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Paragangliomas of the Head and Neck: A Pictorial Essay

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Purpose

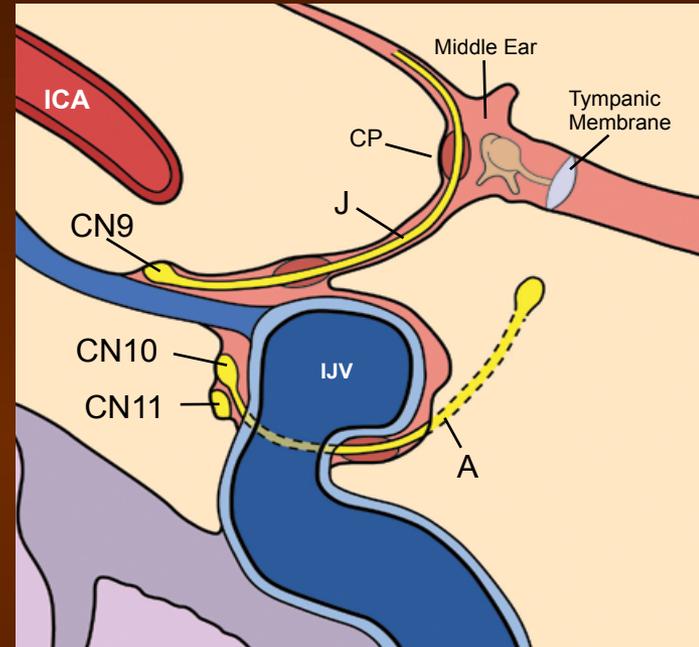
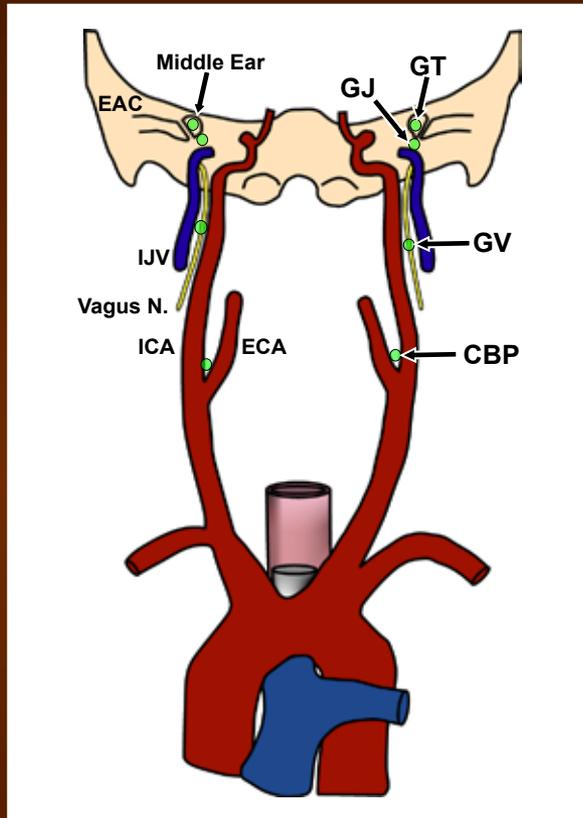
Learn the common locations of paragangliomas of the head and neck and where they originate. Learn the common imaging findings of paragangliomas utilizing CT, MRI, and angiography.

Introduction

Paragangliomas of the head and neck originate most commonly from the paraganglia within the carotid body, vagal nerve, middle ear, and jugular foramen. Also called glomus tumors, they arise from paraganglion cells of neuroectodermal origin frequently located near nerves and vessels. The function of most paraganglia in the head and neck is obscure; one exception is the carotid body, which is a chemoreceptor.

Paragangliomas account for 0.6% of all neoplasms in the head and neck region, and about 80% of all paragangliomas are either carotid body tumors or glomus jugulare tumors. The classic manifestation of a carotid body tumor is a nontender, enlarging lateral neck mass which is mobile, pulsatile, and associated with a bruit. The jugulare and tympanicum tumors commonly cause pulsatile tinnitus and hearing loss and may cause cranial nerve compression. Vagal paragangliomas are the least common and present as a painless neck mass which may result in dysphagia and hoarseness.

