Paragangliomas of the head and neck: imaging findings

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Learning objectives

To describe characteristic features of head and neck paragangliomas, for a better understanding of their imaging appearance, location and post diagnostic management.

Make an overview of their imaging appearance by different imaging techniques for a correct diagnosis and treatment plan.

Background

Head and neck paragangliomas (HNP) are neuroendocrine tumors of the autonomic nervous system. They arise from chromaffin cells of the parasympathetic paraganglia (chemoreceptors), that can be found in numerous locations along the skull base and neck. According to their location, HNP are divided into four common types [Fig. 1 on page 4]:

- carotid body paraganglioma, located at the common carotid artery (CCA) bifurcation
- vagal paraganglioma, located along the vagus nerve
- jugular glomus paraganglioma, located in the jugular foramen (if it extends cranially into the middle ear cavity it is called jugulo-tympanic)
- tympanic paraganglioma, located within the middle ear

Less common sites include the sella turcica, pineal gland, cavernous sinus, larynx, orbit, thyroid gland, nasopharynx, mandible, soft palate, face, and cheek.

HNP are rare tumors, they represent only 0,6% of head and neck tumors and 3% of all paragangliomas. They have no sex predilection, and are mostly found in patients in their 40-60th decade of life.

HNP are slow growing (0,8 mm/year), hypervascular tumors, that very rarely secrete cathecolamines, in spite of their neuroendocrine origin (only 3%). They can be sporadic or hereditary, the former one has been associated with MEN 2a and 2b, as well as type I neurofibromatosis, Von Hippel Lindau disease and Carney-Stratakis dyad. Some studies describe a connection between the sporadic type and hypoxia, based on it's frequency amongst people who live at high altitudes and people with COPD.

HNP can be solitary or multicentric, the former one is more common in the hereditary type and can occur either synchronously or metachronously.
Most HNP are benign, malignant behavior has been described in only 2%-13% of cases. The prevalence of malignancy depends on the site of the primary tumor. Metastasis are most frequent in vagal paragangliomas, both carotid body and jugulotympanic paragangliomas have a lower incidence of malignancy. Metastatic disease is considered the only reliable indicator of malignancy, and it may be observed in the lungs, cervical lymph nodes, liver, and bones. There are no strict histologic criteria within the primary tumor to differentiate benign from malignant paragangliomas. Paragangliomas are highly vascular and characteristically associated with early involvement of blood vessels (carotid artery, jugular bulb) and neural areas (vagus, tympanic plexus), in addition to skull-base and potential intracranial extension.

HNPs are space-occupying tumors consequently the signs and symptoms are induced by the mass effect on the underlying and close anatomic structures, thus there is a variation of the clinical symptoms depending on the tumor's localization:

1. **Carotid body paragangliomas (CBPs)** arise from paraganglia located within the covering of the carotid artery in the neck. 60-70% of CBTs often remain clinically silent before presenting as a painless, slowly enlarging mass in the lateral neck. The initial symptoms can also be represented by a pulsating mass in the neck that is often associated with a bruit. CBTs can be moved horizontally rather than vertically, a finding known as a positive Fontaine’s sign. Large CBTs may induce dysfunction of the vagal nerve and, less frequently, of cranial nerves IX, XI, and XII. Occasionally, in extremely large tumors, Horner’s syndrome or deficits of the facial nerve may result.

2. **Vagal paragangliomas (VPs)** are located more cephalad in the neck (compared to the carotid body tumors), between the jugular vein and the internal carotid artery, sometimes extending to the base of the skull through the jugular foramen or posterior to the mastoid tip. The symptoms of a VP depend on the location of the tumor along the vagal nerve from the skull base to the lower neck. Most VPs arise from the glomus nodosum, that is, the inferior ganglion. Typically, VPs present with an asymptomatic neck mass behind the angle of the mandible. Other symptoms of VPs are pulsatile tinnitus or ringing in the ear heard with each heartbeat. Less than 50% of VPs present with deficits of cranial nerves, which manifest as hoarseness (X), dysphagia (IX), shoulder drop (XI), nasal reflux of fluids, aspiration and hemiatrophy of the tongue (XII). Intracranial extension, which is the main cause of death, occurs in 22% of the cases. Oropharyngeal involvement caused by bulging of the pharyngeal wall into the pharyngeal lumen and medial displacement of the tonsil, may occasionally be observed.

