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CASE REPORT

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Catecholamine-induced cardiomyopathy and multiple organ failure in pheochromocytoma

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ABSTRACT

A 55-year-old Caucasian woman with no significant past medical history presented with chest pain, palpitations, shortness of breath, and nausea. Physical examination was notable for a blood pressure of 182/87 mmHg, heart rate (HR) of 74 beats per minute (bpm), temperature of 98.3ºF, and oxygen saturation of 94% on 15 liters (L) of oxygen per minute. Her initial labs revealed troponin of 0.26 ng/mL (<0.01 ng/mL), blood glucose of 497 mg/dL (70–99 mg/dL), lactic acid of 6.9 mmol/L (0.4–1.9 mmol/L), and white blood cell (WBC) of 21.6 K/uL (4–11.0 K/uL). EKG showed ST elevation in leads V1 and V2. CT Pulmonary angiography with contrast ordered to rule out pulmonary embolism revealed a right adrenal mass measuring 3.5 cm x 4.1 cm. Patient was admitted to the intensive care unit for ST elevation myocardial infarction, hyperglycemia, and sepsis. She was started on heparin, broad-spectrum antibiotics, intravenous fluids, and insulin. Emergent echocardiogram revealed 40–45% ejection fraction with septal, lateral, anteroseptal, and posterolateral hypokinesis. Troponin elevation to 1.00 ng/mL (<0.01 ng/mL) warranted a cardiac angiography which revealed new-onset systolic heart failure with reduced ejection fraction with normal coronary vessels. A relatively rapid improvement in her clinical course suggested that a functioning tumor could be the underlying etiology. Diagnostic work-up for pheochromocytoma showed elevated metanephrine and normetanephrine. Subsequent surgical biopsy of the adrenal mass was consistent with pheochromocytoma. It was a rare case presentation of pheochromocytoma with catecholamine-induced cardiomyopathy and multiple organ failure.

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Pheochromocytoma; catecholamine-induced cardiomyopathy; multiple organ failure; rare complications of pheochromocytoma

1. Introduction

Pheochromocytoma is an endocrine tumor arising from chromaffin cells in the adrenal medulla [1]. Pheochromocytoma classically presents with sustained or paroxysmal hypertension, headache, excessive truncal sweating, and palpitations [1]. We highlight a rare case presentation of pheochromocytoma with catecholamine-induced cardiomyopathy and multiple organ failure.

2. Case Presentation

A 55-year-old Caucasian woman with no significant past medical history presented with chest pain, palpitations, shortness of breath, and nausea. Physical examination was notable for a blood pressure of 182/87 mmHg, heart rate (HR) of 74 beats per minute (bpm), temperature of 98.3ºF, and oxygen saturation of 94% on 15 liters (L) of oxygen per minute. Her initial labs were concerning for troponin: 0.26 ng/mL (<0.01 ng/mL), creatinine (Cr): 1.5 mg/dL (0.6–1.1 mg/dL), white blood cell (WBC): 21.6 K/uL (4–11.0 K/uL), blood glucose: 497 mg/dL (70–99 mg/dL), aspartate aminotransferase (AST): 110 U/L (10–40 U/L), alanine aminotransferase (ALT): 133 U/L (15–37 U/L), lactic acid: 6.9 mmol/L (0.4–-1.9 mmol/L) and pro-BNP of 926 pg/mL (0–124 pg/mL). Electrocardiogram (EKG) showed ST segment elevations in leads V1 and V2 which was new compared to her prior EKG. A CT pulmonary angiography with contrast (CTPA) was negative for pulmonary embolism but did reveal an incidental finding of 3.5 cm x 4.1 cm hypodense right adrenal mass.

Patient was admitted to the intensive care unit (ICU) for ST elevation myocardial infarction (STEMI), hyperglycemia, and sepsis. She was started on heparin drip at 12 Units/kg/hr, metoprolol 12.5 mg two times daily, cefepime 1 g, 30 ml/kg (2.1 L) lactated ringers bolus, and insulin drip at 0.1 Units/Kg/hr. In the ICU, patient had worsening chest pain and palpitations. Her troponin rose from 0.26 ng/mL to 1.00 ng/mL. Emergent echocardiogram showed 40–45% ejection fraction with septal, lateral, anteroseptal, and posterolateral hypokinesis. At this point, the patient was taken for cardiac angiography which surprisingly revealed normal coronary vessels. Considering this patient had no significant past medical history and no signs of infection, her sudden onset of multi-organ failure with cardiomyopathy was proving difficult to elucidate. Myocarditis was