Common Locations of H & N



- GT** = Glomus Tympanicum;
- GP** = Glomus Jugulare;
- GV** = Glomus Vagale;
- CBP** = Carotid Body Paraganglioma

Diagram of the jugular fossa adjacent to the middle ear. Jacobson nerve (**J**), a branch of the glossopharyngeal nerve. Arnold nerve (**A**), a branch of the vagus nerve. Glomus tympanicum occur along Jacobson nerve in the middle ear adjacent to the cochlear promontory (**CP**). Glomus jugulare occur along Jacobson or Arnold nerves within the jugular fossa.

Glomus Tympanicum

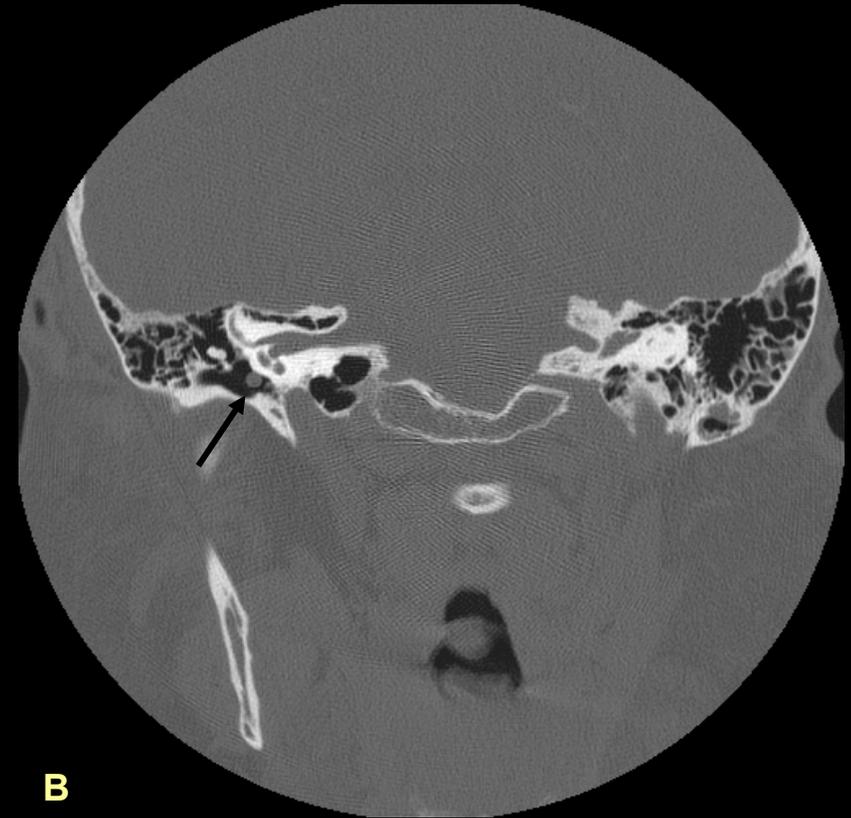
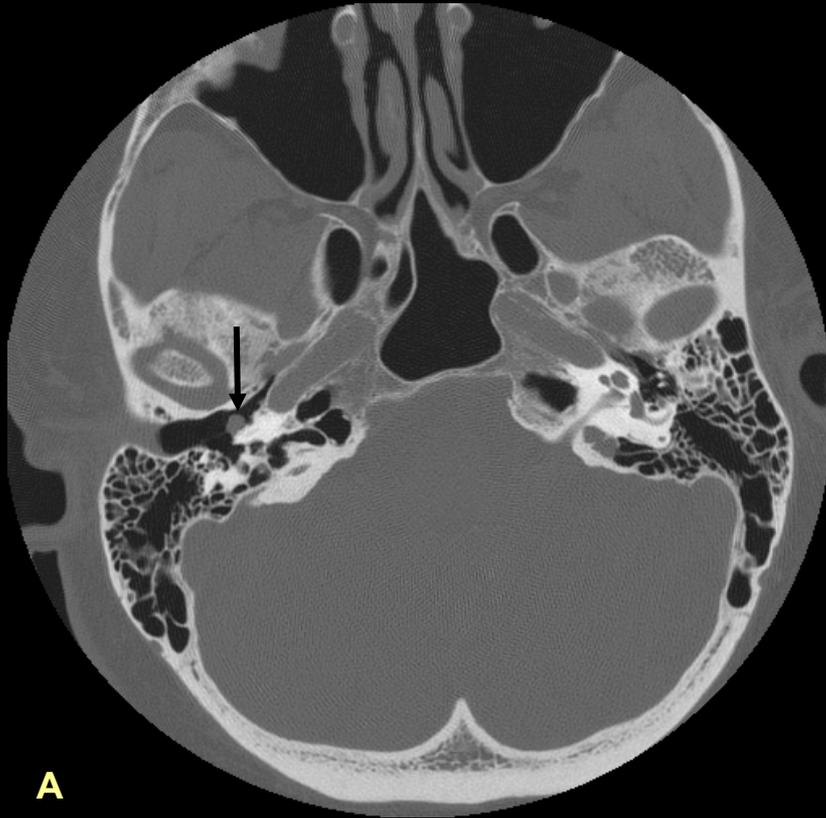
Key Points

