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Two Cases of Pheochromocytoma Associated Takotsubo Syndrome- Importance of Early Recognition and Treatment

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Introduction: Pheochromocytoma is a rare catecholamine producing adrenal medullary tumor that classically presents with hypertension. Takotsubo cardiomyopathy (TC) is a rare but increasingly recognized presentation of pheochromocytoma. We present two cases of TC likely caused by pheochromocytoma.

Case 1: A 60 y/o male who presented with acute onset nausea, vomiting and abdominal pain. Systolic BP was 150-160 mm Hg, and diastolic BP was 100 mmHg. Troponin was elevated (4.15 ng/ml; nl = 0-0.04). An echo-cardiogram showed a reduced left ventricular ejection fraction (LVEF) of 15-20% (nl = 50-75%), a mildly dilated left ventricle, and apical ballooning suggestive of TC. Coronary arteries were normal on cardiac catheterization. CT scan identified a 5.3×5.6×5.7 cm adrenal mass of 50 HU pre-contrast. Further evaluation revealed markedly elevated plasma metanephrine concentration of 858 pg/ml (nl<57) and normetanephrine concentration of 1729 pg/ml (<148). His presentation was strongly suggestive of pheochromocytoma associated TC. He was prepared for surgery with doxazosin 2 mg twice daily followed by metoprolol succinate and metyrosine up to 500 mg/d. Final pathology revealed a 5.3 cm left pheochromocytoma with negative surgical margins. Germline genetic testing was negative for pheochromocytoma/paraganglioma pathogenic gene mutations, and an MIBG scan did not reveal metastatic disease. Echocardiogram one month after adrenalectomy showed a marked improvement in LVEF to 55-60% with no regional wall motion abnormalities, and plasma metanephrine concentrations normalized.
**Case 2:** A 61 y/o female smoker with type 2 diabetes mellitus, hypertension, Graves disease and obesity presented with sudden onset of nausea followed by chest discomfort, bilateral shoulder discomfort and shortness of breath. Troponin was 0.6 ng/ml (nl = 0-0.04), EKG showed ST elevations in leads I, 2, V5 V6 with hyperacute T waves. Coronary arteries were normal on catheterization. She required intubation, and systolic BP ranged from 110 to 200 mm Hg. On echocardiogram the LVEF was 35% to 40% (nl = 50-75), and there was diffuse hypokinesis of the mid to apical LV myocardium with hypercontractility of the LV base, suggestive of TC. The plasma metanephrine concentration was 3.31 nmol/l (nl= 0-0.49), and normetanephrine concentration was 5.02 nmol/l (nl= 0-0.89). CT imaging revealed a 2.9 cm left adrenal mass. She underwent successful left adrenalectomy after preparation with alpha, beta blockade and metyrosine. Germline genetic testing was negative for pheochromocytoma/paraganglioma pathogenic gene mutations. Echocardiogram nine months post-adrenalectomy showed normal EF of 50-55% and resolution of apical hypokinesis.

**Discussion:** We present two pheochromocytoma patients presenting with TC that reversed following appropriate pheochromocytoma management. Few case series report the association of pheochromocytoma and TC. These patients usually present acutely without prodromal stressors commonly found with classic TC.

**Conclusion:** In the setting of TC, pheochromocytoma should be considered.

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