Carotid Body Glomus Tumor (Glomus Caroticum; Carotid Body Paraganglioma)

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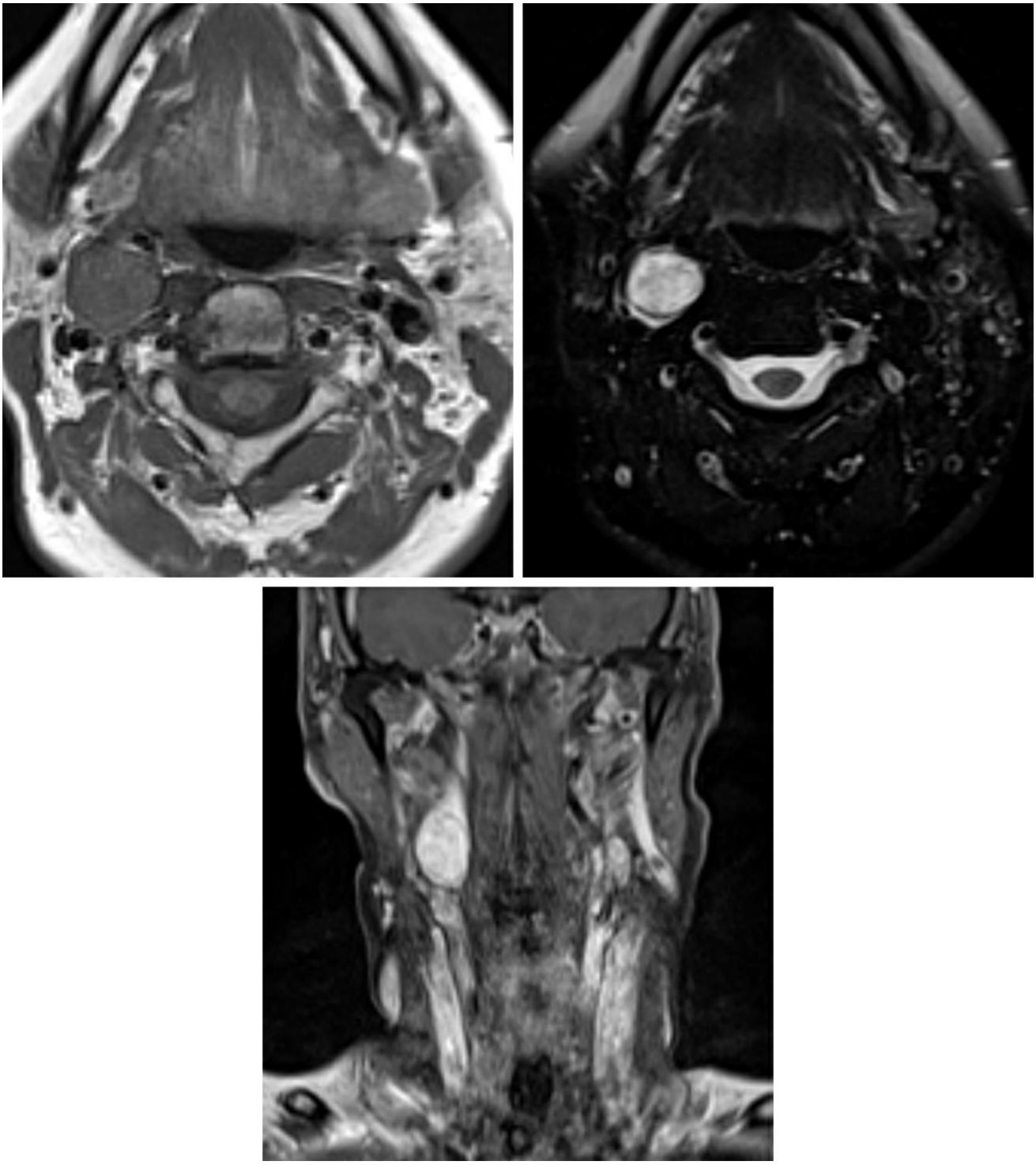

Figure 1: (Top Left) The low-signal-intensity mass splaying the right internal and external carotid artery on T1WI is in a typical location for

carotid body paraganglioma. (Top Right) This lesion tends to be hyperintense on T2WI containing low-signal-intensity flow voids due to its high vascularity. (Bottom) The lesion enhances avidly on postcontrast imaging due to this feature of hypervascularity as well.

