Imaging of head and neck paragangliomas

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Learning objectives

1. To describe and illustrate presentation of head and neck paragangliomas on B-mode sonography and color-coded Doppler sonography, computed tomography, magnetic resonance imaging and digital subtraction angiography.
2. To identify typical imaging appearances of carotid body, glomus jugulare, glomus tympanicum and glomus vagale paraganglioma, based on their characteristic locations and imaging features
3. To describe the possibilities of each diagnostic method

Background

Paragangliomas account for 0.6% of all neoplasmas of the head and neck and 0.03% of all neoplasmas.

Head and neck paragangliomas (also commonly referred to as glomus tumors or chemodectoma) are highly vascular lesions originating from paraganglionic tissue, located at four typical locations: the carotid bifurcation (carotid body tumors), along the vagus nerve (vagal paragangliomas), and in the jugular fossa and tympanic cavity (jugular and tympanic paragangliomas). These lesions are histologically similar to the pheochromocytomas that may develop in the adrenal medulla, but unlike pheochromocytomas, head and neck paragangliomas rarely secrete catecholamines.

In most cases they are benign neoplasms; overall less than 10% of all paragangliomas have been cited to be malignant.
**Clinical presentation:**

The clinical symptoms vary according to size and location of the paraganglioma. Cervical paragangliomas generally present in mid-adult life as asymptomatic, nonfunctional lateral neck masses.

1. **Carotid body paraganglioma** represents the most common form of cervical paraganglioma that accounts for 60%. A carotid body tumor often presents as a painless, slowly enlarging mass in the lateral neck. Physical examination of a patient with a carotid body tumor typically reveals a rubbery, nontender mass along the anterior border of the sternocleidomastoid muscle. The lesion is more freely movable horizontally than vertically ("Fontaine's sign") because of adherence to the carotid artery. The finding of a carotid bruit or a pulsatile character of the tumor strengthens the tentative diagnosis of a carotid body tumor. A careful neurological examination for deficits of the cranial nerves VII, IX, X, XI and XII as well as the sympathetic chain is mandatory.

Shamblin et al. classificated carotid body tumors according to imaging into 3 classes: (Table1):

<table>
<thead>
<tr>
<th></th>
<th>Classification for carotid body tumors by Shamblin</th>
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<tbody>
<tr>
<td>I</td>
<td>Localized tumor</td>
</tr>
<tr>
<td>II</td>
<td>Adherent to and partially surrounding carotid arteries</td>
</tr>
<tr>
<td>III</td>
<td>Large tumor adherent to and totally enveloping carotid arteries</td>
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</tbody>
</table>

2. **Vagal paraganglioma:** A painless neck mass is also the most frequent symptom in patients with vagal paragangliomas. Those tumors most commonly arise from the inferior (nodose) ganglion of the vagus nerve, but it has been demonstrated that they might actually arise at any point along the course of the vagus nerve, most commonly located behind the angle of the mandible. They are most frequently associated with progressive dysphagia and hoarseness with increasing tumor size. Horner’s syndrome (ptosis, myosis, anhidrosis, enophtalmos) may result from cervical sympathetic chain invasion, and syncope can occur after carotid sinus compression. The more superiorly extending vagal paragangliomas can also cause the jugular foramen syndrome by compressing the cranial nerves. Vagal paragangliomas can even extend into the posterior fossa through the jugular foramen, causing the same symptoms as a jugular paraganglioma.
3. **Jugular paraganglioma** originates in the jugular bulb near the skull base and extends both inferiorly in the parapharyngeal space and intracranially. It is not usually accompanied by a palpable neck mass.

The most common symptoms in patients with jugular paragangliomas are pulsatile tinnitus and hearing loss. Audiologic examination reveals significant conductive hearing loss, or mixed conductive and sensorineural hearing loss. Involvement of the inner ear produces vertigo and sensorineural hearing loss. Less common aural signs and symptoms included fullness, bleeding, pain, otorrhea, bruit. Otoscopic examination reveals a characteristic pulsatile, reddish-blue tumor behind the tympanic membrane (Fig 1).

