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Reverse Takotsubo-like Cardiomyopathy and Pheochromocytoma

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Abstract Title: REVERSE TAKOTSUBO - LIKE CARDIOMYOPATHY AND PHEOCHROMOCYTOMA

Introduction

Pheochromocytoma is a rare catecholamine-producing tumor of chromaffin cells in the adrenal gland. Takotsubo Cardiomyopathy is stress induced cardiomyopathy typically with apical ballooning shape of the left ventricle on echocardiography due to apical akinesis. Rarely a reversed variant of Takotsubo cardiomyopathy in which the apex is spared while the base of the heart is akinetic. We present a case of a pheochromocytoma with reverse Takotsubo cardiomyopathy.

Case

34-year-old female presented with lightheadedness, dizziness, flushing, near-syncope, dyspnea on exertion for 1 week. She also reported increasing panic attacks not relieved with home clonazepam. She had a heart rate of 166 and blood pressure of 58/34 that was fluid responsive. Her point of care troponin I was elevated to 1.96 (units), pro-B-type natriuretic peptide (BNP) of 10,773 (pg/mL), an electrocardiogram (ECG) showed no ischemic changes. A Computed Tomography Angiogophraphy (CTA) revealed a right adrenal mass measuring 3.9x2.7cm.

The patient was admitted to the intensive care unit for further management. She was found to have severe orthostatic hypotension. Transthoracic echocardiography showed an ejection fraction of 25-30% with severe global hypokinesis. Wall motion abnormalities included the left ventricular base and midportion, sparing the apex - consistent with reverse Takotsubo cardiomyopathy.

Systolic blood pressures remained consistently above 210 mmhg until the beta blocker was replaced with phenoxybenzamine. Pheochromocytoma was strongly suspected and soon confirmed. Plasma fractioned metanephrines and 24-hour urinary catecholamines and metanephrines were observed at greater than ten-fold the upper limits of normal.

She was eventually transferred to the University of Colorado Hospital for emergent resection of the pheochromocytoma. She underwent laparoscopic right adrenalectomy. Plasma metanephrines and catecholamines normalized post-surgery.

Discussion

Pheochromocytomas are catecholamine producing neuroendocrine tumors with an annual incidence approximately 0.8 per 100,000 person-years. The signs and symptoms include elevated heart rate, hypertension, orthostatic hypotension, palpitations, anxiety, headaches, and hyperglycemia. Pheochromocytoma is rarely associated with cardiomyopathy attributed to catecholamine excess that is similar to stress-induced (Takotsubo) cardiomyopathy. The true incidence of the two together is unknown due to the extreme rarity of such cases. The presence of an adrenal mass along with worsening hypertension secondary to unopposed alpha adrenergic vasoconstriction with beta blockade resulted in high suspicion for pheochromocytoma in this case.

The reverse Takotsubo-like cardiomyopathy observed on echocardiogram was likely a result of catecholamine-induced cardiotoxicity triggering apoptosis. Only 2.2% of patients with Takotsubo had the reverse variant which shows basal akinesis/hypokinesis with apical sparing with a female to male preference. Coronary artery disease is an important consideration in any variant of Takotsubo cardiomyopathy. In our case, a left heart catheterization was not conducted due to the patient's young age, echocardiogram findings and elevated catecholamine levels which made the case for reverse Takotsubos cardiomyopathy far more likely.