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Middle Ear Paraganglioma: Diagnostic Challenges and Surgical Management- A Case Report

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ABSTRACT

Introduction and Purpose. Middle ear paraganglioma (glomus tympanicum) is a rare, benign,

highly vascular neuroendocrine tumor arising on the cochlear promontory. Despite its benign

nature, it may cause local destruction and symptoms such as pulsatile tinnitus, conductive

hearing loss, or a retrotympanic mass. Computed tomography (CT) and magnetic resonance

imaging (MRI) are essential for diagnosis. The aim of this study is to present a clinical case

and underline the role of imaging, embolization, and surgery.

Materials and Methods. A single case was analyzed, including clinical data, otoscopic

findings, CT, angiography, details of embolization and surgery, and histopathological results.

High-resolution CT was used to assess bony involvement, while angiography defined vascular

supply and enabled preoperative embolization. The tumor was surgically removed, and

histopathology confirmed paraganglioma.

Results. The patient presented with pulsatile tinnitus, progressive hearing loss, and intermittent

otorrhea. Otoscopy revealed a pink, pulsatile mass within the external auditory canal. CT

showed a well-defined with limited bony erosion. Angiography demonstrated marked

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vascularity arising from branches of the middle meningeal artery. Embolization effectively reduced tumor perfusion. The mass was completely removed surgically, requiring resection of a diseased incus and reconstruction of the middle ear. Recovery was uneventful.

Conclusion. Middle ear paraganglioma should be considered in patients with pulsatile tinnitus and vascular retrotympanic masses. CT and MRI are essential for evaluating lesion extent, while angiography and embolization reduce intraoperative bleeding and facilitate safer surgery. Surgical excision remains the treatment of choice for class A/B tumors. The present case supports the effectiveness of a multistep approach incorporating imaging, vascular control, and precise surgical technique.

Keywords: glomus tympanicum, middle ear paraganglioma, pulsatile tinnitus, embolization, temporal bone tumor, otologic surgery

Introduction

Paragangliomas constitute a subgroup of neuroendocrine tumors characterized by a highly vascular architecture; these neoplasms originate from the chief cells of the paraganglia of the autonomic nervous system. Glomus tympanicum tumor, also referred to as tympanic paraganglioma, is one of the most common benign tumors of the head and neck region, particularly within the mesotympanum of the middle ear. Although glomus tympanicum tumors are typically benign and only rarely cause autonomic disturbances associated with catecholamine secretion, their continuous growth may become clinically significant due to the close anatomical proximity of the cochlea, jugular bulb, cranial nerves, and the carotid artery [1–3].

This tumor grows slowly and typically occurs sporadically, accounting for approximately 1-3% of all head and neck neoplasms [4]. Chemodectoma is the most common tumor of the middle ear (10–15%) and the second most frequent neoplasm of the temporal bone [2].

The lesions may occur unilaterally, bilaterally, or present as multifocal disease (approximately 10%). A genetic background is identified in 30–40% of patients, and the tumor may coexist with syndromes such as Multiple Endocrine Neoplasia type 2 (MEN II), von Hippel–Lindau disease (VHL), or Neurofibromatosis type 1 (NF-1) [5].

The highest incidence is observed in the fifth decade of life, and the condition occurs significantly more often in women. Approximately 3% of these tumors exhibit hormonal activity [6].

The most commonly reported symptoms include pulsatile tinnitus and hearing loss. Otoscopic examination typically reveals erythema, bulging, and sometimes visible pulsation of the tympanic membrane [2].

A thorough medical history, physical examination, and evaluation of imaging studies are key components in the diagnostic workup of these tumors. Magnetic resonance imaging (MRI) and computed tomography (CT) are complementary modalities for assessing the extent of the lesion prior to surgical planning. CT allows visualization of the tumor mass located on the promontory, its contrast enhancement, and detailed assessment of the bony structures of the middle and inner ear as well as the jugular foramen region. MRI, in turn, enables precise evaluation of the tumor's relationship to soft tissues and vascular structures, as well as potential intra- and extracranial extension [7].

The primary treatment modality is surgical excision of the tumor, performed either through a transcanal approach or via a retroauricular route. The procedure is most commonly conducted using an operating microscope; however, the use of an endoscope offers certain advantages. These include a wider field of view, superior visualization, and the ability to introduce instruments into anatomically confined areas of the middle ear that are typically less accessible under microscopic guidance [8].

Case Description

A female patient was admitted to the Department of Otolaryngology reporting pulsatile tinnitus in the left ear, progressive hearing loss, and intermittent otorrhea. According to the patient, the symptoms had begun several years earlier, but had markedly worsened over the past few months.

In 1981, the patient underwent surgery to remove a lesion from the left ear; histopathological examination dated 12 February 1981 indicated an *angioma*. She also received adjuvant radiotherapy targeted to the left ear as part of her previous treatment.

During the clinical examination, a pink, smooth-walled, soft-elastic, pulsatile proliferative lesion on a broad base was observed within the bony portion of the left external auditory canal,

arising from the postero-inferior canal wall. Due to this mass, the tympanic membrane could not be visualized.

