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A Rare Case of Recurrent Osteoid Osteoma of the Temporal Bone With Concomitant Cholesteatoma

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Abstract

Introduction: Osteoid osteoma (OO) of the temporal bone is very rare. Even more unusual is its recurrence, leading to development of an associated cholesteatoma.
Methods: Case report and literature review
Case: A 63-year old female presented to our practice with recurrent OO, initially resected 8 years earlier at an outside institution via middle fossa craniotomy as it was presumed to be a perigeniculate hemangioma. Hearing was lost at that time, and to exonerate disease, the facial nerve was sacrificed and an interposition nerve graft was required. Final pathology revealed that the diagnosis was actually osteoid osteoma. The patient moved to Arizona and was being followed elsewhere with serial MRI scans; her most recent MRI suggested tumor growth by 4mm in the past year and thus she was referred for surgical management. A translabyrinthine/ transcochlear dissection with ear canal closure was required to remove extensive OO intermixed with cholesteatoma material.
Discussion/Conclusion: There have been few reported cases of OO of the temporal bone, and no reported cases of recurrent OO with concomitant cholesteatoma. Radiological features of OO can mimic other disease processes; thus, histology is required to establish a diagnosis. Complete resection is necessary to prevent recurrence.

Case

A 63 year-old female presented to an outside institution in 2007 with progressive left sided hearing loss. Computed Tomography (CT) and Magnetic Resonance Imaging(MRI) was obtained that demonstrated a mass lesion in the anterior epitympanum. Signal characteristics suggested possible perigeniculate hemangioma, and the patient underwent resection via middle fossa craniotomy. The facial nerve appeared to be involved and was resected, requiring an interposition sural nerve graft for reconstruction. Hearing was lost. Facial nerve function was a House-Brackman (HB) Grade 6/6 for nearly one year; however, there was eventual recovery to a HB Grade 3/6. Review of operative reports indicate that a gross total resection was achieved. Since 2007, the patient moved to Southern Arizona and was followed by an outside neurotologist with annual physical exam and MRI. The most recent MRI found 4 mm of tumor growth in the past year and though not clear why, the patient was referred to our facility for surgical management. Physical exam found complete obstruction of the left external auditory canal (EAC) to the bone-cartilaginous junction by a firm mass lesion. CT (Figure 1) and MRI (Figure 2) were obtained. CT found a ground-glass appearing mass lesion filling the external auditory canal and middle ear – extending from tegmen cranially to hypotympanum caudally and from Eustachian tube orifice anteriorly to the mastoid antrum posteriorly. The lesion was dark on T1-weighted MRI, bright on T2, and enhanced with contrast administration. Interestingly, there was no evidence of restricted diffusion to suggest presence of cholesteatoma. For definitive resection, the patient elected to proceed with a transtemporal craniotomy and ear canal closure. During surgery, disease filled the epitympanum, abutted the superior and horizontal canals, extended to the anterior protympanum, and involved the hypotympanic air cells adjacent to the jugular bulb. Intermixed with OO were areas of cholesteatoma debris; while most deposits of skin were < 2-3 mm in size, several areas of cholesteatoma were clearly larger. To remove all visible disease, a translabyrinthine/transcochlear dissection was performed - removing the horizontal and superior semicircular canals as well as the basal turn of the cochlea. The dome of the jugular bulb and the carotico-jugular spine were dissected. A facial nerve decompression was also performed from the vertical segment to the proximal tympanic segment. Then, the ear canal was closed in two layers, and both AlloDerm as well as abdominal fat were used to obliterate the transtemporal defect. Immediately after surgery, the patient retained HB Grade 3 facial nerve function and was not vertiginous. She continues to heal well from surgery and will undergo annual surveillance MRI scans for several years.

