AMSER Case of the Month July 2025

14 y.o. female presenting with 2 months of painless bilateral neck masses

Arpitha Shenoy, University of Virginia School of Medicine

Dr. Tanvir Rizvi, University of Virginia Department of Radiology

Dr. Ariana Greenwell, University of Virginia Department of Otolaryngology

UVA Health School of Medicine

AMSER

Patient Presentation

HPI: She presented to an outside ED with 2 months of painless bilateral neck masses after being treated by a few courses of antibiotics without improvement at urgent care centers.

She had no inciting illness or trauma. No dysphagia, odynophagia, trismus, Horner's or compressive symptoms. She noted mild voice change and hoarseness.

Medical and Surgical History: None

Family History: No history of head/neck tumors or neuroendocrine tumors.

Physical Exam: Palpable, non-tender, firm mass in the right level II that is minimally mobile laterally. No palpable mass on the left.

Flexible laryngoscopy: Bilateral vocal folds mobile. Visualized sub-glottis unremarkable.

Pertinent Labs

- Plasma Metanephrines
  - Free Metanephrine: <0.20 (Ref Range <0.5 nmol/L)
  - Free Normetanephrine: 0.32 (Ref Range <0.90 nmol/L)
- Invitae Genetic Testing
  - Heterozygous deletion (entire coding sequence) of SDHD (succinate dehydrogenase)

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Neck mass	3149271	US neck	0 mSv O	0 mSv [ped] O	Usually appropriate
		MRI neck without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		MRI neck without IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		CT neck with IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually appropriate
		CT neck without IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	May be appropriate (Disagreement)
		Arteriography cervicocerebral	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate
		MRA neck without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		MRA neck without IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		CT neck without and with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate
		CTA neck with IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually not appropriate
		FDG-PET/MRI skull base to mid-thigh	1-10 mSv ☢☢☢		Usually not appropriate
		FDG-PET/CT skull base to mid-thigh	10-30 mSv ☢☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate

These imaging modalities were ordered by the ER physician

AMSER

Findings (unlabeled):

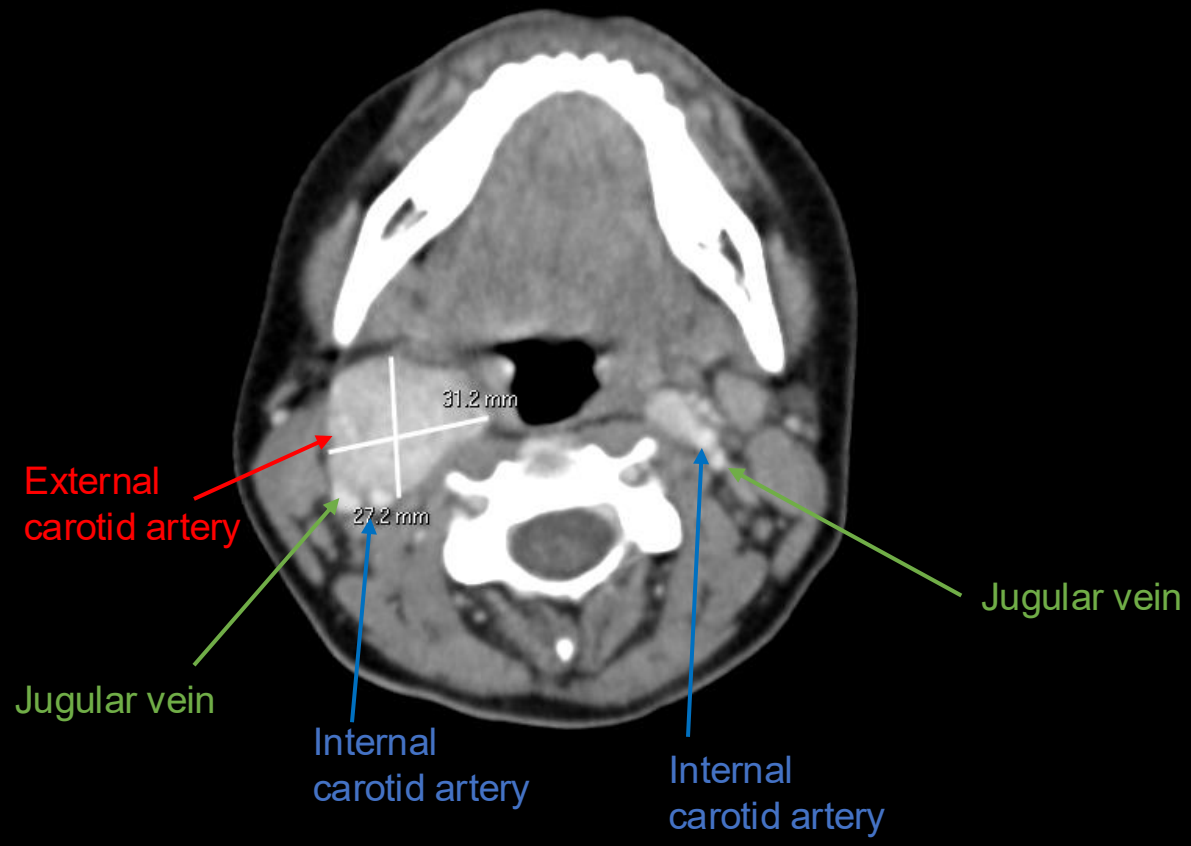
Findings (unlabeled):

