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Case Report

CASE REPORT

A vicious cycle of acute catecholamine cardiomyopathy and circulatory collapse secondary to pheochromocytoma

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Abstract

Acute catecholamine cardiomyopathy is an uncommon, life-threatening manifestation of pheochromocytoma. The massive release of catecholamines from the adrenal medulla and their toxic effects on the coronary vessels and the cardiac myocytes play a significant role in the pathogenesis of cardiomyopathy in patients with pheochromocytoma. Severe manifestations, such as acute catecholamine cardiomyopathy, may be the initial presentation, especially in unsuspected and untreated pheochromocytoma cases. The clinical course of catecholamine-induced cardiomyopathy is unpredictable as patients may rapidly deteriorate into circulatory collapse and multisystem crisis. We report a case of a 25-year-old man who presented with catecholamine-induced cardiomyopathy.

INTRODUCTION

Pheochromocytoma is a rare neuroendocrine tumor of enterochromaffin cells in the adrenal medulla or extra-adrenal sympathetic paraganglia. Hereditary pheochromocytoma accounts for up to 25% of pheochromocytoma cases [1]. Its clinical presentation is highly variable and a high index of suspicion should be maintained. Here, we describe a case of acute catecholamine cardiomyopathy and circulatory collapse in a patient with hereditary pheochromocytoma syndrome.

CASE REPORT

We present the case of a 25-year-old Hispanic male with a past medical history of intermittent asthma who presented to the emergency room with acute onset of headache, nausea and vomiting. He had recently sustained mild trauma to his head, but denied loss of consciousness. He endorsed no known personal or family history of hypertension. Vital signs on presentation were temperature 36.7°C, blood pressure 190/130 mmHg, heart rate ranging from 114 to 165 beats per minute, and respiratory rate of 26–32 breaths per minute, with 100% oxygen saturation at room air. Chest radiograph revealed severe pulmonary edema with normal-sized heart. CT head revealed small right frontal subarachnoid hemorrhage. While in the emergency room, the patient developed sudden onset substernal chest pain, tachycardia and dyspnea resulting in respiratory failure requiring intubation and ICU admission. EKG revealed sinus tachycardia with ST depression in the lateral leads (Fig. 1). Cardiac

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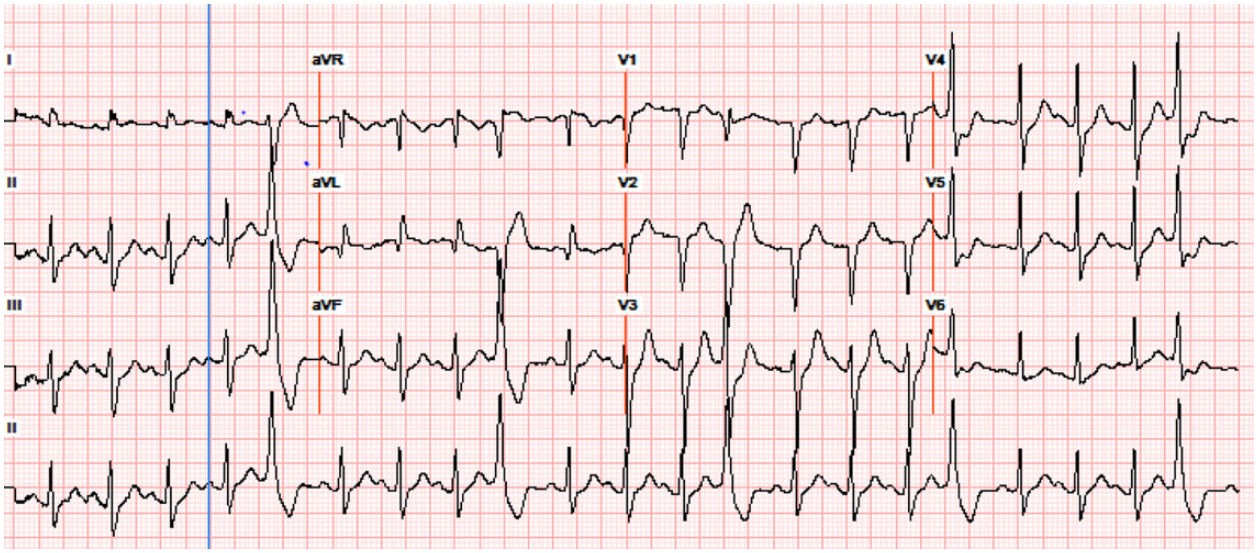

Figure 1: The patient's admission EKG.

