

***SEASONED WITH “SALT AND PEPPER”:
THE TASTE OF HEAD AND NECK
PARAGANGLIOMAS***



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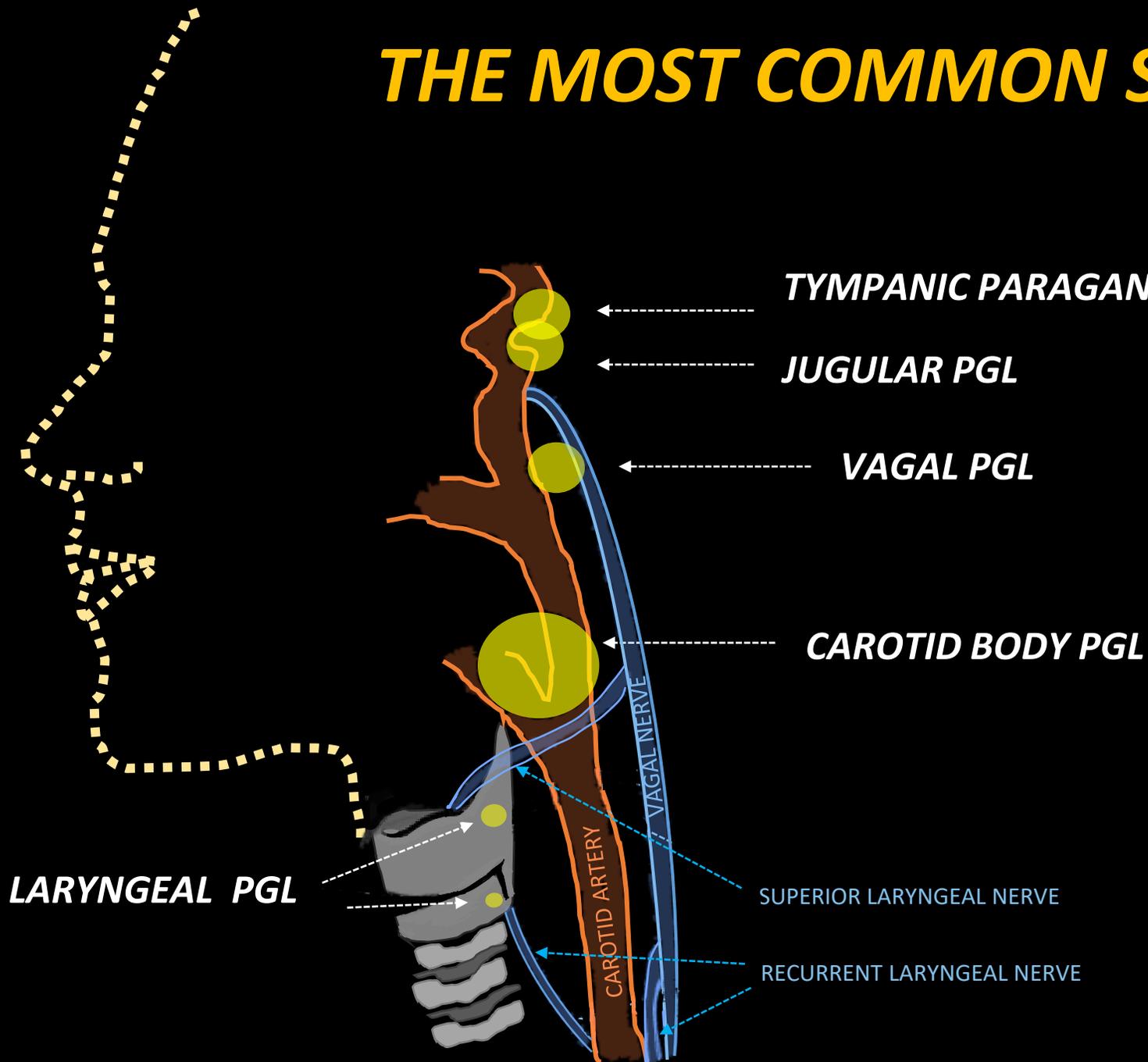
LEARNING OBJECTIVES:

- Be familiar with origin and location of head and neck (HN) paragangliomas (PGLs).
- Recognize CT and MR imaging features of different types of PGLs.
- Identify some differential diagnosis.

Introduction

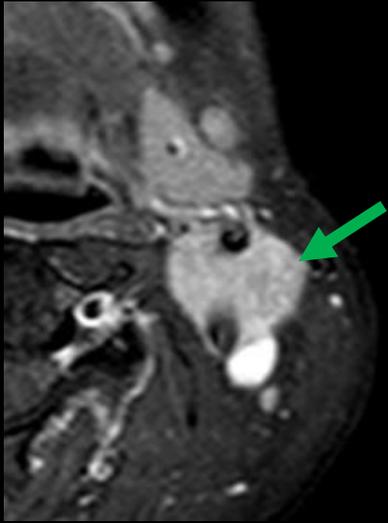
- HN PGLs are uncommon neuroendocrine tumors of neural origin (paraganglion cells), being rarely functional (<1%). Most of them are **benign** (>90%) and only locally invasive.
- Age of onset is around 5th–6th decade, with a female predominance.
- HN PGLs are highly vascularized, showing intense avidity to intravenous contrast on both CT and MR.
- CT has excellent spatial resolution and plays an important role in characterizing bone destruction. MRI, in turn, has superior soft tissue contrast and allows better identification of the vascular anatomy.
- MRI shows a heterogeneous lesion, predominantly hypointense signal on T1WI and isointense to hyperintense signal on T2WI. The typical "Salt and Pepper" appearance on T1WI corresponds to hemorrhage ("salt") and flow voids due to high vascularity ("pepper").

THE MOST COMMON SITES OF HN PGLS

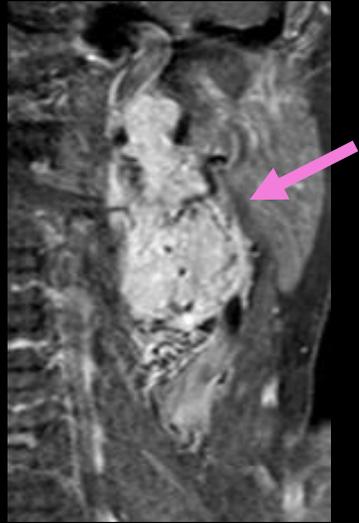


The larger the yellow ball, the more common the site of origin.

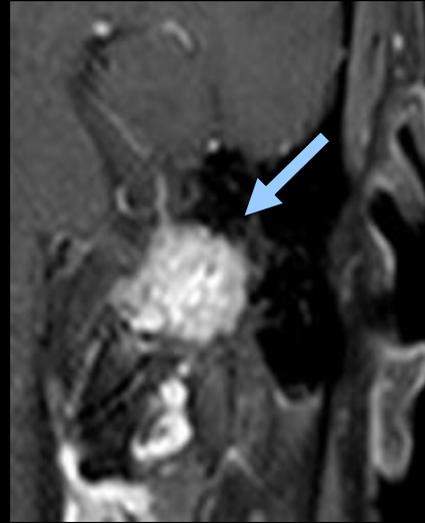
MOST COMMON TYPES OF HN PGL AND IMAGE FEATURES



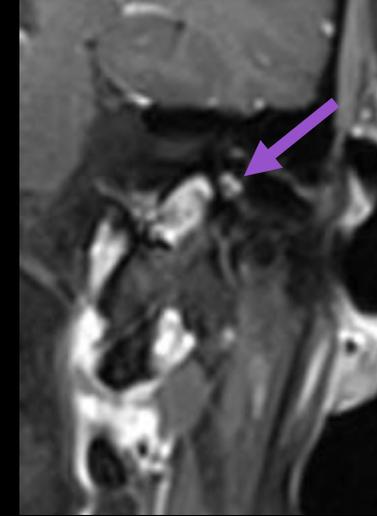
Carotid body



Vagal



Jugular



Tympanic



Laryngeal

Once they belong to the neuroendocrine system, HN PGLs are highly vascularized. Imaging hallmarks are:

- ✓ Highly contrast-enhancing soft-tissue mass in the carotid space, jugular foramen or tympanic cavity on CT.
- ✓ "Salt and Pepper" appearance on standard spin-echo MRI.
- ✓ Intense blush on angiography.

CAROTID BODY PGL

General Aspects

Most common (60%).

Arise from paraganglia of the carotid body, above carotid bifurcation.

