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ORIGINAL PAPER

Case Series of Head and Neck Paragangliomas: Radiological Diagnosis and Clinical Characteristics

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ABSTRACT

Paragangliomas are neuroendocrine tumors arising from extra-adrenal paraganglionic tissue with characteristic anatomic distribution in the head and neck. These include carotid body paragangliomas, tympanic paragangliomas, jugular/jugulotympanic paragangliomas and vagal paragangliomas. This case series compiles radiological features, clinical presentation and treatment strategies from documented imaging and clinical sources. Emphasis is placed on imaging modalities and their diagnostic value in treatment planning and tumor characterization. This summary integrates evidence and case illustrations to emphasize the importance of early recognition and the role of multimodal radiologic assessment.

Keywords: paraganglioma, carotid body paraganglioma, jugulotympanic paraganglioma, tympanic paraganglioma, vagal paraganglioma, radiological diagnosis.

INTRODUCTION

Paragangliomas are tumors originating from extra-adrenal paraganglia of the autonomic nervous system, often located in proximity to the carotid body, jugular foramen, vagus nerve, or middle ear structures. These lesions are derived from neuroectodermal chromaffin cells and classified based on their anatomical location. Head and neck paragangliomas (HNPGLs) constitute about 0.6% of head and neck neoplasms, with carotid body tumors accounting for the majority (60–67%) (1, 2).

These tumors are highly vascular, benign in most cases, and slow-growing. However, due to their location near cranial nerves and major ves-

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sels, clinical symptoms can be significant, necessitating detailed imaging and careful management. This case series reviews representative examples and discusses diagnostic and treatment considerations. ❑

CASE SUMMARIES

Case 1: carotid body paraganglioma

A 40-year-old woman presented with a gradually enlarging non-tender pulsatile neck mass located at the level of the carotid bifurcation on right side. Contrast-enhanced computed tomography (CT) revealed a well-defined enhancing mass splaying the right internal and external carotid arteries, producing the classic "lyre sign". The tumor exhibited intense enhancement. Histopathology confirmed a benign paraganglioma (Figure 1).

Case 2: jugulotympanic paraganglioma

A 64-year-old female reported pulsatile tinnitus in her left ear. Otoscopy revealed a retrotympanic vascular mass. An intensely enhancing lesion was seen in the left jugular fossa involving the pars nervosa and pars vascularis, causing widening of the jugular fossa with associated scalloping and erosion of the jugular spine. The lesion was eroding the posterior wall of the carotid canal. However, no extension was seen into the carotid canal and the internal carotid artery (ICA) appeared normal in caliber and contrast opacification. Medially, the lesion did not extend into the hypoglossal canal. Laterally, the lesion eroded the mastoid bone and extended into the middle ear cavity. The footplate and crus of the stapes were embedded and not separately visualized. Preoperative embolization was performed to reduce intraoperative bleeding. Surgical resection was done *via* an infratemporal fossa approach (Figure 2).

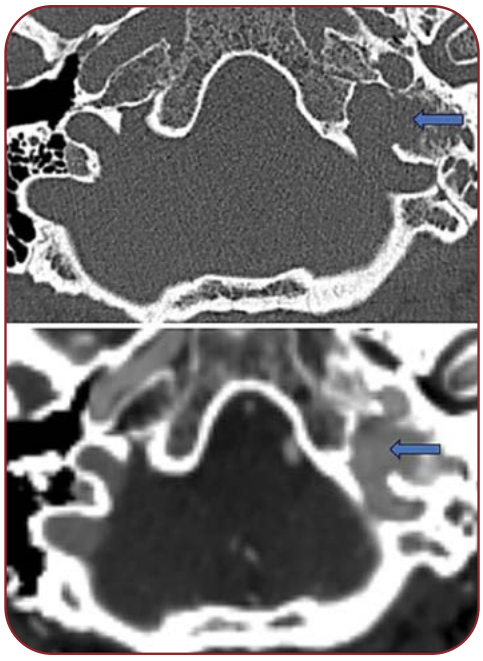

FIGURE 2. Case 2: jugulotympanic paraganglioma. On high resolution computed tomography (HRCT) temporal bone axial images, a relatively well-defined lytic lesion (closed arrow) is seen in the left jugular fossa involving the pars nervosa and pars vascularis, causing widening of the jugular fossa with associated scalloping and erosion of the jugular spine. The lesion is eroding the posterior wall of the carotid canal. On computed tomography (CT) angiography, the lesion shows avid enhancement.

