

Hold Still – A large neck paraganglioma limiting head movement

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Introduction

Paraganglioma is a rare neuroendocrine tumor derived from extra-adrenal autonomic paraganglia cells¹. As opposed to sympathetic paragangliomas and pheochromocytomas, parasympathetic derived paragangliomas of the neck are non-functioning in up to 95% of the cases¹. For functioning tumors, manipulation during surgery can trigger catecholamine release if the patients are not adequately pre-treated with alpha blockade.¹ We present a rare case of a functioning neck paraganglioma that has been neglected for years until head movement was sufficient to induce hyperadrenergic symptoms.

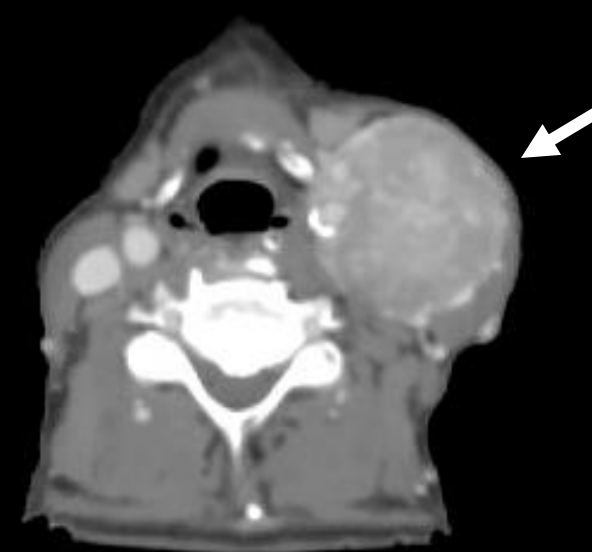
Case report

Seventy nine year old Caucasian farmer, with a history of tobacco and alcohol abuse, presented to the emergency department with an expansive left neck mass. While the mass had been growing for 10 years, he sought medical attention when he could no longer turn his head to the left, which he associated with worsening symptoms. He reported recurrent brief syncopal attacks, intermittent episodes of profuse sweating, headache, palpitations, fluctuating pale and flushed skin. Upon evaluation, he was afebrile, tachycardic and had labile blood pressure readings (high of 340/150 mmHg and a low of 70/40). He was oriented but appeared lethargic and underweight. A solid non-tender neck mass, with an audible bruit, is apparent in left neck level III. A CAT scan of the neck showed a 7.7cm left glomus vagale tumor with partial encasement of the left internal and external carotid artery. **(Image 1)**

He was admitted to ICU with close blood pressure monitoring with intra-arterial line. Due to a high suspicion for catecholamine driven process, he was started on Doxazosin 1 mg every 12 hours which stabilized his blood pressure, heart rate and improved his symptoms. Plasma catecholamines were significantly elevated (metanephrine 437 pg/ml [12-67], normetanephrines 4700 pg/ml [18-101], epinephrine 566 pg/ml [10-200], norepinephrine 18055 pg/ml [80-520], Dopamine 82 pg/ml [[0-20]]). MIBG scan showed intense uptake in the left carotid mass, which was consistent with a paraganglioma. **(Image 2)**

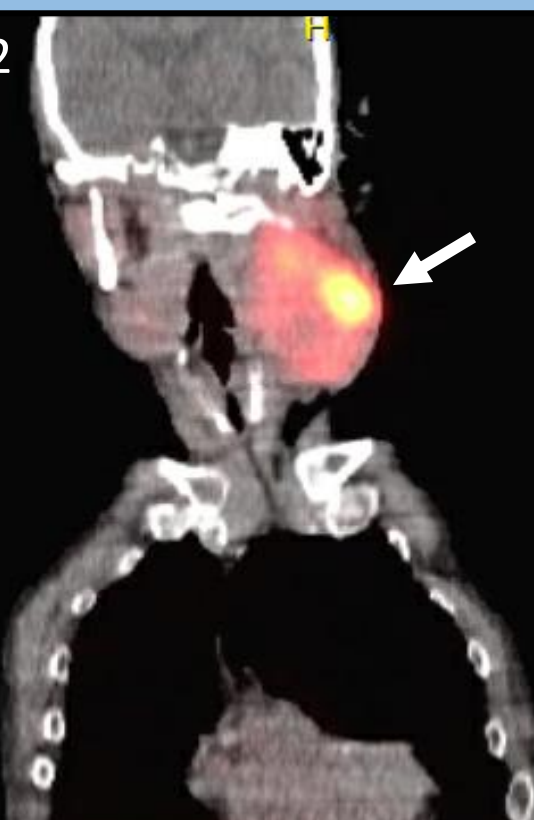
Genetic testing did not show pathologic variants. He was deemed to be at high risk for surgery, given the proximity of the mass to the neuro-vascular bundle. Radiation oncology administered 5 radiation sessions during his inpatient stay. The patient's was able to move his head more freely and was discharged with outpatient follow up.

Image 1



CT with contrast neck; Large soft tissue mass along the left aspect of the neck originating from the left common carotid artery bifurcation, measuring 7.7 cm AP x 5.8 AP x 5.7 transverse centimeters. Partial encasement of the left internal carotid artery and left external carotid artery.

Image 2



SPECT I123 MIBG scan; Intense uptake in the large left carotid mass, compatible with a MIBG avid paraganglioma.

¹ Young WF Jr. Paragangliomas: clinical overview. *Ann N Y Acad Sci.* 2006;1073:21-29. doi:10.1196/annals.1353.002