A Case of Aortic Dissection: Was it a Pheochromocytoma?

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While hypertension is very common, true hypertensive emergencies are rare, and aortic dissection is an unusual clinical presentation of a hypertensive crisis. Aortic dissections occur with an incidence of 3/100,000 cases per year, and are associated with a high morbidity and mortality [1]. Arterial hypertension is the most important risk factor for aortic dissections [2].

We present a case of a 31-year-old African American man who presented to the ER with sudden, severe chest pain during a vacation in France. He had a long-standing history of hypertension and was possibly non-adherent with his prescribed medications, but he did not abuse tobacco, alcohol, or any illegal drugs. He had no other relevant personal or family medical history. His initial blood pressure was 250/130 mmHg. Imaging revealed an acute aortic dissection. An emergent aortic arch replacement was performed, during which the patient suffered profuse intraoperative hemorrhage and hypoxic encephalopathy subsequently. In a comatose state, he was transferred to our hospital for further management. During his hospitalization, he was transitioned to oral blood pressure medications and weaned from the ventilator. He had a thorough evaluation for secondary causes of hypertension. Labs showed normal levels of thyroid stimulating hormone, aldosterone, renin, BUN/Cr, LFTs, and serum electrolytes. Urine electrolytes, EKG and lipids were also normal. RPR was nonreactive. Serum catecholamine testing showed metanephrines of 201 mcg, normetanephrines of 797 mcg, norepinephrine of 797 pg/ml (normal range 70-750), epinephrine of 170 pg/ml (normal range is less than 110) and dopamine of 20 pg/ml (normal range is less than 30). The 24-hour urine sample showed metanephrines of 432 mcg (normal range 74-297 mcg) and normetanephrines of 1,894 (normal range 105-354 mcg). This testing was repeated two weeks later, and levels remained elevated even though the patient was clinically stable and normotensive on a three-drug regimen of antihypertensives. Adrenal CT showed normal adrenal size and no evidence of renal artery stenosis. Whole body nuclear medicine tumor localization showed no scintigraphic evidence for I-123 MIBG avid neuroendocrine tumor. MRI of the cervical spine with and without contrast was performed in order to evaluate for potential neuroendocrine neck tumors, such as paragangliomas, and there were no paraspinal masses seen. He was diagnosed with resistant essential hypertension because the results for secondary hypertension work-up were inconclusive during his hospital stay. He was discharged to a rehab facility with a scheduled follow-up appointment in the endocrinology clinic, where catecholamine testing will be repeated.

We present an unusual case of an acute aortic dissection in a young patient with malignant hypertension. After discovering that he had elevated catecholamines, we suspected he might have secondary hypertension due to a pheochromocytoma, paraganglioma or ganglioneuroma. However, imaging was not diagnostic of adrenal or neuroendocrine tumor. In the setting of hospitalization, patients may experience increased catecholamine levels from the anxiety and pain [3]. Pheochromocytomas have an incidence of 0.3% to 0.95% of the population and are the cause of hypertension in 0.1% of hypertensive patients [4]. Measuring the 24-hour urine metanephrines and normetanephrines yields the highest sensitivity and specificity among the biochemical markers. Other possible pharmacologic tests that could have been performed in this patient to identify pheochromocytomas are the clonidine suppression test and provocative stimulation tests. After diagnosing adrenal or extra-adrenal pheochromocytomas biochemically, it is necessary to localize the tumor with helical or spiral high resolution CT, MRI or MIBG scans make the diagnosis. Furthermore, treatment involves surgical resection or medical management in the setting of catecholamine crisis.
References