A computed tomography (CT) scan was performed. CT imaging of the left temporal bone (Fig. 1) revealed a heterogeneous, well-demarcated soft-tissue lesion occupying the tympanic cavity and the bony segment of the external auditory canal. The lesion filled the middle ear spaces, remodeling and narrowing the canal lumen. Evidence of erosion and bony remodeling of the postero-inferior external auditory canal wall and middle ear structures was present, suggesting a slowly progressive proliferative process.

The auditory ossicles are partially surrounded by the pathological soft-tissue mass, without clear evidence of their destruction. The mastoid air cells show features of sclerosis. No enlargement of the carotid canal or the jugular bulb is observed.

The CT findings are most suggestive of a vascular lesion, such as a glomus tympanicum tumor or another neoplasm of vascular origin.

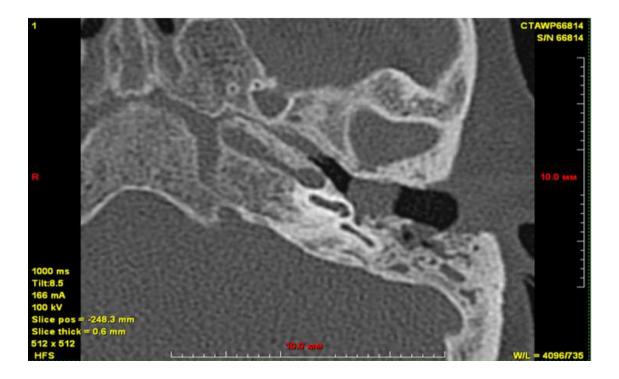


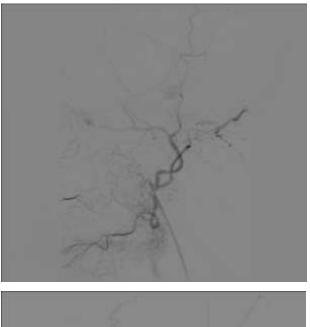
Figure 1. Axial computed tomography scan of the temporal bone (left side)

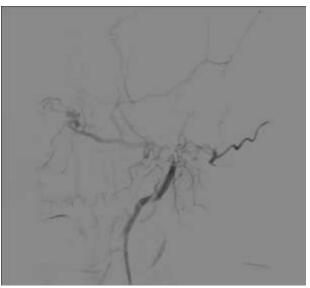
Angiographic evaluation of the external carotid artery revealed pathologically increased vascularity within the region of the tympanic cavity and external auditory canal, corresponding

to a proliferative lesion consistent with a vascular tumor. The vascular supply originated primarily from branches of the left middle meningeal artery, which demonstrated an intense vascular phase with rapid filling of small, tortuous feeding vessels and a characteristic *tumor blush* appearance.

Selective catheterization of the left middle meningeal artery was performed, followed by embolization using calibrated PVA microspheres. This resulted in a marked reduction in tumor vascularity and disappearance of the *tumor blush* on post-embolization control angiography.

Embolization was carried out as a preparatory step prior to the planned surgical treatment to minimize intraoperative bleeding.





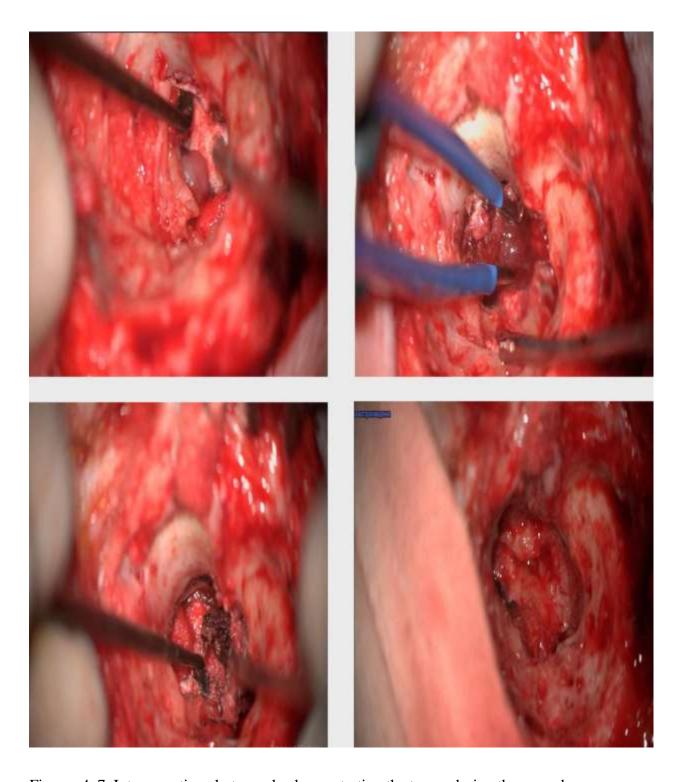
Figures 2–3. External carotid artery angiography

The patient underwent surgery under endotracheal general anesthesia. A postauricular incision was made, the spina suprameatum was exposed, and the antrum was opened. The posterior bony wall of the external auditory canal was removed, and the bony overhang was smoothed.

