Pheochromocytoma is a catecholamine-secreting tumor, with highly variable clinical expression leading to its designation as the great mimicker. A given patient may be completely asymptomatic or present with abdominal, metabolic, and cardiovascular manifestations because of catecholamine excess, such as paroxysmal or sustained hypertension and heart failure or life-threatening arrhythmias. Drugs interfering with adrenergic stimuli may worsen a patient’s condition. In our case report, a patient with a pheochromocytoma presented with acute and sudden heart failure and the delay in diagnosis led to a progression of catecolaminergic myocardial injury until mass resection.

**Case Report**

A 54-year-old woman with no significant past medical history went to the hospital because of nausea and prolonged vomit. On admission, she was treated with antiemetic drugs (metoclopramide) and suddenly developed acute heart failure that progressed to cardiogenic shock. She clinically improved with initiation of inotrope support. Serum laboratory testing was notable for leukocytosis and troponin I elevation (62.5ng/mL). The ECG showed no evidence of ischemia or prior infarct. Echocardiogram and cardiac MRI showed normal left ventricular dimensions and mild systolic dysfunction (left ventricular ejection fraction, 40%) because of akinesia of mid inferolateral wall and hypokinesia of mid anterolateral and inferior walls. Myocardial edema was present in the inferolateral wall (Figure 1A), corresponding with a midwall stria of late gadolinium enhancement (Figure 1B and 1C). No coronary angiography was performed because of a straight nonischemic pattern of myocardial edema and necrosis. The patient was discharged from the hospital with diagnosis of gastroesophageal reflux disease and viral myocarditis and treated with a proton-pump inhibitor and a β-blocker.

A week later, the patient was readmitted to the hospital for lumbar pain and mild dyspnea on exertion. She also had orthostatic hypotension and transient episodes of headache and limb tremors associated with hypertension. An abdominal ultrasound revealed an incidental mass above the right adrenal gland. A catecholamine-producing tumor was suspected and the patient was referred to specialists for further evaluation. The ECG showed T-wave inversion in the lateral leads. Serum laboratory testing showed troponin I elevation. Computed tomographic scan of the abdomen revealed a heterogeneous mass lesion (4x3cm) over the right adrenal gland, consistent with a neuroendocrine tumor. Urine catecholamines levels were high (epinephrine, 5352nmol/24 h; norepinephrine, 3206nmol/24 h) and urine metanephrines were 4-fold higher than the normal ranges. A second cardiac MRI was performed showing hypokinesia of mid segments of the anterior, lateral, and inferior walls with mild systolic dysfunction (left ventricular ejection fraction, 47%). On tissue characterization sequences, tissue edema (Figure 1D) and late gadolinium enhancement were still present, with an increased amount compared with the first cardiac MRI (Figure 1E and 1F).

Given these findings, pheochromocytoma causing myocarditis was considered the most likely diagnosis and the patient was referred for laparoscopic resection of the mass (Figure 2). Pathology examination confirmed the diagnosis and the patient was discharge home on day 16 without symptoms. After 6 months, another cardiac MRI showed persistence of scar areas without edema.

**Discussion**

Pheochromocytoma is a neuroendocrine tumor with variable clinical expressions ranging from hypertension to angina symptoms and heart failure. Moreover, it is often discovered during a workup for an incidentaloma. This case represents a challenging diagnosis of pheochromocytoma, initially missed and later recognized during an evaluation for abdominal pain. The sudden worsening of heart function with drugs, such as metoclopramide, β-adrenoceptor blockers, and steroids, like in this case, could suggest an unknown catecholamine secreting tumor. Several mechanisms have been proposed to explain shock including abrupt cessation of catecholamine secretion by the tumor in a patient with a constricted circulatory volume and desensitized adrenerceptors, after a prolonged period of catecholamine-induced hypertension.
Catecholamines can exert a direct toxic effect on the myocardium through enhanced lipid mobility, calcium overload, free radical production, or increased sarcolemmal permeability. Lyon et al has suggested that high levels of circulating epinephrine may cause a switch from Gs to Gi protein signaling via the $\beta_2$-adrenergic receptor, resulting in a negative inotropic effect, that could be most pronounced at the apex of the heart where the $\beta$-receptor density is greatest. By a comprehensive review of literature high levels of catecholamines can cause myocardial inflammation and necrosis. In a report on 26 patients at the Mayo Clinic who died of complications of pheochromocytoma, 58% had active myocarditis presumed to be the result from excessive circulating concentrations of catecholamines. The lesions found in the myocardium in laboratory animals after injections of catecholamines were similar to an active myocarditis. Catecholaminergic myocardial injury represents a continuum spectrum with transient and completely reversible heart dysfunction during short exposures (possibly the cause of Tako Tsubo syndrome) and progressive patchy necrosis, like myocarditis, when the hormones are secreted for a long time.

In the literature, case reports of myocarditis associated with pheochromocytoma are sporadic and rarely characterized by cardiac MRI. To the best of our knowledge, this case is unique case because the progressive myocardial injury by catecholamines is shown with serial cardiac MRIs.

Disclosures

None.

References


Key Words: catecholamines ■ magnetic resonance imaging ■ pheochromocytoma
Figure 1. T2 weighted (A) and postcontrast images (B and C) of the first cardiac MRI. T2 weighted (D) and postcontrast images (E and F) of the second cardiac MRI. Notice the increasing in edema amount in the second cardiac MRI (asterisks) and the extension of necrotic area between the 2 examinations (arrow heads).

Figure 2. Macroscopic anatomy image of pheochromocytoma after surgical resection.
Heart Failure Due to Adrenergic Myocardial Toxicity From a Pheochromocytoma
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