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Paragangliomas of the head and neck: Imaging assessment

By Alejandro Zuluaga, MD; Daniel Ocazionez, MD; Roy Riascos, MD; Enrique Palacios, MD; Carlos S. Restrepo, MD

Introduction

The parasympathetic nervous system is the site of origin of paragangliomas (PGLs) localized in the head and neck region. These head and neck PGLs are usually benign and hormonally inactive.1

PGLs have been referred to as *chemodectomas* and *non-chromaffin tumors*. Currently they are named by anatomic site: carotid PGL, tympanic PGL, jugular PGL, and vagal PGL. Other PGLs at less common sites are laryngeal PGL, orbital PGL, and aorto-pulmonary PGL. Approximately 1 of every 30,000 head and neck tumors is a PGL.2

Most PGLs present clinically as solitary lesions. However, they can also be multifocal. PGLs may also be components of syndromes such as multiple endocrine neoplasia type 2, von Hippel-Lindau disease, and neurofibromatosis type 1. A familial basis of these tumors has been firmly established, with identification of at least three genetic loci: PGL1, PGL2, and PGL3.3

Although PGLs are usually benign tumors, they occasionally metastasize to the lungs, bones, lymph nodes, and other sites.

Imaging evaluation of PGLs

Imaging plays a vital role in the characterization, diagnosis, and treatment of head and neck PGLs.

Computed tomography (CT) allows preoperative assessment of the extent of tumor and determines whether there is bone invasion at the skull base. It is also valuable for evaluating PGLs involving the temporal bone, demonstrating fine bony detail of the middle ear, jugular foramen, and cerebellopontine angle. There has been concern regarding hypertension in the use of nonionic contrast in these patients; however, studies have shown the use of contrast to be safe.4

Magnetic resonance imaging (MRI) using unenhanced and enhanced fat-suppression sequences provides further information about tumor margins, the soft-tissue anatomy of lesions invading the skull base, and vascular structures. In angiography, these lesions typically show tumor blush in the arterial phase due to high vascularity; the presence of early draining veins is also characteristic of these lesions.

MRI characteristics of all PGLS are similar. A well-defined hypointense mass with areas of signal void is seen on T1-weighted MRIs. Diffuse, marked enhancement is seen following IV contrast administration. T2-weighted MRIs demonstrate a well-defined, heterogeneously hyperintense mass. Punctate vascular flow voids within the tumor are often seen, consistent with the hypervascular nature of this tumor. Olsen et al stated that in PGLs larger than 2 cm, an apparently unique “salt and pepper” pattern can be seen on T1- and T2-weighted images, with serpentine and arborizing flow voids reflecting hypervascularity.5

Contrast-enhanced magnetic resonance angiography has recently been proposed as a complement to MRI sequences in the detection of PGLs.6 Selective angiography is currently used for preoperative embolization and mapping of the vascular supply before surgical resection, rather than for diagnostic purposes alone. Characteristic features include high vascularity with multiple feeding arteries, rapid draining veins, and dense tumor blush.5,7

Scintigraphy using 111 indium pentetreotide is the radiotracer of choice with a sensitivity of 94%. Meta-iodobenzylguanidine (MIBG) is less sensitive and less specific as it only concentrates in functional PGLs.8

Characteristics of PGL types

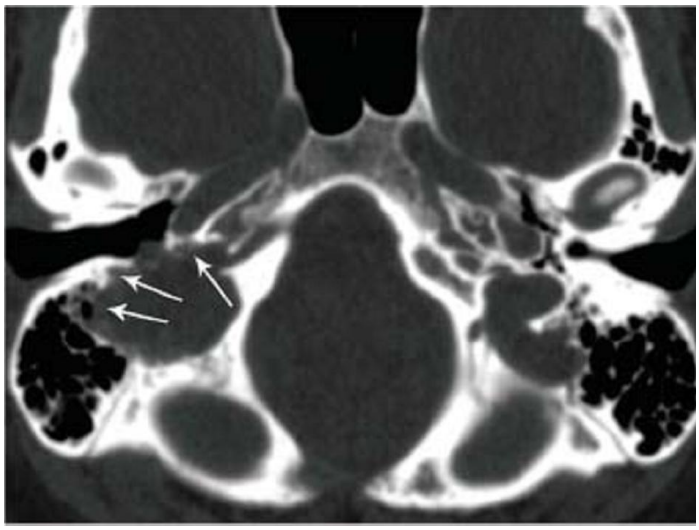
Carotid body PGLs. Carotid body PGLs (CPGLs) are the most common PGLs in the head and neck. These are slowly growing tumors that originate from cells within the carotid body located in the medial aspect of the carotid bifurcation. On contrast-enhanced CT, a CPGL appears as a hypervascular mass located at the carotid bifurcation. Splaying of the internal and external carotid arteries, the "lyre" sign, is a distinguishing feature that suggests the diagnosis of CPGL.9 Small CPGLs show intense enhancement both on CT and MRI. Larger CPGLs show mixed signal and have the typical salt and pepper appearance due to flow voids of intrinsic vessels.5