- Most common tumor of the middle ear.
- Mass arising from the middle ear and NOT involving the jugular foramen.
- Benign tumor arising from glomus bodies found along the inferior tympanic nerve (Jacobson nerve), a branch of the glossopharyngeal nerve on the cochlear promontory.
- Commonly presents in a middle aged (40-60 years of age) female with pulsatile tinnitus (90%), conductive hearing loss (50%), and facial nerve paralysis (5%) with a retrotympanic vascular mass.
- Treatment is tympanotomy for smaller lesions; mastoidectomy for larger lesions.

Glomus Tympanicum

Imaging Characteristics

- CT demonstrates a round mass with flat base on the cochlear promontory projecting into the mesotympanum.
- Larger lesion may resemble “New Jersey” on coronal image when they fill middle ear cavity.
- Focal enhancing mass on cochlear promontory.



59 year old female presents with tinnitus

- A. Thin section axial CT shows a right 3 mm soft tissue mass abutting the cochlear promontory and projecting into the middle ear cavity (arrow).
- B. Coronal reformat CT demonstrates the right middle ear mass (arrow) abutting the cochlear promontory. No adjacent erosions seen.

Carotid Body Paraganglioma

Key Points

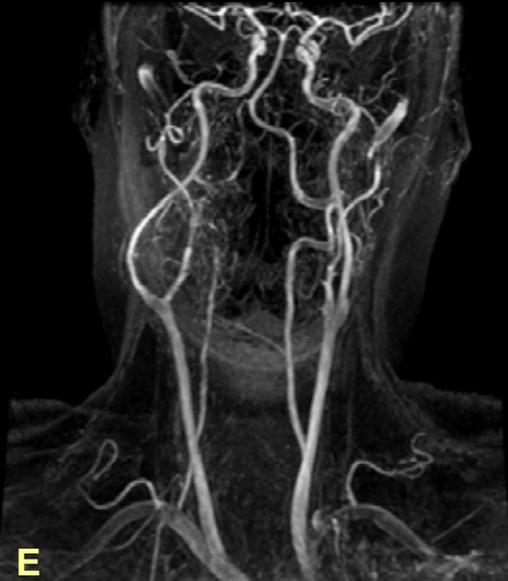
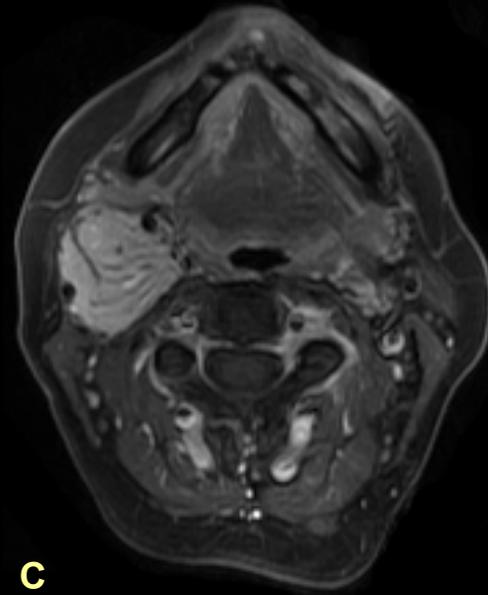
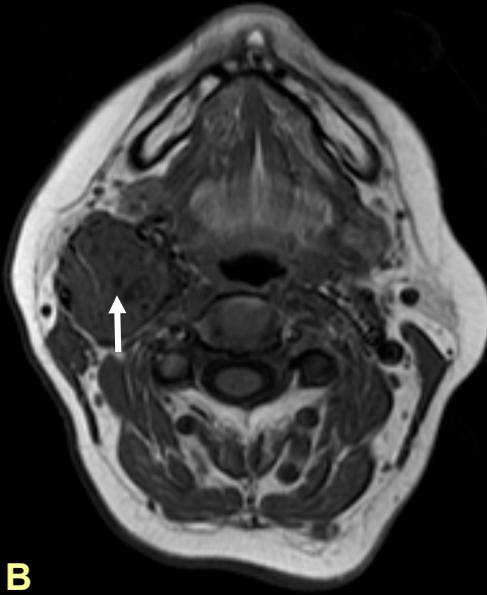
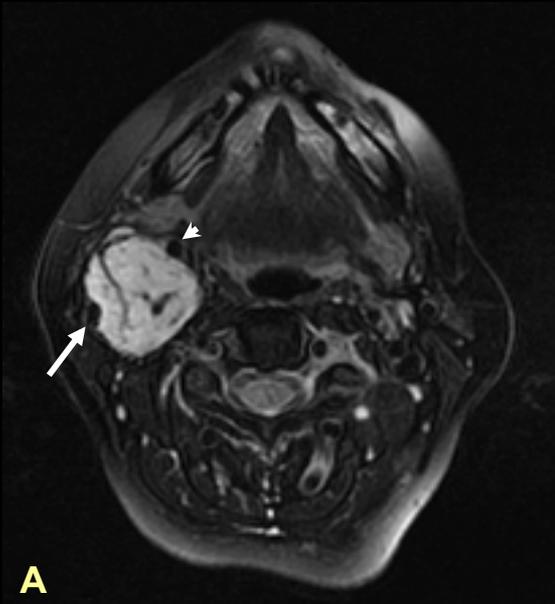
- Most common location for head and neck paragangliomas (60-67%)
- Benign vascular tumor arising in glomus bodies in the carotid body found between ECA and ICA at carotid bifurcation.
- Most common in the 4th and 5th decade. Pulsatile, painless mass at the angle of the mandible.
- Catecholamine-secreting carotid body paraganglioma is rare.
- Treatment is surgical removal.
- Multifocal paragangliomas: may occur with glomus jugulare or vagale paragangliomas.
- Radiologist must look for multiplicity. Look for a 2nd lesion.