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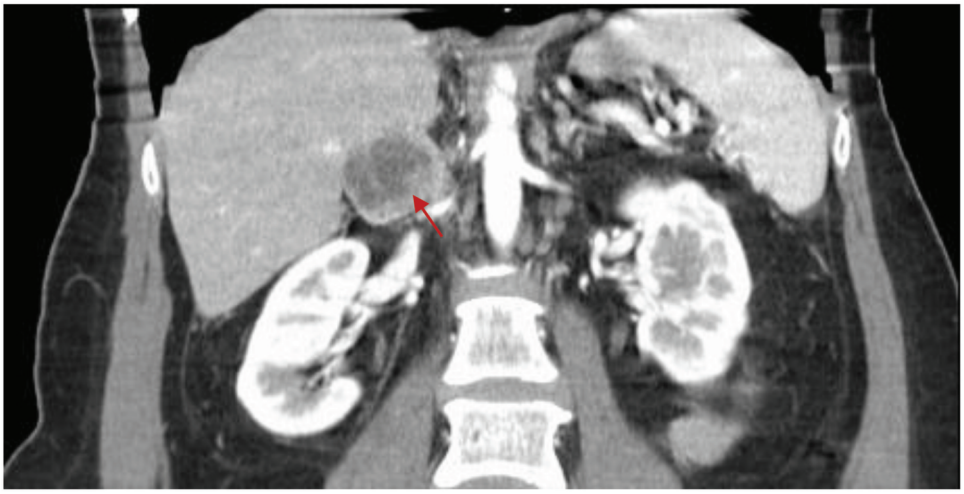

Figure 1. Coronal view of CT Adrenal showing 3.5 cm x 4.1 cm complex right adrenal mass (red arrow).

briefly considered as the underlying etiology for her clinical presentation; however, an erythrocyte sedimentation rate of 0 mm/hr pointed against this diagnosis.

Unexpectedly, without a firm grip on the diagnosis, patient started feeling better during her second hospital day. Her chest pain started to improve, oxygen requirement went down from 15 L to room air, lactic acid trended down from 6.1 mmol/L to 2.0 mmol/L, and blood glucose steadily decreased to 179 mg/dL on the insulin drip. Her AST improved from 133 U/L to 39 U/L and ALT from 110 U/L to 41 U/L. Prompt and unanticipated improvement suggested that a functioning tumor could be the underlying etiology for her clinical presentation. Diagnostic work-up for pheochromocytoma revealed plasma metanephrine: 2.13 nmol/L (0.00–0.49 nmol/L) and normetanephrine: 4.15 nmol/L (0.00–0.89 nmol/L). She was started on doxazosin 2 mg daily and carvedilol 3.125 mg two times daily. CT Adrenal with and without contrast showed 3.5 × 4.1 cm complex right adrenal mass suspicious for pheochromocytoma (Figure 1).

Patient subsequently had a robotic assisted laparoscopic right adrenalectomy with histochemical features consistent with pheochromocytoma. Echocardiogram 2 months after her right adrenalectomy showed resolution of her cardiomyopathy.

3. Discussion

This patient had cardiomyopathy, pulmonary edema, acute kidney injury, acute liver injury, hyperglycemia without a history of diabetes, and elevated lactic acid. This presentation of cardiomyopathy in the setting of multiple organ failure is an extremely rare presentation for pheochromocytoma. She was initially considered to have three separate diagnoses of STEMI, hyperglycemia, and sepsis. Normal coronary vessels on cardiac angiography with cardiomyopathy and unexpected improvement in clinical symptoms over a short time period suggested that a functioning endocrine tumor could be the underlying etiology for her clinical presentation. An incidental finding of an adrenal mass on CTPA was crucial in suspecting the final diagnosis of pheochromocytoma.

Cardiomyopathy in pheochromocytoma can be explained by several mechanisms. The stimulation of beta-1 and beta-2 receptors increases intracellular Ca2 + within the myocardial cells causing positive inotropic and chronotropic effect [2]. Overstimulation during pheochromocytoma causes an increase in myocardial oxygen demand that outweighs oxygen delivery causing 'functional' hypoxia [3]. In addition, oxidation of epinephrine by tyrosine produces adrenochrome causing vasoconstriction of coronary arteries which reduces the oxygen delivery to the myocardium [4]. Adrenochrome also exerts a direct toxic effect on the myocardium by inhibiting oxidative phosphorylation and decreasing calcium binding which causes reduced contractile force and extensive necrotic damage [3].

Several case reports have reported cardiomyopathy [4–6], pulmonary edema [7], and hyperglycemia [8] as isolated complications of pheochromocytoma. However, the concomitant presence of all these complications along with multi-organ failure made the diagnosis and management particularly challenging in this patient.

Pheochromocytoma is a rare tumor with a variety of clinical presentation secondary to the catecholamine effect on different organ systems. Here, we present a rare case of undiagnosed pheochromocytoma that presented with catecholamine-induced cardiomyopathy and multiple organ failure. Since complications from pheochromocytoma are reversible with treatment, it is prudent that clinicians recognize these various clinical presentations early and initiate treatment to prevent adverse outcomes.

Disclosure statement

No potential conflict of interest was reported by the authors.

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