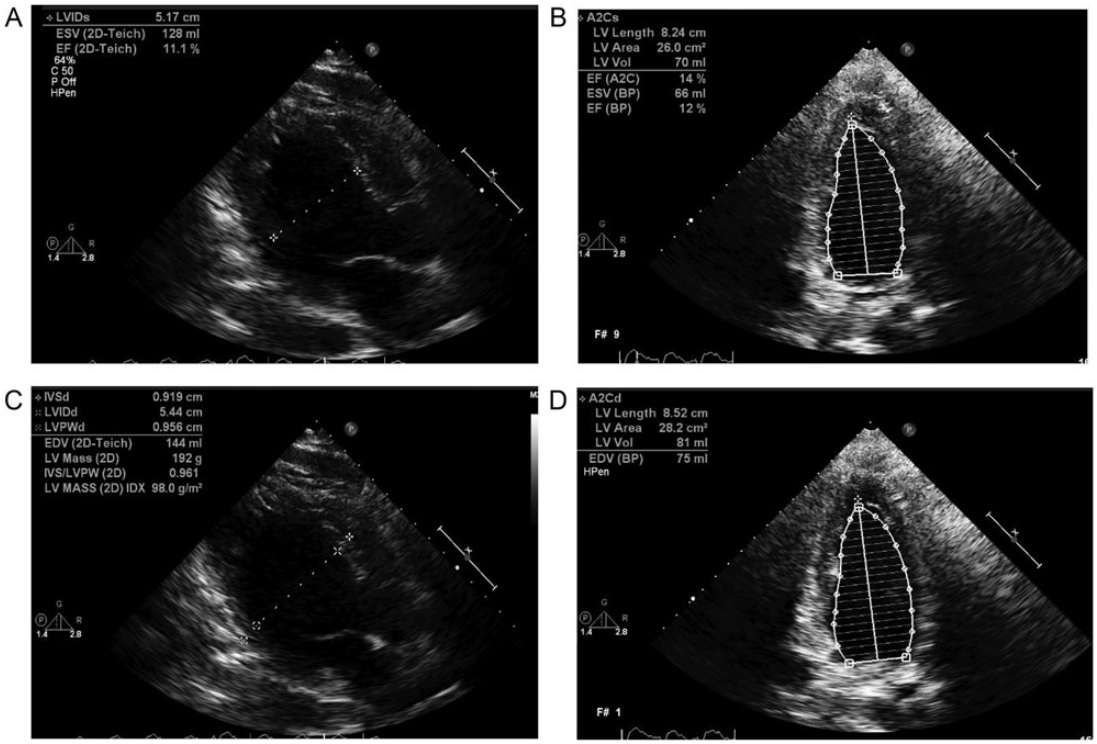

Figure 2: Echocardiographic parasternal long-axis view and two-chamber view of the patient's heart in systole (**A** and **B**, respectively) and in diastole (**C** and **D**, respectively).

enzymes were elevated with troponin I levels peaking at 213.6 μ/l (normal <0.09 μ/l). Transthoracic echocardiogram showed diffuse hypokinesis with ejection fraction of 12% (Fig. 2 and Supplementary Video 1). The patient continued to deteriorate, developed hypotension refractory to vasopressors and ultimately passed away from multiorgan failure.

Plasma-free normetanephrine and metanephrine levels drawn prior to initiation of vasopressors were found to be unmeasurably high (both >20 000 pg/ml). Autopsy report revealed right and left adrenal masses measuring 25 and 100 mm in maximum diameter, respectively (Fig. 3), consistent with pheochromocytoma. An old death certificate was subsequently found at the patient's family home, which revealed that his mother had died of circulatory collapse at our medical center secondary to pheochromocytoma while giving birth to the patient, who himself lost his life from the same disease, in the same hospital, 25 years later. It was ultimately revealed that some estranged maternal relatives had known MEN 2A syndrome. However, the patient and his immediate family were all unaware of these diagnoses.