Carotid Body Paraganglioma

Imaging Characteristics

- Vascular mass splaying the ICA posterolaterally and ECA anteromedially extending from the carotid artery bifurcation cephalad.
- Intense enhancement. Larger high velocity flow voids still visualized.
- T1WI Salt and pepper appearance: “Salt” appearance secondary to subacute hemorrhage. “Pepper” appearance due to flow voids.
- T2WI hyperintense with flow voids.
- Angiography: Prolonged, intense tumor blush between ICA and ECA. Main feeding branch is ascending pharyngeal artery, a branch of the ECA.

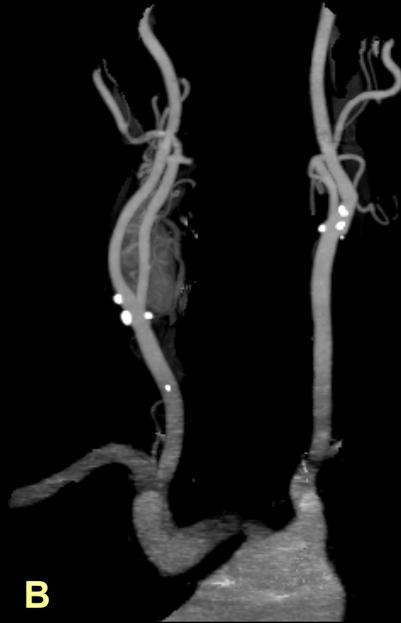
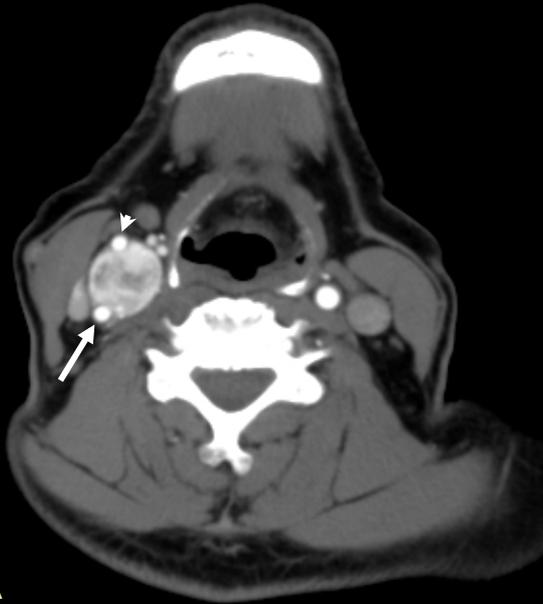
Carotid Body Paraganglioma



54 year old female with a history of right neck mass.

