

## Case Report

# Giant cystic pheochromocytoma masquerading as acute coronary syndrome and transient left ventricular dysfunction: a case report

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## ABSTRACT

Objective of present study to present an atypical manifestation of pheochromocytoma and to illustrate the difficulty in managing an acute coronary syndrome and left ventricular dysfunction during a hypertensive crisis attributable to pheochromocytoma. We present the clinical history, physical findings, lab results and imaging studies of a 62-year-old man with acute coronary syndrome later found to have an adrenal mass. A 62-year-old man was suspected of having myocardial ischemia on the basis of symptoms of paroxysmal chest discomfort, diaphoresis, ST-segment elevation on an electrocardiogram, and elevated levels of cardiac enzymes. Coronary CT angiography was normal. Echocardiography revealed substantial ballooning of the apical, anterior, and inferior cardiac walls, consistent with catecholamine induced cardiomyopathy. He had a history of labile hypertension, headache, diaphoresis and palpitations of 4 years' duration. A right adrenal mass detected on Magnetic resonance imaging and increased plasma catecholamine levels were consistent with a pheochromocytoma. Treatment with prazosin and labetalol was initiated, and he underwent a right adrenalectomy, which confirmed the pheochromocytoma. Pheochromocytomas must be considered in the setting of acute coronary syndrome especially with presence of labile blood pressure. Inadvertent fibrinolysis can be avoided with proper clinical suspicion and timely coronary imaging.

**Keywords:** Cardiomyopathy, Labile hypertension, Pheochromocytoma

## INTRODUCTION

Pheochromocytoma is a rare neuroendocrine tumour with an estimated annual incidence of 1-4/10<sup>6</sup> population.<sup>1</sup> Called the 'great mimic', it is fascinating as well as challenging to the clinician because of its lethal potential if left undiagnosed and at the same time the possibility of curative treatment. Derived from the chromaffin cells of the embryonic neural crest, they produce catecholamines responsible for the classic symptoms of headache, palpitation, anxiety and diaphoresis. The tumor can occur at any age with equal gender distribution.<sup>2</sup>

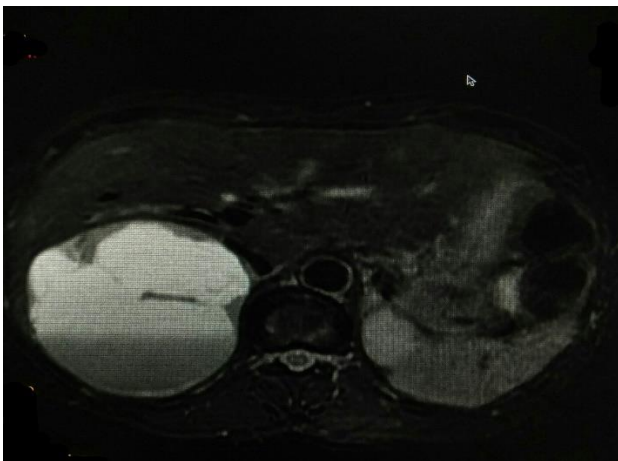
Giant cystic pheochromocytomas are rarer, essentially described as being non-functional. They generally present

with vague abdominal symptoms and are discovered on evaluation as they lack the typical symptoms of pheochromocytoma due to the absence of catecholamine secretion.<sup>3</sup> Here we describe a case of a functional giant pheochromocytoma presenting as ST segment elevation myocardial infarction.

## CASE REPORT

A 62 year old man presented to the emergency department of this tertiary hospital with complaints of chest discomfort, diaphoresis and dizziness of one hour duration. There was no prior history of hypertension, diabetes mellitus, dyslipidemia or prior hospitalisations. Past history was significant only for brief episodes of

dizziness and sweating accompanied by headache which subsided on rest. These happened once in a year and did not require any medical attention except for occasional analgesic use. He was a non-smoker and did not use alcohol. Examination showed a well-built man, with profuse sweating, BP of 100/70mm Hg in supine position, PR of 90 beats/ min, 99% saturation on room air. Bilateral lung bases were clear. Rest of general and systemic examination was within normal limits. Suspecting acute coronary syndrome, an ECG was taken which showed ST segment elevation. Cardiac troponin levels were elevated. Baseline investigations also showed a possible pre-renal acute kidney injury. Aspirin, clopidogrel and atorvastatin were administered. Because of non-availability of primary PCI in our centre and no contraindications for thrombolysis, he was shifted to intensive care unit for thrombolysis. Upon arrival in the ICU, his BP was 230/ 150mm Hg. Postural variation of blood pressure was noted, with orthostatic hypertension accompanied by sweating and headache, and supine normotension to hypotension. Thrombolysis was withheld and immediate echocardiography was performed which showed apical ballooning suggestive of stress induced cardiomyopathy. He was started on heparin infusion for thromboprophylaxis and coronary computed tomography angiography was performed. The coronary vasculature was normal, consistent with takotsubo cardiomyopathy. Clinical suspicion of pheochromocytoma in view of paroxysmal headache, diaphoresis and dizziness prompted an urgent ultrasonography of the abdomen which revealed a large right adrenal mass measuring 12cm in length and 12 cm in width with cystic areas within. Patient continued to have frequent hypertensive crisis episodes, labile blood pressure and was continuously monitored in the ICU. Magnetic resonance imaging of the abdomen confirmed mass in the right adrenal region (Figure 1).