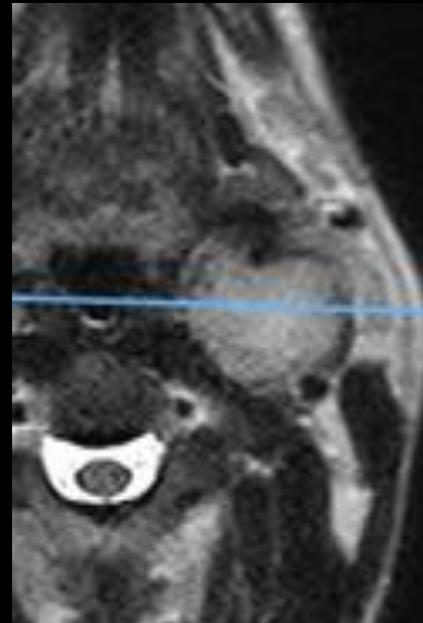
Most cases are unilateral and sporadic. Bilateral/multifocality may occur (usually hereditary).

Increased incidence in patients living at higher altitudes, or in the setting of chronic obstructive lung disease.

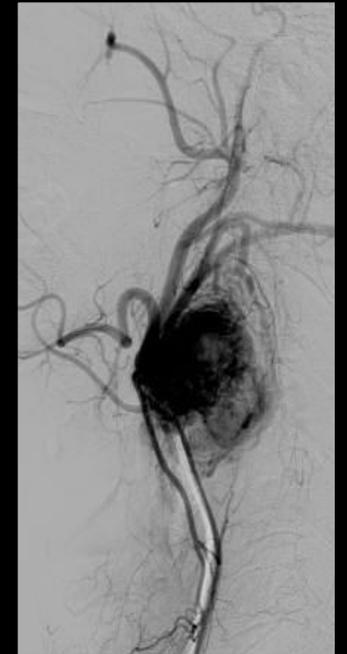
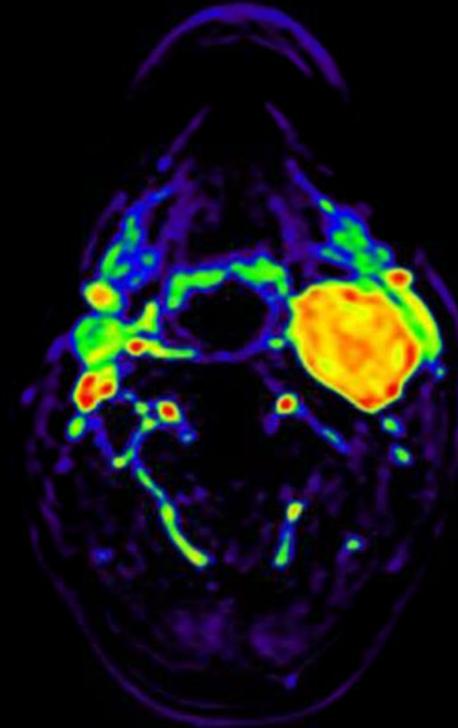
Metastasis risk about 6%.

DYNAMIC CONTRAST ENHANCEMENT (DCE) STUDY USING 3D T1 WI SHOWING HIGH PERMEABILITY

CT ARTERIOGRAPHY SHOWS INTENSE BLUSH DUE TO HYPERVASCULARIZATION



AX T2 WI



CAROTID BODY PGL

Key points

Asymptomatic neck mass near angle of jaw.

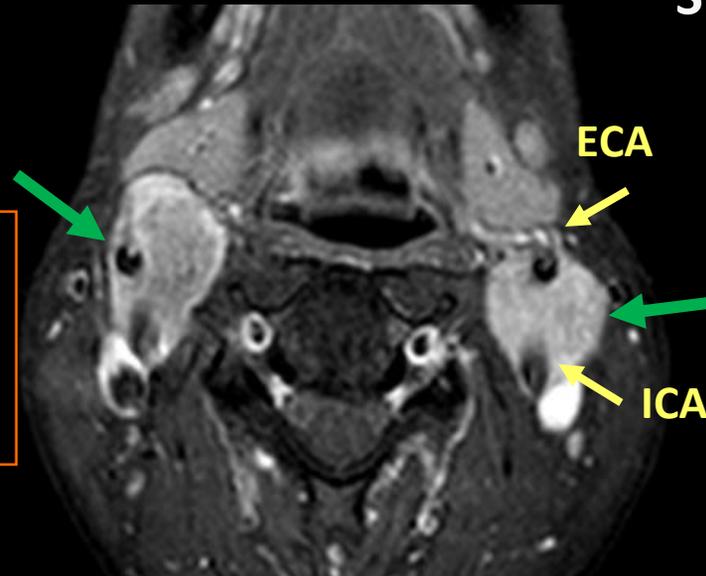
On CT or MR imaging, it presents like a mass above carotid bifurcation (**green arrows**) splaying internal carotid artery (ICA) and external carotid artery (ECA) (**yellow arrows**), resulting in *lyre sign*.

Association with Von Hippel Lindau Syndrome (6%)



**FAMILIAL
BILATERAL PGL**

**MR ANGIOGRAPHY + GD CORONAL
MIP RECONSTRUCTION SHOWS
TUMOR BLUSH IN THE ARTERIAL
PHASE (ORANGE ARROWHEADS).**

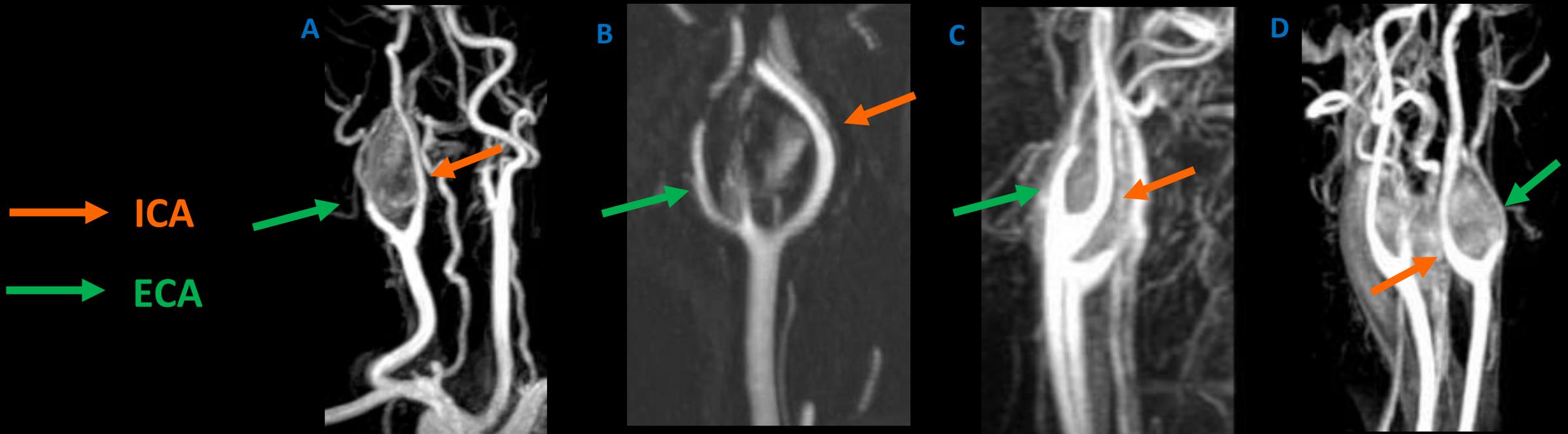


AX T1 WI FS Gd+

**LOOK FOR MORE LESIONS IN FAMILIAL
TUMORS!!!!**

- ≈ 40% OF CASES ARE HEREDITARY (USUALLY MULTICENTRIC / BILATERAL).
- IN THOSE CASES, THE PEAK OF PREVALENCE IS EARLIER (30-35 YEARS).

THE TYPICAL LYRE SIGN



Mass effect classically results in splaying of **ICA** and **ECA**, resulting in the characteristic “**lyre sign**”

COME CLOSER TO SEE THE “SALT AND PEPPER”...