- A. Axial T2WI shows hyperintense mass with flow voids situated between the ICA (arrow) and ECA (arrowhead).
- B. Axial T1WI shows low signal isointense mass with “pepper” (arrow) due to flow voids.
- C. Axial T1WI post gadolinium demonstrates avid enhancement of the right carotid body mass.
- D. Coronal T1WI post gad demonstrates the enhancing mass situated at the carotid bifurcation deviating the ICA posteriolaterally (arrow).
- E. Coronal 3D time of flight SPGR demonstrates splaying of the right ICA and ECA at the bifurcation.

Carotid Body Paraganglioma



65 year old male with clinical suspicion of right carotid stenosis

- A. Axial CECT demonstrates an avidly enhancing mass situated between the ICA (arrow) and ECA (arrowhead).
- B. Projection image shows patency of the right ICA and ECA with the mass situated at the bifurcation.
- C. Coronal oblique MIP shows the mass splaying the ICA (arrow) and ECA (arrowhead).
- D. Volume rendering better depicts the right carotid body mass with splaying of the ICA and ECA.

Glomus Vagale

Key Points

- Rarest of major head and neck paragangliomas (2.5%).
- Benign vascular tumor from glomus bodies associated with nodose ganglia of vagus nerve.
- Painless, pulsatile posterolateral pharyngeal mass. Vagal neuropathy, vocal cord paralysis with hoarseness, horner syndrome possible.

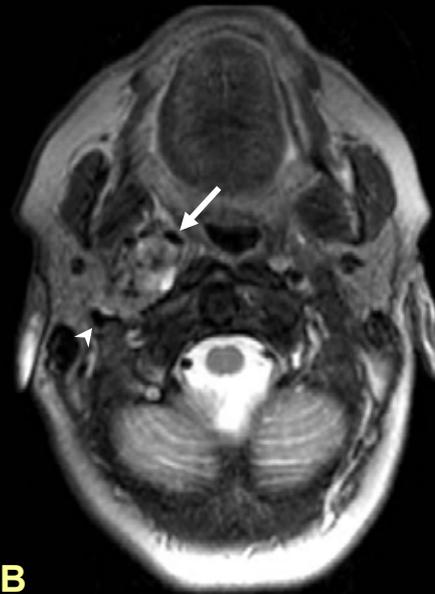
Glomus Vagale

Imaging Characteristics

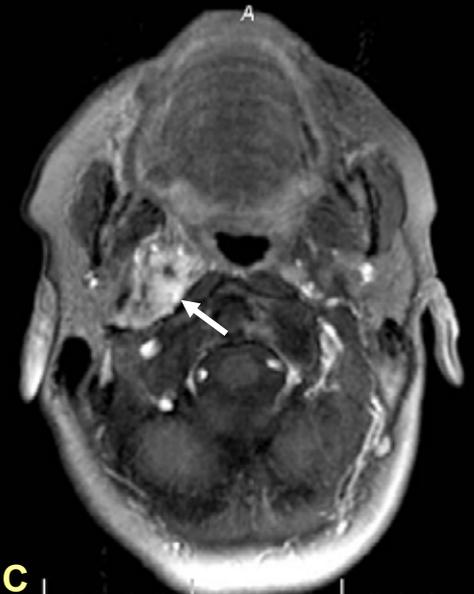
- Avidly-enhancing ovoid carotid space mass.
- Anteromedial displacement of ICA and posteriolateral displacement of IJV. No widening of the carotid bifurcation.
- MRI imaging findings similar to carotid body paraganglioma.



