Carotid body paraganglioma, diagnosis imaging

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Learning objectives

The purpose of this poster will be: Review the imaging techniques that are used for patients with suspected paragangliomas. Describe the imaging hallmarks of paragangliomas to confirm this diagnosis and illustrate the importance of the imaging studies to predict operative morbidity and mortality, because imaging depict the location and extent of tumor involvement and help determine the surgical approach.

Background

Numerous terms have been used to describe these tumors: glomus tumor, chemodectoma or paraganglioma.

Paragangliomas are very slow-growing benign neoplasms of the head and neck region arising from sympathetic or parasympathetic nerve tissue.

The carotid body paraganglioma or carotid body tumor, which originates from paraganglionic cells within the carotid body, is the most common paraganglioma of the head and neck.

The carotid body is situated within or outside the adventitial layer of common carotid artery at the bifurcation level. The carotid body is a chemoreceptor organ which detects changes in arterial partial pressures of oxygen and carbon dioxide, pH, and other factors.

A higher prevalence of carotid body paragangliomas has been noted in some patients with chronic obstructive pulmonary disease and in certain populations living at high altitudes; this is believed to be secondary to chronic hypoxia in combination with genetic factors.

The carotid body tumor can occur at any age, with a peak of incidence in the 45-50 year-old age group. It is uncommon in the pediatric population.

Familial paragangliomas have an overall prevalence of 7%, with approximately 90% of cases arising from the carotid body, and a higher prevalence in younger patients with an average age of 38 years.

Bilateral carotid body tumors occur in 5% of sporadic and 30% of familial paragangliomas.
The typical clinical presentation of a carotid body tumor is a painless, slowly growing lateral neck mass.

**Imaging findings OR Procedure details**

**Ultrasonographic**

A preliminary Ultrasonographic (US) study of the lateral neck is performed (5-MHz) for tumor localization, followed by a high-resolution study (7.5-10-MHz) for tumor characterization.

The characteristic appearance of a carotid body tumor on gray-scale US scans is a round-to-oval, well-defined, heterogeneously hypoechoic solid mass in the lateral neck with splaying of the common carotid bifurcation (Fig. 1 on page 7).

Duplex Doppler imaging and color Doppler imaging, demonstrate the intrinsic hypervascularity of the cervical paraganglioma (Fig. 2 on page 8) (Fig. 3 on page 9).

The differential diagnosis for a carotid body tumor on gray-scale US scans includes salivary gland tumor, lymphadenopathy, carotid artery pseudoaneurysm, branchial cleft cyst, and nerve sheath tumor. Use of duplex and color Doppler techniques can facilitate the diagnosis by demonstrating the hypovascular nature of all of these masses.

**CT**

On CT scans, the carotid body tumor manifests as a well-defined soft-tissue mass within the carotid space of the infrahyoid neck.

Paragangliomas, including carotid body tumors, typically show homogeneous and intense enhancement after intravenous administration of contrast material because of their hypervascularity. (Fig. 4 on page 9). However, large tumors are frequently heterogeneous, with areas of both focal thrombi and hemorrhage.

Splaying of the internal and external carotid arteries is a distinctive imaging feature on CT, MR and angiography, which is highly suggestive of a carotid body tumor. (Fig. 5 on page 10) (Fig. 6 on page 11)
Approximately 8% of carotid body tumors extend into the suprahypoid neck and present clinically as a parapharyngeal space mass (Fig. 7 on page 12).

**MR**

MR imaging has proved to be superior to other imaging techniques to provide definition of location, extent, and characterization of paragangliomas.

Accurate assessment of tumor margins and invasion of adjacent structures are also essential for proper staging and therapy. For this task, MR imaging using unenhanced and enhanced fat-suppressed spin-echo (SE) sequences is widely accepted as the method of choice, in addition to high-resolution CT. MR imaging also better demonstrates tumor involvement of the internal carotid artery and internal jugular vein compared with that seen with CT. MR imaging can also depict paragangliomas that are smaller than 5 mm, whereas CT demonstrates only lesions greater than 8 mm.