Cranial nerve deficits in jugular paragangliomas usually present with a Vernet i.e. jugular foramen syndrome which includes paralysis of cranial nerves IX-XI. This syndrome is pathognomonic for glomus jugulare and includes: hoarseness due to vocal cord paralysis (X), difficulty in swallowing, (IX), and weakness and atrophy of the trapezius and sternocleidomastoid muscles (XI). Collet Sicard syndrome is present in larger tumors with the XIIth cranial nerve involvement. Collet Sicard syndrome includes Vernet syndrome with involvement of the XII cranial nerve) causing ipsilateral atrophy of the tongue. Less commonly, larger glomus tumors produce facial nerve palsy (due to VII cranial nerve involvement) (Fig.2), or Horner syndrome.

4. **Tympanic paraganglioma** is usually relatively small at diagnosis, since symptoms are produced at an early stage. As they originate from the middle-ear cavity they usually become symptomatic as a pulsatile tinnitus that might be accompanied by a conductive hearing loss.

The two established classifications of these tumors are based on imaging and depend mainly on tumor size with special emphasis on intracranial extension as a decisive factor for resectability. Paragangliomas of the temporal bone are usually staged according to the classification by Fisch (Table 2) and Glasscock and Jackson (Table 3). All these classifications distinguish a group of complex tumors that present challenges to treatment and are associated with adverse outcomes.

Table 2. *Fisch classification of paragangliomas of the temporal bone*

<table>
<thead>
<tr>
<th></th>
<th>Description</th>
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<tbody>
<tr>
<td>A</td>
<td>Tumors arising along the tympanic plexus on the promontory; confined to the middle ear</td>
</tr>
<tr>
<td>B</td>
<td>Tumors arise in the canalis tympanicus and invade the hypotympanum and mastoid</td>
</tr>
<tr>
<td>C</td>
<td>Tumors originate in the dome of the jugular bulb and invade the petrous bone and pyramid; subgrouping C1-4 is based on the</td>
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degree of erosion of carotid canal from the carotid foramen to the cavernous sinus

<table>
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<tr>
<th>D</th>
<th>Tumors with intracranial extension (posterior fossa); subdivided according to depth of invasion</th>
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<tbody>
<tr>
<td>De:</td>
<td><em>extradural</em></td>
</tr>
<tr>
<td>De1</td>
<td>- displacement of the dura less than 2 cm</td>
</tr>
<tr>
<td>De2</td>
<td>- displacement of the dura more than 2 cm</td>
</tr>
<tr>
<td>Di:</td>
<td><em>intradural</em></td>
</tr>
<tr>
<td>Di1</td>
<td>- intradural invasion less than 2 cm</td>
</tr>
<tr>
<td>Di2</td>
<td>- intradural invasion more than 2 cm</td>
</tr>
<tr>
<td>Di3</td>
<td>- inoperable</td>
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Table 3. **Glasscock-Jackson classification of Glomus Jugulotympanicum Tumors**

**Glomus Tympanicum**

| 1       | Small mass is limited to the cochlear promontory                                            |
| 2       | Mass completely fills the tympanic cavity                                                  |
| 3       | Mass fills the tympanic cavity and the mastoid                                            |
| 4       | Mass fills the tympanic cavity, extending into the mastoid, the external auditory canal, or anterior to the carotid |

**Glomus Jugulare**

<p>| 1       | Mass is confined to the jugular bulb, middle ear and mastoid                               |</p>
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<tbody>
<tr>
<td>2</td>
<td>Mass extends below to the internal auditory canal (with or without intracranial extension)</td>
</tr>
<tr>
<td>3</td>
<td>Mass involves the petrous apex (with or without intracranial extension)</td>
</tr>
<tr>
<td>4</td>
<td>Mass extends into the infratemporal fossa and clivus with intracranial extension</td>
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**II Therapy**

The different therapeutic options for paragangliomas include surgical extirpation, conventional radiotherapy and stereotactic radiosurgery. Surgery is the treatment of choice for most patients with head and neck paragangliomas (Fig 3,4.). Whenever surgical extirpation is too risky, conventional radiotherapy and stereotactic radiosurgery may be considered as alternative treatment options. Boedecker and al. support preoperative embolization in patients with glomus jugulare paragangliomas, class-II and III, carotid body tumors and vagal paragangliomas with a diameter larger than 3 cm. In selected cases a "wait and scan" policy or permanent embolization may be discussed.

