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Carotid Body Tumor in Review

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Introduction

Carotid Body Tumors (CBTs) are rare neoplasms derived from neural crest cells. Diagnosis is straightforward as they typically present as asymptomatic, pulsatile neck masses due to their high vascularity. However, the diagnosis can be easily overlooked due to their low occurrence and confusion with lymphadenopathy.

The clinical incidence of carotid body tumors reported by various studies range from 1/30,000 [2] to as little as 1/1,000,000. CBTs originate from paraganglia cells, which are divided into chromaffin paraganglia and non-chromaffin paraganglia. There are 3 types of CBTs: hyperplastic, familial and sporadic. Hyperplastic CBT is more prevalent in patients with COPD, congenital heart disease or other forms of chronic hypoxia. The incidence can be 10 times higher in high altitude areas. Familial CBT presents earlier in age and is more likely to be malignant. Mutations related to the pathogenesis have been implicated in overexpression of hypoxia inducible genes. The genes linked to familial CBTs are related to Succinate Dehydrogenase (SDH) function.

The first step in diagnostic studies should be Doppler US, which would show high vascularity and narrow down the differential to include vascularized lymph node, metastasis, or lymphoma. Contrast-enhanced CT is great at visualizing CBTs, which would present with characteristic splaying of the Internal and External Carotid Arteries and bright, rapid enhancement. MRI with contrast has similar findings as well as a "salt and pepper" appearance due to a mix of slow flow and signal voids respectively.

Images

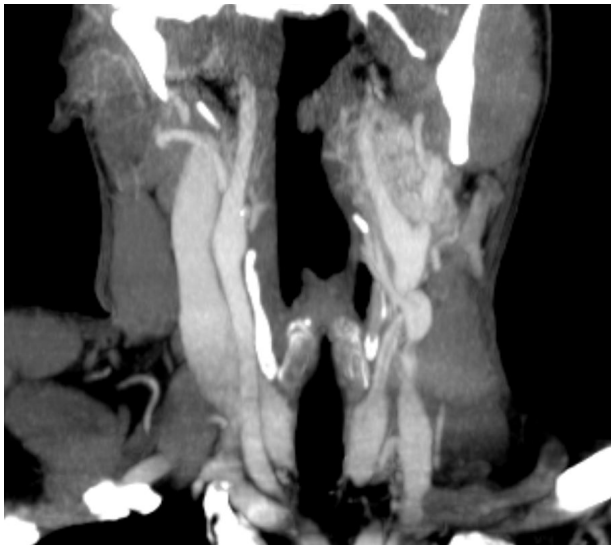

Figure 1. *Coronal CT image of the neck shows characteristic splaying of the external carotid artery (ECA) and internal carotid artery (ICA) by a left hypervascular CBT.*

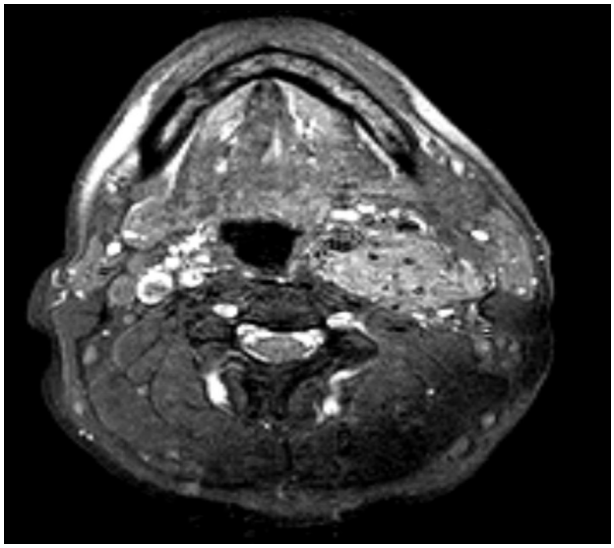

Figure 2. *Axial T1-weighted MRI of neck post fat saturation reveals flow voids within the carotid mass.*

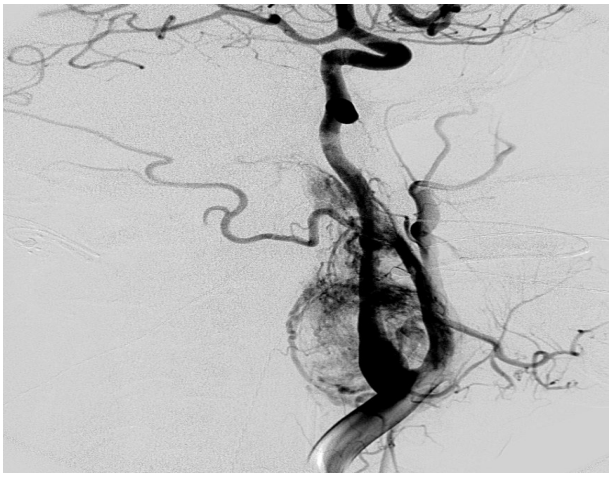

Figure 3. *Lateral DSA shows a mass posterior to the carotid bifurcation with significant vasculature.*

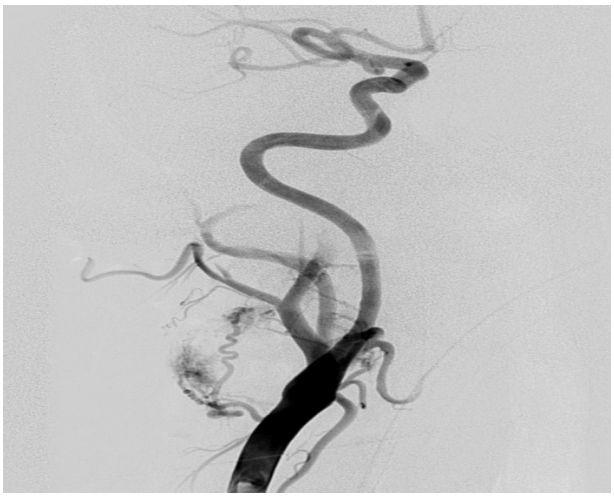

Figure 4. *Post particle embolization demonstrates 80% reduction in vascularity of the mass.*

Video related to the discussion is available at:

https://youtu.be/biUiHv37RhA

Discussion

Our patient is a 49-year-old male with no significant past medical history who presented with a high pitched voice and hoarseness, found to have a left neck mass, most concerning for a carotid body tumor. The patient was initially taken to IR for embolization of the tumor, prior to definitive surgical excision through a modified radical neck dissection. The patient was discharged home, with follow-up with ENT.

Surgery is the ideal treatment. Some sources suggest follow up with radiation for any residual tumor or recurrences, and if surgery cannot be done for any reason, radiotherapy alone should be performed. Due to inadequate data on this disease, there has not been a study done comparing the efficacy of surgery versus radiotherapy. The biggest risk for surgery is cranial nerve damage, which can range from 11% to 49%. When evaluating neck masses, CBTs should be considered because, despite their rarity, it is an incredibly simple diagnosis to make and treat.

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