On MRI, they typically have a low to intermediate signal intensity on T1-weighted MR images and high signal intensity on T2-weighted MR images. (Fig. 8 on page 13) (Fig. 9 on page 14) (Fig. 10 on page 15) (Fig. 11 on page 16)

The most characteristic MR finding of paragangliomas is the presence of multiple serpentine and punctate areas of signal void within the tumor matrix. These areas of signal void are believed to be caused by high velocity flow of the intratumoral vessels. The adjacent high- and low-intensity regions of the tumor have been described as having a "salt-and-pepper" appearance. (Fig. 12 on page 17). The salt component representing high-signal regions is because of slow flow or hemorrhage and the pepper component corresponds to the multiple signal voids of tumor vessels on both T1- and T2-weighted images. This feature is limited to paragangliomas that are greater than 1 cm in diameter and is not considered diagnostic, as it has also been reported in other hypervascular lesions (metastatic hypernephroma, metastatic thyroid carcinoma).

As with CT, a homogeneous and intense pattern of enhancement is noted following the intravenous administration of contrast material. (Fig. 13 on page 18) (Fig. 14 on page 19).

Paragangliomas have a typical temporal contrast enhancement, this characteristic contrast enhancement can be useful in the diagnostic work-up of lesions that may mimic paraganglioma.
In combination with conventional MR imaging (unenhanced and enhanced spin-echo imaging), Contrast-Enhanced-MRAngiography (CE-MRA) yields an excellent diagnostic value for the assessment of head and neck paragangliomas. (Fig. 15 on page 20) (Fig. 16 on page 22) (Fig. 17 on page 23)

**Angiography**

Once it has been decided that an operation is indicated, detailed information about the vascular supply of the tumors is required.

Digital subtraction angiography (DSA) is the imaging reference standard for assessing the vascular architecture of the tumor and it is required preoperatively in larger paragangliomas for surgical planning and often for preoperative embolization.

DSA should be performed in all patients with selective injection of the common carotid, external carotid, and vertebral arteries.

The typical angiographic appearance of a paraganglioma is that of a hypervascular mass with enlarged feeding arteries, intense tumor blush, and early draining veins.

Carotid body tumors typically cause splaying of the external carotid artery and internal carotid artery. (Fig. 18 on page 25) (Fig. 19 on page 27) (Fig. 20 on page 29)

The most common feeding vessels to any head and neck paraganglioma are the ascending pharyngeal artery (via the musculospinal artery) and the ascending cervical artery. With progressive tumor growth, other sources of arterial supply may be recruited from the facial, lingual, thyroid, posterior auricular, occipital, and deep cervical arteries.

Digital subtraction angiography demonstrates the vascular supply (feeding vessels and collateral supply) of a paraganglioma, the relationship of the mass to the internal carotid artery and internal jugular vein (IJV), and the patency of the IJV (which is frequently thrombosed in larger paragangliomas).

This method is invasive with a complication rate of 0.5%-1% which could be too high for a diagnostic examination. Consequently, some investigators have discussed the utility of more specific methods in MR imaging to differentiate paragangliomas from other tumors or vascular abnormalities; these investigators say that in the future, Contrast Enhanced MR Dynamic Angiography has the potential to replace DSA as the imaging reference standard for the diagnosis of paraganglioma.
DIFFERENTIAL DIAGNOSIS

- **Masses in the Carotid Space of the Neck**: These include nerve sheath tumors, nodal metastasis, abscess, and venous thrombosis as the most common considerations.

- **Nerve sheath tumors**: Displace the carotid arteries anteromedially and the IJV posteriorly. On nonenhanced CT scans, they exhibit homogeneous attenuation when small, whereas larger tumors may be heterogeneous with areas of cystic degeneration. At MR imaging, schwannomas appear isointense relative to soft tissue with short TR/short TE sequences and hyperintense with long TR/long TE sequences. On both contrast-enhanced CT and MR images, they enhance intensely and homogeneously, secondary to the pooling of contrast material in the interstitial spaces.

- **Metastases**: Metastases from renal and thyroid malignancies may closely mimic paragangliomas in their CT and MR imaging appearances. However, these uncommon lesions infiltrate more into the surrounding soft tissues and do not follow the typical routes of spreading of paragangliomas.

- **The vagal paraganglioma** appears similar to the carotid body tumor with some exceptions. These masses displace both the ECA and ICA anteromedially, separating these vessels from the IJV. In addition, extension into the suprahypoid carotid space is seen in approximately two-thirds of vagal paragangliomas.

