Hemangioma of the middle ear mimicking glomus jugulotympanicum: A case study and review of the literature

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ABSTRACT

OBJECTIVES: Hemangiomas are benign vascular tumors that present frequently in the skin and soft tissues of the head and neck, but only rarely in the middle ear cavity. In this case report, we present the imaging, surgical, and pathological findings of a middle ear hemangioma mimicking a glomus jugulotympanicum, followed by a review of the literature.

STUDY DESIGN: Case Study

METHODS: We report the case of a 49-year-old male who presented with a two-year history of left sided hearing loss with intermittent tinnitus. Physical exam was remarkable for an erythematous mass behind the left tympanic membrane. Contrast-enhanced CT and MRI revealed an enhancing mass involving the left middle ear cavity and cochlear promontory with likely contiguous erosive change of the left jugular foramen, suggesting a diagnosis of glomus tympanicum or jugulare.

RESULTS: Left tympanoplasty with canaloplasty and transmeatal subtotal resection of the tumor was performed, with intraoperative findings suggestive of a glomus jugulare with a broad base in the hypotympanum. However, the specimen stained negative for s100 and chromogranin, ruling out paraganglioma and demonstrated morphologic and immunohistochemical features consistent with a diagnosis of hemangioma. The postoperative period was uneventful, and the patient will undergo surveillance at this time.

CONCLUSIONS: Hemangiomas arising primarily in the middle ear are rare and not easily distinguishable from glomus jugulotympanicum. Therefore, hemangioma should be considered in the differential diagnosis of a vascular middle ear mass.

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INTRODUCTION

Hemangiomas are benign, vascular tumors that are relatively common in the cutaneous tissues of the head and neck, but they have rarely been reported in the middle ear. We present the imaging, surgical, and pathological findings of a middle ear hemangioma mimicking a glomus jugulotympanicum, followed by a review of the literature.

CASE PRESENTATION

A 49-year-old male presented with a complaint of left sided hearing loss and intermittent tinnitus that had progressed over a two-year period. He reported no otologic history aside from extensive noise exposure while working in construction. He denied otalgia, otorrhea, imbalance, and vertigo. Otoscopy revealed an erythematous mass behind the left tympanic membrane. Rinne test was equivocal in the left ear and positive in the right, and Weber lateralized to the left ear.

A pure tone audiogram showed a mild conductive downsloping hearing loss to a severe mixed hearing loss of the left ear and a downsloping mild to moderately-severe sensorineural hearing loss of the right ear (fig. 1). Contrast-enhanced CT and MRI revealed an enhancing soft-tissue mass filling the left middle ear cavity with likely contiguous erosive change of the left internal jugular foramen, consistent with a preoperative diagnosis of glomus jugulare or tympanicum (fig. 2).

The patient underwent left tympanoplasty including canaloplasty with subtotal CO2 laser-assisted resection of the tumor. The tumor was isolated to the posterior-interior quadrant of the middle ear space and appeared to arise from a broad vascular base in the hypotympanum. After significant debulking, it was determined that the tumor could not be fully resected using a transmeatal approach and the procedure was terminated. At the end of the case, the ossicular chain, chorda tympani, and facial nerve were intact.

Final pathological evaluation was consistent with a benign hemangioma. H&E staining revealed numerous closely spaced vascular channels with intervening cells (fig. 3). Immunohistochemical staining was negative for s100 and chromogranin, ruling out paraganglioma and consistent with hemangioma. The postoperative course was uneventful and the patient has elected to undergo surveillance at this time.

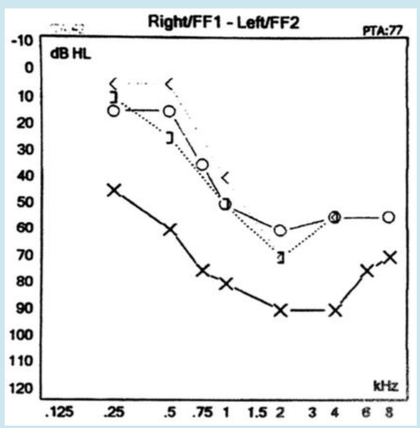

Figure 1. Preoperative audiogram showing a mixed left-sided hearing loss (X) and right-sided sensorineural hearing loss (O).

