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Huge mastoid congenital cholesteatoma in a 52-year-old patient

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Abstract. *Huge mastoid congenital cholesteatoma in a 52-year-old patient*. Congenital cholesteatoma may arise in various locations within the temporal bone. The rarest site of origin is the mastoid process. We report an unusual case of a 52-year-old man with mastoid congenital cholesteatoma that manifested as a persistent ear discharge. The preoperative suspicion was based on the imaging findings and the patient's history. A simple mastoidectomy was conducted and the cholesteatoma was completely removed while using facial nerve monitoring. Although rare, mastoid congenital cholesteatoma can be considered as an alternative in the differential diagnosis of persistent otorrhea.

Introduction

Congenital cholesteatoma is a rare lesion of the temporal bone, which arises from ectopic epidermoid remnants. The reported sites of origin include the middle ear, the petrous apex, the external auditory canal, the cerebellopontine angle, and the mastoid process. The middle ear is the most common site of origin, and the mastoid is the rarest.[1] The diagnosis is based on an absence of previous middle ear disease or middle ear surgery, and the presence of an intact tympanic membrane.[2] We report an unusual case of congenital cholesteatoma isolated to the mastoid process that manifested as a persistent ear discharge.

Figure 1
Axial computed tomography scan of the temporal bone showed an expansile, lytic lesion in the left mastoid process that had eroded the bony plate of the posterior fossa, the bony plate covering the sigmoid sinus, and the posterior wall of the external auditory canal.

Case Report

A 52-year-old man presented in our department with a two-week history of left ear otorrhea. He had already received amoxycillin but the otorrhea did not improve. There was no history of previous ear discharge, tympanic membrane perforation, ear infections, ear operations, or other otological complaints. Microscopic examination showed a posterior wall protrusion with a small opening through which pus was leaking out. The tympanic membrane was found to be normal. Audiometric evaluation showed a symmetric mild sensorineural hearing loss at high frequencies. Based on these findings, a furuncle was suspected. The patient got intravenous antibiotic therapy (fusidic acid) supplemented with topical antibiotic treatment (mupirocin). Despite a transient improvement, some days later he again presented

Figure 2
Axial T2-weighted magnetic resonance imaging showed a hyperintense mass in the left mastoid.

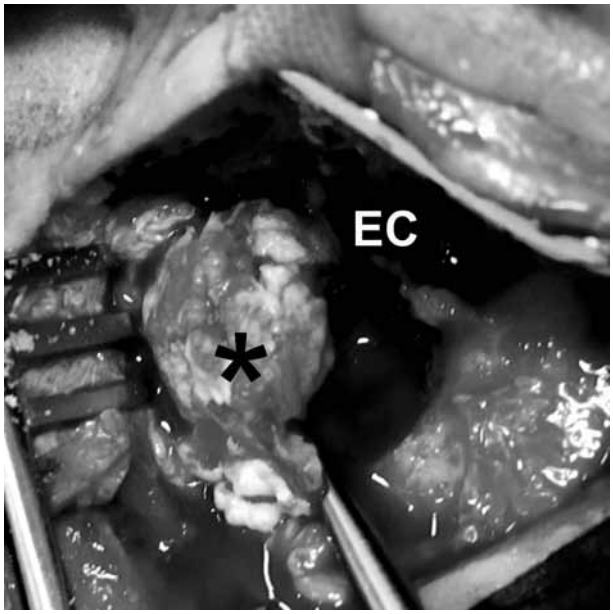

Figure 3
The mastoid process was filled with cholesteatoma (*). EC: ear canal.

to our department with recurrent ear discharge.

A computed tomography (CT) scan was ordered and showed that the whole left mastoid cavity was occupied by a large, lytic mass that had eroded the bony plate of the posterior fossa, the bony plate covering the sigmoid sinus, and the posterior wall of the external auditory canal (Figure 1). Moreover, the mass occluded the left sigmoid sinus. The lesion did not extend into the middle ear, and the inner ear was normal. Subsequent magnetic resonance imaging (MRI) of the brain confirmed these findings (Figure 2). Thus, it became apparent that the opening in the posterior meatal was a small fistula communicating with the mastoid cavity. Based on the imaging findings and the patient's history, a mastoid congenital cholesteatoma was suspected.