**Images for this section:**
Fig. 1: Otoscopic finding of patient with glomus jugulare paraganglioma presented by reddish-blue pulsating mass behind the tympanic membrane
Fig. 2: Clinical aspect of patient with glomus jugulare paraganglioma presented by facial nerve palsy on the left side (with patient’s consent).
Fig. 3: Intraoperative finding of glomus jugulare paraganglioma
Fig. 4: Pathohystological finding of glomus jugulare paraganglioma
Imaging findings OR Procedure details

Evaluation by an imaging procedure is absolutely necessary to establish the

diagnosis of a head and neck paragangliomas and for treatment planning. Diagnostic
imaging can be considered in two clinical situations:

- patients who present with clinical symptoms suggestive of a paraganglioma, and
- individuals from families with hereditary paragangliomas. Patients with a positive family
  history are at a higher risk of having multicentric disease.

1. **B-mode sonography in combination with color-coded Doppler sonography** is an
   inexpensive, non-invasive, readily available diagnostic tool that is frequently used as
   first-line diagnostic procedure for cervical paragangliomas. The role of color Doppler
   ultrasound in the diagnostic work-up of paragangliomas is limited because only a limited
   area of the neck can be investigated. Ultrasound is the best in detection and follow-up
   of cervical paragangliomas and in detection of small paragangliomas. This study is
   performed with 5-10MHz transducer. It is necessary to detect tumor localisation, borders
   and relationship with surrounding structures. In general, skull base masses are poorly
   evaluated by ultrasound.

   **Carotid body tumors** usually present as solid, well-defined, hypoechoic tumors in B-
   mode sonography (Fig. 1a).

   Typically, there is a splaying of the carotid bifurcation (Fig. 1b), external carotid artery is
   usually displaced anteriorly and both the internal carotid artery and internal jugular vein
   are displaced posteriorly. Using B-mode sonography alone, it is often impossible though,
   to differentiate a carotid body tumor from other solid masses in the area of the carotid
   bifurcation. Color and power Doppler imaging demonstrate the intrinsic hypervascularity
   of the cervical paragangliomas (Fig. 2a,b).

   By using of high-resolution transducer, small vessels can be demonstrated within the
   tumor matrix (Fig. 1a).

   Depending on the site of origin, **vagal paragangliomas** often can not be assessed in
   their full extend by B-mode sonography and color-coded Doppler sonography. Large
   vagal paragangliomas may also lead to a splaying of the carotid bifurcation and might
   therefore be misinterpreted as a carotid body tumors. Delineation of high cervical vagal
   and jugular paragangliomas is cumbersome and concomitant lesions in the skull base
   region remain undetected, while such lesions are important in future treatment planning.

2. **CT scanning** with thin sections are superior at demonstrating the extent of bone
   destruction in **jugular and tympanic paragangliomas** (Fig. 6, 8).
The extend of temporal bone destruction is important to classify those tumors. The preoperative classification of jugular and tympanic paragangliomas is essential, since the operative approach will be chosen depending on the tumor stage. The patterns of spread of the glomus jugulare paragangliomas tumors are predictable and follow the paths of least resistance. High resolution CT of the temporal bone will show expansion and a moth eaten pattern of erosion of the jugular foramen in case of jugular paragangliomas (Fig. 6).

Tumor expansion will occur superiorly and subsequently into the tympanic cavity, causing destruction of the ossicular chain. Further extension laterally will destroy the bony canal of the facial nerve with infiltration of the nerve itself. Finally, intracranial posterior fossa extension can occur (Fig. 7b).

The tympanic paraganglioma can be depicted clearly as a small mass at the cochlear promontory (Fig. 8). Ossicular destruction is not common (Fig. 8b) and is especially present in larger lesions.

The CT appearance reflects the hypervascular nature of the head and neck paragangliomas resulting in homogeneous and intense enhancement after administration of intravenous contrast material (Fig 3, 7). In addition, the typical localization of each type of paraganglioma contributes to the specific diagnosis.

