Paraganglioma Presenting with Epistaxis: A Diagnostic Dilemma

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ABSTRACT

Paraganglioma is a benign tumour which is highly vascularised and mostly occurs in the middle ear. Although rare, patients typically in middle-aged group and present with pulsatile tinnitus and conductive hearing loss, but they may also experience symptoms such as vertigo, bleeding, and ear pain. These tumours are diagnosed on the basis of clinical history, thorough examination, and certain imaging modalities, where MRI is superior to other scans in reporting the extent, size, location, and nature of the tumour. Out of various treatment options, the most effective option for definitive management is surgery. This case reports an unusual presenting complaint of paraganglioma, *i.e.*, epistaxis experienced by a middle-aged woman. This case is a compelling example of the necessity for a broad differential diagnosis, comprehensive evaluation, and a high index of suspicion when encountering atypical presentations of diseases that are uncommon or rare causes of frequently occurring symptoms in otolaryngology.

Key Words: *Paraganglioma, Benign, Epistaxis*.

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INTRODUCTION

Paraganglioma, or glomus tumour, is a rare middle ear tumour arising from non-chromaffin sympathetic paraganglia cells, mostly in middle-aged women.[1-3] Typical symptoms include hearing loss, tinnitus, vertigo, ear pain, and occasional bleeding.[4,5] While a case reported haemoptysis and epistaxis from the USA,[6] the sole epistaxis is unreported in Asia. Diagnosis involves history, examination, and CT/MRI, with MRI superior for delineation.[7] Treatment includes observation, radiation, or surgery, with surgical removal being the most effective option, either *via* open or endoscopic approaches.[8] Unusual presentations, such as nosebleeds, can lead to misdiagnosis without thorough assessment. This highlights the consideration of rare origins of common symptoms.

CASE REPORT

A 55-year woman with hypertension, diabetes mellitus, and a history of cerebrovascular accident (CVA) presented as an outpatient in otolaryngology clinic with complaint of occasional bleeding from the left nose for 4 years, which initially started with frequency of 2-3 episodes per year, then it progressed to 4-5 episodes per month after two years.

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Episodes were usually controlled by conservative measures. She also has decreased hearing from the left ear for 17 years, mild-to-moderate and progressive in nature, not associated with tinnitus, vertigo, ear pain, or any other signs/symptoms.

On general physical examination, she had left-sided facial weakness (House-Brackman Class III), which was present after a CVA that occurred 12 years back. Nasal examination was normal with no active bleeding at the time of presentation. Examination of the left ear revealed a scar from a past surgery on the left mastoid area, indicating a mastoidectomy performed 17 years ago, though no record of the procedure was available. A polypoidal mass, reddish-pink in colour with irregular surface and fleshy texture, was identified in the left external auditory canal upon otoscopy, completely occluding the view of the tympanic membrane. The right ear examination was normal. On the tuning fork test, Rinne's was negative in the left ear, and Weber's was lateralised to the right side. Pure tone audiometry showed mild-to-moderate mixed hearing loss in the left ear. There was no record of previous audiometry.

A CT scan of temporal bone with contrast reported a soft tissue mass in the left ear measuring 2.7 × 2.1 × 2.6 cm (TR × AP × CC) in dimensions (Figure 1) involving the left external acoustic meatus and mastoid air cells, with erosion of adjacent bone and attenuation of ossicles. A biopsy was taken under general anaesthesia due to the potential risk of bleeding. Histopathological examination led to the diagnosis of paraganglioma. According to the Modified Fisch classification, it was a Class-B2 tumour, involving the middle ear with extension to the hypotympanum and the mastoid.

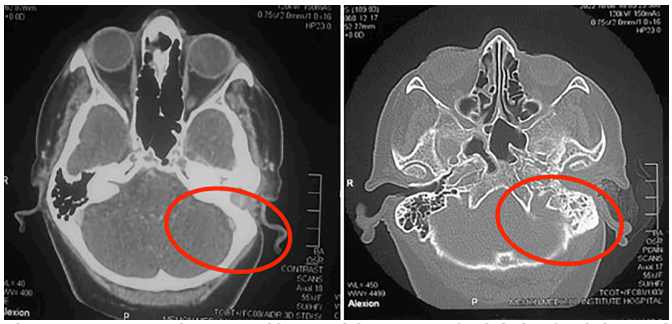

Figure 1: CT scan of temporal bone with contrast (axial view). Right: Red outlined circle shows soft tissue mass occupying left external auditory canal and middle ear cavity with erosion of ossicles; left: Mastoid air cells are sclerotic.

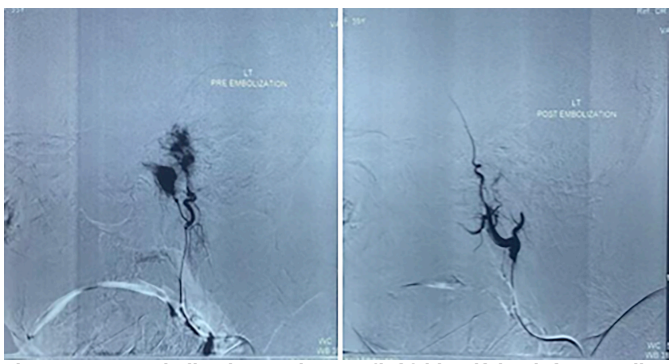

Figure 2: Pre-embolisation angiogram (left) identifying left ascending pharyngeal and posterior auricular arteries as the main feeding vessels. Post-embolisation angiogram (right) shows no flushing.