AXIAL T1 WI IMAGE SHOWS THE TYPICAL PGLS APPEARANCE OF “SALT” (HEMORRHAGIC FOCI) AND “PEPPER” (FLOW VOIDS)

Most carotid body PGLs are asymptomatic. However, if the tumor keeps on growing within the carotid space, it may **compress** the adjacent nerves (most commonly the **vagus**), resulting in symptoms such as **dysphagia, hoarseness and those related to the Horner’s syndrome.**

JUGULAR PGL

General aspects

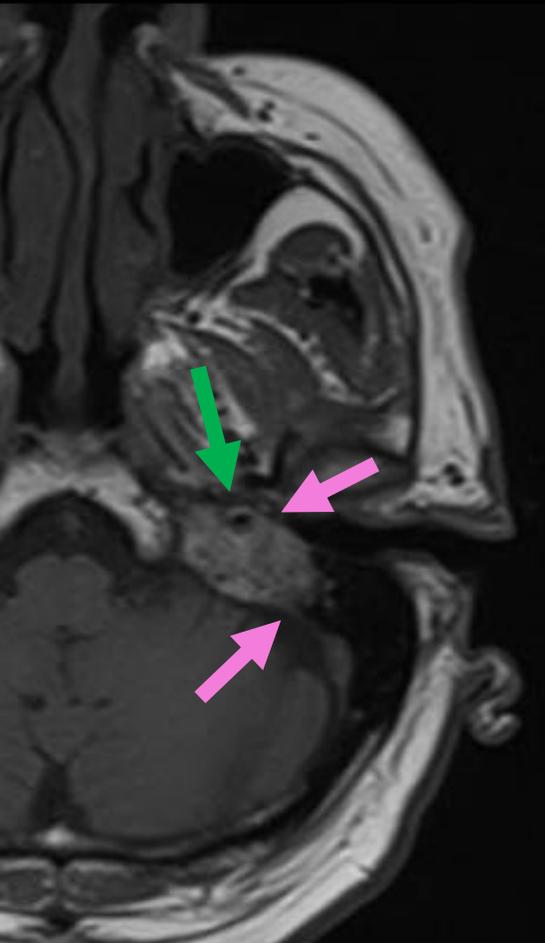
2nd most common HN PGL
(most prevalent tumor within the jugular fossa).

Arises from the paraganglia associated with the
adventitia of the jugular bulb.

Tends to grow aggressively beyond the limits of the
jugular foramen, causing bone erosion and reaching
adjacent foramina, as well as the eustachian tube.

LOOK FOR BONE EROSION (PERMEATIVE MARGINS)

**THE SHOWN CASE DEMONSTRATES LEFT ICA ENCASEMENT
(GREEN ARROW) AND VENOUS (IJV AND SIGMOID SINUS)
COMPRESSION (PINK ARROWS).**



AX T1 WI



Axial CT-section reconstructed
with a bone algorithm

JUGULAR PGL

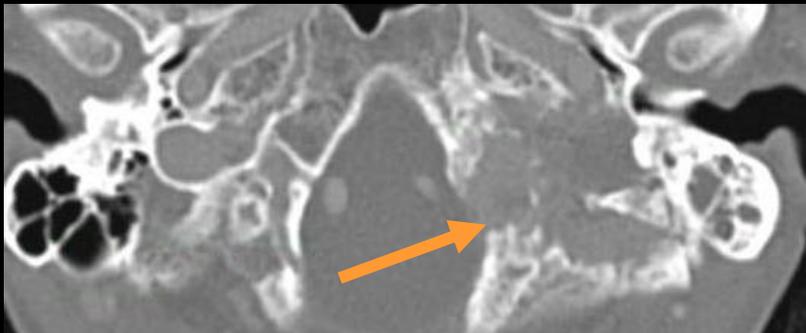
Key points

Unilateral tinnitus and/or hearing loss (51% of patients).

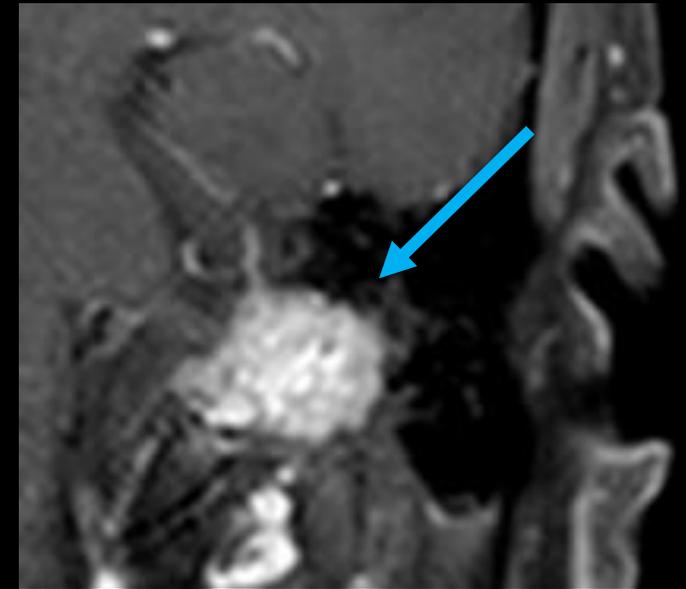
MR is specially important in detecting intracranial extension.



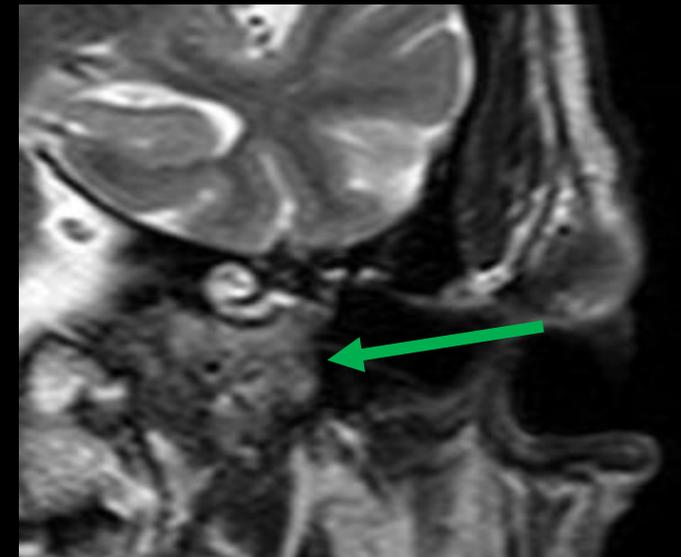
**Due to frequent bone involvement,
dedicated temporal bone CT is
mandatory in jugular PGL**



Axial CT-section reconstructed with a bone algorithm

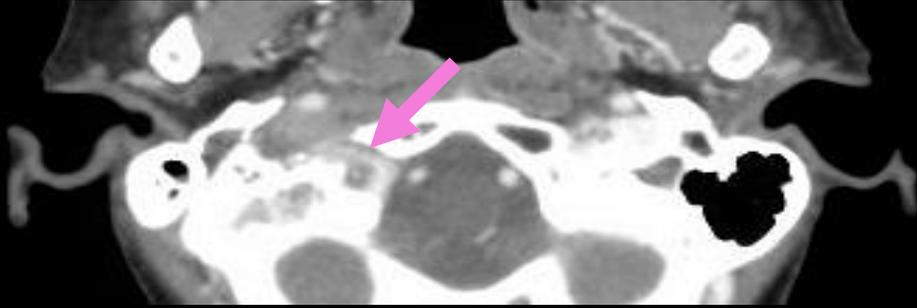


COR T1 WI FS Gd+



COR T2 WI

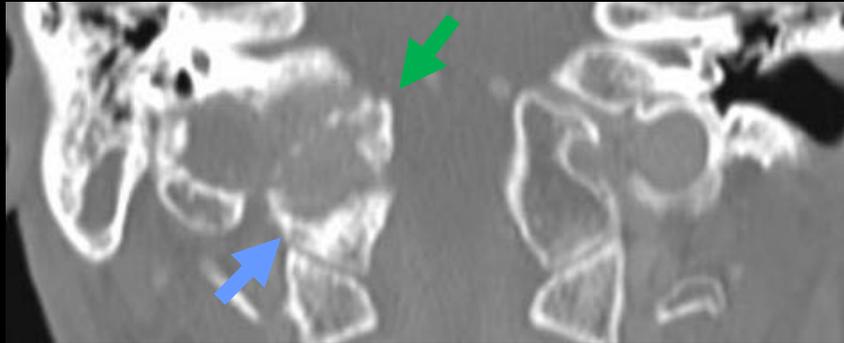
JUGULAR PGL AND BONE DESTRUCTION



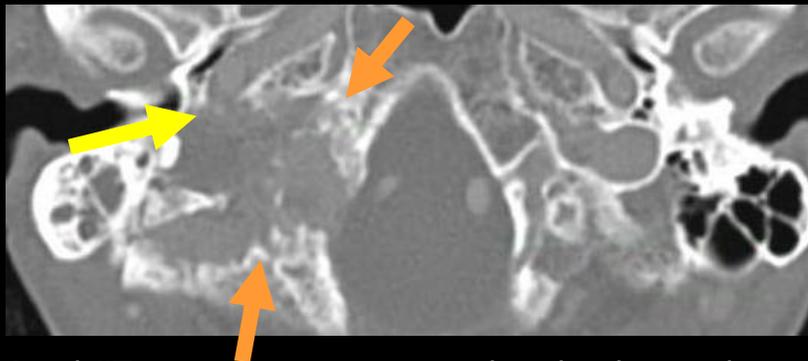
Jugular foramen mass and permeative-destructive changes in the adjacent bone.

