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Case Report

Paraganglioma masquerading as acute myocardial infarction and cardiogenic shock

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ABSTRACT

Paragangliomas, extra-adrenal pheochromocytomas, are rare catecholamine-secreting tumor. A 34-year-old lady admitted with diagnosis of ST elevation acute myocardial infarction with cardiogenic shock. Left ventricular function, severely depressed, returned to normal after initial stabilization. Coronary angiogram was normal. A para-aortic paraganglioma was diagnosed during the patient's work-up with biochemical studies, computed tomography of abdomen and functional radioisotopes imaging and was eventually surgically resected.

This case shows that acute myocardial infarction may be the initial manifestation of these neuroendocrine tumors. Hypertensive emergency, much less elevated blood pressure may not be present at time of presentation.

1. Introduction

Paraganglioma is a rare catecholamine-secreting tumor that arises in the sympathetic or parasympathetic nervous system and usually presents with benign manifestations, whose typical clinical presentation includes the triad of headache, palpitations, and diaphoresis. However, a wide range of signs and symptoms may be present, including a variety of life-threatening cardiovascular syndromes, such as hypertensive crisis, shock or profound hypotension, acute heart failure, acute myocardial infarction (AMI), arrhythmia, cardiomyopathy, myocarditis, dissection of an aortic aneurysm, and acute peripheral ischemia.[1] Failure to identify a paraganglioma in these situations may be fatal.

We describe a challenging diagnosis of paraganglioma with many cardiovascular manifestations, which could have been missed due to the absence of typical symptoms.

2. Case report

A 34-year-old woman was transferred to our tertiary care hospital from a local hospital with complaints of retrosternal

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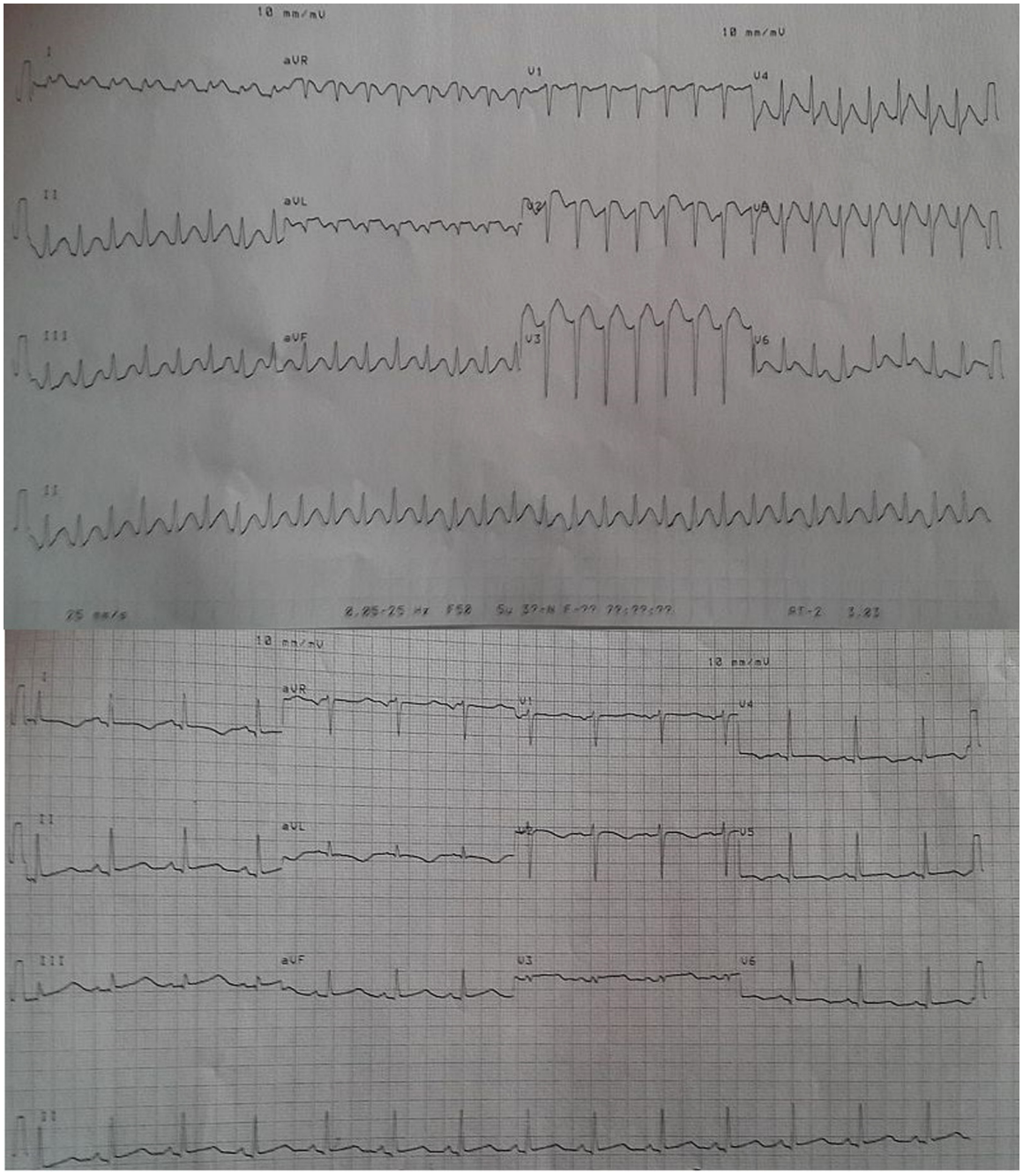