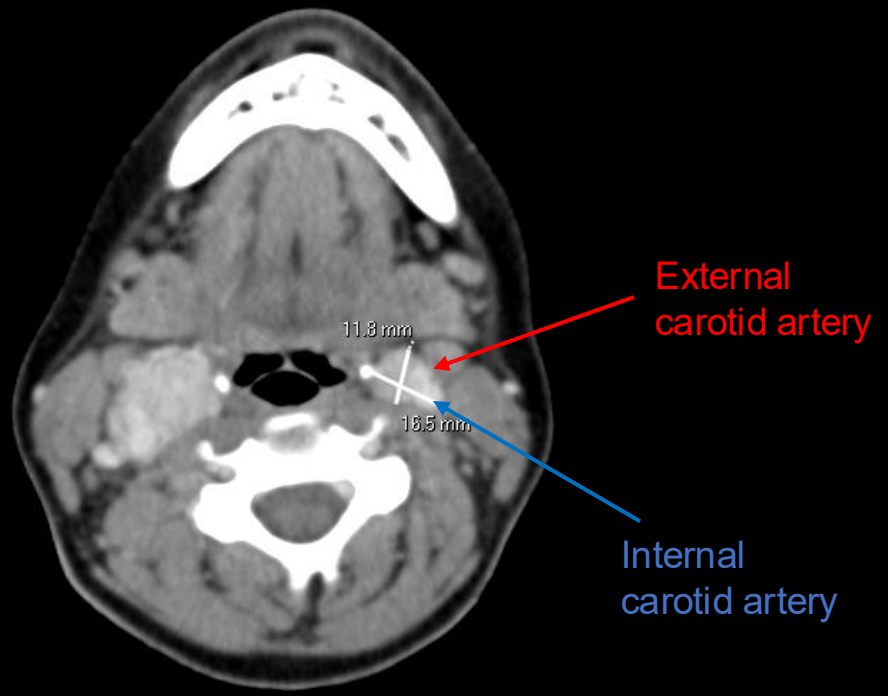
Findings (unlabeled):