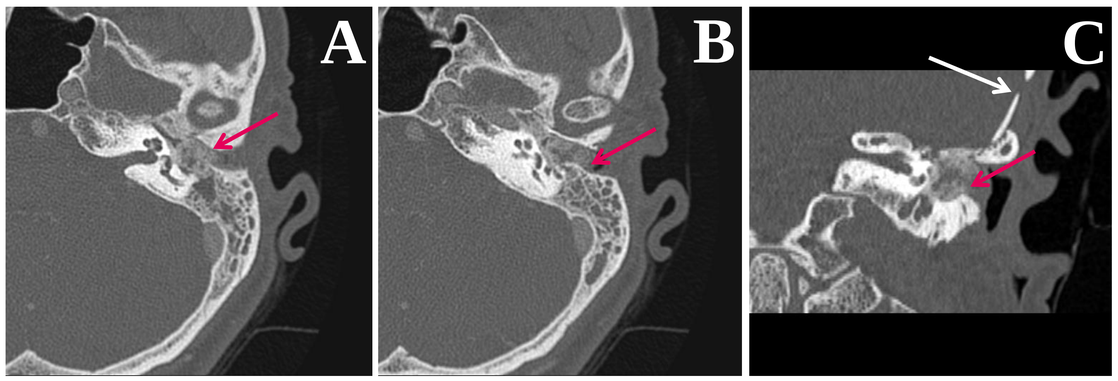

Figure 1.
A. Axial CT of the left temporal bone reveals OO (red arrow) from Eustachian tube orifice extending into mastoid antrum, with chronic mastoiditis
B. Axial CT of left temporal bone shows OO (red arrow) abutting cochlea, extending through tympanic membrane to bony-cartilaginous junction of EAC
C. Coronal CT of left temporal bone shows evidence of previous middle fossa craniotomy (white arrow) and OO (red arrow) extending from tegmen superiorly to hypotympanum inferiorly

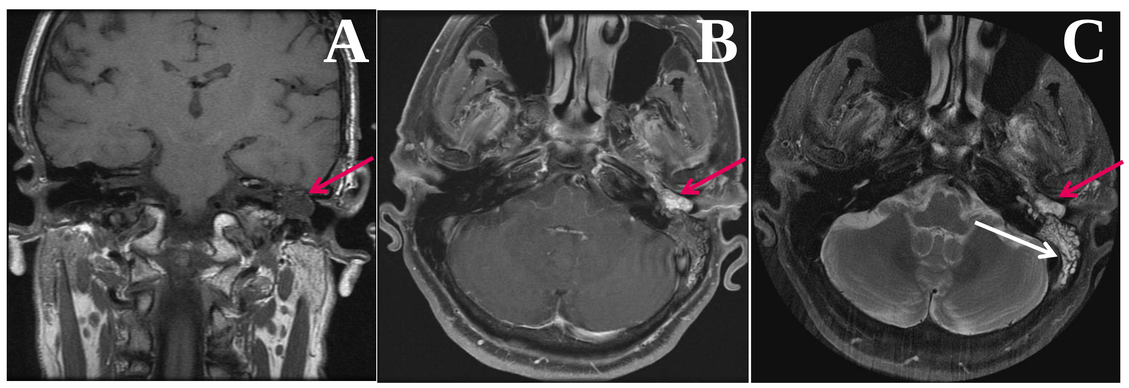

Figure 2.
A. Coronal T1 pre contrast MRI of the temporal bones reveals a non-enhancing mass in middle ear (red arrow) extending into EAC
B. Axial T1 post contrast MRI of the temporal bones reveals an enhancing lesion in middle ear (red arrow) extending into EAC
C. Axial T2 weighted MRI of the temporal bones reveals a bright lesion in the left middle ear extending into EAC (red arrow), with bright fluid signal in adjacent mastoid air cells (white arrow)