**Figure 1: Magnetic resonance imaging of the abdomen.**

Though 24 hr urinary fractionated metanephrines are described as the screening test, we chose to do plasma catecholamine levels for a faster diagnosis and high likelihood of a positive result in view of continuing

symptoms and adrenal mass on imaging. This was found to be highly elevated. The ST segment changes on the ECG normalised by day 5 of hospitalisation. Diagnosis of giant pheochromocytoma with catecholamine induced cardiomyopathy was made.



**Figure 2: Post-operative resected specimen.**

### Management

The patient was started on prazosin on small doses and titrated up for control of hypertensive crisis. Labetalol was added once he was stable on the alpha blocker. Heparin infusion was continued. Hypertensive crises reduced in frequency and were well controlled. The Surgical and anaesthesia team was contacted and a right adrenalectomy under general anaesthesia was planned. The procedure went uneventfully, the resected mass measured cm with areas of cystic change (Figure 2). Postoperatively the patient required no anti-hypertensives. He remains on follow up, with no further recurrence of symptoms. Repeat echocardiography showed complete resolution of left ventricular dysfunction.

### DISCUSSION

Pheochromocytomas are rare neuroendocrine tumors being responsible for less than 1% of cases of investigated secondary hypertension.<sup>4</sup> It is often said that a physician sees only one case in his lifetime, owing both to the rarity as well as the myriad presentations. Typical symptoms, including the triad of headache, labile hypertension and diaphoresis, raise the suspicion of pheochromocytoma, but the absence of these symptoms can present a considerable diagnostic challenge. Tumors that produce only norepinephrine generally manifest with uncontrolled hypertension; however, the tumors that produce both epinephrine and norepinephrine manifest with episodic hypertension. Because pheochromocytomas are not innervated, catecholamine release is not precipitated by neural stimulation. Multiple mechanisms have been postulated as triggers for catecholamine release including direct pressure, medications, and changes in tumor blood flow.

The symptom with highest sensitivity of about 80% for pheochromocytoma, a history of hypertension was conspicuously absent in our case. This highlights the unique nature of paroxysmal hypertension in pheochromocytoma and the fact that blood pressure can be entirely normal in the inter crisis period.<sup>5</sup> While orthostatic hypotension is well described in pheochromocytoma with a sensitivity varying from 21-50%, this patient had orthostatic hypertension. Orthostatic hypertension is an underappreciated clinical sign, owing to lack of knowledge among physicians and varying definitions.<sup>5</sup> The magnitude of increase in sphygmomanometric pressure required for a diagnosis of orthostatic hypertension is an increase in systolic BP of 20 mmHg or more with no defined change in diastolic blood pressure. Pheochromocytoma is cited as a reason for the same.<sup>6</sup>

Cardiac complications as the presenting symptom of pheochromocytoma is rare, with acute myocardial infarction contributing only 4%.<sup>7</sup> Patients with cardiac complications are found to have similar demographics and cardiac risk factors as rest of pheochromocytoma patients. As is consistent with our patient, those with cardiac complications are found to have larger tumors, higher catecholamine levels and lower ejection fraction.<sup>7</sup> Prognosis is excellent with good cardiac recovery following complete surgical resection.

Giant cystic pheochromocytomas are rare, a recent literature review summarising all cases of pheochromocytoma over 10 cm diameter reported about 37 cases worldwide and only 6 from India.<sup>8</sup> Most of the cases lacked typical symptoms of pheochromocytoma.

Our case is unique in many respects. The late presentation after having paroxysmal symptoms for 4 years without hypertension, initial presentation as ST elevation myocardial infarction with labile blood pressure, subsequent development of catecholamine induced cardiomyopathy and the particularly large tumor which made surgical resection difficult were all challenging.

## CONCLUSION

Pheochromocytoma should be considered in the differential diagnosis of acute coronary syndrome. This case report stresses the importance of targeted history taking even in acute emergencies. In this era of stress on

faster management of acute coronary syndrome, failure to recognise this condition could result in unnecessary invasive interventions.

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