Advanced stages result in a “moth-eaten” appearance due to bone erosion.

Axial CECT-section reconstructed with a soft tissue algorithm



The hypervascular mass is not so evident on CT. However, there is invasion of the hypoglossal canal (**pink arrow**) and erosion of the adjacent occipital condyle (**blue arrow**), jugular tubercle (**green arrow**), posterior carotid wall (**yellow arrow**) and jugular foramen walls (**orange arrows**).



Axial CT-section reconstructed with a bone algorithm

TYMPANIC PGL

General aspects

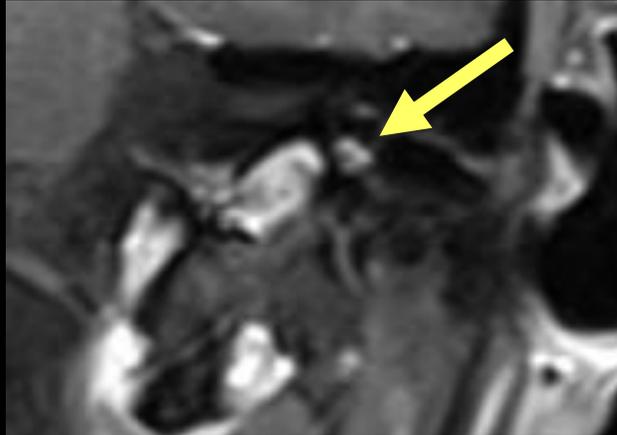
Marked female predilection (80–90%) and later age of onset (60 yo).

Arises from the tympanic branch of the glossopharyngeal nerve.

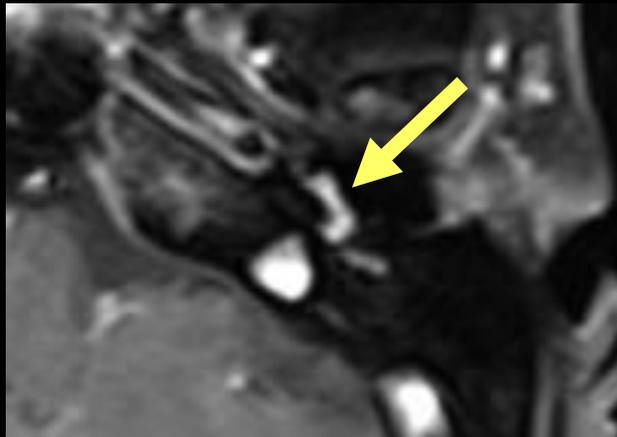
Bone involvement is rare and the ossicles are typically spared.

Metastases risk about 2-4%.

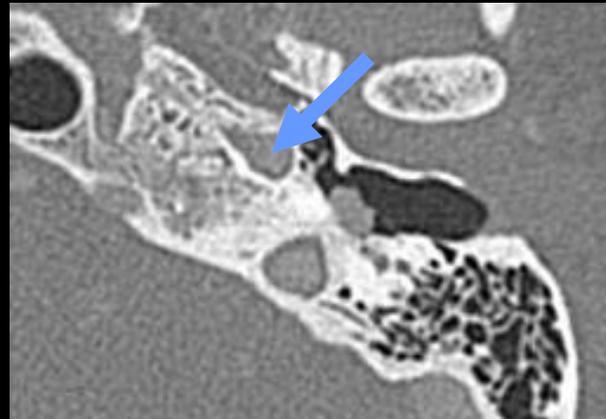
EXPANSIVE LESION OVER THE COCHLEAR PROMONTORY (BLUE ARROW). THERE IS NO BONE INVOLVEMENT.



EXPANSIVE AND IRREGULAR LESION WITH INTENSE GD ENHANCEMENT (YELLOW ARROWS).



COR T1 WI FS Gd+



Axial CT-section reconstructed with a bone algorithm

TYMPANIC PGL

Key Points

Like jugular PGL, tympanic PGL frequently presents with pulsatile tinnitus and/or unilateral hearing loss, but are typically smaller and less aggressive.

Otосcopy may show a blueish pulsatile mass within the middle ear.



Axial CT-section reconstructed with a bone algorithm

Source: Case courtesy of Dr Mohammad Taghi Niknejad, Radiopaedia.org, rID: 54275



Axial CT-section reconstructed with a bone algorithm

Source: Tiago, RSI ET AL. Rev Bras Otorrinolaringol 2007;73(1):143.

Temporal bone CT shows a soft tissue mass confined to the tympanic cavity, centered over the cochlear promontory.

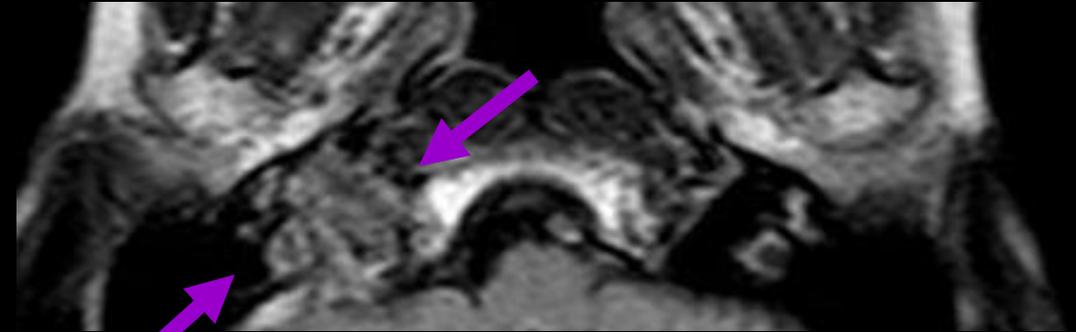
PGL highly vascular nature helps to differentiate them from common tympanic lesions such as cholesteatomas on both CT and MR.

JUGULOTYMPANIC PGL

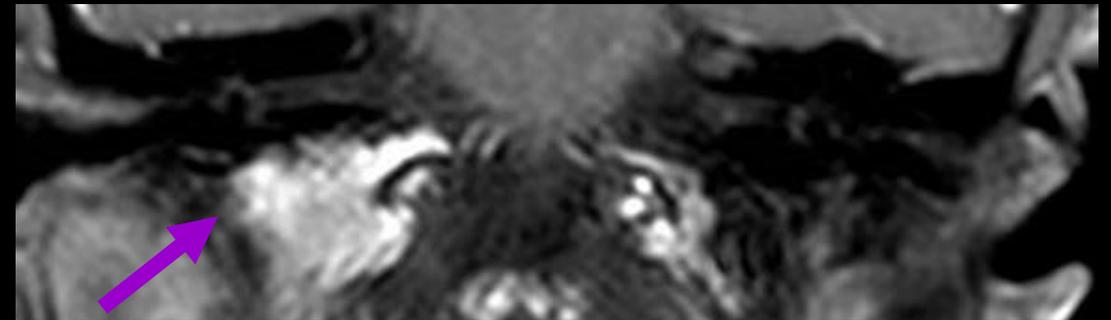


When there is invasion of the middle ear by a Jugular PGL, the term "jugulotympanic" should be used.

ANY BONE EROSION IS CONSIDERED DIAGNOSTIC OF JUGULAR OR JUGULOTYMPANIC RATHER THAN TYMPANIC PGL



AX T1 WI



COR T1 WI FS Gd+



Axial CT-section reconstructed with a bone algorithm

VAGAL PGL

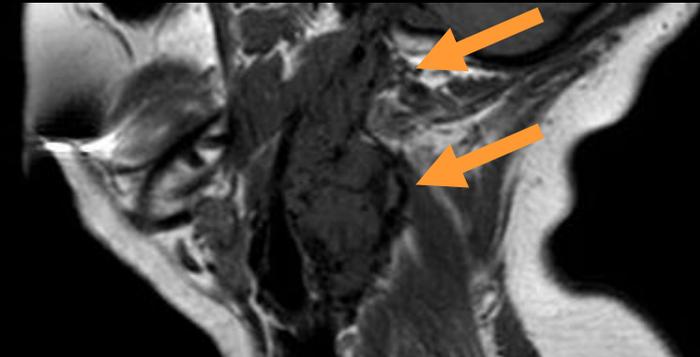
General aspects

Uncommon HN PGL (5-10%).