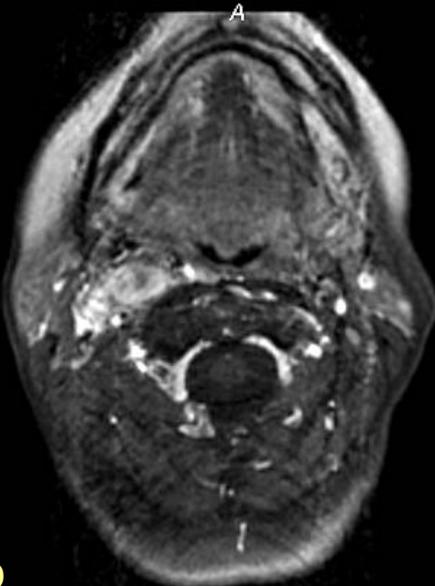
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B



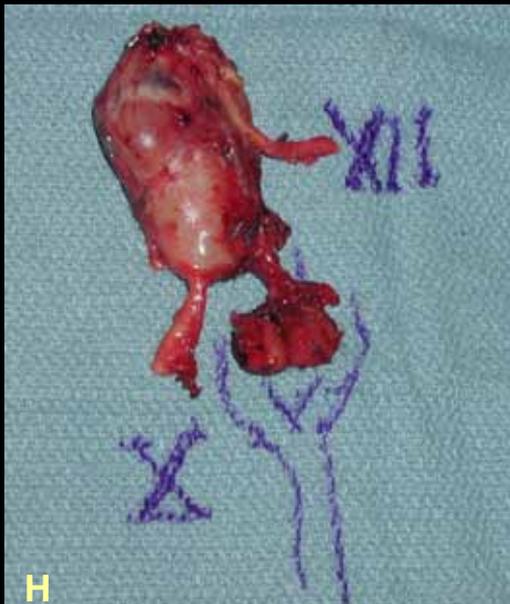
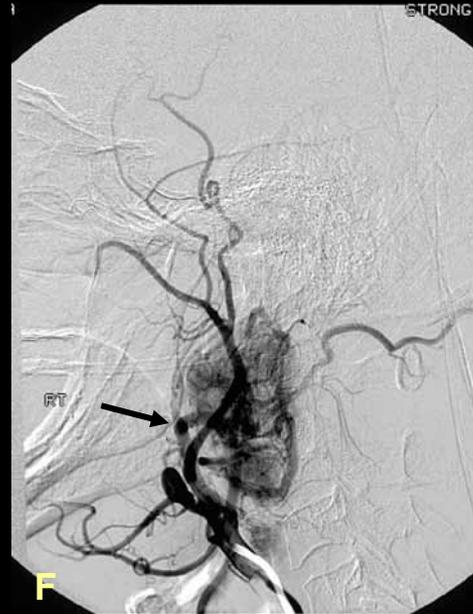
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D

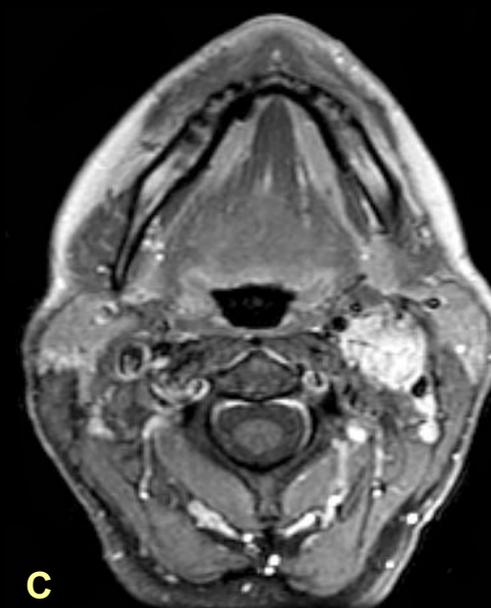
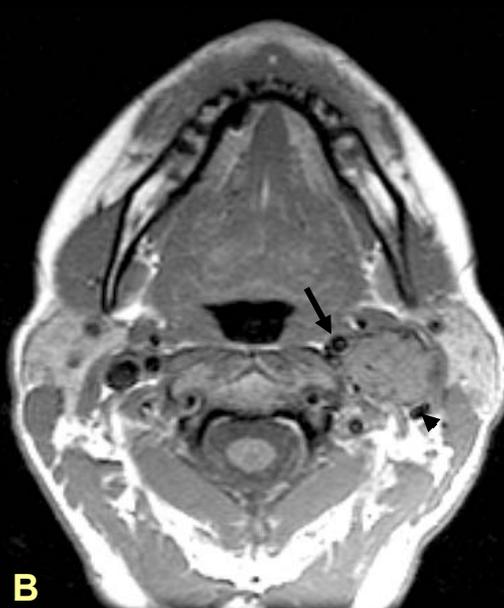
54 year old female with dysphasia

