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A case of carotid bifurcation vascular malformation masquerading as carotid body tumor

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Background

- The differential diagnosis for a carotid space mass can often be narrowed expeditiously using several pathognomonic radiographic features and laboratory tests.
- In particular, the splaying pattern of the great vessels and the MRI contrast and enhancement characteristics of the mass are used to predict the structure of origin.
- Nevertheless, surgical exploration can yield an unexpected finding. Here we present a rare case of a carotid bifurcation vascular malformation.

The Case

- A 60-year-old woman with hypertension presented to the emergency department with one month of new-onset intermittent left facial paresthesia and sharp occipital pain.
- As part of her initial work up, CT angiography revealed a 1.6 x 2.5 x 2.3 cm mass with internal calcifications at the level of the left carotid bifurcation extending superiorly between the internal and external carotid arteries (Figure 1). The patient was referred to neurosurgery and otolaryngology for management of the neck mass, with high suspicion for carotid body tumor.
- Subsequent MRI showed the mass to be enhancing, T1 hypointense, and T2 heterogeneously hyperintense with vascular signal voids – features supportive of carotid body tumor (Figure 2a-c).
- The patient recalled that her mother had a peri-brainstem mass that had been excised, which she thought may have been a paraganglioma. Genetic testing for succinate dehydrogenase mutations was sent and returned negative. Additionally, twenty-four hour urine fractionated metanephrine testing was within normal range.
- The decision was made to proceed with surgery. Intraoperatively the mass was found to be rather amorphous in nature, and frozen section suggested that the mass was in fact a vascular malformation. This was confirmed on final pathology, which showed a lesion comprising tortuous, bizarrely shaped, inter-anastomosing vasculature of variable caliber, including arteries and veins, with no neoplasm identified (Figure 3).

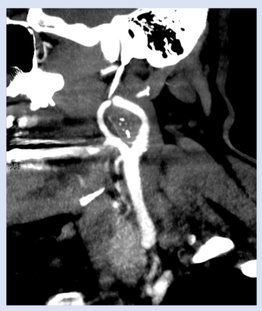

Figure 1. CT angiography. Maximum intensity projection CT angiography image of the left carotid bifurcation showing a calcified mass splaying the internal and external carotid arteries.

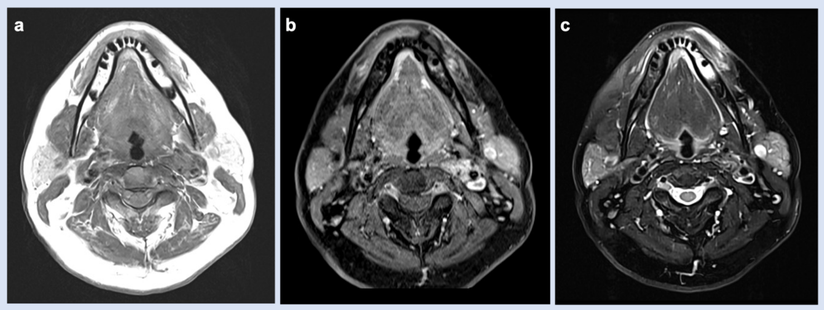

Figure 2. MRI. T1 pre-contrast (a), T1 fat-saturated postcontrast (b), and T2-weighted (c) MR images showing an enhancing mass at the left carotid bifurcation, containing multiple vascular signal voids.

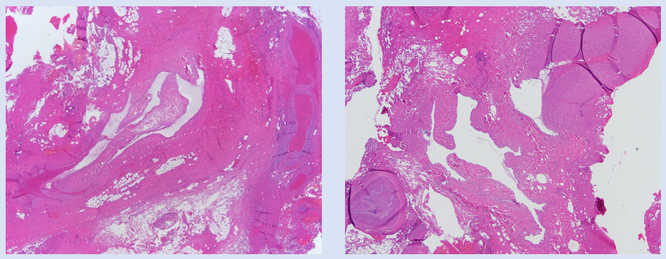

Figure 3. Pathology. Histologic examination showing a lesion comprising tortuous, bizarrely shaped, inter-anastomosing vasculature of variable caliber, including arteries and veins.

Literature Review

- There are six prior reports of carotid sheath vascular malformation, all described as slow-flow venous malformations or cavernous hemangiomas (Table 1). Of these, two have been described at the carotid bifurcation. Though surgical intervention remains an option for these entities, as they often do not involute and can continue to grow in size, other less invasive interventions (e.g. sclerotherapy, embolization) are available.
- Additionally, it is noted in the literature that phleboliths or calcifications may be a radiologic feature more prevalent in slow-flow venous malformations than in other entities such as paraganglioma. Further, phleboliths are best visualized on CT and on MRI can have a similar appearance to flow voids, as is demonstrated in the present case (Figure 1, 2).

Table 1. Literature Review of Carotid Sheath Vascular Malformations

Paper	Case Presentation	Imaging Findings	Pathology Findings
Antonopoulos 2009	51F with growing 3.8cm neck mass over three months. Non-pulsatile, mobile, painless	Ultrasound – Solid well-defined nodule with mixed echogenicity in left anterior part of neck. CT – Ovoid mass in carotid sheath close to left thyroid lobe with significant uptake MRI – Diffusely enhancing well defined soft tissue mass w low signal on T1 and high signal on T2 MRA – No uptake by mass or feeding vessels	FNA – Attempted but discontinued due to bleeding Multiple vascular spaces of thick-walled blood vessels, and the diagnosis was 'cavernous hemangioma'
Zagzag et al 2009	64M with incidentally noted 5.5cm right neck mass. Mobile, nontender, non-pulsatile, and without detectable bruits	CT – Non-enhancing. Splaying CC and IJ with an enhancing vessel at its posterior aspect. A low density area at posterolateral aspect suggesting cystic change or necrosis. No MRI presented	FNA x2 were nondiagnostic Sclerosing cavernous hemangioma
Massi et al 2014	71F with headache, hypertension, recurrent right periorbital pain. 2.4cm mass that is non-pulsatile, no bruit	MRI and CT - Oval vascularized mass close to bifurcation of left carotid artery.	Cavernous hemangioma – large thin-walled blood vessels, lined by endothelial cells, separated by fibrous tissue
Gao et al 2020	60F with painful 3.4cm neck mass for 6 months. No bruits, non-pulsatile, mobile in lateral plane but not cephalocaudal	Ultrasound – Solid, well-defined mass with mixed echogenicity and no blood flow signal. Centered at bifurcation with splaying of ICA and ECA CT – Ovoid mass centered at bifurcation with splaying of ICA and ECA MRI – T1 isointense, T2 hyperintense	Intraoperative – 2 feeding vessels from ECA Final pathology – Vascular malformation with thin-walled blood vessels, lined by endothelial cells and separated by fibrous tissue
Goksel et al 2020	31F with 4.8cm mass	MRI, MRA – inferior neck mass (no other description)	Sclerosing cavernous hemangioma
Suthar et al 2024	58M with 4.1cm left neck mass slowly growing over four years. Asymptomatic	MRI – Homogeneously T1 isointense, T2 hyperintense. Avid contrast enhancement. Splaying of IJ and ICA, but minimal splaying of ICA and ECA. CTA – A well-circumscribed hypodense soft tissue mass, punctate calcifications	Dilated congested, thin walled blood vessels lined by single layer of flat, bland-appearing endothelial cells. No atypia or mitotic activity. Diagnosis: slow-flow vascular malformation

CC: common carotid; ICA: internal carotid artery; ECA: external carotid artery; IJ: internal jugular vein

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