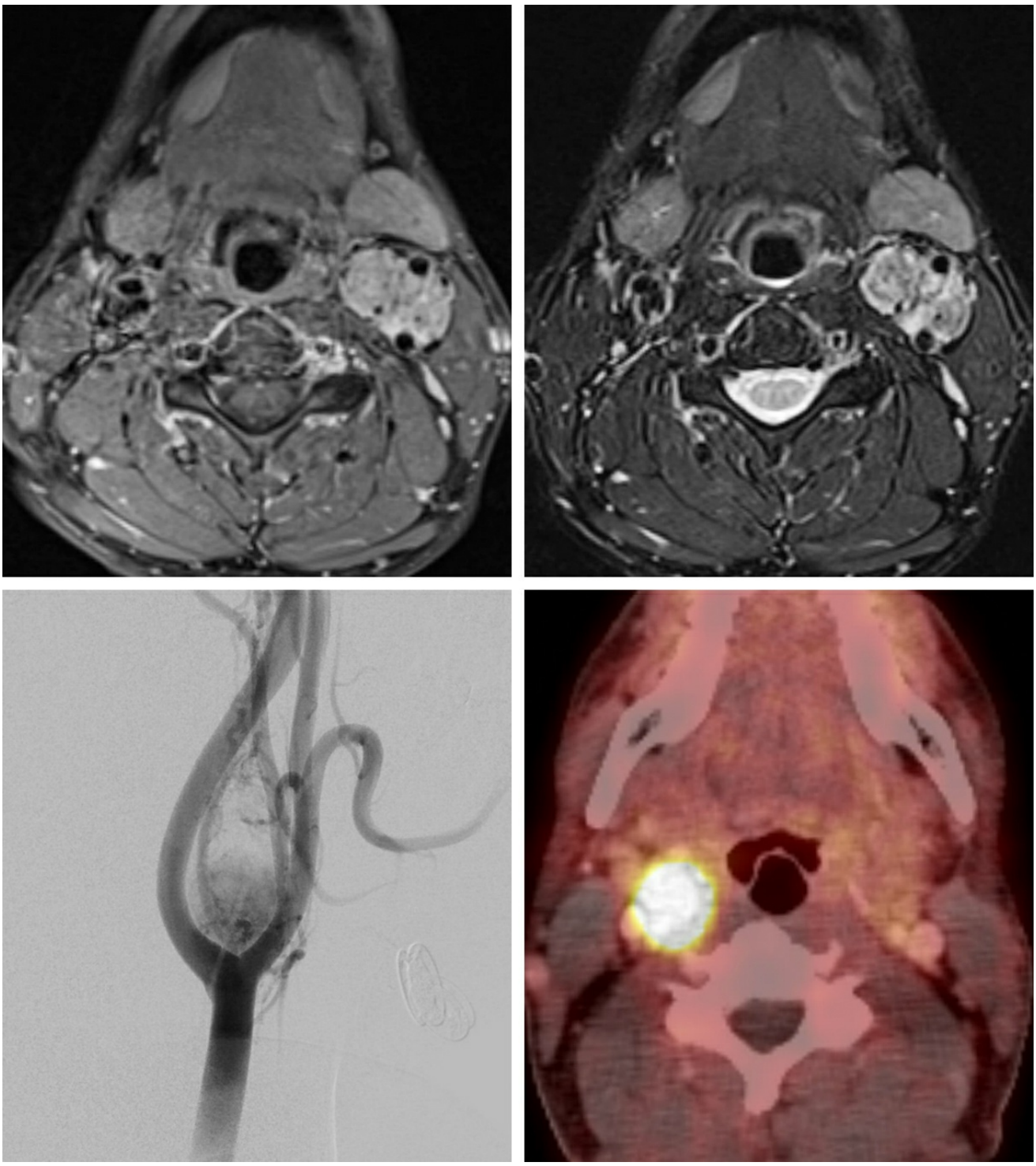

Figure 2: T1FS postcontrast (top left) and T2FS (top right) images demonstrate an avidly enhancing mass with associated internal flow voids splaying the left internal and external carotid arteries in a patient with multiple paragangliomas. Digital subtraction angiography (bottom left) performed before embolization demonstrates a hypervascular mass