3. MRI plays an important role in evaluation of head and neck paragangliomas. The recommended MR pulse sequences should include a T1 weighted spin echo and a T2 weighted turbo spin echo sequence. T2 weighted fat suppressed and contrast-enhanced fat-suppressed T1-weighted sequences can offer additional diagnostic information for improved depiction of paragangliomas in the skullbase region but have not proven to be as effective as a pre- and post-contrast enhanced 3D Time of Flight (TOF) MR angiography sequence. The MR appearance is that of a lesion exhibiting low signal intensity on T1 (Fig. 9a, 10b, 11a, 13a) and a high signal intensity on T2 weighted images (Fig. 4b, 9c, 11b, 12b, 13b). Multiple serpentine and punctate areas of signal void characterize the typical paraganglioma with all MRI sequences; these areas are variably distributed throughout the mass and represent flow voids in the larger intratumoral vessels. Multiple areas of high and low signal intensity, the so-called "salt and pepper appearance", originally described by Olsen and al. can be seen within the lesion on T1w and T2w images (Fig. 11,12). The"pepper" component represents the multiple areas of signal void interspersed with the "salt" seen as hyperintense foci (due to slow flow or hemorrhage) on both T1w and T2w images (Fig. 11,12). This feature is especially seen in larger paragangliomas. On a 3D TOF MR angiographic sequence these areas of high flow are seen as areas of high signal intensity within the tumor. 3D TOF MR angiography is more susceptible in showing these high flow areas within the tumor than are conventional spin echo MR sequences (Fig. 5,13d).

The combination of imaging features with the typical localization, typical vessel displacement, enlarged feeding vessels, and intra tumoral flow signal makes the diagnosis paragangliomas highly likely, and the combination of these features almost
rules out other causes. When paragangliomas are localized in the region of the skull base, coronal MR images will give information on the extension of the tumor in and around the jugular foramen, but even the coronal and sagittal reconstructions of a 3D TOF MRA sequence can provide this information. Contrast enhanced MR angiography has been used to study the tumor hemodynamics of paragangliomas. Paragangliomas show a rapid and intense homogeneous enhancement following the intravenous administration of contrast material (Fig. 4c, 9b,d; 10c, d; 12a, 13c). These features could be established in tumors as small as 10 mm in diameter.

4. The digital subtraction angiography is definitive imaging procedure to confirm the diagnosis of a head and neck paraganglioma and it is still the gold standard in detection of small paragangliomas. Angiography shows the specific vascular supply of the paraganglioma. The arteriographic findings of glomus jugulare tumors are typically a hypervascular mass and with an intense characteristic tumor "blush" (Fig. 14 a, b). Large feeding vessels and early draining veins are commonly encountered, constituting an early arterovenous shunting. Glomus jugulare tumors are predominantly supplied by the external carotid artery system, including mainly the ascending pharyngeal artery. The ascending pharyngeal artery can be considered "the artery of the paraganglioma", because its branches can supply tympanic, jugular, vagal, carotid, and even laryngeal paragangliomas.

In addition the supply of paragangliomas is derived from the occipital artery, posterior auricular artery, muscular branches of the vertebral artery and deep cervical artery and thyrocervical trunk can also contribute to the vascularization. The highly vascular nature of the tumor is reflected in enlargement of the feeding arteries, the intense staining of the tumor and a rapid venous drainage.

Embolization can be performed with DSA for preoperative devascularization of the tumor (Fig. 14 c, d). The primary aim of preoperative embolization is to reduce tumor vascularity which can lead to a decreased intraoperative blood loss. This fact obviously provides a better operative field for the surgeon, thus reducing the possibility of a nerve or vessel injury.

Images for this section:
Fig. 1: Carotid body tumor. a) B mode sonography shows solid, well-defined, hypoechoic tumoral mass with many anechoic, small blood vessels (arrows), b) color doppler sonography demonstrate splaying of carotid bifurcation

Fig. 2: Carotid body tumor - a) color and b) power doppler sonography demonstrate hypervascularity of the carotid body paraganglioma with multiple small blood vessels into tumoral tissue (arrows)
Fig. 3: Left carotid body tumor- postcontrast MDCT images: a) sagittal and b) axial images demonstrate strong postcontrast enhancement of the tumoral mass that splays the internal and external carotid arteries (arrows)
Fig. 4: Left-sided carotid body paraganglioma: different presentations on the MRI sequences a) T1w fat-suppressed axial image shows isointense tumor (arrow) b) T2w axial image shows hyperintense tumor (arrow) c) T1w fat-suppressed postcontrast axial image and d) T1w postcontrast coronal image demonstrates strong tumor enhancement (arrows)
Fig. 5: Left sided carotid body tumor: 3D-TOF MR angiography after contrast administration a) axial and b) coronal images clearly show the highly vascular nature of the lesion and the tumor in relation to the carotid arteries (arrows)