In the tympanic cavity, pink, pulsatile lesions were observed, filling its lumen and partially extending into the external auditory canal. The mass was firmly adherent to the tympanic membrane and the walls of the tympanic cavity. The lesion was removed using blunt dissection and electrocautery, together with a carious incus that had been destroyed. Bleeding was minimal; the tumor pedicle was coagulated.

The opening of the Eustachian tube was reinforced with a fragment of mucosa. A T-plasty of the external auditory canal skin was performed, and the tympanic cavity was packed. Skin sutures and an aseptic dressing were applied. The intraoperative course was uneventful.

Histopathological diagnosis: paraganglioma.



Figures 4–7. Intraoperative photographs demonstrating the tumor during the procedure.



Figure 8. Postoperative photograph of the excised lesion.

In the postoperative period, the patient remained in satisfactory condition, and no bleeding was observed. Skin sutures were removed on postoperative day 7, and the packing on day 14. The patient was discharged in good overall condition.

The first descriptions of paragangliomas within the temporal bone were provided by Stacy R. Guild, who in 1941 reported the presence of an oval lesion located in the jugular bulb and on the promontory of the middle ear, extending along Jacobson's nerve-a branch of the glossopharyngeal nerve. He termed this lesion "glomus jugularis" [9].

Four years later, Harry Rosenwasser published a case report of a 36-year-old man with a middle ear tumor extending into the external auditory canal. During surgery, he noted tight adhesion of the mass in the hypotympanum region and very profuse bleeding. Histopathological examination revealed neoplastic paraganglionic cells consistent with a "carotid body tumor" [9].

The diagnosis of glomus tympanicum is based on clinical evaluation, audiological testing, and imaging studies. Imaging techniques such as computed tomography and magnetic resonance imaging play a pivotal role in early detection. On radiological assessment, paragangliomas

typically present as highly vascular tumors with intense contrast enhancement. Imaging is also valuable for monitoring tumors within the middle ear. Temporal bone CT is frequently used to evaluate progression [10].

Glomus tumors are slow-growing, benign lesions and represent the most common primary neoplasms of the middle ear. According to the literature, the initial symptoms typically include pulsatile tinnitus synchronized with the heartbeat, hearing loss (conductive, sensorineural, or mixed), and otalgia. The most characteristic otoscopic finding is a pulsatile mass visible behind the tympanic membrane, usually located in the inferior quadrant [11].

In contrast to certain other head and neck paragangliomas-such as carotid body tumors, jugular paragangliomas, or vagal paragangliomas-which often require multidisciplinary management involving oncologists, neurosurgeons, vascular surgeons, and radiologists, the treatment of middle ear paragangliomas relies primarily on single-specialty care consisting of otologic microsurgery [11].

Analysis of available data indicates that extended tympanotomy techniques allow complete excision of most middle ear paragangliomas (classes A and B) without significant deterioration of auditory function.

In cases where the tumor extends into the mastoid process-into the infralabyrinthine space (class B)-or when there is concern regarding the integrity of the inferior wall of the tympanic cavity or the lateral wall of the internal carotid artery canal, a more advanced surgical approach using radical petrotympanotomy is preferred. In our experience, this method provides optimal access to the key anatomical structures of the middle ear (the inferior wall of the tympanic cavity), the mastoid process, and the temporal bone (the lateral wall of the internal carotid artery canal) when neoplastic progression is suspected [7,12].

Conclusions

Middle ear paragangliomas (glomus tympanicum), although rare, should be considered in the differential diagnosis of any vascular lesion within the ear-particularly in patients presenting with pulsatile tinnitus and progressive hearing loss. A thorough medical history, otoscopic evaluation, and high-resolution imaging studies (CT and MRI) are essential for accurate diagnosis and assessment of tumor extent. These imaging modalities provide complementary

information: CT enables detailed evaluation of bony structures, whereas MRI delineates the

involvement of soft tissues.

Angiography combined with preoperative embolization serves as a valuable preparatory tool in

the management of vascular tumors of the middle ear. In the present case, embolization of the

left middle meningeal artery significantly reduced tumor vascularity, contributing to a safer

surgical procedure with minimal intraoperative bleeding.

Surgical treatment remains the method of choice for class A and B tympanic paragangliomas.

Proper selection of the operative technique-including radical tympanotomy or, when necessary,

extended petrotympanotomy-allows complete tumor resection while minimizing the risk of

injury to critical otologic structures. Histopathological confirmation is a necessary component

of the diagnostic and therapeutic process; in this case, the final diagnosis of paraganglioma was

consistent with the clinical and radiological findings.

The presented case highlights the importance of a multistep, integrated approach-combining

imaging assessment, vascular evaluation, and surgical management-to achieve favorable

therapeutic outcomes with a low risk of complications.

Author's contribution:

Conceptualization, supervision and project administration- AK, TS

Methodology- TS

Software, validation-AK, TS

Formal analysis, investigation, resources-TS, KS

Writing original draft preparation- AK, TS, DW, WŁ, WB, DP, KS

Writing review, editing and visualization- AK, WB, TS, KS

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