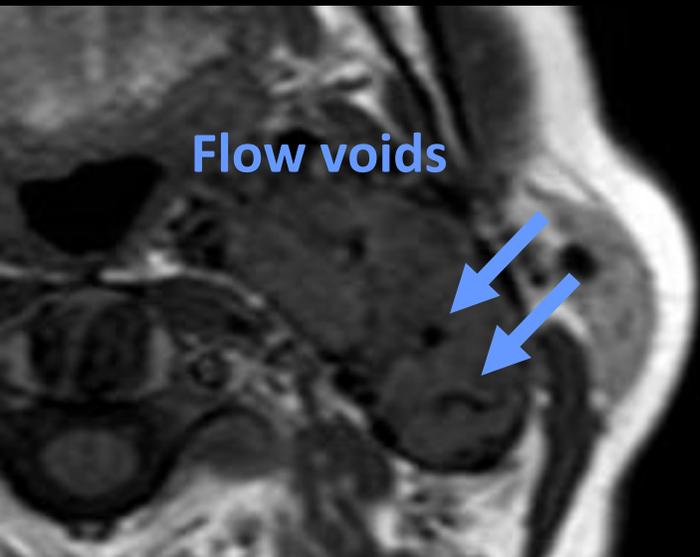
Bilateral / Multifocal in 20-40%.

Typically seen where the vagus exits the jugular foramen, in the suprahyoid neck (paraganglia associated to the inferior nodose ganglion).

Metastases risk about 16%.

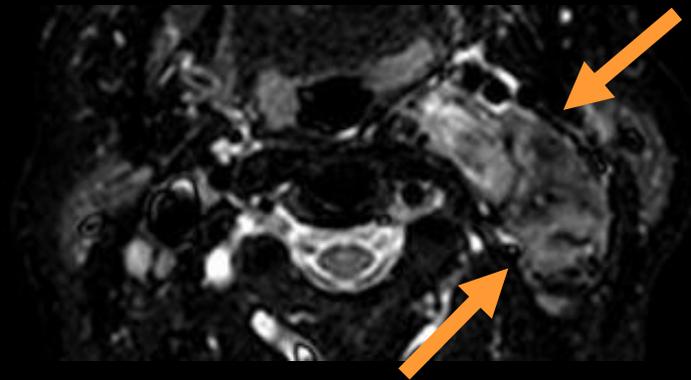


SAG T1 WI



Flow voids

AX T1 WI

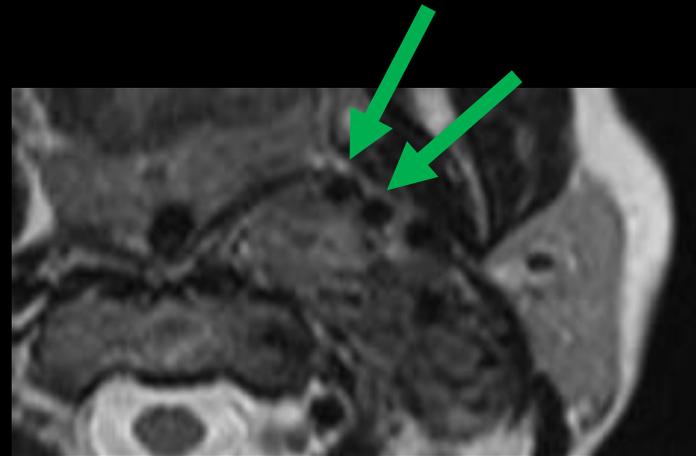


AX STIR

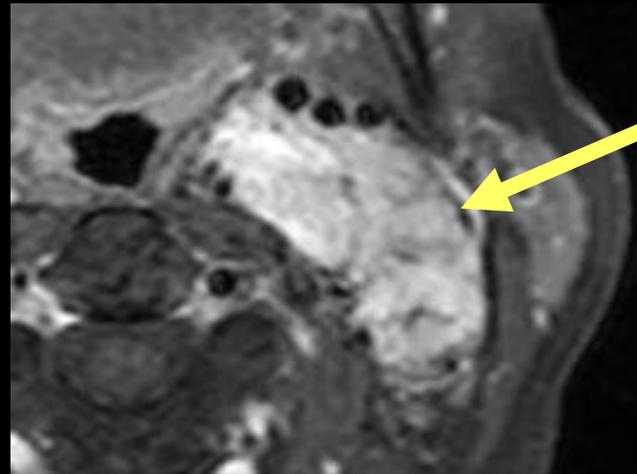


AX T1 WI FS Gd+

VAGAL PGL



AX T2 WI



AX T1 FS Gd+

Key Points

Typically presents as a slow-growing painless neck mass (similar to carotid body PGL) in the cranial aspect of the carotid space (**purple arrows**).

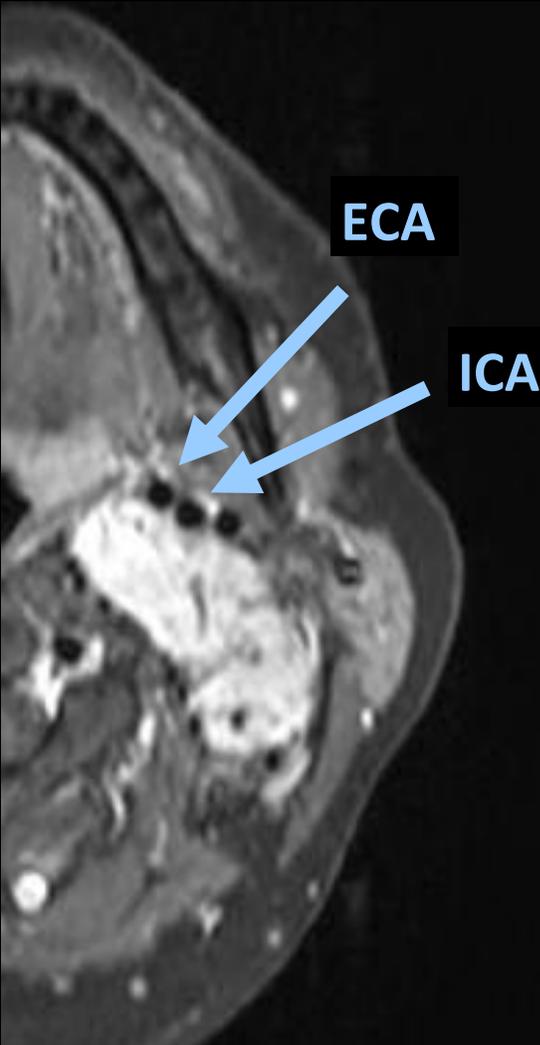
Therefore, vagal nerve palsy and hoarseness are the most common symptoms (35%).

Due to the posterior and lateral path of the vagus nerve with respect to ICA and ECA, both arteries are **displaced anteromedially together** (**green arrows**). IJV, in turn, is compressed posterolaterally (**yellow arrow**)

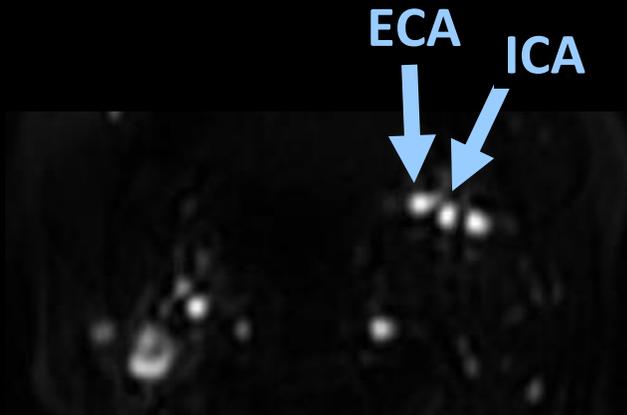
Coronal reconstruction, 3D
Phase-contrast MRI angiography.

VAGAL x CAROTID BODY PGL

Despite many clinical and radiological similarities between vagal and carotid body PGLs, ICA and ECA displacement patterns are different.