- A. Axial T2WI demonstrates a heterogeneously hyperintense mass in the right carotid space (arrow). Also note deviation of the right pharyngeal space medially (arrowhead).
- B. Axial T1WI demonstrates the heterogeneous mass situated in the right carotid space at the skull base displacing the right internal carotid artery anteriomedially (arrow) and the right jugular vein posterolaterally (arrowhead). The mass has a “pepper” appearance due to the flow voids.
- C. Axial T1WI post gadolinium demonstrates avid enhancement of the right carotid space mass (arrow).
- D. Axial T1WI post gadolinium at a lower level shows the enhancing mass.



54 year old female with dysphasia (continue)

- E. Conventional angiogram of the right common carotid demonstrates a hypervascular mass (arrow) just above the bifurcation with displacement of the ICA anteromedially (arrowhead).
- F. Selective right external carotid artery angiogram reveals an enlarged posterior auricular artery (arrow) mainly supplying this vascular mass.
- G. Post embolization angiogram of the right common carotid demonstrates significant reduction in the vascularity of this mass.
- H. Gross image of the surgically removed glomus vagale which was adherent to the 10th and 12th cranial nerves.



44 year old female with enlarging left neck mass

- A. Axial T2WI demonstrates a heterogeneous mass situated in the left carotid space at the skull base (arrow).
- B. Axial T1WI demonstrates intermediate signal intensity of the left carotid space mass displacing the left internal carotid artery anteromedially (arrow) and the left jugular vein posterolaterally (arrowhead).
- C. Axial T1WI post gadolinium demonstrates avid enhancement of the left carotid space mass.
- D. Coronal MRA demonstrates the left carotid space mass with displacement of the left ICA medially.

Glomus Jugulotympanicum

Key Points

- Glomus Jugulotympanicum describes a paraganglioma involving both the jugular foramen and middle ear cavity.
- Jugular foramen mass extends superiolaterally into the floor of the middle ear cavity.
- Tumor spread vector is superiolateral.

Glomus Jugulotympanicum

Imaging Characteristics

- Bone CT demonstrates mass in the jugular foramen with “permeative-destructive” changes along the superolateral margin of the jugular foramen. Mass invading the adjacent middle ear.
- MRI imaging findings similar to glomus jugulare.

Glomus Jugulotympanicum



55 year old female with known right carotid space paraganglioma. 2nd lesion.

- A. Thin section axial CT of the left temporal bone demonstrates a soft tissue mass (white arrow) within the middle ear cavity abutting the tympanic membrane. There are surrounding erosions of the petrous bone (black arrow).
- B. Coronal reformat CT shows expansion of the left jugular foramen with erosions of the petrous bone (arrow).
- C. Axial T2WI shows an intermediate signal mass (arrow) in the left middle ear which correlates with the findings on the axial CT.
- D. Axial T2WI shows a heterogeneous mass (arrow) arising from the left jugular foramen and extending superiolaterally into the left middle ear.

Glomus Jugulotympanicum



55 year old female with known right carotid space paraganglioma. 2nd lesion. (cont.)

- E. Axial T1WI post gadolinium demonstrates enhancement of the left jugular foramen mass (arrow).
- F. Coronal T1WI post gadolinium shows the enhancing mass (arrow) within the left jugular foramen.
- G. Conventional angiogram of the left common carotid artery demonstrates a blush (arrow) representing the vascular glomus jugulotympanicum in the region of the left middle ear/petrous temporal bone.

Glomus Jugulare

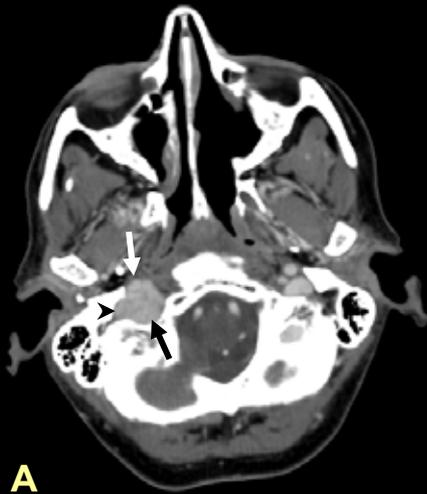
Key Points

- Considered 2nd most common head and neck paraganglioma.
- Mass arising from the jugular foramen and NOT involving the middle ear.
- Arising in the jugular foramen from the tympanic branch (Jacobson nerve) of the glossopharyngeal nerve or the auricular branch (Arnold nerve) of the vagus nerve.
- Commonly presents in a middle aged (40-60 years of age) female with pulsatile tinnitus and retrotympanic vascular mass.
- Cranial neuropathy involving 9, 10 and 11th cranial nerves.