Fig. 1 – Pictures of ECGs: (A) at admission and (B) after 2 days.

chest pain, diffuse sweating, shortness of breath, nausea, and vomiting since morning.

On arrival in Emergency Department, she was found to be in cardiogenic shock, severe lactic acidosis with pulmonary edema requiring intubation, assisted ventilation, and inotropic support. On examination, she was severely dyspnoeic with sinus tachycardia and hypotension. Electrocardiogram (ECG) revealed ST-segment elevation in anterolateral leads (Fig. 1A) and strongly positive cardiac enzymes. Her serum creatinine level was 1.9 mg/dL. 2D-Echocardiogram (ECHO) showed global hypokinesia and severe left ventricle dysfunction with ejection fraction of 30%.

An Initial diagnosis of STEMI (ST-segment elevation myocardial infarction) with cardiogenic shock was made and an emergency coronary angiogram was performed. This showed widely patent coronary arteries (Fig. 2).

She was kept on ventilator support for next 2 days. Her ST-changes became normal on ECG (Fig. 1B). Review 2D-echo revealed LV function improved spontaneously to mild dysfunction with EF 55% and no regional wall motion

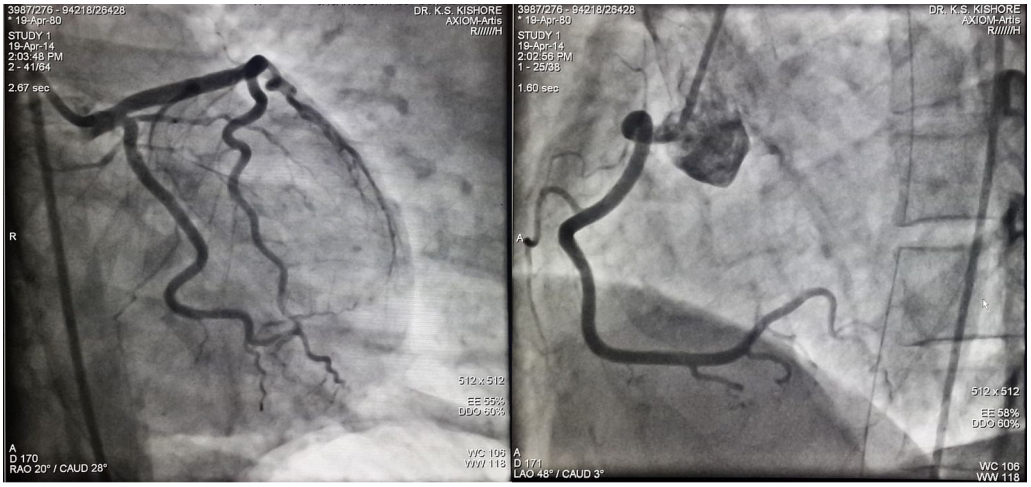

Fig. 2 – Coronary angiogram images showing widely patent coronary arteries.

abnormality. She was extubated and history was reviewed with patient which revealed an episode of severe headache and palpitation 2 years back when she was found to have severe hypertension (220/120 mm Hg) and was started on antihypertensives. But she had discontinued after a week when her symptoms resolved. She was found to have normal blood pressure recordings, and she was not investigated further. She gave history of on and off episodes of increased fatigability, sweating, and palpitation associated with fluctuating blood pressure but was never investigated for same.

