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Tympanic Paraganglioma: A Case Report and Review of the Literature

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Abstract **Case Report**

Tympanic paragangliomas are rare, slow-growing neuroendocrine tumors arising from the paraganglionic cells of the middle ear. Although benign, they may cause significant morbidity due to local invasion. We present a case of a 54-year-old female with a tympanic paraganglioma, detailing the clinical presentation, imaging findings, histopathological features, and surgical management. A review of the literature highlights diagnostic challenges and treatment approaches, emphasizing the role of imaging and histopathology in optimizing patient outcomes.

Keywords: Tympanic Paraganglioma, Middle Ear Tumor, Neuroendocrine Tumor, Temporal Bone, Glomus Tympanicum, Imaging, Histopathology.

INTRODUCTION

Paragangliomas (PGLs) of the head and neck region are uncommon neuroendocrine tumors that arise from the paraganglia of the autonomic nervous system. Tympanic paragangliomas (TPGs) originate from the glomus bodies within the middle ear and are considered the most common primary neoplasms of the temporal bone, after vestibular schwannomas. The clinical presentation varies, with pulsatile tinnitus being the most frequent symptom. Diagnosis relies on a combination of clinical examination, imaging, and histopathological confirmation. Treatment options include surgical excision and, in selected cases, radiation therapy. This report presents a case of TPG in a 54-year-old female and discusses current diagnostic and therapeutic strategies.

CASE PRESENTATION

Clinical History

A 54-year-old female with no significant past medical history presented with a 2-year history of progressive pulsatile tinnitus in the left ear, accompanied by mild hearing loss. There were no associated vertigo, facial nerve involvement, or otalgia. Otoscopic examination revealed a reddish retrotympanic mass occupying the middle ear space. The tympanic membrane was intact, with no signs of effusion or infection.

Imaging Findings

Magnetic resonance imaging (MRI) with contrast showed a 8 mm well-defined high T2, avidly enhancing soft tissue mass lateral to the right cochlear promontary without bone erosion notable. A classic "salt-and-pepper" appearance, characteristic of paragangliomas, was observed on gradient-echo imaging. Digital subtraction angiography (DSA) revealed a hypervascular lesion supplied primarily by the ascending pharyngeal artery.

Histopathology

Surgical excision was performed via a transcanal approach. Histological analysis revealed a well-defined tumor composed of chief cells arranged in a classic "Zellballen" pattern, surrounded by a delicate capillary network. Immunohistochemical staining was positive for synaptophysin and chromogranin A, confirming the neuroendocrine nature of the tumor. S100 protein highlighted sustentacular cells, further supporting the diagnosis of tympanic paraganglioma.

Treatment and Outcome

Complete surgical excision was achieved with no intraoperative complications. Postoperatively, the patient experienced transient conductive hearing loss, which improved over six months. There was no recurrence at the 18-month follow-up, as confirmed by

imaging. Given the benign nature and successful resection, no adjuvant therapy was required.

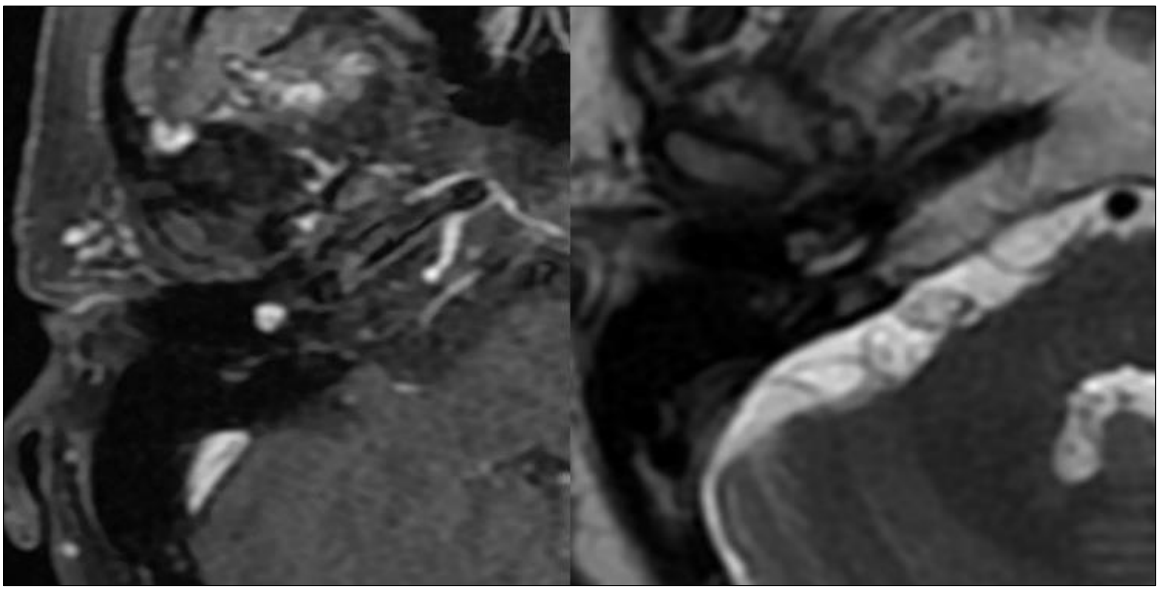

Figure 1 (a, b, c): Well-defined high T2 (Figure B), avidly enhancing (Figue A) soft tissue mass lateral to the right cochlear promontary without bone erosion

DISCUSSION

Tympanic paragangliomas are slow-growing, highly vascular tumors originating from the paraganglia within the middle ear. The differential diagnosis includes other vascular middle ear lesions, such as aberrant internal carotid artery, facial nerve hemangioma, and high-riding jugular bulb. Clinical suspicion, combined with characteristic imaging findings, is essential for accurate diagnosis.

The Fisch classification system categorizes glomus tympanicum tumors based on their extent, with Type A lesions confined to the middle ear and Type B lesions extending beyond the tympanic cavity. In this case, the tumor was classified as Fisch Type A, allowing for a minimally invasive surgical approach.

Surgical resection remains the treatment of choice for localized tympanic paragangliomas, with endoscopic and microscopic techniques offering excellent visualization and preservation of middle ear structures. In cases of extensive or unresectable disease, stereotactic radiosurgery is a viable alternative.

Recent studies suggest that genetic screening for SDHB, SDHC, and SDHD mutations should be considered, particularly in cases with a positive family history or multiple paragangliomas. Although this patient did not undergo genetic testing, future guidelines may emphasize its role in comprehensive patient management.

CONCLUSION

Tympanic paragangliomas, while rare, should be considered in the differential diagnosis of pulsatile tinnitus and retrotympanic masses. Early recognition, imaging, and histopathological confirmation are key to optimizing treatment outcomes. Surgical excision remains the mainstay of treatment, with favorable long-term prognoses in localized cases. Further research is needed to elucidate the genetic basis of these tumors and refine management strategies.

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