



Primary Extracranial Meningioma of Middle Ear

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Abstract

The differential diagnosis of middle ear tumor is numerous and variable. In this case report, we describe a patient who presented with conductive hearing loss and was found to have a middle ear meningioma. This is one of the few reports of this rare middle ear tumor, and it serves to remind otolaryngologists that meningioma can sometimes occur in the middle ear.

Introduction

Meningiomas are one of most common benign tumors of the central nervous system [1]. They account for approximately 13% to 26% of all intracranial neoplasms; however, less than 2% of meningiomas have extracranial extension or temporal bone involvement [1]. Due to the rare occurrence of middle ear meningiomas, the diagnosis of these tumors can be challenging [1]. We present a rare case of primary extracranial meningioma located in the middle ear to highlight its clinical, imaging, immunohistochemical, and histological features. This case report was exempt from IRB review per institutional policy.

Case Presentation

A 65-year-old female was referred for evaluation of an enlarging, painful mass in her left middle ear. She had a five-year history of left-sided hearing loss and pulsatile tinnitus that occurred during strenuous physical activity. She denied otorrhea, vertigo, or any neurological or constitutional symptoms. She did not have any previous history of ear surgery, recurrent ear infections, or exposure to loud noise.

Binocular microscopy showed normal right external auditory canal and right tympanic membrane. Left external auditory canal was also found to be clear; however, an intact left tympanic membrane was revealed. Cranial nerve examination was normal bilaterally.

Audiometry demonstrated a left conductive hearing loss. Speech Reception Threshold (SRT) was 5dB on the right and 25dB on the left. Speech discrimination score remained 100% on the right and 100% on the left.

A contrast-enhanced temporal bone computed tomographic revealed a 9mm lobulated mass in the left middle ear with extension into the hypotympanum. There was no bony erosion. The ossicular chain was intact but was being pressed by the middle ear mass.

The patient underwent complete surgical resection of the mass. Histological examination showed whorls and lobules of cells with indistinct cellular borders forming syncytium (Figure 1). No nuclear pleomorphism and mitotic activity was identified. Immunoperoxidase stains were positive for epithelial membrane antigen (EMA) and CD56 (Figure 2). S-100 protein, chromogranin, and synaptophysin were negative. These findings were consistent with meningioma.

Postoperatively, the patient reported improvement in her hearing, which was confirmed by her 3-month postoperative audiogram. The pulsatile tinnitus had resolved. The patient had shown no clinical evidence of recurrence during a follow-up period of 12 months.

Discussion

Extracranial meningiomas are classified into primary or secondary types based on their origins [1]. Primary meningiomas arise from ectopic arachnoid cells and therefore are not associated with an intracranial mass, whereas secondary tumors result from direct extensions of intracranial masses [1]. In this case, the patient experienced a left conductive hearing loss and pulsatile tinnitus without

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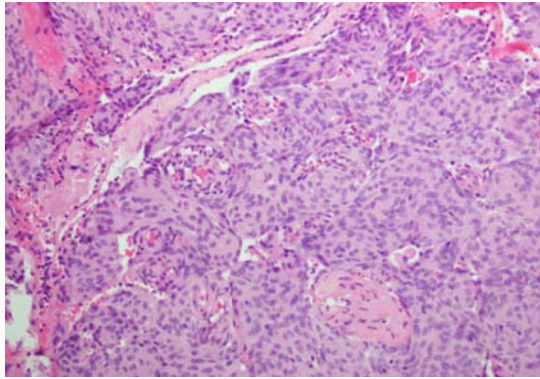


Figure 1: Histological examination demonstrating whorls and lobules of cells with indistinct cellular borders forming syncytium.

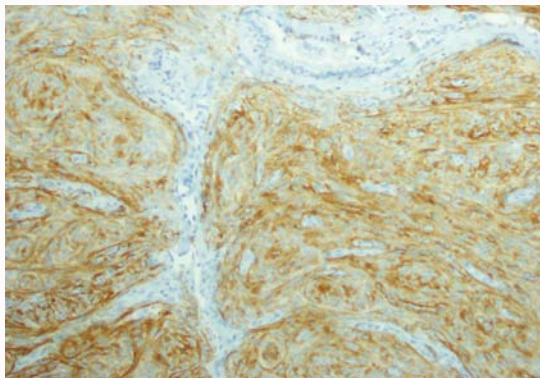


Figure 2: Positive immunohistochemical staining for EMA.

any symptoms of cranial nerve involvement, which is consistent with the diagnosis of primary middle ear meningioma. This suggests that meningiomas should be considered in the differential diagnosis of middle ear mass. Other differential diagnosis may include schwannomas, paragangliomas, middle ear adenomas, metastatic carcinomas, cholesteatomas, glomus tumors, retrotympanic vascular tumors, and lipoma [2].

Imaging modalities may aid the diagnosis of primary extracranial meningioma. While CT may reveal the presence of calcification and bony hyperostosis, MRI has a higher sensitivity than CT and may show an enhancing dural-based soft-tissue mass with a characteristic dural tail [3]. In this case, the CT did not reveal calcification or bony hyperostosis.

Immunohistochemistry and histology are crucial to differentiate extracranial meningiomas from other differential diagnosis. In this case, the tumor was characterized by positive immunostaining for EMA, which is a characteristic finding of meningioma [1]. While meningiomas exhibit similar immunohistochemical profiles, they may express a variety of different histological patterns. The most common histological pattern is meningotheial [4]. Moreover, cells are arranged in syncytial sheets with indistinct cytoplasmic margins, whorl formation, and clear to faintly eosinophilic cytoplasm [4]. Occasional psammoma bodies may be present, but mitotic activity and necrosis are absent [4]. These findings in this case confirms the diagnosis.

As demonstrated in this case, patients with symptomatic meningiomas or consistently growing tumors should be managed with complete surgical excision. Because meningiomas are slow-growing tumors, long-term follow-up is necessary to exclude the recurrence [4]. The recurrence rate for meningiomas is reported to vary widely from 7% to 84% [4]. Nevertheless, the prognosis remains excellent, and to date, there are no published cases indicating that recurrence can reduce life expectancy [5].

Conclusion

Meningioma can rarely present in the middle ear. This should be kept in the differential diagnosis when evaluating patients with middle ear mass. Patients with growing and symptomatic middle ear meningioma should undergo surgical excision and postoperative surveillance to monitor tumor recurrence.

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