R neck mass

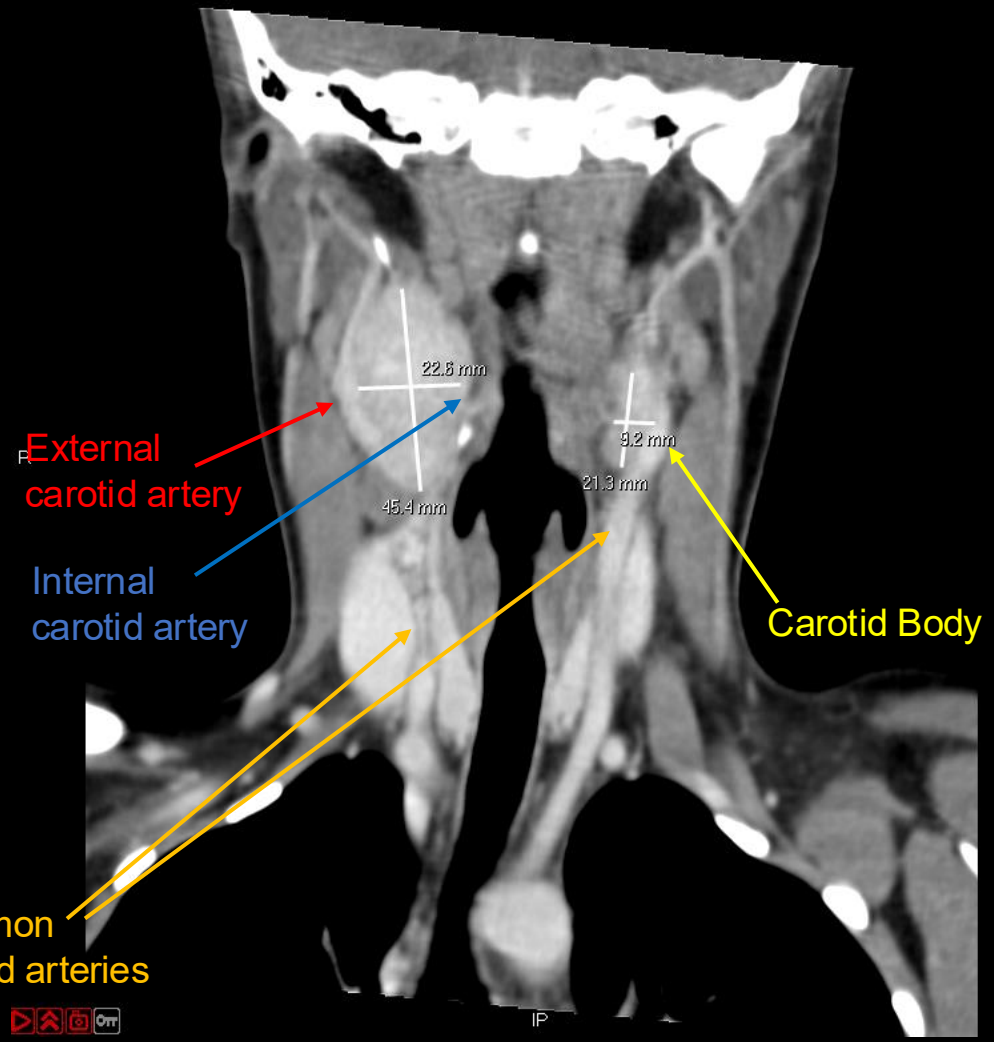
L neck mass

Findings: (labeled)





Findings: (labeled)