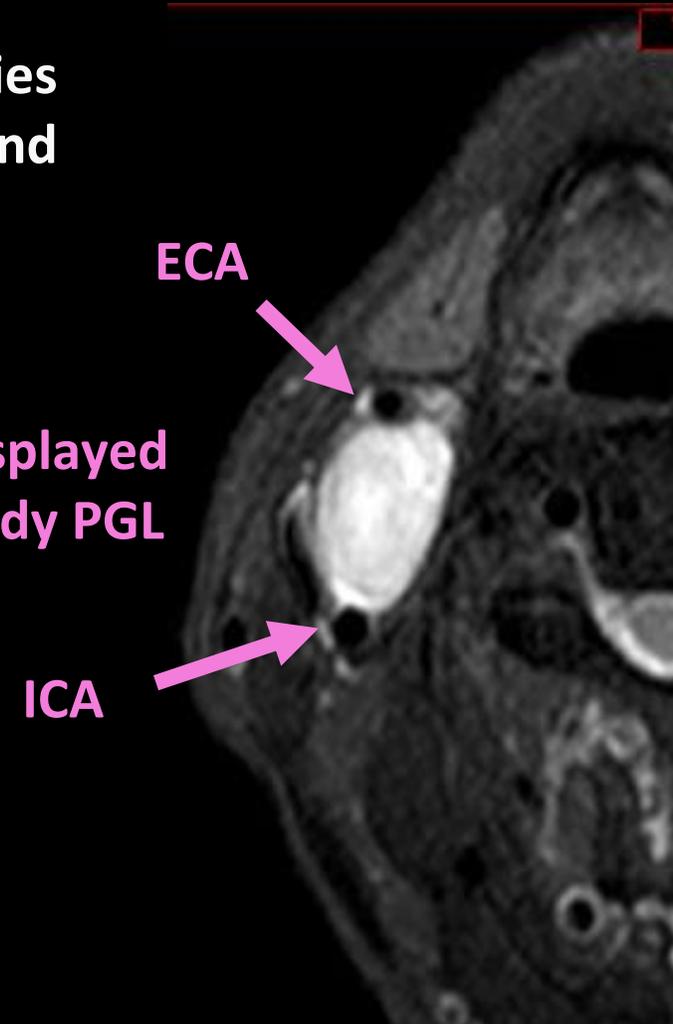


Both ICA and ECA displaced anteromedially by vagal PGL



TOF Axial reconstruction

ICA and ECA splayed by carotid body PGL

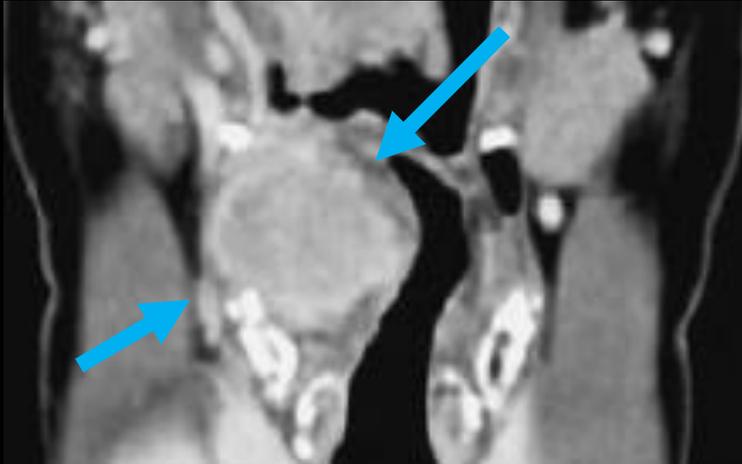


AX T2 WI FS

AX T1 WI Gd+

LARYNGEAL PGL

General aspects

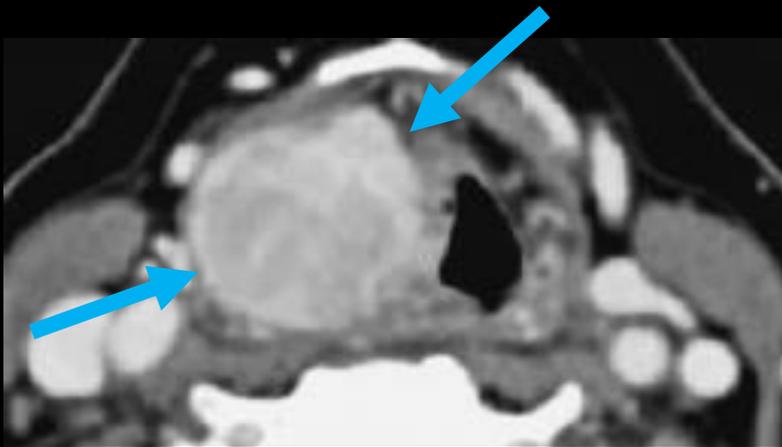


Very rare type of HN PGL (\cong 100 verified cases in the literature to date).

Superior laryngeal paraganglia tissue accounts for 90% of laryngeal PGL.

Important differential diagnosis with carcinoid tumors.

Metastasis risk about 2%.



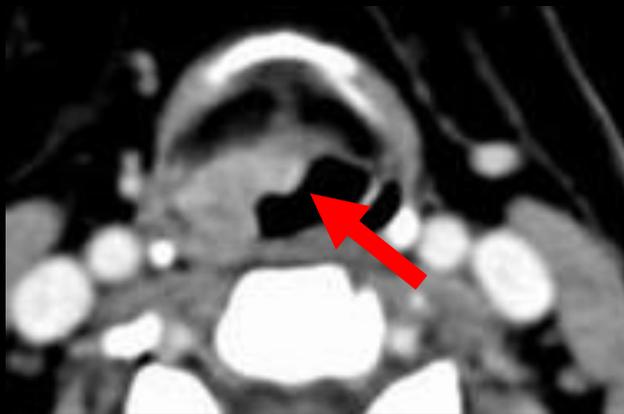
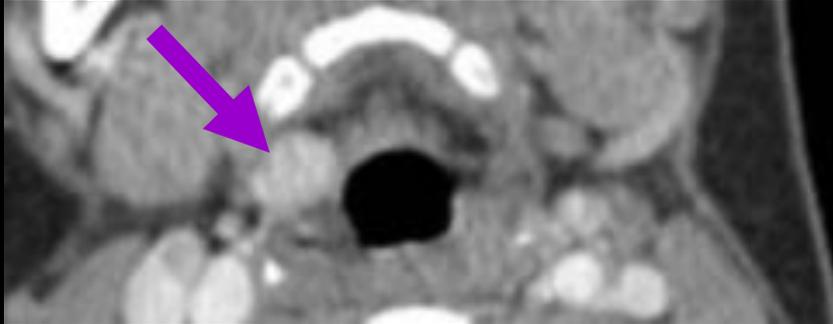
Coronal and Axial CT images reconstructed with a soft-tissue algorithm

LARYNGEAL PGL

Key Points

Typically presents as a supraglottic mass, usually in the right aryepiglottic fold (82%) (purple arrow).

Patients may present hoarseness, wheezing sound and/or hemoptysis.



The case shown presents a typical laryngeal carcinoid. But laryngeal PGL, although rare, must be considered as a differential diagnosis when a supraglottic hypervascular mass is seen.

Soft tissue algorithm axial reconstruction of mastoid CT

Source:

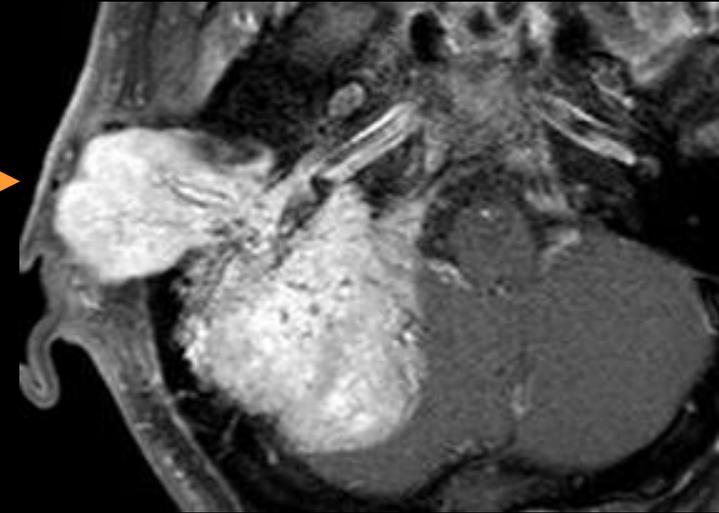
(A) Dogan S. et al. Hindawi Publishing Corporation Case Reports in Radiology Volume 2015. <http://dx.doi.org/10.1155/2015/342312>

(B) Kayhan, F.T., Başaran, E.G. Typical carcinoid tumor of the larynx in a woman: a case report. *J Med Case Reports* 4, 321 (2010). <https://doi.org/10.1186/1752-1947-4-321>

COMPLICATIONS

CONCERNING INTRINSIC COMPLICATIONS OF PGLS ARE MAINLY RELATED TO LOCAL INVASION

Giant Jugulotimpanic PGL protruding laterally through the external auditory canal and posteromedially into the posterior fossa, compressing the right cerebellar hemisphere.