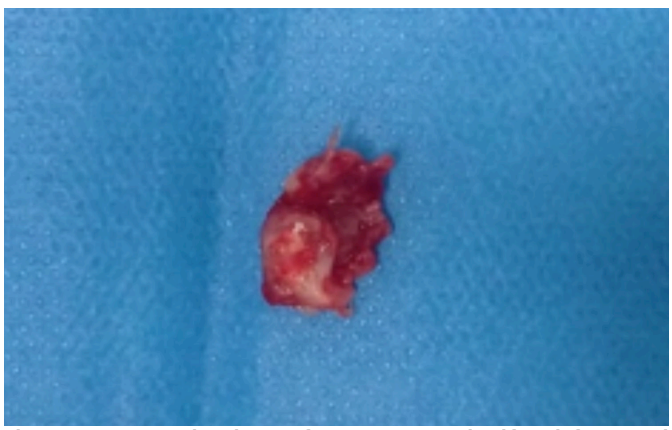

Figure 3: Postoperative picture of 2 × 2 cm mass excised from left external auditory canal and middle ear cavity *via* post-auricular approach.

Selective angiograms were performed, which revealed 80% of the tumour supply from left ascending pharyngeal and posterior auricular arteries and 20% supply from branches of the internal maxillary artery. Subsequently, feeding vessels were super selectively cannulated using a 2.7 Fr microcatheter. Post-embolisation angiogram revealed complete embolisation of the tumour blush. (Figure 2).

The patient underwent a canal wall down mastoidectomy under general anaesthesia. Intraoperatively, a 2 × 2 cm mass was excised without any complications (Figure 3). No middle ear ossicles were seen, and the floor of the ear was intact. Final histopathology of the excised sample confirmed the diagnosis of paraganglioma. Postoperative recovery was uneventful.

DISCUSSION

Glomus tympanum tumours are paragangliomas in the middle ear that arise from glomus bodies or paraganglia, which are specialised organelles distributed along the course of autonomic nerves throughout the body. Paraganglia are frequently located in the temporal bone, specifically in the mesotympanum, often alongside Jacobson's nerve, which is a branch of the glossopharyngeal nerve.[4,5]

These are rare benign tumours with the incidence of about one per 1.4 million people per year.[8] This patient reported with approximately four-year history of unusual presentation of nasal bleeding. This highlights the need for a high index of suspicion and the inclusion of rare pathologies in the differential diagnosis, especially when standard treatments for common conditions do not yield the expected results and possibly increase the costs and adverse outcomes for affected patients. Possible pathophysiology for a nasal bleed can be dribbling of blood from the middle ear to the nose *via* the Eustachian tube. CT scan and MRI of the temporal bone are imaging modalities for the diagnosis of paraganglioma to see the extent and size of the tumour, but MRI is better than CT scan as it differentiates the tumour from inflammatory tissue and blood.[7] Clinically, paragangliomas are slow-growing but locally destructive lesions. So, surgical excision is considered to be an effective treatment option, with different surgical approaches depending on the location of the disease. Due to high vascularity, preoperative selective embolisation is considered to reduce the risk of bleeding.[9] The purpose of this case report is to highlight the unusual presentation by disseminating the understanding of diverse manifestations of paraganglioma and refining the approach to diagnosis and management.

PATIENT'S CONSENT:

Informed consent was obtained from the patient to publish the data concerning this case.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

ARS: Literature search and write-up.
YA: Write-up and editing.
MM: Write-up and data collection.
MI: Supervision and project idea.
SA: Supervision of the project, submission, and final drafting.
All authors approved the final version of the manuscript to be published.

REFERENCES

1. Appannan VR, Md Daud MK. Glomus tympanicum. *Malays Fam Physician* 2018; **13(1)**:45-8.
2. Sweeney AD, Carlson ML, Wanna GB, Bennett ML. Glomus tympanicum tumours. *Otolaryngol Clin North Am* 2015; **48(2)**:293-304. doi: 10.1016/j.otc.2014.12.004.
3. Malik NZ, Baumann A, Curtis D, Shaw A, Jeyakumar A. An unusual presentation and diagnosis of a type 4 middle ear

paraganglioma: A case report. *Cureus* 2023; **15(2)**:e35358. doi: 10.7759/cureus.35358.

4. Chou YL, Huang BR, Wang CH. En bloc surgical removal of an asymptomatic glomus tympanicum tumour. *J Chin Med Assoc* 2011; **74(11)**:520-2. doi: 10.1016/j.jcma.2011.09.009.
5. Dimakis C, Beka D, Papageorgiou E, Tsetsos N, Poutoglidis A, Gortsali A, *et al*. A case report of glomus tympanicum complicated with facial nerve palsy. *Iran J Otorhinolaryngol* 2022; **34(125)**:327-31. doi: 10.22038/IJORL.2022.64737.3217.
6. Akpan I, Orabueze I, Ibizugbe U, Akhdar G, Agyeman WY. A Rare differential of epistaxis. *Cureus* 2024; **16(5)**:e59981. doi: 10.7759/cureus.59981.
7. Tuan AS, Chen JY, Mafee MF. Glomus tympanica and other intratympanic masses: Role of imaging. *Operat Tech Otolaryngol Head Neck Surg* 2014; **25(1)**:49-57. doi: 10.1016/j.otot.2013.11.007.
8. Daneshi A, Asghari A, Mohebbi S, Farhadi M, Farahani F, Mohseni M. Total endoscopic approach in glomus tympanicum surgery. *Iran J Otorhinolaryngol* 2017; **29(95)**:305-11.
9. Texakalidis P, Charisis N, Giannopoulos S, Xenos D, Rangel-Castilla L, Tassiopoulos AK, *et al*. Role of preoperative embolization in carotid body tumour surgery: A systematic review and meta-analysis. *World Neurosurg* 2019; **129**:503-13.e2. doi: 10.1016/j.wneu.2019.05.209.