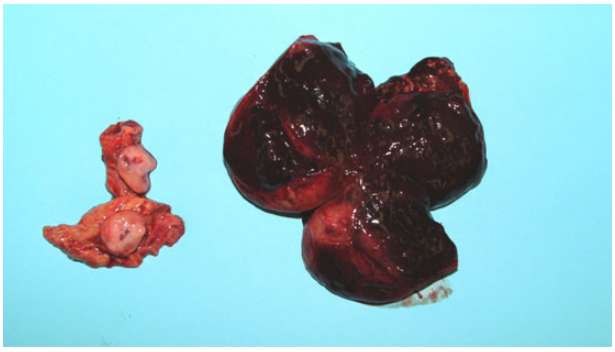

Figure 3: The patient's right and left adrenal glands, respectively (image not to scale).

DISCUSSION

Acute catecholamine cardiomyopathy is a life-threatening cardiac manifestation of pheochromocytoma characterized by surges in catecholamine levels causing acute myocardial injury and dysfunction. The mechanism of injury includes direct cardiac myocyte damage, increased myocardial oxygen demand and decreased oxygen supply [2]. Only 11% of patients with pheochromocytoma or functional paragangliomas will develop acute catecholamine cardiomyopathy [3]. Patients with unsuspected tumors may be left untreated until excess catecholamine levels rise to near-lethal levels. Consequently, in >90% of acute catecholamine cardiomyopathy cases, multisystem crisis and circulatory collapse will be the first clinical manifestation of pheochromocytoma. The successful treatment of multisystem crisis requires prompt diagnosis, intensive preoperative medical therapy to achieve blood pressure control and volume expansion, followed by emergency tumor removal.

Preoperative blood pressure control is achieved using α- and β-blocking agents. Phenoxybenzamine, a non-selective α-blocker, is recommended for 10–14 days to achieve effective α-blockade. Other selective α_1-blockers, such as doxazosin or prazosin, can also be used. β-Blockers, such as metoprolol and atenolol, are required to prevent catecholamine-induced tachycardia, but should never be used without an α-blocker as the patient may develop hypertensive crisis due to unopposed α-adrenergic stimulation [4]. Oral calcium channel blockers may be added to α- and β-blockers if blood pressure control is suboptimal. Some patients, particularly undiagnosed and untreated cases, may present with acute hypertensive crisis. Initial blood pressure control can be achieved by intravenous administration of nitroprusside, phentolamine or nicardipine [5].

MEN 2A syndrome is a subclass of the MEN 2 syndrome, an autosomal dominant disease caused by rearranged during transfection (RET) proto-oncogene germline mutations. The syndrome is characterized by medullary thyroid carcinoma, primary parathyroid hyperplasia and pheochromocytoma. Medullary thyroid carcinoma is found in 90% of all MEN 2A cases. Some patients may present with one or two clinical features of MEN 2A. In these patients, a positive RET proto-oncogene mutation or a first-degree family history of MEN 2A is required for the diagnosis of MEN 2A [6]. Our patient did not have hypercalcemia and his autopsy did not reveal any abnormalities suggestive of hyperparathyroidism or medullary thyroid cancer. Besides his family history of MEN2A, his only presenting entity was pheochromocytoma.

It is recommended that patients with genetic cancer susceptibility conditions, such as medullary thyroid cancer or pheochromocytoma, and all patients with a personal history or family history of MEN 2 syndrome, should be offered germline RET mutation screening along with pre- and post-test genetic counseling [6, 7]. The present case underscores the importance of genetic testing and counseling of patients and families with MEN2A to avert future tragedies.

Guarantor: Pavlos Msaouel MD, PhD.

SUPPLEMENTARY MATERIAL

Supplementary material is available at *Oxford Medical Case Reports* online.

CONFLICT OF INTEREST STATEMENT

None declared.

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