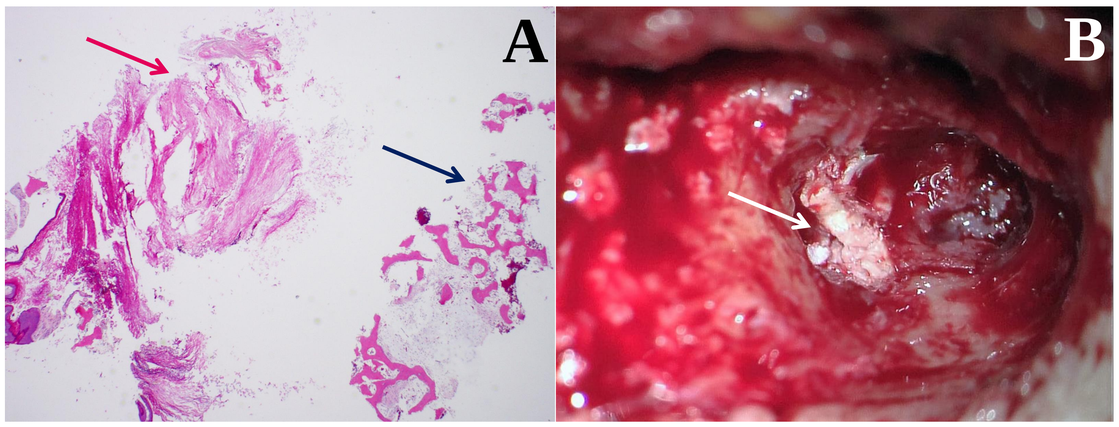

Figure 3.
A. Histopathology reveals keratin debris and squamous epithelium of cholesteatoma (red arrow) adjacent to characteristic fibrovascular stroma and woven bone of OO (black arrow) (H&E stain)
B. Intraoperative imaging taken during operation for recurrent OO reveals cholesteatoma in EAC (white arrow)

Discussion

Osteoid osteoma, first described by Jaffe in 1935, is a benign osteoblastic lesion characterized by a central nidus of vascular connective tissue stroma surrounded by atypical, immature bone. OO treatment requires complete surgical excision; particularly, failure in completely removing its nidus results in recurrence. Based on a review of outside records and operative notes, we speculate that the vascular connective tissue nidus of OO in this patient was misinterpreted as a perigeniculate hemangioma and thus treated using a middle cranial fossa approach. Both OO and hemangioma have similar appearances on MRI; both are hypointense on T1, hyperintense on T2, and enhance with gadolinium. Also, both hemangioma and OO are brownish-red, mottled lesions and may appear similar on gross inspection. It was thought that a gross total resection had been achieved at the patient's first operation. However disease around the Eustachian tube orifice is difficult to access via middle fossa craniotomy and this remnant of disease likely persisted. As the lesion grew, it filled the epitympanum, protympanum, mesotympanum, hypotympanum, and mastoid antrum. It also grew laterally through the tympanic membrane, thereby trapping both ear drum and ear canal skin to cause cholesteatoma formation. The natural history of osteoid osteoma growth as well as best treatment practices are unknown. Only 6 cases have been reported in the temporal bone. Lateral growth, skin trapping, and cholesteatoma formation may also explain the lesion's 4mm growth on MRI within the year prior to patient presentation at our institution.
In modern times, both CT and MRI play critical roles in developing a differential diagnosis for lesions in the temporal bone. For example, newer diffusion weighted imaging (DWI) protocols have proven successful in differentiating cholesteatoma from granulation tissue, fluid, fibrous tissue, and other soft tissue lesions as cholesteatomas demonstrate restricted diffusion. While details regarding mechanism are not clear, DWI images show cholesteatomas as hyper-intense relative to brain and CSF. Current theories suggest that this may be related to T2 shine through effect. The newest DWI techniques are capable of detecting cholesteatoma lesions as small as 2mm. For this patient, however, DWI performed prior to the operation at our facility did not indicate presence of cholesteatoma. This unexpected imaging aberration may be secondary to (1) most foci of cholesteatoma intermixed with tumor being </= 2 mm or (2) the presence of bone may hinder the detection of cholesteatoma due to artifact from the bone itself. Both factors may have been at plan. By direct visualization, most areas of cholesteatoma material were < 2mm; however, there were clearly other areas > 5mm (figure 3) in size but surrounded by OO and clearly missed on DWI imaging.
Ultimately, an aggressive surgical approach with removal of otic capsule bone and ear obliteration was required to fully exonerate disease. Future surveillance will require both CT and MRI. Because the latter may have poor sensitivity for cholesteatoma formation, a second look operation may be required and has been discussed with the patient.

Conclusion

Osteoid osteoma of the temporal bone is a rare disease entity with only about 6 cases reported. Grossly, it appears reddish-brown within mottled bone. CT typically demonstrates a ground-glass appearing mass, while MRI is dark on T1, bright on T2, and enhances with gadolinium. Histopathology exhibits a nidus of vascular connective tissue stroma surrounded by immature bone. Imaging characteristics for osteoid osteoma are similar to other disease processes; therefore, histology is required for definitive diagnosis. Because OO originates from a central nidus, the entire nidus must be removed in order to achieve cure. With lateral growth through the tympanic membrane, skin trapping and cholesteatoma formation is possible, and due to its slow growth and potential for recurrence, long term surveillance is required.

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