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Tumor Biology
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Hold Still – A Large Neck Paraganglioma Limiting Head Movement
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Background: A head and neck paraganglioma is a rare diagnosis with most cases found incidentally or diagnosed due to mass effect symptoms. Only 3.5% of them are found to secrete catecholamines leading to episodic symptoms. Herein, we present a rare case of a functioning neck paraganglioma with head movement induced hyperadrenergic paroxysms.

Clinical Case: A 79-year-old Caucasian man, with previous left neck mass found incidentally on imaging after a motor vehicle accident, presented to the ER with syncope. While the mass had been growing for 10 years, he sought medical attention when he could no longer turn his head to the left due to worsening symptoms. He reported recurrent brief syncopal attacks along with intermittent episodes characterized by profuse sweating, headaches, palpitations, 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 mmHg). He was oriented to time and place but appeared lethargic and underweight. A solid non-tender neck mass, with an audible bruit, was apparent at the Level II of the left neck. A CT scan of the neck showed a 7.7cm left glomus vagale tumor with partial encasement of the left internal and external carotid artery. He was admitted to the intensive care unit for close blood pressure monitoring via an intra-arterial line. Due to high suspicion for a catecholamine driven process, he was started on Doxazosin 1 mg every 12 hours which stabilized his blood pressure and improved his symptoms. CT scan of the abdomen and pelvis showed a 1.1 cm left adrenal nodule with an intensity of 4 Hounsfield units. A trial of Hyoscyamine was done to help with hypotension and symptoms of dizziness and bradycardia. Labs showed plasma catecholamines that were significantly elevated to ten times the upper limit of normal. An MIBG scan showed uptake in the left carotid mass. He was a poor surgical candidate because of the proximity of the mass to the neurovascular bundle and was treated with radiation during hospitalization. The patient was discharged home on Doxazosin with improvement in his blood pressure. Genetic testing was negative for a pathogenic variant but detected a variant of uncertain significance (VUS) in CDKN2A, c.373G>C (p.Asp125His). Unfortunately, three months later the patient was admitted with an intracranial hemorrhage secondary to a fall and was transitioned to hospice care. There was no change in the tumor size noticed during that admission.

Conclusion: To our knowledge, this is the first case to describe hypertension paroxysms triggered by movements of the neck in the setting of a functional neck paraganglioma.