Carotid Body Tumors

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Our Patient

- AD is a 48 year old male with 7–8 year history of a left neck mass
- No previous imaging studies
- On exam:
  
  “Firm, well-circumscribed, round L submandibular mass. No tenderness to palpation, no bruit.”
- CT scheduled and ENT consulted
January 2002: Axial CT w/ Contrast

Mandible

External Carotid a.

Internal Carotid a.

Internal Jugular v.

Submandibular g.

Mass

SCM

External Jugular v.

Vertebral a.

BIDMC PACS
January 2002: Axial CT w/ Contrast

- Hypervascularity demonstrated by prompt, intense enhancement
- Located at carotid bifurcation splaying the internal and external carotids
- Airway displaced to right
- No bony destruction
- No lymphadenopathy
- Well-preserved planes
- Approximately 5cm
CT January 2002

Extends from carotid bifurcation superiorly to C1
Carotid Space Anatomy

- Internal carotid artery
- Internal jugular vein
- Sympathetic chain
- Cranial nerves IX, X, XI, XII
- Lymph nodes

http://www.bartleby.com/107/143.html
DDX Carotid Space Masses

Carotid Artery
- Ectasia
- Aneurysm
- Pseudo-aneurysm
- Dissection
- Encasement by direct spread of SCC

Jugular Vein
- Asymmetric enlargement
- Thrombosis
- Thrombo-phlebitis
- Meningioma (from jugular foramen)

Inflammatory
- Abscess

Cranial Nerves
(X, XI, XII, sympathetic chain)
- Neurogenic tumor
- Neuroblastoma
- Paraganglioma

Lymph Nodes
- Metastatic cervical adenopathy
- Lymphoma
## Narrowing the DDX

### Carotid Artery
- Ectasia
- Aneurysm
- Pseudo-aneurysm
- Dissection
- Encasement by direct spread of SCC

### Jugular Vein
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### Inflammatory
- Abscess
Narrowing the DDX con’t

Cranial Nerves
(X, XI, XII, sympathetic chain)
- Neurogenic tumor
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Lymph Nodes
- Metastatic cervical adenopathy
- Lymphoma

- Hypervascularity
- No calcification or necrosis
- No lymphadenopathy

It must be a paraganglioma – a carotid body tumor!
Paragangliomas

- Rare tumors that arise from specialized neural crest cells associated with autonomic ganglia.
- 4 extradrenal locations
  - Group I: Great vessels of chest and neck
  - Group II: Vagus nerve
  - Group III: Aorticosympathetic chain
  - Group IV: Visceral organs
Paragangliomas

Head and Neck

- Carotid body paraganglioma
- Vagal paraganglioma (nodose ganglia)
- Glomus tympanicum—middle ear along tympanic plexus
- Glomus jugulare—jugular bulb

Glenner, GG and Grimley PM. Tumors of the Extra-Adrenal Paraganglion System. 
Bethesda, MD: Armed Forces Institute of Pathology, 1974
Paragangliomas

- 1/30,000 head&neck tumors are paragangliomas
- 2–3% head/neck paragangliomas have functional hormone secretion
- Usually benign—6% CBTs reported to be malignant
- Familial form (10–25%)—present younger and with multiple tumors
Carotid Body Tumors

- CB sits in adventitia at bifurcation of common carotid a.
- Regulates respiration and maintains arterial gases (chemoreception)
- Hyperplasia seen in chronic hypoxic states – altitude, COPD, cyanotic heart disease
- Presentation: avg. age = 45, slow-growing, asymptomatic or mass-related effects, 10% present with CN palsy
Imaging Studies

- CT
- MRI/MRA
- Ultrasound
- Angiography
- Radionuclide imaging
CT

- Thin section scanning from thoracic inlet to skull base in patients with CB or vagal paragangliomas or other palpable neck mass
- Examines integrity of associated soft tissues
- Detection of multiple lesions
- 3D reconstruction visualizes associated vasculature.
CT May 2002

- No significant interval change to large enhancing mass
- Displacement of airway to right
3-D CT Reconstruction

MRI

- Aids in lesion diagnosis and localization
- Differentiates mass from surrounding inflammatory changes, fluid or vascular structures
- More sensitive for delineating encroachment and encasement of vessels
- Images middle ear structures and bony erosions
- Coronal sequences
T1 MRI w/o Contrast

- Paraganglioma
- Carotid arteries
- Trachea
MRI: T2

- Well-defined mass
- Heterogeneous hyperintensity
- Punctate flow voids.
- “Salt and pepper” pattern: due to high vascularity with associated areas of hemorrhage, slow-flowing blood, and tumor cells.
MRI: T1 Coronal

- Paraganglioma
- Carotid Bifurcation
- Common Carotid
- Aortic arch
• Splaying of internal and external carotid arteries
• No aneurysm or stenosis
MRA

- Noninvasive
- Delineates displacement of vasculature
- Demonstrates tumor vascular supply
MRA-Normal

Common Carotid a.

Internal Carotid a.

External Carotid a.

Vertebral a.

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BIDMC PACS
Ultrasound

- Delineates tumor margins, size and location
- Doppler: demonstrates hypervascularity of paragangliomas
- Surveys neck for other lesions
- Differentiates CBTs from vascular anomalies and pseudoaneurysms
- Can obtain US guided fine needle aspiration
Well-defined, hypoechoic heterogeneous mass at carotid bifurcation measuring 5.7x4.2x4.1 cm
Hypervascular mass, splaying of internal and external carotid arteries
Ultrasound-Guided Fine Needle Aspiration
Angiography

- Demonstrates the primary arterial supply and collateral vessels of tumor
- Reveals relationships with neck blood vessels
- Presurgery transcatheter arterial embolization

Radionuclide Imaging

- Pentetreotide = octreotide radiolabelled with $^{111}$ indium–DTPA binds somatostatin type 2 receptors in paragangliomas

- Uses: follow recurrent disease, locates multiple lesions, detects familial paragangliomas

Metastatic CBT with Lytic Bone Lesions

Familial Paraganglioma

- T1 weighted MRI
- T2 weighted MRI
- Angiography

Vagal Paraganglioma

A. T1: lesion posterolateral to carotids
B. T2: Salt and pepper appearance
C. Coronal T2
D. Arteriogram: tumor blush in relation to carotids, no splaying as in CBTs.

Glomus Jugulare

- Destructive lesion in skull base involving middle ear and hypoglossal canal

A. T1 w/o contrast
B, C. T1 w/ contrast

Glomus Tympanicum

- High resolution axial CT of right temporal bone

Gross Pathology

Well-defined neoplasm with a pseudocapsule

Glenner, GG and Grimley PM. Tumors of the Extra-Adrenal Paraganglion System. Bethesda, MD: Armed Forces Institute of Pathology, 1974
Histology

Our Patient:
no malignant cells

• Small nests of bland cells with centrally placed hyperchromic nuclei that form clusters called Zellballen (cell balls).

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Our Patient…

• Followed by ENT
• Will have surgical resection of CBT
References

Glenner GG and Grimley PM. Tumors of the Extra-Adrenal Paraganglion System. Bethesda, MD: Armed Forces Institute of Pathology, 1974


http://brighamrad.harvard.edu/Cases/bwh/hcache/74/full/html
http://www.bartleby.com/107/143.html


Acknowledgements

- Gwendolyn Dole, MD
- Pamela Lepkowski
- Larry Barbaras and Cara Lyn D’Amour
- Gillian Lieberman, MD