Vagal PGLs. Vagal PGLs (VPGLs) usually originate from the first 2 cm of the extracranial course of the vagus nerve, commonly at the level of the inferior vagal or ganglion nodosum. Because VPGLs originate in the poststyloid compartment of the parapharyngeal space, these lesions rarely extend to the jugular foramen. VPGLs tend to displace both the internal and external carotid arteries anteriorly and medially, as opposed to the CPGLs that usually widen the carotid bifurcation.

Jugulotympanic PGLs. Glomus tympanicum and glomus jugulare (tympanic and jugular) PGLs, also known as *jugulotympanicPGLs* (JTPGLs), are the second most common tumors involving the temporal bone, and the isolated tympanic PGL (TPGL) is the most common tumor of the middle ear. TPGLs and jugular PGLs (JPGLs) are, by location, closely related to the jugular foramen. They arise from cells of the tympanic branch of the glossopharyngeal nerve (Jacobson's nerve), the auricular branch of the vagus nerve (Arnold's nerve), and from the intravagal paraganglia inferior to the jugular foramen. TPGLs include those in the tympanic cavity and the mastoid. JPGLs include those involving the jugular bulb and the jugular fossa.

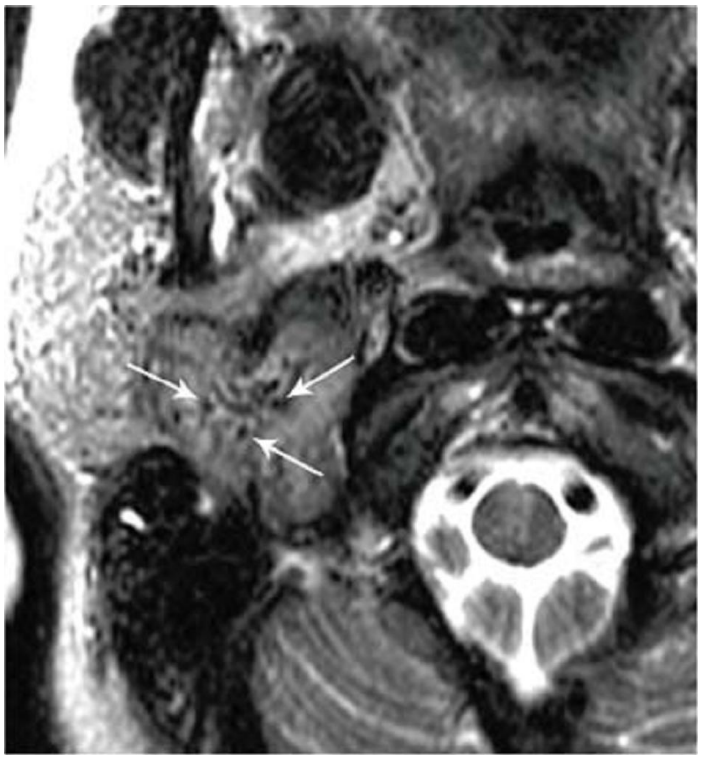
On imaging studies, localized bone destruction at the jugular foramen is a common finding. There also may be enlargement of the foramen. PGLs in this area tend to grow along the sites of least resistance within the temporal bone (i.e., fissures, air cell tracts, vascular channels, and foramina). CT findings include an irregular, destructive process, also described as a "moth-eaten" appearance (figure 1).

Figure 1. High-resolution axial CT of the temporal bone in a 59-year-old man with a right JTPGL (Fisch type C) shows demineralization of the bony margins and destruction of the fine cortical line, demonstrated along the lateral aspect of the vascular portion of the jugular foramen on the right side (arrows); this is known as the *moth-eaten* pattern. Compare with the normal, opposite side.