Case 3: tympanic paraganglioma

A 51-year-old woman experienced progressive hearing loss and tinnitus on the right side. An 8 mm intensely enhancing lesion was seen in the region of the mesotympanum and hypotympanum overlying the cochlear promontory. On CT

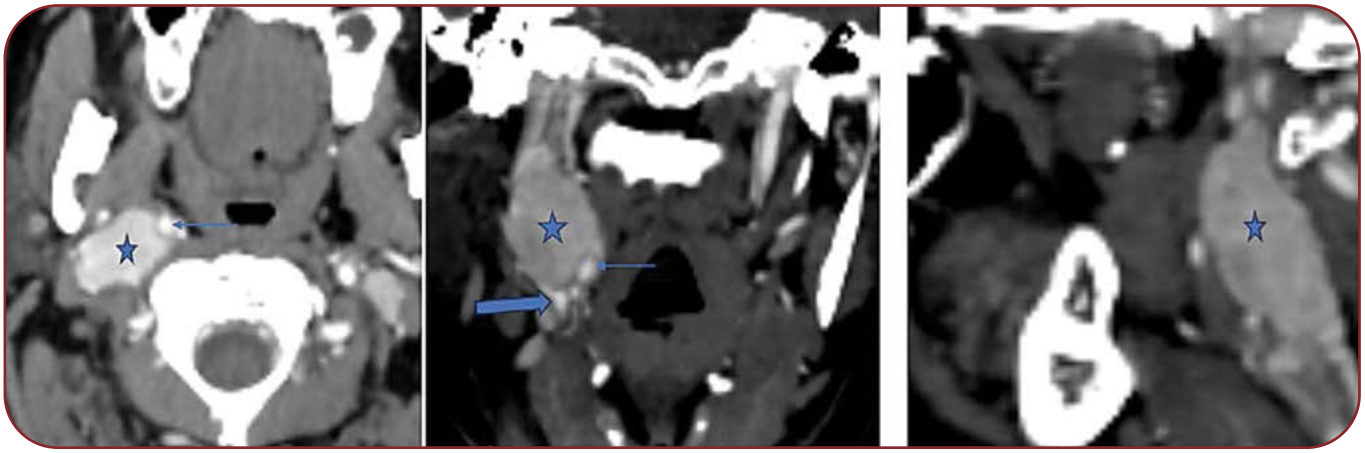

FIGURE 1. Case 1: carotid body paraganglioma. Computed tomography (CT) angiography neck axial and sagittal images showing a well-defined intensely enhancing lesion (star) epicentered in the right carotid sheath at the carotid bifurcation. The right internal carotid artery (ICA) (thin arrow) is seen along the medial aspect of the lesion. Coronal images show the lesion splaying the ICA (thin arrow) and external carotid artery (ECA) (thick arrow).