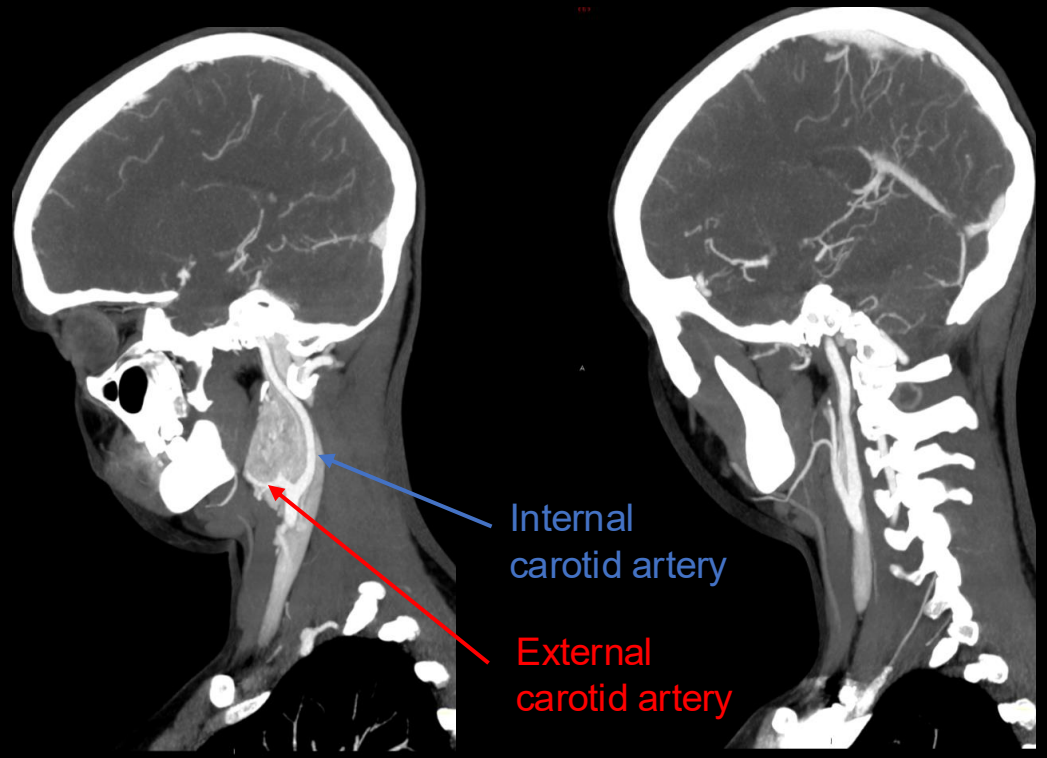


Heterogeneous enhancing lesions are seen within the bilateral carotid spaces.

The larger lesion on the right measures up to 4.7 cm with approximately 180 degrees encasement of the proximal cervical ICA and apparent complete encasement of the proximal right ECA.

The smaller lesion on the left abuts the medial carotid bulb with less than 180 degrees encasement.

Findings: (labeled)



R neck mass

L neck mass

On the right, the mass appears to splay the internal and external carotid arteries

Final Dx:

Bilateral Carotid Body Tumors

Case Discussion: Overview

- Head and Neck Paraganglioma Classifications
  - Other names: Glomus tumor, chemodectoma
  - **Carotid Body (Glomus Caroticum):** at the carotid body
  - Jugulotympanic: in the middle ear/jugular fossa
  - Glomus Vagale: along the vagus nerve
- Prevalence: 500 to 1600 cases in the United States per year [1]

Case Discussion: Pathophysiology

- Cell of origin: paraganglion cells; neural crest. These cells can differentiate into parasympathetic or sympathetic cells. In the head and neck they tend to be parasympathetic and thus do not secrete catecholamines [1]
- Behavior: slow-growing, locally destructive, benign, highly-vascular (biopsy is contraindicated). [2,3]
- Associations:
  - 20-50% genetic association (*SDH*, MEN2, *NF1, VHL*)
  - 10% multiple
  - <5% malignant
  - 1-3% catecholamine secretion
  - Chronic hypoxia (high altitude, COPD, cyanotic congenital disease) [1]