She was shifted to general ward where she complained of giddiness on standing and was found to have paroxysmal hypertension and postural hypotension. She was evaluated for secondary causes of hypertension considering her young age. Ultrasound abdomen revealed a well-defined mass lesion near the hilum of left kidney (Fig. 3). Doppler study revealed no segmental renal artery stenosis and normal renal veins on both sides. Contrast-enhanced CT scan of abdomen revealed well defined, well enhancing left paraaortic retroperitoneal mass lesion inferior to the level of left renal vessels (probably paraganglioma?) (Fig. 4).

Endocrinologist's opinion was taken and 24-h urine metanephrines levels came out positive. She underwent Gallium-68 Dotanoc and 18-fluoro-2-deoxyglucose (FDG) Positron Emission Tomographic (PET) scan which revealed a FDG avid mass lesion in left paraaortic region (Fig. 5) and confirmed the diagnosis of left paraaortic paraganglioma.

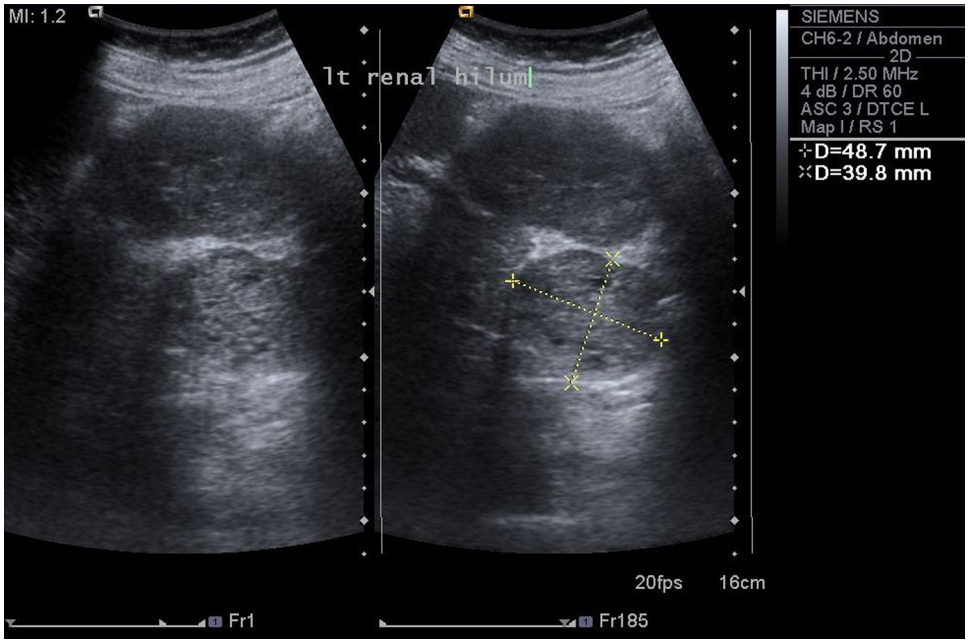

Fig. 3 – USG abdomen image showing well-defined mass lesion near the hilum of left kidney.

