CASE STUDY

Tympanic Paraganglioma With Extension Into the Eustachian Tube and Nasopharynx: A Case Report

Miriam Portero de la Torre,a,* Rafael Sánchez Gutiérrez,b Inmaculada Gallardo Muñoz,a Francisco A. Bravo Rodrígueza

a Servicio de Radiodiagnóstico, Hospital Universitario Reina Sofía, Córdoba, Spain
b Servicio de Otorrinolaringología, Hospital Universitario Reina Sofía, Córdoba, Spain

KEYWORDS
Tympanic paraganglioma; Eustachian tube; Nasopharynx; Epistaxis

PALABRAS CLAVE
Paraganglioma timpánico; Trompa de Eustaquio; Nasofaringe; Epistaxis

Abstract
Tympanic paragangliomas usually present as a vascular middle ear mass, with the most common presenting symptoms being pulsatile tinnitus and hearing loss. We report an unusual case of a recurrent tympanic paraganglioma extending along the Eustachian tube and nasopharynx, presenting with recurrent epistaxis.

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Introduction
Paragangliomas (also known as “glomus” or “chemodectomas”) are rare tumours derived from ectodermal cells of the neural crest. The most frequent locations in the head and neck region are the carotid body, jugular foramen, vagus nerve and middle ear (tympanic).1

Tympanic paragangliomas are usually located immediately lateral to the middle ear promontory and may extend to the mastoid or the external auditory canal. Rare extensions to the Eustachian tube have also been reported.2–5

We report the case of a recurrent tympanic paraganglioma extending to the Eustachian tube and nasopharynx.

Clinical Case
The patient was a 62-year-old female suffering recurrent epistaxis of 1-month evolution. Four years earlier she had been intervened, through left mastoidectomy, due to a left

* Corresponding author.
E-mail address: miriam_portero@hotmail.com (M. Portero de la Torre).

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Tympanic mastoid paraganglioma with a millimetric component in the Eustachian tube which was controlled. During those 4 years the patient remained asymptomatic (even missing some of the scheduled reviews) until the onset of bleeding episodes.

We performed a computed tomography (CT) scan with intravenous contrast, which revealed a hypervascular mass in the left nasopharynx, extending to the Eustachian tube and middle ear and eroding the bony walls (Fig. 1).

Diagnostic angiography showed a hypervascular lesion with 3 compartments: 1 in the middle ear, irrigated by the inferior tympanic artery (ascending pharyngeal branch), another in the petrous apex, irrigated by the caroticotympanic artery (a branch of the internal carotid artery) and another with caudal extension, irrigated by branches of the ascending pharyngeal and accessory meningeal arteries (Fig. 2).

The findings were indicative of tympanic paraganglioma, extending into the Eustachian tube and nasopharynx.

In a second therapeutic stage, we conducted a preoperative therapeutic angiography in order to reduce the risk of bleeding during surgery. We closed the vascular supply from the middle meningeal branches, internal maxillary, stylomastoid and pharyngeal trunk of the left ascending pharyngeal artery by embolisation with Contour® polyvinyl alcohol particles of 150–250 microns (Boston Scientific, Cork, Ireland) and fibrous coils of 2 × 10 mm (Boston Scientific, Cork, Ireland).

Subsequently, through endoscopic sinus surgery (a less invasive technique than the external approach), we performed laser ablation of the nasopharyngeal tumour and excised the tumoural portion located in the middle ear and Eustachian tube (this resection was partial because the radical treatment entailed unassumable sequelae).

The anatomopathological diagnosis was paraganglioma.

After the intervention, the patient underwent radiosurgery of the nasopharyngeal component. In the last control CT scan (2 years after surgery), this component had become reduced to half its original size and the patient remained asymptomatic.

Discussion

Paragangliomas are rare tumours. The peak age of onset is between the fifth and sixth decades of life, with a higher prevalence among women.¹

Paragangliomas have capsules with septa and a stroma composed of sinusoidal spaces which are anastomosed together, thus giving them their characteristic hypervascularity with pathognomonic arteriographic signs and haemorrhagic character (which justifies preoperative embolisation).²

Tympanic paragangliomas originate from glomus bodies of the cochlear promontory, along the tympanic branch of the glossopharyngeal nerve (nerve of Jacobson). Examination usually reveals a hypotympanic or mesotympanic mass, with the most common symptoms being pulsatile tinnitus and unilateral hearing loss. CT and magnetic resonance imaging (MRI) scans show a mass in the medial wall of the middle ear, which may extend to the mastoid or external ear canal. Angiography shows characteristic findings, revealing a hypervascular mass with compartmentalised vascularisation by specific hypertrophied arteries.²

The differential diagnosis should include undifferentiated carcinoma, which shows less enhancement after contrast administration, haemangioma and Kaposi sarcoma, which are vascular tumours, although there are no reported cases of Eustachian tube extension, and juvenile nasopharyngeal angiofibroma, which is a highly vascular tumour, almost exclusively found in adolescent males.

Only 4 cases of tympanic paraganglioma extending into the Eustachian tube and nasopharynx have been published,²⁻⁵ all of them occurring in women aged between

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**Figure 1**  CT scan with intravenous contrast. The axial (A) and coronal (B) sections show a hypervascular mass located in the left nasopharynx (white arrow), Eustachian tube (black arrow) and middle ear. (C) Reconstruction of the lesion in the major axis (white arrow), showing its extension from the middle ear to the nasopharynx.
50 and 65 years and beginning with epistaxis, as in our patient. In the cases described, the diagnosis was obtained by CT and/or MRI, supplemented in some cases by angiography and biopsy. Treatment was surgical resection or radiotherapy (1 patient underwent ligation of the external carotid artery prior to radiotherapy). Only 1 of these cases underwent preoperative embolisation by angiography.

Our case illustrates a very unusual extension of tympanic paraganglioma, with characteristic findings on CT scans and angiography. The latter was useful in both diagnosis and preoperative treatment of the lesion.

In summary, the diagnosis of paraganglioma should be considered in patients with epistaxis and CT and/or MRI findings of hypervascular lesions in the middle ear extending to the Eustachian tube and nasopharynx. In such cases, angiography will enable better characterisation of the lesion and its vascular supply. Embolisation prior to biopsy or surgical resection will diminish the risk of uncontrollable bleeding.

Conflict of Interests

The authors have no conflicts of interest to declare.

References