splaying the left internal and external carotid arteries. After embolization, a whole-body ^{68}Ga-DOTANOC PET/CT (bottom right) was performed to assess for additional sites of disease. ^{68}Ga-DOTANOC localizes to organs and tumors that express specific subtypes of somatostatin receptors (neuroendocrine tumors, carcinoid, paragangliomas, etc). The selected image demonstrates a ^{68}Ga-DOTANOC avid mass in the right carotid space compatible with an additional glomus caroticum paraganglioma.

BASIC DESCRIPTION

- Benign, hypervascular neuroendocrine tumor of neural crest origin

PATHOLOGY

- Glomus caroticum (GC) arise from glomus bodies within the carotid body at the carotid bifurcation
- Composed of chemoreceptor cells of neural crest origin
- Arterial supply from the ascending pharyngeal artery
- Sporadic >> familial
    - Associated with NF-1, MEN-2, and von Hippel-Lindau (VHL), and multiple paraganglioma syndromes
    - Medullary thyroid carcinoma, adrenal pheochromocytomas, multiple paragangliomas, renal and pancreatic tumors
    - Multiple tumors are more common in familial cases
- May also develop as a response to chronic hypoxia (chronic obstructive pulmonary disease/chronic lung disease, high altitude)
- Chief cells rests (zellballen) and sustentacular cells 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

- Slight male gender predilection
- Common presenting signs/symptoms
    - Pulsatile, painless mass at the angle of the mandible with gradual enlargement
    - CN 10 and CN 12 neuropathy
    - Hormonally active tumors (catecholamine secretion) are rare: palpitations, flushing, hypertension
- Treatment
    - Surgical resection based on Shamblin classification: tumor size and degree of contact with ICA
    - Higher classification predicts surgical morbidity (CN neuropathy)
    - ±Presurgical embolization to reduce bleeding
    - Serial imaging follow-up with smaller, asymptomatic tumors

IMAGING FEATURES

- General
    - Lobulated, enhancing mass centered within the carotid bifurcation
    - Splays the internal and external carotid arteries
        - Internal carotid artery displaced posterolaterally
        - External carotid artery displaced anteromedially
        - Jugular vein displaced posteriorly
    - Single or multiple tumors
    - Variable size
    - Hallmark “salt-and-pepper” magnetic resonance imaging (MRI) appearance
        - T1 hyperintense “salt” due to subacute hemorrhage, hypointense “pepper” due to arterial flow voids (more commonly seen in larger tumors)
- CT

    - Well-marginated soft tissue mass centered within carotid bifurcation
    - Avid enhancement on contrast-enhanced CT
- MRI
    - T1WI: heterogenous 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: internal carotid artery (ICA)-external carotid artery (ECA) splaying
- Nuclear Medicine
    - ^{123}I-MIBG: radiopharmaceutical localizes to catecholamine producing tumors. Sensitivity for paraganglioma 57%–78%
    - ^{111}I-Ocrtreotide: radiopharmaceutical localizes to tumors and tissue expressing somatostatin receptors. Sensitivity for paraganglioma 94%.
    - ^{68}Ga-DOTANOC and ^{68}Ga-DOTATATE positron emission tomography (PET)/CT: emerging diagnostic PET/CT agent used for detection of somatostatin expressing tumors (neuroendocrine tumors, paraganglioma, etc).

IMAGING RECOMMENDATIONS

- Contrast-enhanced CT or MRI without and with intravenous contrast with catheter angiography
- Evaluate for multiple tumors
- Imaging tumor surveillance if familial

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

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