Carotid Paraganglioma

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BACKGROUND

- Paraganglion tissue 1st described by Von Haller in 1743
- Tissue contains catecholamine + tryptophan granules
- Histologically and functionally = adrenal medulla
- Located at:
  - carotid, aorticopulmonary, para-aortic and coccygeal bodies
  - urinary bladder, gall bladder, heart
- 3 tissue components
  - 1. Type I/chief cells (catecholamine granules)
  - 2. Type II/sustentacular cells
  - 3. Capillary network
BACKGROUND

• Tumors of this tissue are called paragangliomas
• Nomenclature has been confusing!
  – glomus tumors, chemodectomas, non chromaffin tumors, chromaffin tumors…
  – based on staining, function and adjacent structures
• Glenner and Grimely in 1974 standardized naming based on adrenal vs. extra-adrenal sites
  – now there are pheochromocytomas of the adrenal and paragangliomas, further specified by site
  • note: many authors still use the original terminology
BACKGROUND

- 10% multicentric
- 7-9% familial
- 2-15% malignant
- 90% are adrenal pheochromocytomas
- 10% extra-adrenal:
  - 85% in abdomen
  - 12% in thorax
  - 3% in head and neck
    1. Carotid body
    2. Jugular foramen
    3. Middle ear cavity
    4. Course of vagus nerve
BACKGROUND

• Carotid body described by Von Haller in 1743
  – Medial to carotid bifurcation (5x3x1.5mm)
  – Functions as baroreceptor, \( O_2 \), pH receptors

• Carotid paragangliomas soft, non-tender, slowly enlarging neck mass
  – possible dysphagia, hoarseness, tongue parasthesia
  – 40-60 y/o Male=Female
  – Saldana et al noted increased incidence with COPD patients and people living at high altitudes…chemoreceptor hyperplasia
PATIENT

HPI: *The patient is a 49 y/o man presenting with complaint of non-painful, left sided, neck swelling of approximately 5 years. He believes it is increasing in size. No other symptoms.*

PMH: heroin abuse with CVA secondary to OD

MEDS: ASA 325mg PO qd

Serax PO qhs

NKDA

PE: revealed soft, large, left sided neck mass with no other focal findings
CT IMAGING OF PARAGANGLIOMAS

- 2.5-3.0 mm axial slices from thoracic inlet to skull base
- Excellent for imaging bony detail
- Win et al. describes “… a well marginated ovoid mass… which splays the internal and external carotid arteries at the level of the bifurcation and demonstrates intense homogenous enhancement following the intravenous administration of iodinated contrast.”
- May appear similar to schwannoma but can differ using dynamic bolus CT or MRI
- Must consider ionizing radiation and contrast
CT WITH CONTRAST

MANDIBLE

MYLOHYOID MUSCLE

EXTERNAL CAROTID A.

INTERNAL CAROTID A.

INTERNAL JUGULAR V.

EXTERNAL JUGULAR V.

5cm MASS

Axial CT w/ contrast courtesy of Mike Stella, MD BIDMC
CT FINDINGS

Axial CT + contrast

Axial CT + contrast

Axial CT bone window


http://brighamrad.harvard.edu/education/online/tcd/tcd.html

CAROTID PARAGANGLIOMA
METASTASIS TO BONE
3D CT RECONSTRUCTION

MRI IMAGING OF PARAGANGLIOMAS

• Allows imaging of lesion, surrounding nerves and vessels without ionizing radiation
• Allows multiplanar imaging without repositioning patient
• Not as good as CT at imaging bone and ear structures
• T1 imaging shows a lesion intensity \( \geq \) than muscle and \( > \) than muscle on T2 and following gadolinium
• Punctate, serpentine or channel-like, hypointense flow voids should be noted… creating a “salt and pepper appearance”
• Mass effect is noted
• Time-of-flight magnetized bolus of blood allows clear imaging of vessels for brief periods of time
SPLAYING OF EXTERNAL AND INTERNAL CAROTID A.A.
TRACHEAL DEVIATION TO THE RIGHT
“SALT AND PEPPER” TUMOR APPEARANCE
CORONAL T1 MRI

NOTE: TUMOR IS HYPERINTENSE COMPARED TO MUSCLE

TUMOR

CAROTID BIFURCATION

COMMON CAROTID A.
T2 WEIGHTED MRI SHOWING HYPERINTENSE LESIONS


http://brighamrad.harvard.edu/education/online/tcd/tcd.html
MRA vs. CONVENTIONAL ANGIOGRAPHY

• In MRA no contrast is given. Magnetized blood can be visualized only temporarily. Only rapidly filling vessels will be imaged and many tumor “feeders” will not be seen. Thus the lack of tumor “blush” is normal on MRA. Large vessels will be imaged showing mass effect.