Case Discussion: Presentation and Diagnosis

- Presentation:
  - Neck mass (55%); Rubbery, nontender, horizontally mobile (positive Fontaine's sign), Hypertension (42%), Cranial nerve palsies (16%), Incidentalomas (10%) [3,4]
- Imaging:
  - Ultrasound: solid, well-defined, hypoechoic, splaying of the carotid bifurcation (Lyre Sign)
  - CT with contrast: densely enhancing due to hypervascularity, Lyre sign (as above)
  - CT Angiography: dense capillary staining
  - Shamblin Group System:
    - group I: <180 degrees of encasement of the ICA
    - group II: 180-270 degrees of encasement of the ICA
    - group III: >270 degrees of encasement of the ICA [5]
- Histopathology (on excision):
  - thin capsule
  - round or polygonal epithelioid cells in nests or trabecular patterns (Zellballen) [1]

Case Discussion: Treatment Options

- Observation
  - <1cm
  - Asymptomatic
  - No evidence of metastasis
- Surgical Resection
  - >2cm
  - Symptomatic
  - Catecholamine-secreting
- Radiation therapy
  - Non-resectable or morbid surgery [3,6,7,8,9]

Patient is scheduled for surgery in the coming months

Case Discussion: Key Considerations

- Paragangliomas are rare, highly vascular tumors that arise from neural crest chromaffin cells
- Early diagnosis with imaging and genetic testing is key
  - A classic radiographic sign for carotid body tumors is splaying of the carotid bifurcation
    - These tumors can be further categorized under the Shamblin group system

References:

1. Ikram A, Rehman A. Paraganglioma. [Updated 2024 Feb 29]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK549834/
2. Majid Khan, Joseph Schulte, S. James Zinreich and Nafi Aygun. Cummings Otolaryngology: Head and Neck Surgery, 8, 90-140.e1
"Paraganglioma: Causes, Symptoms & Treatment." Cleveland Clinic, 19 Dec. 2024, my.clevelandclinic.org/health/diseases/22394-paraganglioma.
3. Raza Pasha, and Justin S Golub. Otolaryngology-Head and Neck Surgery: Clinical Reference Guide, Fifth Edition. 5th ed., Plural Publishing, 2017.
4. Erickson D, Kudva YC, Ebersold MJ, Thompson GB, Grant CS, van Heerden JA, Young WF Jr. Benign paragangliomas: clinical presentation and treatment outcomes in 236 patients. J Clin Endocrinol Metab. 2001 Nov;86(11):5210-6. doi: 10.1210/jcem.86.11.8034. PMID: 11701678.
5. Sharma R, Gaillard F. Carotid body tumour. Radiopaediaorg. Published online September 24, 2008. doi:https://doi.org/10.53347/rid-4629
6. Ifeoluwa, A., Lázár, I., Szövördi, É., & Karosi, T. (2017). Management of carotid body tumor in pediatric patients: A case report and review of the literature. International Journal of Pediatric Otorhinolaryngology, 93, 47–52. https://doi.org/10.1016/j.ijporl.2016.12.017
7. Kuchakulla, M., Shah, A. H., Armstrong, V., Jernigan, S., Bhatia, S., & Niazi, T. N. (2019). Multimodal management of pediatric carotid body tumors: A systematic review and case illustrations. Journal of Neurosurgery: Pediatrics, 23(3), 325–332. https://doi.org/10.3171/2018.8.peds18393
8. Alanezi, T., Bin Onayq, A. I., & Al-Omran, M. (2024). Pediatric carotid body tumors: A case report and systematic review. Journal of Vascular Surgery Cases, Innovations and Techniques, 10(6), 101584. https://doi.org/10.1016/j.jvscit.2024.101584
9. Fagan JJ, Vander Poorten V. (2014). Resection of extratemporal paragangliomas including carotid body and glomus vagale tumors. In The Open Access Atlas of Otolaryngology, Head & Neck Operative Surgery. Retrieved from https://vula.uct.ac.za/access/content/group/ ba5fb1bd-be95- 48e5-81be586fbaeba29d/Resection%20of%20extrate mporal%20paragangliomas%20including %20carotid%20body%20and%20glomus% 20vagale%20tumours.pdf