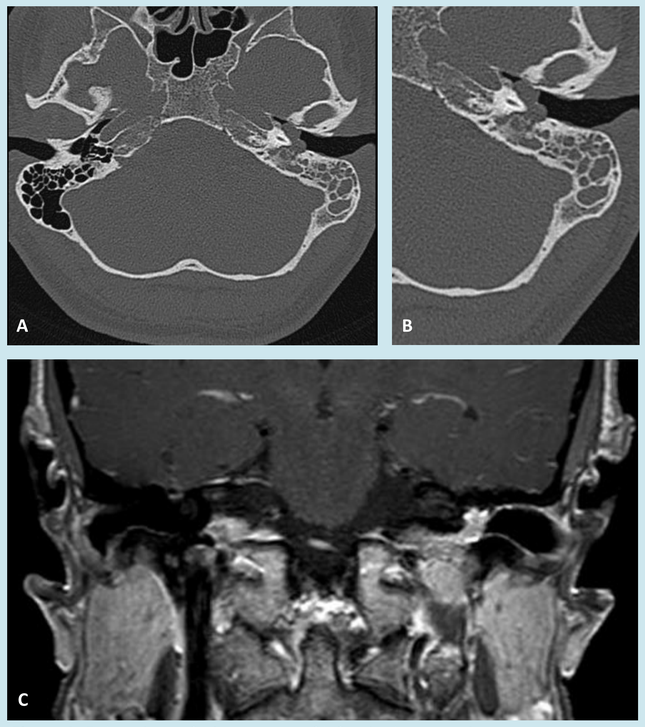

Figure 2. Enhancing soft tissue mass of left middle ear cavity as seen on computed tomography (A, B) and magnetic resonance imaging (C).

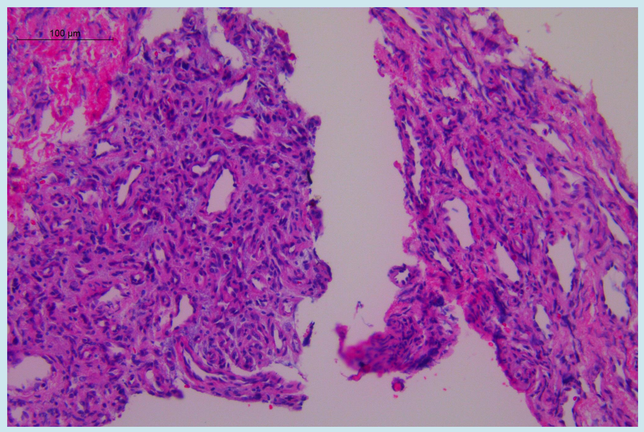

Figure 3. Benign vasoformative lesion showing vascular spaces lined by endothelial cells at 20x magnification.

DISCUSSION

Though relatively common in the soft tissues of the head and neck, hemangiomas only rarely present in the middle ear, where there is a paucity of vasoformative tissue. A 1981 retrospective review of 1430 intra-temporal tumors revealed only three hemangiomas[1]. Given their rarity and their ability to mimic more common otologic tumors, preoperative diagnosis can be difficult.

Our literature review revealed 23 previously reported cases of hemangioma involving the middle ear in the English literature; however, only seven of these were limited to the middle ear. The remaining 16 cases extended to involve the tympanic membrane (TM) and/or the external auditory canal (EAC) as well.[2-8] The clinical presentation of a middle ear hemangioma is variable but a patient may experience pulsatile tinnitus, aural fullness, conductive hearing loss, recurrent bloody otorrhea, recurrent otitis media, otalgia, and dizziness.[2-6] Otoscopic exam may reveal a reddish-blue mass behind or involving the TM or a fleshy polypoid mass in the EAC.[2,5]

DISCUSSION (continued)

High resolution temporal bone CT scan is the imaging modality of choice for vascular middle ear tumors and is excellent for the identification of bony destruction. A hemangioma will appear as a soft tissue lesion that enhances with contrast. MRI may provide more detail for preoperative planning and is helpful for facial nerve analysis. On MRI, a hemangioma will appear moderately intense on T1, highly intense on T2, and will be enhancing with gadolinium. Angiography may prove valuable if excessive intraoperative bleeding is expected, and it can help to differentiate a hemangioma from a glomus tumor, which will clear contrast much more quickly than a hemangioma.[6]

Treatment of choice in the majority of cases is surgical resection, though there is a solitary report of an EAC hemangioma treated successfully with radiation therapy.[8] Surgical approach depends on involvement of the TM, ossicles, or other structures beyond the middle ear cavity. For those confined to the middle ear, a transcanal approach with tympanoplasty is most often performed. For those extending beyond the middle ear, both canal wall-up and wall-down mastoidectomy have been described. The utility of the CO2 laser in middle ear hemangioma resection was described once before in 2010; we also found it a valuable tool for hemostasis, tissue vaporization, and tumor shrinkage in this case.[7]

Given the nonspecific clinical presentation and imaging characteristics, middle ear hemangioma is often diagnosed preoperatively as glomus tympanicum or glomus jugulare, as in our case. Therefore, a full differential diagnosis of middle ear lesions should be considered. In 1983, Dayal et al. described a group of lesions mimicking glomus jugulotympanicum as follows: 1) Vascular lesions including high jugular bulb, aberrant intratemporal internal carotid artery, and arteriovenous malformation, 2) Neoplastic lesions including meningioma, hemangioma, rhabdomyosarcoma, melanoma, carcinoma, and pyogenic granuloma, and 3) Inflammatory lesions including cholesterol granuloma and aural polyp.[9]

CONCLUSION

Though a rare entity, hemangioma of the middle ear should be considered in the differential diagnosis of a vascular middle ear lesion.

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