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An incidental but fatal nor-epinephrine secreting carotid body paraganglioma presenting as hypertensive emergency: A rare clinical entity

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Introduction: Pheochromocytomas (PCCs) and paragangliomas (PGLs) are neuroendocrine tumors derived from the chromaffin tissue. Timely diagnosis of these tumors is extremely important as they are associated with great cardiovascular morbidity and mortality. Vagal PGLs account for 5-25% of head and neck PGLs. Majority of these tumors are non-functional and present due to mass effect with only about 5% secreting catecholamines.

Objective: Here we present the case of a fatal norepinephrine secreting carotid body paraganglioma diagnosed incidentally during workup for a suspected stroke.

Case Report: A 58-year-old African American lady, presented to the emergency room for sudden onset of right eye blindness and left eye blurry vision, associated with a 2 day history of a temporal headache. She was noted to be in hypertensive crisis. ESR was normal. A CT scan head ruled out hemorrhage. Carotid Doppler incidentally noted a vascularized mass at the bifurcation of the right internal and external carotid arteries. CTA neck characterized this to be a large 7.5 cm avidly enhancing hypervascular mass in the right carotid space, suspicious for carotid body paraganglioma. Plasma and 24 hour urine nor metanephrines were significantly elevated at 5245 pg/ml (0-145 pg/ml) and 10,471 (82-500 microgram/24 hr) respectively. 24 hour urine metanephrines and cortisol were normal. Chromogranin was elevated at 43 nmol/L (0-5nmol/L). CT scan abdomen did not show evidence of pheochromocytoma. Past history was significant for uncontrolled hypertension, CKD 3 and cardiomyopathy. No family history of pheochromocytoma or MEN II. Antihypertensive regimen included labetalol, clonidine, Isosorbide mononitrate, nifedipine and hydralazine. In addition to elevated blood pressure, exam was pertinent extensive hypertensive changes in the retina, with no papilledema, and a palpable mass in the right neck. Her vision improved subsequently. She did admit to occasional headaches, episodes of sweating, tremors, and a throbbing sensation in the head. An increase in labetalol and nifedipine improved hypertension control and she was discharged with plan for surgery. Further workup included 18FDG PET scan and I123 MIBG scan which showed accumulation within the documented parapharyngeal PGL with no evidence of additional disease. She was referred for genetic testing, which she did not pursue. Phenoxybenzamine was initiated few weeks pre operatively and she was optimized for surgery. Considering the increased vascularity, tumor embolization was performed pre operatively to limit surgical blood loss. Right neck dissection was performed and pathology confirmed a catecholamine secreting PGL. Post operatively, she continued to require anti-hypertensives. Further hospital course was eventful for a right MCA stroke that she developed post-operative day 3, possibly due to dissection of internal carotid artery and she subsequently had a cardiac arrest and died from multi organ failure.

Conclusion: This is not only a rare case of a functional carotid body paraganglioma, but also illustrates the high morbidity and mortality associated with these conditions. Long standing uncontrolled hypertension continues to be an under evaluated entity among hospitals and primary care settings. Timely diagnosis and treatment may have prevented this fatality.

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