Fig. 6: Left sided glomus jugulare paraganglioma: skull base MDCT (bone window) a) axial and b) coronal images demonstrate permeative destruction and enlargement of the jugular foramen in association with loss of the skull base’s density (arrows)
**Fig. 7:** Left sided glomus jugulare paraganglioma: postcontrast MDCT a) axial and b)sagittal images show strong tumoral enhancement with extension in posterior cranial fossa

**Fig. 8:** Right sided tympanic paraganglioma: MDCT of the skull base (bone window) a) axial and b) coronal images demonstrate small, oval mass at right cochlear promontory without ossicle chain destruction
**Fig. 9:** Right sided glomus tympanicum paraganglioma: different presentations on MRI sequences a) T1w axial image shows hypointense tumor mass b) T1w postcontrast axial image shows postcontrast enhancement of the tumor mass c) T2w axial image demonstrates hyperintense tumor d) T1w postcontrast coronal image demonstrates postcontrast tumoral enhancement
**Fig. 10:** Left sided glomus jugulare paraganglioma: different presentations on MRI sequences a) T1w axial fat suppressed image shows isointense tumor b) T1w coronal image demonstrates hypointense tumor c) T1w axial postcontrast fat suppressed and d) T1w coronal postcontrast images show strong tumoral enhancement
**Fig. 11:** Glomus jugulare paraganglioma: MRI sequences a) T1w axial image - hypointense tumoral mass with multiple flow voids (arrows) b) T2w axial image - hyperintense tumoral mass with multiple flow voids and hyperintense foci which represent high and low blood flow - "salt and pepper" appearance (arrows)

**Fig. 12:** Left sided glomus vagale paraganglioma: MRI a) T1w axial postcontrast fat suppressed image - tumoral enhancement (arrow) b) T2w axial image - "salt and pepper" appearance (arrow)
**Fig. 13:** Left sided glomus jugulare paraganglioma: Different MRI sequences a) T1w axial image-hypointense tumor presentation b) T2w axial image-hyperintense tumor presentation c) T1w postcontrast coronal image - strong tumoral enhancement d) contrast 3D-TOF MRA shows the highly vascular nature of the lesion.
**Fig. 14:** Left sided glomus jugulare: DSA- sagittal view a,b) DSA imaging before embolization demonstrates characteristic intense tumor blush and intratumoral shunts c,d) after selective embolization-significant reduction in vascular blush indicative of reduced arterial supply to the tumor
Conclusion

1. A proposed diagnostic algorithm in management of head and neck paragangliomas includes B-mode sonography with color-coded Doppler sonography, computed tomography, magnetic resonance imaging and digital substraction angiography. Paragangliomas of the head and neck are highly vascular lesions with hypervascularity demonstrated on each diagnostic modalities. Intense enhancements always noted following contrast administration. Imaging hallmarks of head and neck paragangliomas include an hypervascular, enhancing soft tissue mass in the carotid space, jugular foramen or tympanic cavity noted by all diagnostic procedures.

2. Imaging studies depict the location and extent of tumor involvement, help determine the type of head and neck paraganglioma. Carotid body tumors are typically located at carotid bifurcationa with splying of external and internal carotid artery; jugular, sometimes vagal paragangliomas demonstrate temporal bone infiltration with erosion and enlargement of the jugular foramen; and tympanic paragangliomas are usually present on cochlear promontory in the middle ear cavity.

3. Each imaging modality has its own role in diagnostic management of paragangliomas. Color Doppler sonography is first imaging step and it is useful in diagnostic of carotid body tumors and paragangliomas which extended to the neck. CT is sensitive in evaluation of bony destructions which demonstrate jugular, sometimes vagal and tympanic paragangliomas. MRI is the important imaging technique for defining the relationship of the lesion to the adjacent anatomic structures and it shows the typical "salt and pepper" presentation of paragangliomas. Magnetic resonance angiography provides information concerning the vascular supply of the lesion and displacement of adjacent vessels. The definitive imaging procedure to confirm the diagnosis of a head and neck paraganglioma represents the DSA, which is the gold standard in detection of small paragangliomas. Angiography shows the specific vascular supply of the paraganglioma and it is required preoperatively in larger paragangliomas for surgical planning and preoperative embolisation.

Personal Information

Sladjana Petrovic, MD, PhD
References