AX T1 WI FS Gd+



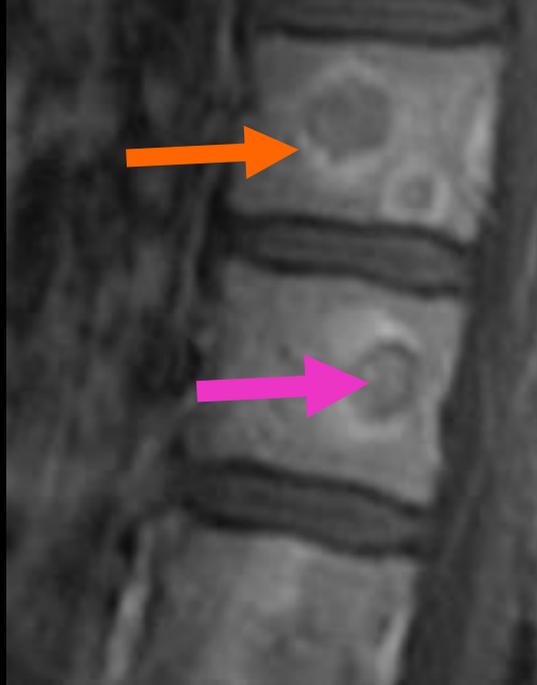
An expansile carotid PGL growing medially and narrowing the aerodigestive lumen.

Axial CT-section reconstructed with a soft-tissue algorithm

COMPLICATIONS

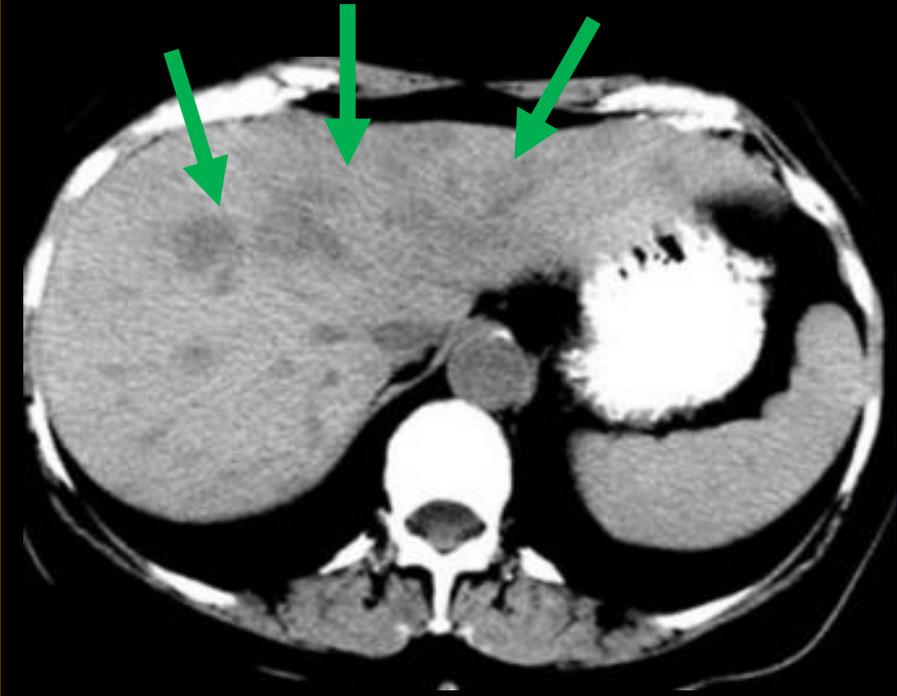
Malignant transformation is diagnosed when there is metastatic disease (mainly for jugular and carotid PGLs).

Distant metastasis are very rare, but may occur to the bone, lung and liver.



Multiple nodular vertebral lesions with a subtle hyperintense halo suggesting fat (**orange arrows**), and a central hypointense area with slightly enhancement (**pink arrow**) on T1 W1 Gd+ Patient presented both carotid body and vagal PGL.

Source: Duet M. et al JMRI, 24:428– 433 (2006)



Axial TC image shows hypodense nodular hepatic areas (**green arrows**) in a patient with carotid body PGL.

Source: Moskovic et al doi:10.1186/1758-3284-2-23.

COMPLICATIONS

Most complications of PGL are related to treatment, especially surgical (which is the only definitive one).

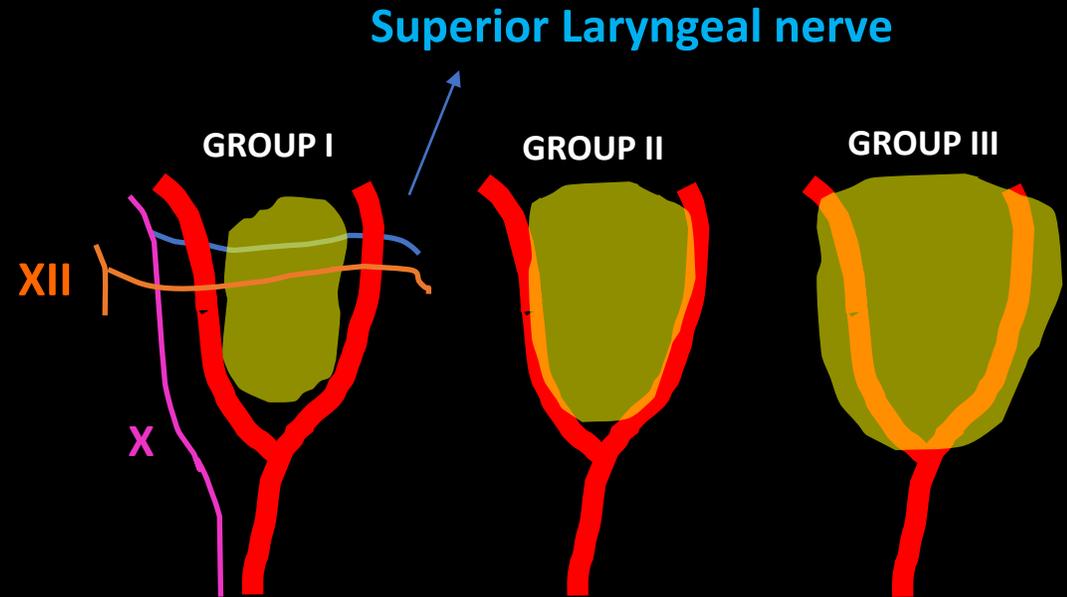
Early postoperative nerve deficit (56%), although mostly temporary.

Vagal nerve palsy (even without being sectioned).

Perforation of the tympanic membrane.

Neurosensorial hearing loss.

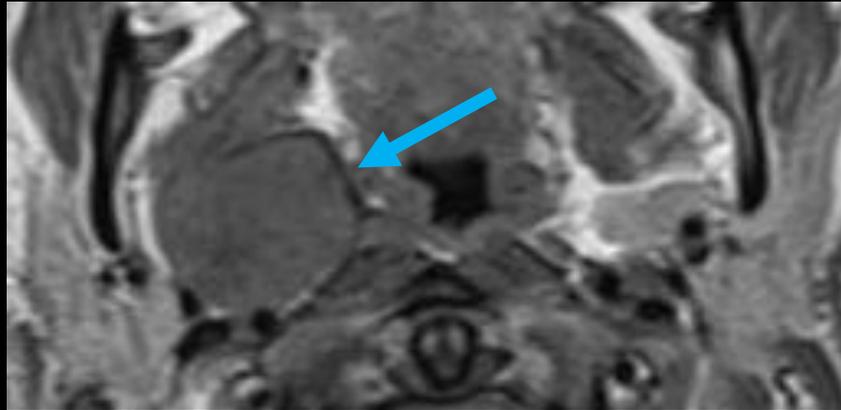
Iatrogenic cholesteatoma.



For carotid body PGL, tumor size and Shamblin (1970) classification are predictive of resection neurovascular complications. Group III lesions present the worse surgical prognosis.

BE AWARE TO SOME OF DIFFERENTIAL DIAGNOSIS...

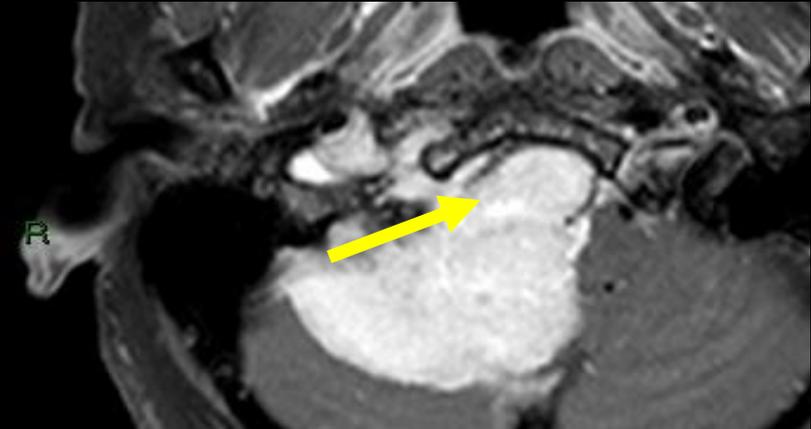
Vagal PGL or Schwannoma?



