Glomus Vagale Paraganglioma

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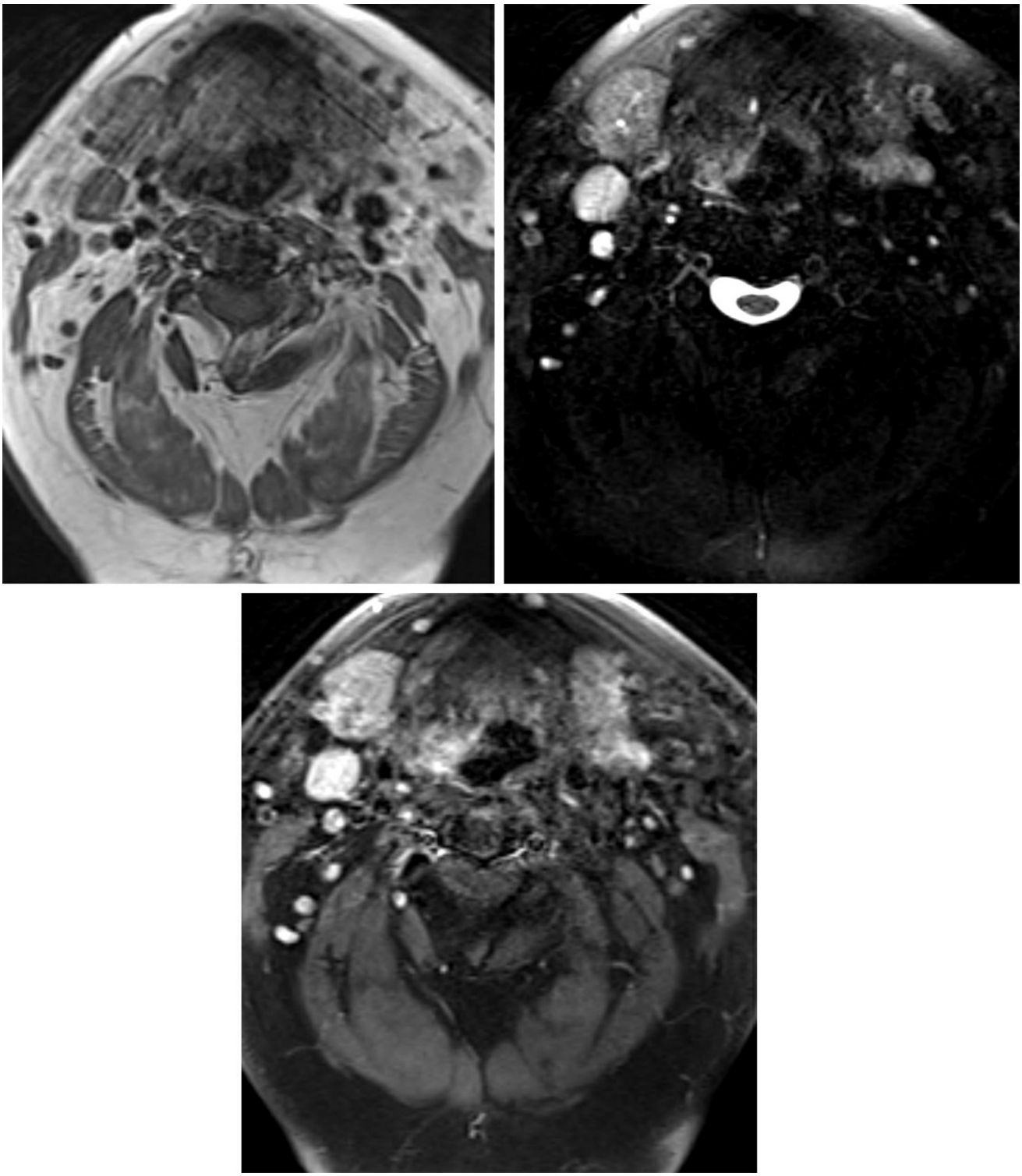

Figure 1: (Top Left) T1WI demonstrates a low-signal-intensity mass in the typical location of the vagus nerve along the carotid space. The lesion can be easily mistaken for a lymph node. As with other paragangliomas, these

lesions are typically hyperintense with salt-and-pepper vascular flow voids on T2WI (top right) and avidly enhancing on postcontrast T1WI (bottom).

BASIC DESCRIPTION

- Benign, hypervascular neuroendocrine tumor of neural crest origin
- Less common than glomus caroticum (carotid body tumor) and glomus jugulare

PATHOLOGY

- Arises from glomus bodies within cranial nerve X (CN X) nodose ganglion
- Composed of chemoreceptor cells of neural crest origin
- Arterial supply from the ascending pharyngeal artery
- Familial or sporadic
    - Associated with neurofibromatosis type 1 (NF-1), multiple endocrine neoplasia type 2 (MEN-2), and von Hippel-Lindau (VHL) disease, and multiple paraganglioma syndromes
    - Medullary thyroid carcinoma, adrenal pheochromocytomas, multiple paragangliomas, and renal and pancreatic tumors
- Chief cells rests (zellballen) within fibromuscular stroma are characteristic microscopic features
- Neurosecretory granules on electron microscopy

CLINICAL FEATURES

- Usually afflicts middle-aged adults (40–50 years old); younger at presentation if familial
- Female gender predilection
- Common presenting signs/symptoms
    - Pulsatile, painless lateral neck mass
    - CNs IV to XII neuropathy (CN X most commonly); vocal cord

paralysis, hoarseness

- Treatment: surgical resection versus observation; high surgical morbidity with loss of CN X function (vocal cord paralysis)
    - If bilateral, only one side is resected
- Prognosis: must outweigh risks and benefits of surgery; progressive CN X neuropathy if untreated; rare malignant potential

IMAGING FEATURES

- General
    - Lobulated, enhancing mass centered within the nasopharyngeal/suprahyoid carotid space ~2 cm below the jugular foramen
    - Displaces the internal carotid artery anteromedially, jugular vein posterolaterally, and parapharyngeal fat anterolaterally
        - No splaying of the internal carotid artery (ICA) and internal jugular vein (IJV); splaying suggests carotid body paraganglioma
    - Single or multiple
    - Variable size
    - Right-sided position more common than left
    - Hallmark “salt-and-pepper” MRI appearance
        - T1WI hyperintense “salt” due to subacute hemorrhage, hypointense “pepper” due to arterial flow voids (more commonly seen in larger tumors)
    - ±Adjacent permeative destruction of skull base
- CT
    - Well-marginated soft-tissue mass centered within the suprahyoid carotid space ~2 cm from the jugular foramen
    - Avid enhancement on contrast-enhanced CT
    - ±Adjacent permeative-destructive bony changes
- MRI
    - T1WI: heterogeneous signal, ±hyperintense areas of subacute hemorrhage (“salt”) is an uncommon finding, hypointense flow

voids (“pepper”)
    - T2WI: heterogeneously hyperintense, hypointense flow-voids
    - T1WI+C: avid early enhancement
    - MRA: anteromedial displacement of internal carotid artery

IMAGING RECOMMENDATIONS

- MRI without and with intravenous contrast from base of skull to carotid bifurcation; ±CT to evaluate for adjacent bony changes
- Evaluate for multiple tumors
- Imaging tumor surveillance if familial

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

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