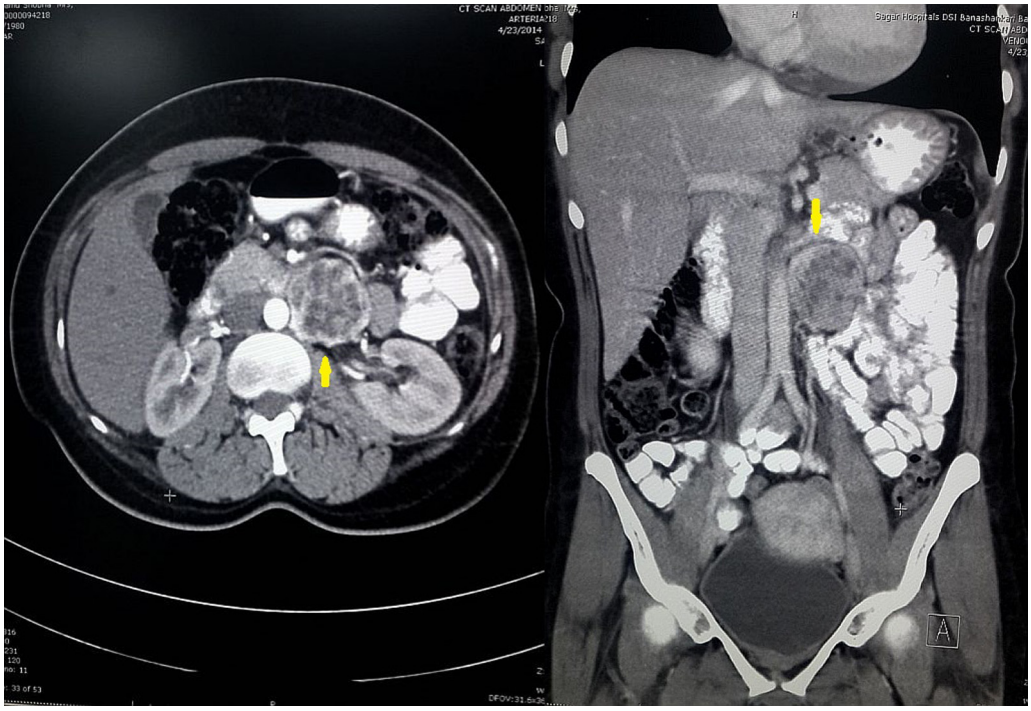

Fig. 4 – Contrast-CT Abdomen: transverse and coronal films showing well defined, well enhancing left paraaortic retroperitoneal mass lesion.

She was put on treatment with alpha blocker, and she further underwent surgery for the removal of the paraganglioma successfully. Biopsy was morphologically consistent with extra-adrenal paraganglioma.

3. Discussion

Extra-adrenal pheochromocytomas, better known as paragangliomas, are neuroendocrine tumors that produce, store, and secrete catecholamines. These neoplasms may occur in all age groups but are most common in young and middle-aged adults.

Pheochromocytoma crises resembling AMI have been reported.[2] Angina and acute myocardial infarction may occur in the absence of coronary artery disease due to severe coronary spasm, direct myocardial damage by catecholamines, and increased oxygen uptake as a result of tachycardia and increased afterload contributing to ischemia.

The case presented herein demonstrates myocardial stunning secondary to severe catecholamine excess, with the rapid recovery of the LV function. The patient presented with features of acute STEMI with acute heart failure and initially demonstrated profound myocardial dysfunction with a severely depressed LVEF.

Case reports of catecholamine-induced cardiomyopathy and myocardial stunning, either from paraganglioma or sudden emotional stress, have shown that cardiac dysfunction can be reversible. Reported recovery times as documented by serial echocardiography have varied from 3 weeks to over a year.[3,4] Our patient improved within 48 h of presentation, and her LVEF returned to baseline by day three of hospitalization. To the best of our knowledge, our patient had the shortest reported length of time for the recovery of LV function from a release of excess catecholamines.

Acute heart failure or pulmonary edema requires immediate treatment, because the prognosis for patients with paraganglioma presenting with acute heart failure is poor, and death due to pulmonary edema may occur within 24 h of the onset of such complaints.[5] Cardiac changes were found to be reversible in most cases after the institution of appropriate medication or excision of the pheochromocytoma.[6]

Biochemical diagnosis of paraganglioma is done by measurements of fractionated metanephrines and catecholamines in urine and/or plasma. Imaging studies provide information about the location of the tumor, vascularization and relationship to adjacent organs. The methods used are computed tomography (CT) and magnetic resonance imaging (MRI). If the CT or abdominal and pelvic MRI is negative in the presence of clinical and biochemical evidence of a catecholamine-secreting tumor, the next step is to apply functional studies with radioisotopes images using metaiodobenzylguanidine (MIBG), Gallium-68 Dotanoc or 18-fluoro-2-deoxyglucose (FDG) - PET.[7] The definitive diagnosis is achieved at the time of resection. A biopsy (incisional or fine needle aspiration) is contraindicated in a patient with suspected paraganglioma because it can trigger a severe hypertensive crisis.