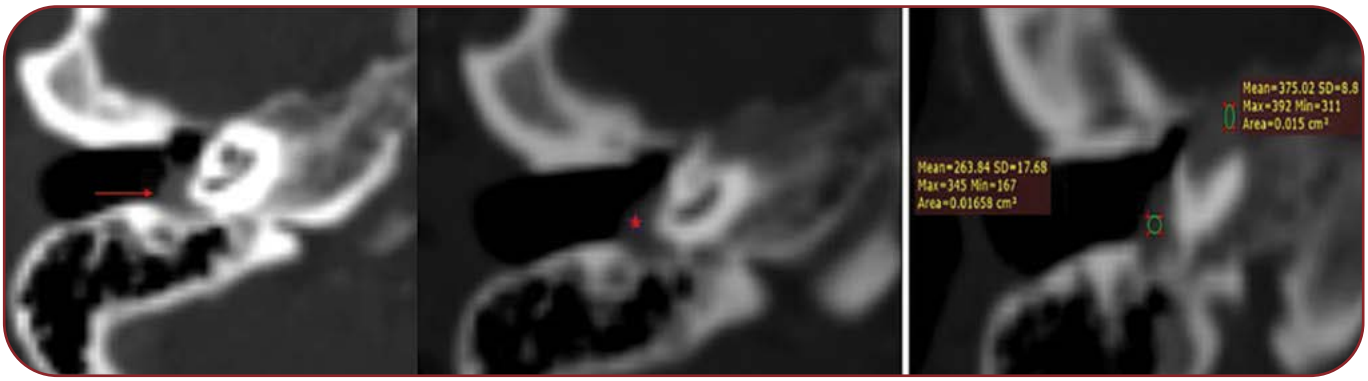

FIGURE 3. Case 3: tympanic paraganglioma. On high resolution computed tomography (HRCT) temporal bone axial images, an 8 mm lesion (red arrow and star) is seen in the region of the mesotympanum and hypotympanum overlying the cochlear promontory. On computed tomography (CT) angiography, the lesion shows avid enhancement (ROI).

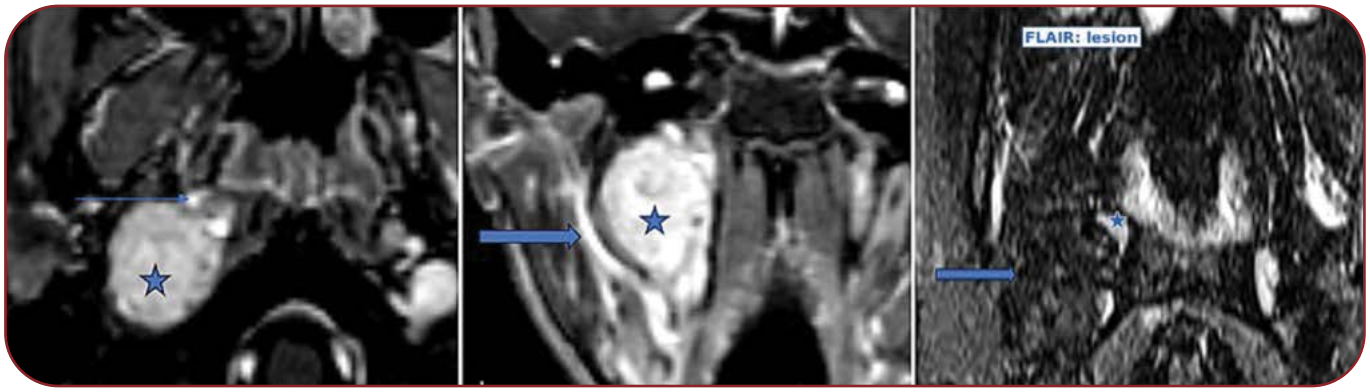

FIGURE 4. Case 4: vagal paraganglioma. Contrast-enhanced magnetic resonance imaging (CEMRI) axial and coronal images of the skull base revealed a well-defined lobulated mass lesion (star) epicentered just inferior to the right jugular fossa. The lesion shows intense homogeneous enhancement on post-contrast images. It displaces the internal carotid artery (thin arrow) anteromedially and the internal jugular vein (thick closed arrow) posterolaterally, without widening of the carotid bifurcation. The T2-weighted/FLAIR image demonstrates heterogeneous hyperintensity with internal flow voids (salt-and-pepper appearance). In the rightmost FLAIR image, the blue arrow and star indicate the same vagal paraganglioma. FLAIR=fluid-attenuated inversion recovery

angiography, no obvious arterial supply to the lesion could be identified. The patient underwent tympanotomy and the tumor was excised with preservation of hearing. Postoperative follow-up was uneventful (Figure 3).

Case 4: vagal paraganglioma

A 15-year-old female presented with a gradually enlarging mass on the right side of her neck. Magnetic resonance imaging (MRI) revealed a well-defined lobulated mass lesion epicentered just inferior to the right jugular fossa. The lesion appeared isointense on T1-weighted images. On T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences, it was heterogeneously hyperintense with a few internal punctate flow voids, giving a salt-and-pepper appearance. The lesion showed intense homogeneous enhancement on post-contrast images. The lesion displaced the internal carotid artery anteromedially and the internal jugular vein posterolaterally, without widening of the carotid bifurcation. These imaging findings were suggestive of a vagal paraganglioma. Angiography confirmed vascularity supplied predominantly by the ascending pharyngeal artery. The patient underwent staged embolization and tumor excision, with partial symptom resolution postoperatively (3) (Figure 4).