THERAPY

**Radiation Therapy**

In patients with unresectable tumors, multiple tumors, residual tumor following surgery, or tumor involvement that occludes the internal carotid artery, 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 as a means of palliation.

**Surgical Management**

Surgery, which is the curative method of treatment for carotid body paragangliomas, requires complete excision of the tumor, with preservation of vital neurovascular structures.
The main factors influencing surgical treatment of carotid body paragangliomas, apart from age and operative risk, are multifocality and the possibility of impairment of cranial nerves and injury to internal carotid artery.

**Preoperative embolization** has been acclaimed by many investigators as a useful adjunctive tool in the surgical management of paragangliomas.

To completely embolize a paraganglioma, all the feeding vessels must be occluded. Most arteries can be embolized by using polyvinyl alcohol particles. The decrease in tumor size is estimated at about 80%.

Complications of preoperative embolization include postembolization fever and ear pain. They are attributed to tumor ischemia, are transient, and suggest successful treatment. Major complications include cerebral ischemia and cranial nerve palsy. Other major risks of preoperative embolization include transient aphasia, carotid sinus syndrome, and the sequela of catecholamine secretion.

**Images for this section:**
Fig. 1: Gray-scale US scans: Heterogeneously hypoechoic solid mass in the lateral neck with splaying of the common carotid bifurcation.
Fig. 2: Color Doppler imaging demonstrates the intrinsic hypervascularity

Fig. 3
Fig. 4: Contrast-enhanced axial CT image demonstrates an intensely enhancing right carotid space mass.
Fig. 5: Contrast-enhanced coronal CT image demonstrates an intensely enhancing right carotid space mass that splays the ECA from the ICA.
Fig. 6: Contrast-enhanced axial CT image demonstrates a carotid space mass that splays the external carotid artery (ECA) from the internal carotid artery (ICA).
Fig. 7: Contrast-enhanced sagital CT image of the suprhyoid neck shows the mass extending superiorly within the carotid space.
Fig. 8: Axial T1-weighted TSE MR image shows a left-sided neck mass that is isointense relative to muscle at the level of the common carotid bifurcation.
Fig. 9: Axial T1-weighted TSE MR image shows the mass that is isointense relative to muscle. The ECA is splayed from the ICA.
Fig. 10: Axial T2-STIR shows intensely hyperintense well-circumscribed left-sided cervical mass
Fig. 11: Axial T2-STIR shows intensely hyperintense mass
**Fig. 12:** Axial T2-weighted image shows heterogeneous high signal intensity of tumor in left carotid space. Multiple flow voids combined with hyper intense areas demonstrate the "Salt-and-pepper" appearance in tumor.
Fig. 13: Axial T1-weighted TSE images after gadolinium injection, showing a left-sided carotid body tumor splaying the ICA and ECA.
**Fig. 14:** Coronal T1-weighted TSE image after intravenous administration of contrast material shows an intense pattern of enhancement.
Fig. 15: Splaying of the carotid bifurcation can be noticed on the coronal view unenhanced 3D time-of-flight MR angiogram

Fig. 16
Fig. 18: Lateral angiogram obtained after a right CCA injection reveals splaying of the ECA from the ICA by a hypervascular mass that extends to the bifurcation.
Conclusion

- The carotid body tumor, the most common paraganglioma of the head and neck, is a highly vascular neoplasm originating from paraganglionic tissue located at the carotid bifurcation.

- Paraganglioma-associated morbidity occurs because of local tumor growth compromising neural and vascular structures in the neck. Such morbidity may be an indication for treatment, which includes surgical resection with or without preceding embolization.

- Diagnostic radiology plays an important role in the diagnosis and surgical planning.

- Meticulous preoperative planning and careful patient selection are essential for a successful therapeutical outcome. First, the tumor has to be detected and characterized, and because paragangliomas are multiple in 30% of patients, the presence of concomitant tumors should also be studied. In addition, tumor extension has to be determined, especially the relation of the tumor to the surrounding vascular structures.

- Imaging hallmarks of paragangliomas of the head and neck include an enhancing soft-tissue mass in the carotid space, salt-and-pepper appearance at standard spin-echo magnetic resonance imaging; and an intense blush at angiography.

- Radiation treatment is included as a palliative adjunct for the exceptional paraganglioma not suitable for surgery.

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References