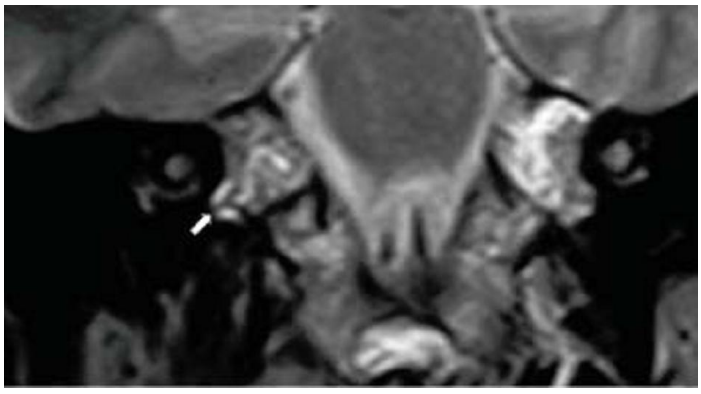
Given the proximity of the hypotympanum and the jugular fossa, a large percentage of JTPGLs involve both regions (figure 2). Oldring and Fisch described five types of JTPGLs:10

Figure 2. Axial T2-weighted MRI in a 59-year-old man with a right JTPGL (Fisch type C) demonstrates a destructive lesion in the right skull base. The lesion is heterogeneously hyperintense with punctate central flow voids (arrows), representing the hypervascular nature of this tumor. The lesion has the classic "salt and pepper" appearance of a PGL.



- Type A: tumors limited to the middle ear cavity (figure 3)
- Type B: tumors limited to the tympano-mastoid area
- Type C: tumors involving the infralabyrinthine area
- Type D1: tumors with intracranial extension less that 2 cm (figure 4)
- Type D2: tumors with intracranial extension greater than 2 cm^2

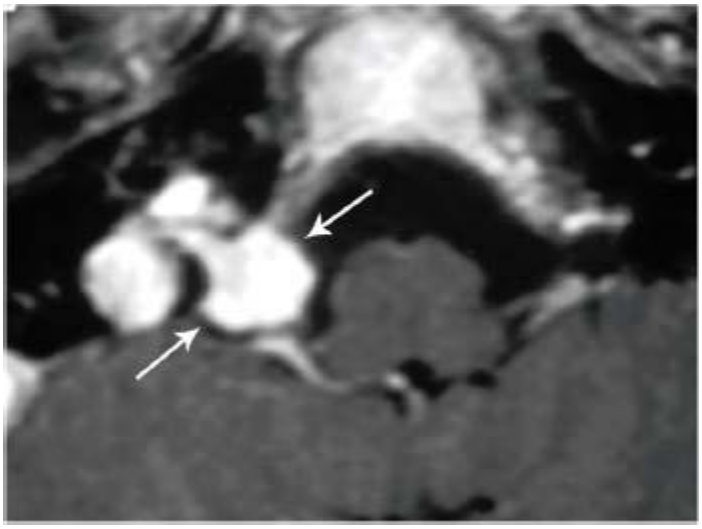
Figure 3. T2-weighted coronal MRI in a 62-year-old woman with a right tympanic PGL (Fisch type A) shows the hyperintense right tympanic PGL lateral to the cochlea (arrow).



Glomus tympanicum typically appears as an enhancing soft-tissue mass adjacent to the promontorium. The mass can grow and fill the typmpanic cavity, involving without destroying the middle ear ossicles.11

Figure 4. Axial T1-weighted MRI with contrast in a 58-year-old man with a right JPGL (Fisch type D1) shows the lesion's intracranial extension of less than 2

cm (arrows).



Treatment of PGLs

Surgical resection remains the treatment of choice. With PGLs at the skull base, such as those involving the jugulotympanic area and extending into the cerebellopontine angle, preoperative embolization is performed to facilitate resection. In selected cases, and particularly with recurrence, radiation therapy is used.

In conclusion, PGLs are important tumors of the head and neck. Recognition of their anatomic landmarks and imaging manifestations allows for correct diagnosis and characterization.

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From CediMed, Antioquia, Colombia (Dr. Zuluaga); the Department of Radiology, The University of Texas in Houston (Dr. Ocazionez); the Department of Radiology, The University of Texas Medical Branch, Galveston (Dr. Riascos); the Department of Radiology, The University of Texas Health Science Center at San Antonio (Dr. Restrepo); and the Department of Radiology, Tulane University Hospital and Clinics, New Orleans (Dr. Palacios).
Corresponding author: Roy Riascos, MD, The University of Texas Medical Branch, 301 University Blvd., Galveston, TX 77555.
Email: riascosr@gmail.com

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