3. **Jugular paragangliomas (JPs)** or glomus jugular tumors originate from paraganglia found in or around the jugular bulb. When these tumors increase in size, occlusion of venous flow can be observed. Symptoms are often represented by pulsatile tinnitus. Conductive hearing loss is seen with progression of the tumor, which can cause impairment of vibration of the
ossicles or invades the bones behind the eardrum. Dizziness is reported by patients when the tumor has invaded the inner ear (due to sensorineural hearing loss). Occasionally, JPs can cause deficits of other cranial nerves and create dysfunctional swallowing and huskiness of the voice. However, due to the relatively slow growth, the swallowing mechanism and vocal cord function of the opposite side may initially compensate and mask the disease symptoms. When these tumors grow, they can also invade the facial nerve leading to facial paralysis, or they can encompass the hypoglossal nerve, leading to paralysis of half of the tongue. Further growth can lead to compression of the brain and/or brainstem.

4. **Tympanic paragangliomas (TPs)** are mostly small-sized tumors originating in the middle ear, in the paraganglia found in the cochlear promontory's mucosa. These tumors become symptomatic as pulsatile tinnitus in the vast majority of patients. Hearing loss is initially present in about half of patients.

Diagnostic imaging can be considered in two clinical situations: patients who present with clinical symptoms suggestive of a paraganglioma, and in individuals from families with hereditary paragangliomas.

The diagnosis of HNPs is based on the clinical manifestations and imaging appearance.

The differential diagnosis has to be made with other masses that may arise in the carotid space of the suprahyoid and infrahyoid neck. The most common considerations are nerve sheath tumors (shwanomas, neurofibromas), nodal metastasis, abscesses, venous thrombosis and ICA aneurysm. There are rarer possibilities such as lipoma, liposarcoma and salivary gland tumors.

Surgical resection is the main treatment method of HNPs, the outcome varies on many factors that may influence the ideal result of total tumor removal and minimal postoperative complications. If surgery is the chosen course of treatment, preoperative embolization can be performed. Another treatment method is represented by stereotactic surgery. In patients with unresectable tumors, residual tumor following surgery, or tumor involvement that occludes the ICA, radiation therapy may serve as an excellent palliative modality. Patients who refuse surgery or those who are not suitable surgical candidates can also be offered radiation therapy. In bilateral vagal paragangliomas, bilateral resection is not an option because it usually entails bilateral vagal nerve paralysis with unacceptable morbidity and mortality. In these cases, adjunctive radiation therapy of at least one lesion with surgical extirpation of the other is recommended. The management decisions of HNPs are based on imaging studies.

Images for this section:
**Fig. 1:** The drawing represents the most common locations of paraganglia of the head and neck. The carotid body paraganglion is a discrete mass found at the CCA bifurcation. Other locations can be visualized along the path of the vagus nerve, the jugular foramen, and the middle ear. The paraganglia tend to occur along the path of either vessels or nerves.

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Findings and procedure details

Imaging techniques along with the clinical presentation represent the diagnostic methods for HNP and the grounds for an adequate treatment plan. Imaging can also determine the benign or malignant nature of a paraganglioma, as well as identify if the lesion is single or multicentric.

HNPs produce characteristic findings on different diagnostic imaging techniques.

The main imaging techniques of HNPs are represented by ultrasound, CT, MRI, CT and MRI angiography and DSA (preoperative embolisation).

Which study is performed first or whether a second imaging study is required depends on the availability of the specific imaging modality at a particular institution and the preferences of the referring physician and the radiologist.

Carotid body paragangliomas (CBPs):

B-mode sonography and color duplex sonography often represent the first imaging choice in CBP, mostly because they are inexpensive, noninvasive, and based on non ionizing radiation.

A CBP is usually sonographically seen as a solid, well-defined, hypoechoic mass with splaying of the carotid bifurcation (distinctive imaging feature also visible on CT, MRI and angiography): the external carotid artery (ECA) being displaced anteromedially, and the internal carotid artery (ICA) posterolaterally.[Fig. 2 on page 11] In most cases, CBPs show intense, increased color Doppler signal.[Fig. 3 on page 11]

The ultrasound exam, needs to be followed and completed with a CT or MRI exam.

Computed tomography (CT) is an excellent imaging technique that can effectively diagnose a CBP, document the extent of the tumor, and the possible bone invasion. It has a better spatial resolution and less movement artifacts. The typical CT appearance of a carotid body tumor is a welldefined soft-tissue mass located within the carotid space of the infrahyoid neck. Most characteristic feature is splaying of the internal and external carotid arteries. Because of their hypervascularity, CBPs exhibit early, homogeneous and intense enhancement after injection of contrast material. Large tumors are frequently inhomogeneous, with areas of necrosis and hemorrhage. [Fig. 4 on page 12 ; Fig. 5 on page 13 ; Fig. 6 on page 14 ; Fig. 7 on page 15]

MRI provides a superior definition of location, extent, and characterization of carotid body paragangliomas (CBPs). It can make a better assessment about the involvement of the
ICA and IJV compared to CT. It is also worth noting that MRI is more effective than CT in identifying small synchronous paragangliomas, especially those smaller than 5 mm, whereas CT demonstrates only lesions greater than 8 mm.