Consequently, a simple mastoidectomy was conducted and the mass was completely removed (Figure 3). Surgery confirmed the radiological extent of the mass. The facial nerve was surrounded by the lesion. However, with the intra-operative use of facial nerve monitoring, the mass was removed without causing any damage to the nerve (Figure 4). Histopathological examination of the lesion confirmed the diagnosis of cholesteatoma.

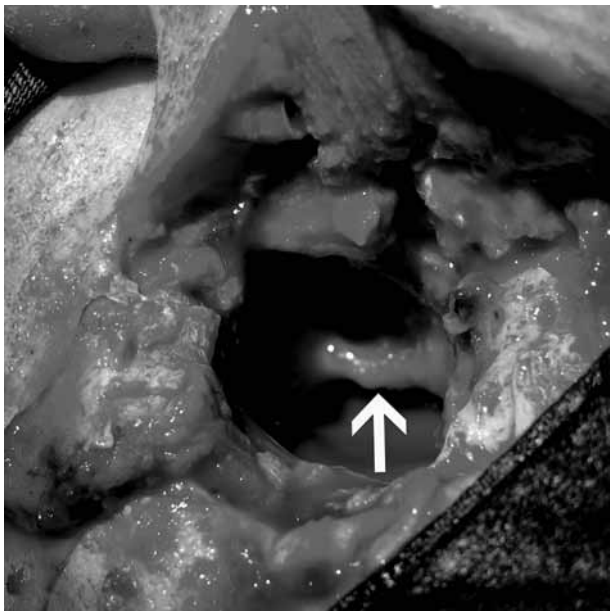

Figure 4
Appearance after total surgical resection of the cholesteatoma. The mastoid segment of the facial nerve (white arrow) was not damaged.

Upon follow-up one month later, the ear discharge had stopped and the patient's hearing remained the

same. The first post-operative CT scan displayed no sign of recurrence. The mastoid cavity was empty and free of disease (Figure 4). A second surgical procedure is planned to close the opening in the fistula with a cartilaginous graft. Publication of the patient's data was approved by the Ethics Committee of General Hospital of Volos.

Discussion

Congenital cholesteatoma is far less frequent than its acquired counterpart. It may originate in various locations within the temporal bone. Its occurrence in the mastoid process is the least common site of origin.[1] To the best of our knowledge, there are 17 literature reports on mastoid congenital cholesteatomas.[3]

Mastoid congenital cholesteatoma manifests with a variety of symptoms and signs. The most common presentation is either neck pain or ear pain.[3] However, our patient experienced persistent ear discharge through a formed fistula that connected the mastoid cavity with the external auditory canal. A literature search identified only one previous article reporting a mastoid congenital cholesteatoma with a similar clinical presentation.[4]

A point that should be underlined is that our patient presented with complete atelectasia of the sigmoid sinus as a result of compression by the mass. Many authors reported extensive erosion of the bony plate covering the sigmoid sinus. However, there is only one report of sigmoid sinus occlusion.[5]

Another rare feature of our case was the fact that the disease occurred relatively late in adulthood. Of the 17 reported cases of congenital cholesteatoma isolated to the mastoid region, the mean age of presentation was 31.5 years.[3] There are few reports of patients in whom the diagnosis was made at an older age.

Finally, it should be emphasized that CT and MRI are invaluable in the diagnosis of mastoid congenital cholesteatomas.[6] The differential diagnosis of temporal bone lesions includes an anomalous sigmoid sinus, cholesterol granulomas, and other possible tumours such as paragangliomas, endolymphatic sac tumours, and meningiomas.

Surgical management of mastoid congenital cholesteatomas is primarily directed by the imaging findings because the clinical presentation can belie the extent of disease. If the disease is limited to the mastoid, a mastoidectomy approach can be applied. On the other hand, if the disease is more extensive with exposure of dura, sigmoid sinus, or the facial nerve, a transtemporal approach can be planned.

Conclusion

In conclusion, the possibility of a mastoid congenital cholesteatoma can be included in the differential diagnosis of persistent otorrhea. The appropriate radiologic examinations can enable an accurate preoperative diagnosis and optimal treatment planning.

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