Glomus Jugulare

Imaging Characteristics

- Bone CT demonstrates a mass in the jugular foramen with “permeative-destructive” changes of adjacent bone.
- T1WI greater than 2 cm demonstrates characteristic “salt and pepper” appearance.
- T2WI shows mixed hyperintense mass with flow voids.
- Intense enhancement.



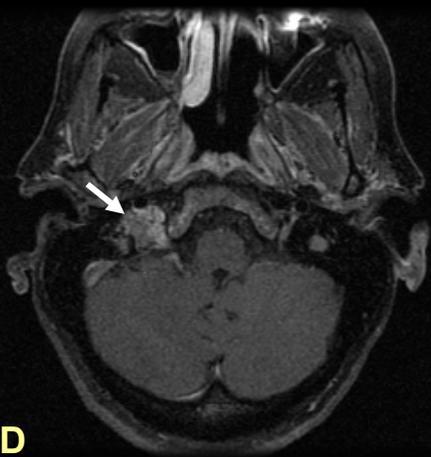
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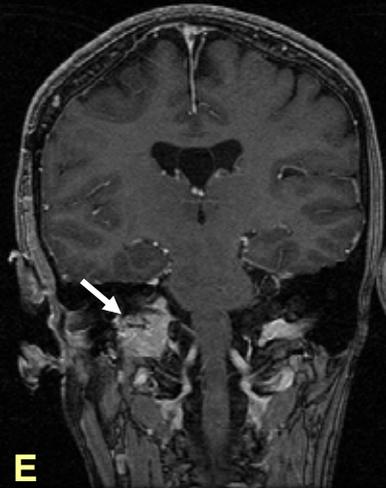
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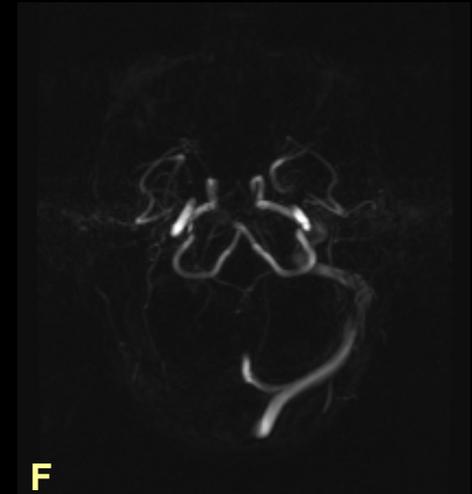
C



D



E



F

55 year old woman who presents with pulsatile tinnitus in the right ear.

- A. Axial CECT shows enhancing mass (black arrow) within the right jugular foramen which expands and erodes the adjacent petrous bone. Adjacent right jugular vein compressed laterally (arrowhead). Right carotid artery intact (white arrow).
- B. Coronal reformat NECT shows the mass (arrow) expanding the right jugular foramen when compared to the left.

- C. Axial T2WI shows a heterogenous isotense mass (arrow) within the right jugular foramen.
- D-E. Axial and coronal gadolinium enhanced T1WI shows the enhancing mass (arrow) in the right jugular foramen.
- F. 2D axial phase contrast image does not demonstrate signal in the right transverse and sigmoid sinus. This represents slow flow as contrast images demonstrate flow.

Take Home Points

- Carotid body paraganglioma and glomus jugulare make up 80% of paragangliomas of the head and neck.
- MRI classic appearance of paragangliomas is “salt and pepper”. “Salt” due to hemorrhage (rare) and “pepper” due to flow voids.
- Paragangliomas are hypervascular masses therefore avidly enhancing.
- Carotid body mass splays the ICA posteriolaterally and the ECA anteromedially at the carotid bifurcation.
- Glomus vagale displaces the ICA anteromedially and the IJV posterolaterally.
- Glomus tympanicum classically found at the cochlear promontory arising from Jacobson nerve.
- Glomus jugulare arises from Jacobson nerve or Arnold nerve within the jugular foramen and not involving the middle ear.

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