Pheochromocytoma Presenting as Acute Myocardial Infarction

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Abstract

Pheochromocytomas are rare neuroendocrine tumors arising from chromaffin cells of the sympathetic nervous system. Pheochromocytoma can have diverse clinical presentations, which makes the diagnosis often difficult. We present a case of adrenal pheochromocytoma presenting as acute ST elevation myocardial infarction. Catecholamine surge in patients with pheochromocytoma can cause myocardial infarction in the absence of atherosclerotic coronary artery disease. Pheochromocytoma presenting as acute myocardial infarction is very rare and it occurs in young individuals. This is a rare case report of pheochromocytoma in an elderly male presenting with acute STEMI successfully treated by resection of tumor.

Keywords: Pheochromocytoma; Myocardial infarction; Catecholamines; Hypertension

Case Report

A 62 year old farmer presented with severe epigastric discomfort, vomiting and profuse sweating. He was hypertensive and diabetic for the last four years on irregular treatment with amlodipine and metformin. His blood pressure was 190/110 and heart rate was 60/minute. Precordial examination was unremarkable except for a loud fourth heart sound. Initial ECG showed tall peaked T waves in precordial leads suggesting hyperacute changes of myocardial infarction. Subsequent ECGs showed loss of R waves with coved ST elevation and T wave inversion in leads V2, V3 and V4. There were T wave inversions in leads I,II,III,aVF, V5 and V6 (Figure 1).

His echocardiogram showed severe hypokinesia of apical interventricular septum and apex with normal left ventricular systolic function. He was subjected to an immediate coronary angiogram with intention for primary percutaneous revascularization. Surprisingly his diagnostic coronary angiogram showed normal epicardial coronary arteries and no evidence of atherosclerosis (Figure 2). So he was treated conservatively with antiplatelets, intravenous nitroglycerine and statins. His subsequent ECGs showed settling of ST segment with deep T wave inversions in precordial leads typical of serial evolutionary changes in non Q myocardial infarctions. Cardiac enzymes were estimated which showed serum troponin I level of 9 ng/ml, the normal being less than 0.05 ng/ml.

Figure 1: 12 lead ECG showing loss of R waves with coved ST elevation and T wave inversion in leads V2, V3 and V4. T wave is inverted in leads I,II,III,aVF, V5 and V6.

Figure 2: Coronary angiogram RAO caudal view showing normal left coronary artery and its branches.
the episodes along with sinus tachycardia which prompted us to suspect an alternative diagnosis in this case. His thyroid function tests were normal. In view of the labile hypertensive changes, we also considered the possibility of pheochromocytoma and proceeded with an ultra sonogram of abdomen which showed a mass in the region of right suprarenal gland. 24 hr urine metanephrine levels were elevated (14.5 mg, normal value < 1 mg/24 hrs). A contrast enhanced CT scan of abdomen confirmed the right suprarenal mass. It was a well encapsulated mass with a fluid level (Figure 3). Both kidneys and left suprarenal gland were normal. A diagnosis of pheochromocytoma was made, which presented as acute myocardial infarction due to the sympathetic surge. He was started on prazosin 2 mg three times daily which controlled the blood pressure and his symptoms got ameliorated. Echocardiogram after one week showed no regional wall motion abnormality. Subsequently he underwent excision of the pheochromocytoma. There were wide BP fluctuations during surgery. Blood pressure surges during surgery were treated with controlled infusion of sodium nitroprusside. A large cystic right suprarenal mass with multiple areas of hemorrhage was noted during surgery. Left suprarenal gland was normal. Histopathology showed sheets of chromaffin cells consistent with pheochromocytoma and multiple areas of hemorrhage and necrosis (Figure 4). Patient is asymptomatic during follow up and is doing well.

Discussion

Pheochromocytomas most commonly arise from adrenal gland (90%) and are benign in 90% of cases. They produce characteristic systemic manifestations by secretion of catecholamines, most commonly norepinephrine. Extra adrenal pheochromocytomas are more likely to be malignant (30%). Cardiac manifestations of pheochromocytoma include hypertension, myocardial hypertrophy, myocarditis, cardiomyopathy, pulmonary edema, cardiogenic shock, arrhythmias and rarely myocardial infarction. Hypertension is the most frequent (>70%) cardiovascular manifestation of pheochromocytoma [1]. Hypertension is paroxysmal only in 50% of the cases and is persistent in the rest which makes diagnosis difficult. Quantitative 24-hour urinary metanephrine levels are the most reliable screening procedures for pheochromocytoma. Imaging modalities include computed tomography which can easily detect adrenal lesions and MRI which has got high sensitivity and specificity especially for extra adrenal-tumors.

Figure 3: Contrast enhanced CT scan of abdomen showing a well encapsulated mass with a fluid level in the region of right suprarenal gland (marked by arrow).

Figure 4: Histopathology showing sheets of chromaffin cells with multiple areas of hemorrhage and necrosis.

Pheochromocytomas have been rarely associated with acute myocardial infarction. Significant coronary atherosclerosis is present in less than half of these cases. The mechanism of myocardial infarction or segmental myocardial dysfunction associated with pheochromocytoma has been linked to coronary spasm or a direct toxic effect induced by catecholamines [2]. Catecholamines increase left ventricular work by inducing left ventricular hypertrophy from hypertension and increase in heart rate. Changes in coronary arteries include thickening of media potentially impairing blood flow to myocardium and coronary vasospasm [3]. Catecholamines can cause cardiac myocyte apoptosis [4]. Prolonged vasospasm can induce endothelial injury and promote platelet aggregation. The myocardial stunning-like phenomenon during pheochromocytoma crisis has also been reported. The myocardial regional wall motion abnormality is often reversible as in our case. Anti-ischemic therapy with beta-blockers can be detrimental before establishing alfa receptor blockade in these patients. Our patient did not receive beta blockers on account of initial low heart rate though he had tachycardia during catecholamine surge.

Angiography carries high-risk in patients with pheochromocytoma as radiocontrast media can precipitate hypertensive crisis or congestive heart failure due to catecholamine release from the adrenal medulla [5]. Antihypertensive agents advised include nonselective adrenergic antagonists (like labetalol), alfa antagonists (like prazosin or phentolamine), or vasodilatory agents like nitroglycerine or sodium nitroprusside. Hemorrhage into the pheochromocytoma can precipitate catecholamine crisis and myocardial infarction [6]. In our case also, hemorrhage into the mass might have precipitated the catecholamine surge accounting for the myocardial infarction.

Summary

This case illustrates an uncommon presentation of pheochromocytoma crisis presenting as acute ST elevation myocardial infarction in an elderly man. A normal coronary angiogram indicates
that coronary vasospasm or direct myocardial toxicity is the mechanism. A high index of suspicion for pheochromocytoma is needed in such cases as it is a lethal but potentially curable disease.

References