On MRI, CBPs typically have a low to intermediate signal intensity on T1-weighted MR images and high signal intensity on T2-weighted MR images. The most characteristic MR finding of paragangliomas is the "salt-and-pepper" appearance where the pepper is represented by presence of multiple serpentine and punctate areas of signal void within the tumor matrix, due to high-velocity flow of the intratumoral vessels (on both T1- and T2-weighted images). The salt component represented by high-signal regions is a consequence of slow flow or hemorrhage. The enhancement is pattern is early, homogeneous and intense [Fig. 8 on page 17, Fig. 9 on page 19].

Magnetic resonance angiography (MRA) provides excellent visualization of the major head and neck vasculature and can demonstrate vessel displacement, gross tumor involvement, and possible compromised blood flow. It may be useful in defining flow-related enhancement of lesions greater than 1.5 cm. While three-dimensional time-of-flight angiography (TOF) appears superior to other MRA techniques in identifying some tumor feeders, its sensitivity is not high enough to demonstrate detailed tumor vascular supply, which is best defined by digital subtraction superselective angiography.

Angiography plays an important role in the evaluation of CBPs if surgery is being contemplated. It provides great detail of the vascular anatomy, it delineates the tumor’s blood supply, displacement of vessels, potential vessel compromise by tumor invasion, and it can also assess the intracranial circulation if internal carotid artery sacrifice is necessary [Fig. 10 on page 20]. It can also reveal previously undiagnosed synchronous paragangliomas. Superselective angiography allows safe preoperative embolization of the tumor, hopefully avoiding proximal vessel occlusion or accidental migration of embolization material into the cerebral or systemic circulation.

On DSA, we can notice the splaying of the carotid bifurcation, and an intense tumor blush.

**Vagal paragangliomas (VPs):**

These tumors are usually confined to the parapharyngeal space, but large tumors may extend higher up through the jugular foramen into the posterior fossa or extend down toward the carotid bifurcation. However, in contrast to carotid body tumors, vagal paragangliomas rarely fill the carotid bifurcation. US, CT, MRI and DSA findings of the vagal paraganglioma are similar to those of the carotid body tumor: a well-defined ovoid parapharyngeal mass with intense contrast enhancement and possibly a salt-and-pepper appearance (on MRI). However, vagal paragangliomas, unlike the carotid body tumors, tend to displace both the external carotid artery and internal carotid artery anteromedially, separating these vessels from the IJV and commonly extend into the suprahyoideal neck. [Fig. 11 on page 21] The ascending pharyngeal and occipital arteries commonly
supply this tumor. Less frequently, the lingual, facial, and deep cervical arteries may also provide feeding vessels.

VPs demonstrate the highest rate of malignancy (16%), of all the HNPs. [Fig. 12 on page 21]

Jugular paragangliomas (JPs) or gloms jugular tumor:

For JPs, both MR and CT are recommended. CT-angiography depicts the anatomy more accurately than MR-angiography because of the better spatial resolution. However, contrast-enhanced MR-angiography is better suited for screening and detection of multiple lesions, a better assessment of the surrounding soft tissues. Both cross-sectional methods can be used for operative navigation.

On high-resolution CT scans of the temporal bones and skull base, expansion and erosion of the jugular foramen are characteristic features of JPs [Fig. 13 on page 22, Fig. 14 on page 23]. Early in the disease, the jugular fossa is enlarged and its margins are irregular. Progressive growth of the tumor produces a typical moth-eaten pattern of erosion of the jugular foramen and destruction of the surrounding bony labyrinth. The tumor spreads along the paths of least resistance, thus it is initially directed superiorly, possibly involving the structures of the middle ear (hypotympanum, mesotympanum, sinus tympani and the ossicular chain), in this case the tumor is also called jugulo-tympanic paraganglioma. Inferior spread of the tumor produces infiltration of the IJV and infratemporal fossa. As the tumor spreads laterally, it may destroy the facial nerve canal and infiltrate the facial nerve.

Tympanic paragangliomas (TPs) or gloms tympanum tumor:

CT is superior to the other imaging techniques in the evaluation of TPs,. The glomus tympanicum tumor manifests as a small discrete mass arising from the mucosa of the cochlear promontory and is confined to the tympanic cavity[Fig. 15 on page 24]. Ossicular destruction is not typical, although encasement is frequent in larger lesions. Uncommon patterns of tumor spread include lateral extension into the mastoid air cells and anterior extension into the eustachian canal and nasopharynx.

MRI is rarely used, it can offer a better delineation of the humoral extent.