• Conventional angiography will show the tumor “blush” and is important if embolization is to be attempted pre-op to minimize intra-op bleeding.
NORMAL ANATOMY

INTERNAL CAROTID A.

EXTERNAL CAROTID A.

VERTEBRAL A.

CAROTID BIFURCATION

MRA courtesy of Mike Stella, MD BIDMC

http://www.bartleby.com/107/
2D MRA

NORMAL

ABNORMAL

ABNORMAL

SPLAYING OF INTERNAL AND EXTERNAL CAROTID A.A.

MRA courtesy of Mike Stella, MD BIDMC
3D MRA

NORMAL

ABNORMAL

SPLAYING OF INTERNAL AND EXTERNAL CAROTID A.A.

3D MRA courtesy of Mike Stella, MD BIDMC
NOTE “TUMOR BLUSH” AND SPLAYING OF INTERNAL AND EXTERNAL CAROTID A.A.
UTRASOUND

• Gray scale ultrasound is used to delineate tumor size, margins and location
• Typically a well-defined, hypoechoic heterogeneous, mass is noted splaying the ICA and ECA
• Hypervascularity is noted on color doppler ultrasound
GRAY SCALE ULTRASOUND

HYPOECHOIC
WELL DEFINED MASS

Left sagittal ultrasound courtesy of Mike Stella, MD BIDMC
COLOR DOPPLER ULTRASOUND

HYPERVASCULAR MASS SPLAYING INTERNAL AND EXTERNAL CAROTID A.A.

Left transverse color doppler ultrasound courtesy of Mike Stella, MD BIDMC
RADIONUCLIDE IMAGING

- Octreotide is labeled with $^{111}$Indium-labeled-DTPA (pentetreotide)
- Bind somatostatin type 2 receptors common to paragangliomas
- advocated if suspect multicentricity in familial disease or to image postoperatively
PENTETREOTIDE SCINTIGRAPHY

33 y/o with familial h/o paragangliomas. Presented with B neck masses...B paragangliomas noted

74 y/o with hoarseness thought to be hemangioma...showing L neck paraganglioma

DIFFERENTIAL DIAGNOSIS CAROTID SPACE MASS

- **Inflammatory**
  - abscess
- **Pseudotumor**
  - carotid artery ectasia
- **Vascular**
  - ICA dissection
  - carotid aneurysm/thrombosis
  - jugular vein thrombosis
- **Benign tumor**
  - carotid body, jugular, vagal paraganglioma
  - schwannoma
- **Malignant tumor**
  - squamous cell carcinoma
  - NHL

Abscesses appear as homogenous fluid filled lesions/no hypervascularity

Ectasia would be seen on MRA and angiography

All the vascular lesions would be elucidated on MRA and angiography

Malignancy is always possible...here there is a circumscribed, non-invasive appearance coupled with slow growth on Hx

Schwannomas have a hyperintense appearance on CT but do not exhibit “salt and pepper” appearance on MRI

Paraganglioma is most likely given MRI appearance and vascular “blush”...the lesion is isolated to the carotid bifurcation
TREATMENT

- External embolization of the tumor preoperatively with 2mm microcoils
- Excision of left carotid body tumor with interposition nonreverse vein graft from common carotid to the internal carotid artery
REFERENCES

- Jeroen C. Jansen, MD, Robert J. Baatenburg de Jong, MD, PhD, Jaap Schipper, MD, PhD, Andel G. L. van der mey, MD PhD, Adrian P.G. van Gils, MD, PhD. Color Doppler Imaging of Paragangliomas in the Neck. *Journal of Clinical Ultrasound* 1997; 25(9): 481-485.
- [http://brighamrad.harvard.edu/education/online/tcd/tcd.html](http://brighamrad.harvard.edu/education/online/tcd/tcd.html)
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