Diagnostic imaging modalities

Ultrasound – A useful approach as an initial modality, especially in evaluating cervical masses. It can identify hypervascular lesions with low-resistance arterial flow. However, the limited sensitivity for skull base lesions reduces its utility as a standalone tool (1, 4).

Computed tomography and magnetic resonance imaging – Computed tomography provides

bony detail and enhancement characteristics, while MRI is superior for delineating soft tissue extension and cranial nerve involvement. The salt-and-pepper appearance on MRI is a hallmark sign – reflecting hemorrhagic components (salt) and flow voids (pepper) (5, 6). Head and neck paragangliomas typically show marked enhancement after gadolinium administration.

Angiography – It is used less frequently for primary diagnosis, but essential for surgical planning and preoperative embolization. It reveals intense tumor blush and feeding arteries, aiding in precise localization (3, 7).

Nuclear imaging – Metaiodobenzylguanidine (MIBG) scintigraphy, fluorodeoxyglucose-positron emission tomography (FDG-PET) and octreotide scans help evaluate multifocality or metastasis, particularly in functional or hereditary tumors (8, 9). ❑

DISCUSSION

Head and neck paragangliomas are a diverse group of highly vascular tumors derived from paraganglionic tissue. These tumors, particularly tympanic and jugulotympanic paragangliomas, are more common in females and typically manifest in middle-aged adults. The clinical presentation depends largely on the site and size of the tumor. Paragangliomas of the head and neck, though rare, require high clinical suspicion and comprehensive imaging evaluation for accurate diagnosis and effective management. Multimodality imaging, particularly MRI and angiography, plays a crucial role in the characterization and surgical planning of these tumors. Early diagnosis and multidisciplinary treatment planning are essential to optimize outcomes, minimize complications and preserve neurological function. Head and neck paragangliomas can present variably depending on their location:

- Carotid body paragangliomas present as lateral neck masses at the angle of the mandible and are the most common subtype. Catecholamine secretion is rare but should be excluded prior to intervention (10).
- Jugular and jugulotympanic paragangliomas commonly cause auditory symptoms. Their proximity to cranial nerves IX–XI poses surgical challenges.
- Tympanic paragangliomas are the most frequent middle ear paragangliomas, typically diagnosed by their location and retrotympanic appearance.
- Vagal paragangliomas are rare, often large, and may extend to the skull base. Hoarseness and vagal nerve palsy are typical late symptoms (3, 9).

Though mostly benign, paragangliomas may be multicentric or malignant. Up to 10% may be familial and associated with syndromes such as MEN II or Von Hippel-Lindau disease. Genetic testing should be considered in patients with multiple paragangliomas or a family history of neuroendocrine tumors. Mutations in succinate dehydrogenase (SDH) genes are commonly implicated. Familial cases often demonstrate bilateral and multicentric disease, warranting lifelong surveillance. Hence, genetic testing and family screening may be warranted in select cases (2, 8, 9).

Management

Treatment is individualized, depending on tumor size, location, symptoms and patient comorbidities. Options include: a) observation – in elderly or asymptomatic patients; b) surgical resection – mainstay for accessible lesions with acceptable morbidity; c) preoperative embolization – decreases intraoperative bleeding, especially in jugular, jugulotympanic and vagal paragangliomas (5); and d) radiation therapy – used for unresectable lesions or recurrence. ❑

CONCLUSION

Paragangliomas of the head and neck are rare but clinically significant tumors. Radiological imaging plays a crucial role in diagnosis, characterization, and treatment planning. Multimodal imaging with CT, MRI and angiography enables precise localization and vascular assessment. While surgery remains the definitive treatment in most cases, embolization and radiation therapy are valuable adjuncts. Awareness of classic imaging features and clinical patterns is essential for early recognition and optimal management. ❑

Patients' written informed consent: Patients' written informed consent for publication of anonymized clinical and imaging information was obtained.

Conflicts of interest: none declared.

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