Images for this section:
Fig. 1: The drawing represents the most common locations of paraganglia of the head and neck. The carotid body paraganglion is a discrete mass found at the CCA bifurcation. Other locations can be visualized along the path of the vagus nerve, the jugular foramen, and the middle ear. The paraganglia tend to occur along the path of either vessels or nerves.

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Fig. 2: A transverse US image of a 56 year old female, at the carotid artery bifurcation shows a hypoechoic mass located at the CCA bifurcation, encasing both the external and internal carotid arteries.

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**Fig. 3:** A transverse color Doppler image, of the same patient, at the level of the carotid bifurcation shows a highly vascularized mass between the internal and external carotid artery.

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Fig. 4: Axial unenhanced [FIG 4], axial enhanced [FIG 5], enhanced coronal reconstruction [FIG 6] and a VRT reconstruction of a 69 year old woman with a carotid body tumor. Where we can see a well delineated, hyperenhancing mass occupying the left carotid space, located between the left internal carotid artery and external carotid artery.

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**Fig. 5:** Axial unenhanced [FIG 4], axial enhanced [FIG 5], enhanced coronal reconstruction [FIG 6] and a VRT reconstruction of a 69 year old woman with a carotid body tumor. Where we can see a well delineated, hyperenhancing mass occupying the left carotid space, located between the left internal carotid artery and external carotid artery.

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Fig. 6: Axial unenhanced [FIG 4], axial enhanced [FIG 5], enhanced coronal reconstruction [FIG 6] and a VRT reconstruction of a 69 year old woman with a carotid body tumor. Where we can see a well delineated, hyperenhancing mass occupying the left carotid space, located between the left internal carotid artery and external carotid artery.

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Fig. 7: Axial unenhanced [FIG 4], axial enhanced [FIG 5], enhanced coronal reconstruction [FIG 6] and a VRT reconstruction [FIG 7] of a 69 year old woman with a carotid body tumor. Where we can see a well deliniated, hyperenhancing mass occupying the left carotid space, located between the left internal carotid artery and external carotid artery.

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Fig. 8: Axial T2-weighted image of a 35 year old male, shows heterogeneous high signal intensity of the tumor (arrow) in left carotid space associated with the characteristic "salt and pepper" appearance.

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Fig. 9: Coronal T1-weighted image of the same patient shows isointense tumor (arrow) at left common carotid bifurcation. Also note the multiple flow voids (arrowheads) within tumor.

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Fig. 10: Lateral angiography image of the same patient. The left common carotid artery shows splaying of internal carotid artery and external carotid artery by hypervascular mass (arrow).

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**Fig. 11:** Sagittal reconstructed CT image of a patient with vagal paraganglioma, where we can see a well-defined ovoid parapharyngeal mass with intense contrast enhancement, located above the carotidian bifurcation.

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**Fig. 12:** Axial thoracic CT image, of the previous patient, in lung window depicts multiple pulmonary metastases.

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Fig. 13: Axial CT image of a 59 year patient, in parenchymal [FIG 13 ]and bone windows [FIG 14], where we can notice a glomus jugular tumor, associated with the expansion and erosion of the left jugular foramen.

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**Fig. 14:** Axial CT images of a 59 year patient, in parenchymal [FIG 13] and bone windows [FIG 14], where we can notice a glomus jugular tumor, associated with the expansion and erosion of the left jugular foramen.

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Fig. 15: Axial thin-section CT scan of temporal bone in a 51 year old patient in bone window shows a 5 mm soft-tissue mass (arrow), adjacent to the cochlear promontory, filling the hypotympanum of the left middle ear cavity.

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Conclusion

Paragangliomas are uncommon lesions of the head and neck region. They are usually seen in four locations: the carotid body tumor at the CCA bifurcation, the gloms jugular or vagal paraganglioma at the jugular foramen and the gloms tympanic tumor in the middle ear. HNPs usually have similar clinical manifestations as nontender, slowly enlarging, soft-tissue lesions of the neck and skull base. Because of their location, cranial nerve paralysis are a common symptom, particularly for those lesions arising near the skull base. Imaging techniques play an important role in the diagnosis and management of these tumors. CT and MR are considered "golden standard". HNPs typically have a salt-and-pepper appearance with prominent flow-voids on MR imaging. Intense enhancement is almost always noted following contrast material administration both on MRI and CT. Jugular and tympanic paragangliomas are better evaluated by CT scans. Angiography is required preoperatively in larger paragangliomas for surgical planning and often for preoperative embolization. Most paragangliomas are benign. The prognosis is directly related to the location of the tumor: Patients with paragangliomas arising at the carotid body have the best outcome, whereas those with skull base tumors have a less favorable prognosis because of the increased difficulty in achieving total resection.

Personal information

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