In our case, elevated levels of urine and plasma catecholamines supported the diagnosis. CT scan and MIBG uptake

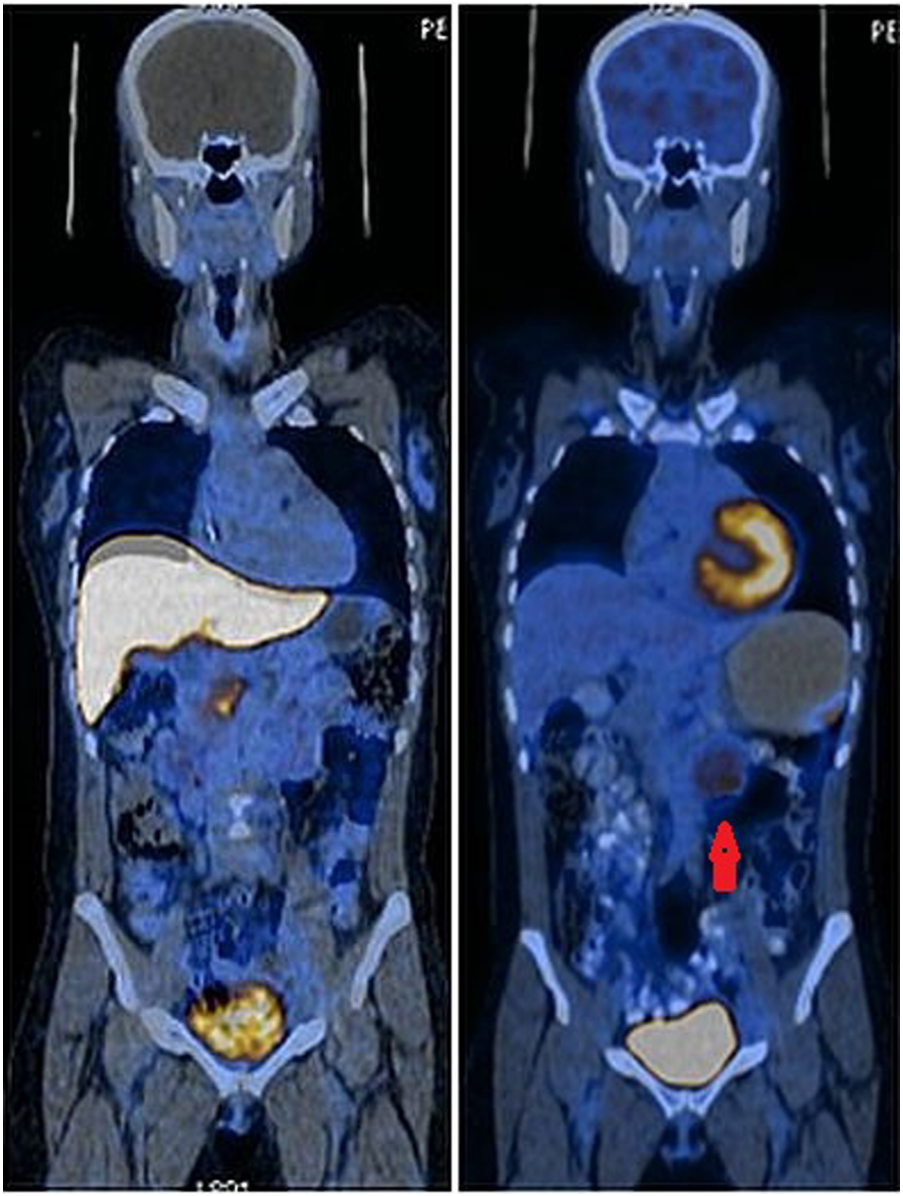

Fig. 5 – Fusion Images of Ga-68 Dotanoc and FDG PET scan & CT scan showing the FDG avid Paraaortic mass in left paraaortic region.

scan corroborated it. The histopathological examination of the resected specimen confirmed the diagnosis.

4. Summary

Paraganglioma may lead to important emergency situations. Our case shows that cardiac chest pain leading to an acute myocardial infarction may be the initial manifestation of a paragangliomas in the absence of prior symptoms or significantly elevated blood pressure. This is a good example of how challenging the diagnosis of pheochromocytoma can be, especially when it lacks the typical clinical picture, and is manifested by confounding cardiovascular system events.

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