AX T1 WI

Well circumscribed mass homogeneous and isointense on T1WI (**blue arrow**) pushing both ICA and IJV posteriorly and the parapharyngeal fat anteriorly. Location anterior to the vessels and absence of flow-voids may help differentiate between PGL and the correct diagnosis: sympathetic chain Schwannoma.

Jugular PGL or Meningioma?

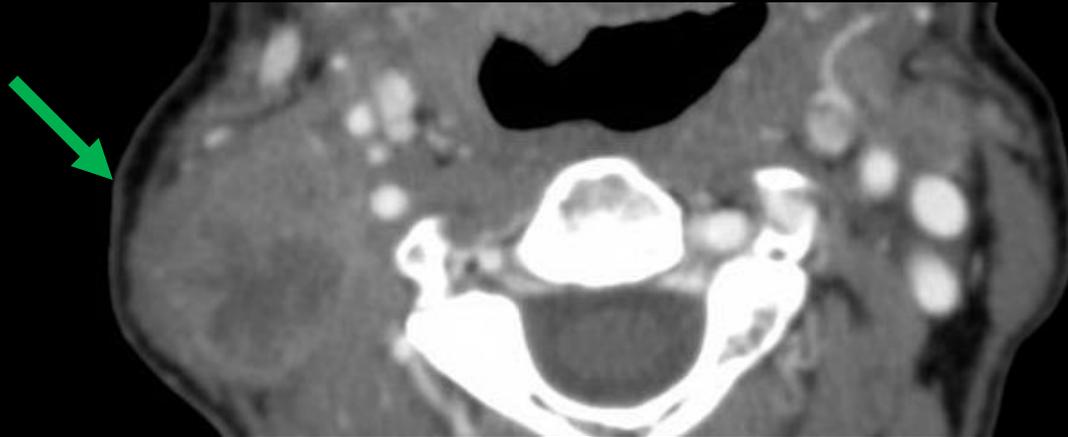


AX T1 FS Gd+

Lobulated mass growing from the posterior fossa to the jugular foramen (**yellow arrow**). No flow voids, homogeneous contrast enhancement and dural attachment. Diagnosis: meningioma.

BE AWARE TO SOME OF DIFFERENTIAL DIAGNOSIS...

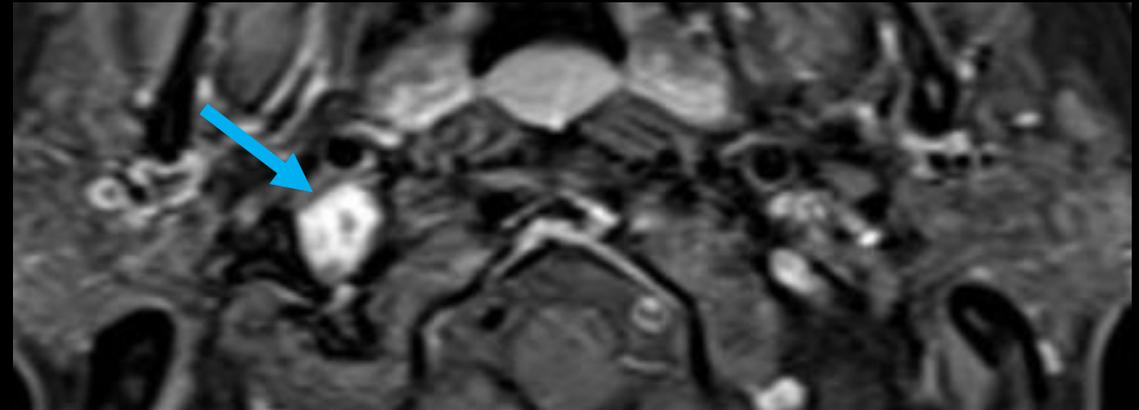
Carotid Body PGL or Lymph Node Enlargement?



Axial CT-section reconstructed with a soft-tissue algorithm

Right cervical necrotic mass (green arrow) compressing the IJV and pushing both ICA and ECA anteromedially. Level IIA lymph node enlargement.

Jugular PGL or Vagal Schwannoma?

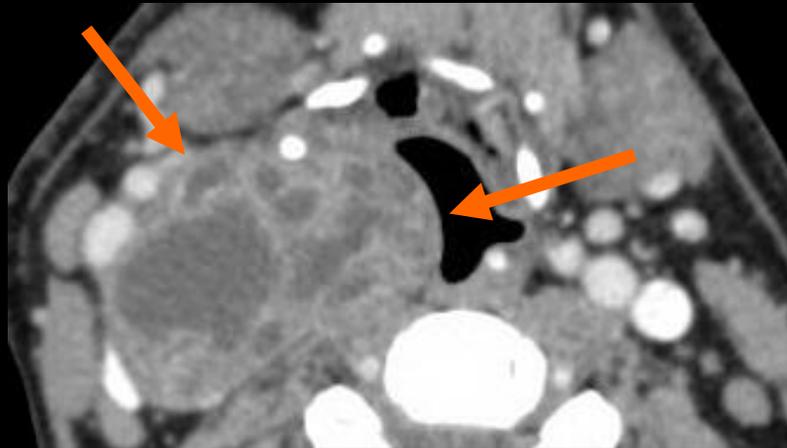


AX T1 WI Gd+

Well circumscribed mass with intense contrast enhancement close to the right jugular foramen (blue arrow). Small vagal Schwannoma.

BE AWARE TO SOME OF DIFFERENTIAL DIAGNOSIS...

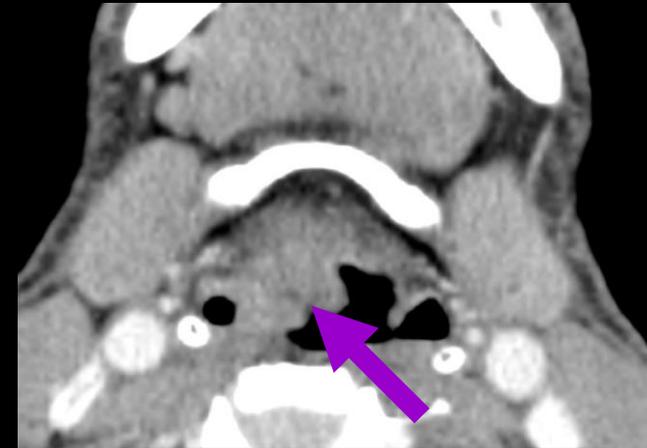
Carotid Body PGL or Schwannoma?



Axial CT-section reconstructed with a soft-tissue algorithm

Predominantly cystic mass (**orange arrows**) located deep to the carotid space. Diagnosis: sympathetic chain Schwannoma

Larynx PGL or Supraglottic Squamous Cell Carcinoma?



Axial CT-section reconstructed with a soft-tissue algorithm

Poorly circumscribed supraglottic mass (**purple arrows**) with a moderate contrast enhancement. Diagnosis: Squamous Cell Carcinoma.

HIGHLIGHTS

PGL	Frequency	Beyond brightness	Clinic aspects
Carotid body	60%	Location above the carotid bifurcation, splaying the ECA and ICA (LYRE SIGN)	Usually ASSINTOMATIC
Jugular	30%	PERMEATIVE BONE DESTRUCTION	PULSATILE TINNITUS and/or HEARING LOSS symptoms
Tympanic		SMALL LESION OVER THE COCHLEAR PROMONTORY	
Vagal	10%	HIGHEST METASTATIC RISK (16%)	NERVE COMPRESSION SYMPTOMS (IX-XII)
Larynx	Rare	SUPRAGLOTTIC SUBMUCOSAL MASS	HOARSENESS, STRIDOR or DYSPNEA

CONCLUSIONS

- Although rare, HN PGLs should be considered when a mass is seen within the carotid space, jugular foramen, middle ear or along the course of the vagus nerve.
- Knowledge of their common locations and imaging features may guide the radiologist to accurately diagnose these tumors.
- These highly vascularized masses demonstrate intense avidity for intravenous contrast on both CT and MR, and often exhibit a characteristic “salt and pepper” appearance on the latter.
- When the radiologist is able to recognize the main PGLs imaging aspects, proper management decisions can be taken.

Main References

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