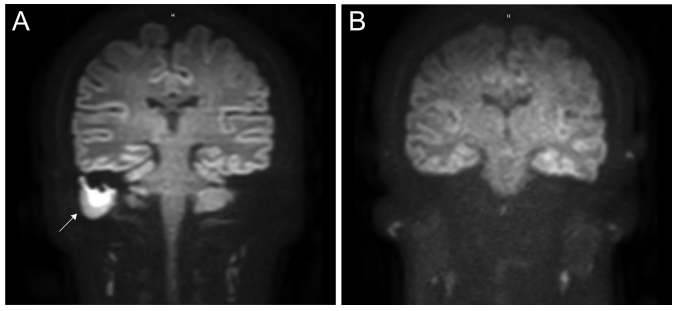

Figure 2. Coronal b1000 non-EP DW MR images. (A) A preoperative scan shows a 24 mm large diffusion-restricted mass in the right mastoid region, compatible with a large cholesteatoma (arrow). (B) A postoperative scan performed at the same level 1 year postoperatively shows no diffusion restriction: no residual cholesteatoma is present.

An intact tympanic membrane and no otologic history are classic features to distinguish congenital from acquired cholesteatomas. It is known that ventilation tube insertion sporadically may cause ingrowth of epithelial cells in the middle ear. In a large retrospective analysis by Golz et al[6] (5575 ears), however, this very uncommon complication was only seen at or near the site of tube insertion. As the middle ear, attic, and entire antrum were completely free of pathology (Figure 1b), an iatrogenic cholesteatoma could be excluded in this case.

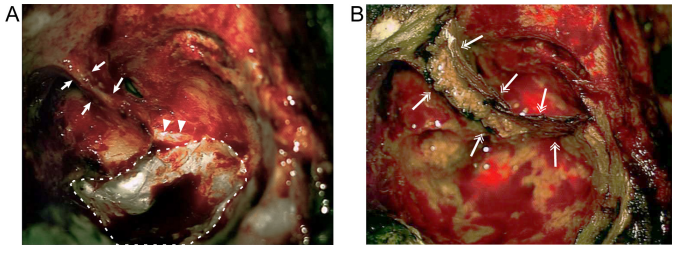

Figure 3. (A) A preoperative image shows the congenital cholesteatoma in the mastoid process (dotted line). The matrix has been dissected from the denuded facial nerve (arrowheads). Only a small remnant of the posterior canal wall is present (arrows). (B) A peroperative image shows the reconstruction of the posterior canal wall with large cortical bone chips and bone paté (double arrows).

Main Points

- Congenital cholesteatomas of the mastoid process are rare lesions that often remain undiagnosed until adulthood.
- Possible presenting symptoms are retroauricular swelling, ear or neck pain, hearing loss, dizziness, and a stenosis of the external auditory canal.
- Resection can be challenging due to involvement of the sigmoid sinus, posterior fossa dura, facial nerve, and otic capsule.
- Reconstruction of the external auditory canal with bone chips and bone dust is a feasible treatment option to preserve the normal ear physiology.

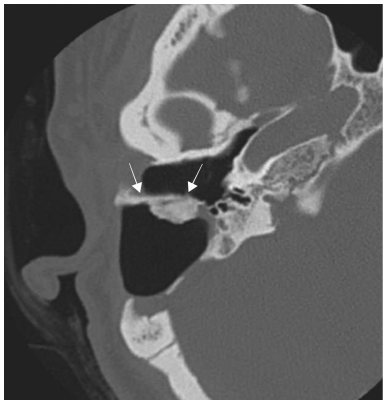

Figure 4. A postoperative axial MSCT image shows the reconstruction of the bony posterior canal wall with cortical bone chips (arrows). The mastoid cavity is free of pathology. MSCT, multislice computed tomography.

Determining the exact site of origin is not always possible. Due to extension, it may be difficult to differentiate a CC originating from the middle ear, mastoid process, or petrous apex. To fit the criteria of a pure CC of the mastoid process there may be no involvement of the middle ear, attic, or aditus ad antrum. In a literature review by Giannuzzi et al in 2011, only 9 cases fulfilled these conditions.[3] Whereas a CC of the middle ear often causes early conductive hearing loss, a CC of the mastoid may be a silent lesion for years. They may be found incidentally, for example on head and neck imaging studies performed for other reasons, or after substantial destruction of the surrounding structures. The clinical presentation is variable. Possible (nonspecific) symptoms may be retroauricular swelling, ear or neck pain, hearing loss, dizziness, or a stenosis of the external auditory canal.[3-5,7-9]. In this case, the latter feature led to the diagnosis. Nagato et al published a similar case of an ear canal stricture caused by a CC of the mastoid process, but without involvement of the sigmoid sinus, posterior fossa dura, facial nerve, or posterior semicircular canal.[9]

On MSCT imaging, a CC of the mastoid typically presents as an expansile soft tissue mass in a normally aerated mastoid, with an erosion of the surrounding structures. Lesions that may appear similar on CT are cholesterol granulomas, temporal bone tumors, and histiocytosis.[3] The well-established technique of non-EP DWI helps to confirm the diagnosis of a CC of the mastoid with high specificity. Interestingly, intradiploic epidermoid cysts—which are mainly described in the neurosurgical literature—have similar histological and radiological characteristics compared to congenital cholesteatomas.[10] These cysts originate from the diploë and may occur at different locations within the skull. As they are also derived from ectodermal remnants during embryonic development, Clark et al concluded that intradiploic epidermoid cysts and congenital cholesteatomas are actually one and the same disease.[11]

As for other cholesteatomas, surgery is the preferred management strategy. Although Sepehri et al[12] reported a case of a CC of the mastoid in a very old person, which was treated by watchful waiting, surgery is usually necessary to avoid further destruction. Frequent intraoperative findings are exposure to the sigmoid sinus, posterior fossa dura, and third segment of the facial nerve. These features normally can be clearly assessed preoperatively on CT imaging. Usually, removal of the cholesteatoma matrix from these structures is feasible by gentle dissection. Giannuzzi et al[3] reported 2 cases of sigmoid sinus involvement which required obliteration of the sinus. In our case, this was not necessary as there was a clear cleavage plane between the sinus and the cholesteatoma matrix. The dehiscence of the posterior semicircular canal in our case did not cause any symptoms of dizziness and could be treated successfully by sealing. Fowler et al reported a similar case of a CC of the mastoid with involvement of the posterior canal and typical symptoms of a third-window phenomenon. In their case, the inner ear function could be preserved by occluding the canal.[13] Destruction of the posterior canal wall in the CC of the mastoid is often managed by performing a canal wall down procedure.[3,5,14] In our opinion, canal wall down surgery and its subsequent problems mostly can be avoided, especially in cases with intact ear canal skin.

Similar to acquired cholesteatomas, the procedure for congenital cholesteatomas of mastoid origin can be combined with obliteration of the mastoid cavity with bone dust or hydroxyapatite. A potential benefit could be a lower risk of residual cholesteatoma. On the other hand, re-exploration in these cases may be more challenging. The most important reason to perform obliteration, namely prevention of recurrent disease, is not a concern, as in most congenital cholesteatoma cases there are no ventilation problems.

Informed Consent: Verbal informed consent was obtained from the patient who agreed to take part in the study.

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

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