Glomus Jugulare Paraganglioma

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Figure 1: (Top Left) CT demonstrates a typical permeative lucent appearance of this glomus jugulare tumor in the left petrous apex. These lesions tend to be low signal intensity on T1WI (top right) and

hyperintense with a salt-and-pepper appearance on T2WI (bottom).

BASIC DESCRIPTION

- Benign neuroendocrine tumor of neural crest origin arising near the jugular foramen

PATHOLOGY

- Arises from glomus bodies, which function as chemoreceptors
  - Located within jugular bulb, cranial nerve IX (CN-IX) tympanic branch and CN-X auricular branch
- Classically spreads through the middle ear in a superior–lateral vector
  - May involve CN-VII mastoid segment
- Arterial supply from the ascending pharyngeal artery
- Familial or sporadic
  - Associated with multiple endocrine neoplasia type 1 (MEN 1) syndrome, neurofibromatosis type 1 (NF-1), and multiple myocutaneous neuromas
  - Patients are at increased risk of thyroid malignancy
- Chief and sustentacular cells within fibromuscular stroma are characteristic microscopic features
- Neurosecretory granules on electron microscopy

CLINICAL FEATURES

- Usually afflicts middle-aged and older adults (40–60 years old)
- Female gender predilection (male/female ratio, 1:4)
- Common presenting signs/symptoms
  - Pulsatile tinnitus
  - Cranial neuropathy involving CN-IX to CN-XII
- Treatment: surgical resection and radiation; radiosurgery; ±presurgical tumor embolization

IMAGING FEATURES

- General
  - Lobulated solid mass of variable size; often large at presentation
  - Hallmark “salt-and-pepper” MRI appearance
    - T1 hyperintense “salt” due to subacute hemorrhage; T1 hypointense “pepper” due to arterial flow voids (more commonly seen in larger tumors)
    - Adjacent bony changes: permeative-destructive
  - Involvement of middle ear common; might invade jugular vein or sigmoid sinus
- CT
  - Soft tissue mass centered near the jugular foramen
  - Avid enhancement on contrast-enhanced CT
  - ±Adjacent permeative-destructive bony changes
- MRI
  - T1WI: hyperintense “salt” due to subacute hemorrhage, hypointense “pepper” due to arterial flow voids (more commonly seen in larger tumors)
  - T2WI: hyperintense, hypointense flow voids (“pepper”)
  - DWI: hyperintense signal that might represent “T2 shinethrough,” hypercellularity, or increased density of axons
  - T1WI+C: avid enhancement
  - MRV: might show jugular vein and/or sigmoid sinus involvement/occlusion
- PET/CT
  - Avid fluorodeoxyglucose (FDG) uptake, which can be useful in metastatic evaluation or evaluating treatment response

IMAGING RECOMMENDATIONS

- MRI without and with intravenous contrast, temporal bone CT to evaluate for adjacent bony changes; consider MRV and PET/CT

For more information, please see the corresponding chapter in Radiopaedia.

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