CASE REPORT

Phaeochromocytoma presenting with ST segment elevation myocardial infarction

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SUMMARY

Phaeochromocytoma is a rare endocrine disorder with different cardiovascular presentations. In this brief report, we discuss a case of a 59-year-old woman who presented with acute ST segment elevation myocardial infarction secondary to phaeochromocytoma. Coronary angiogram showed non-obstructive coronary artery disease.

BACKGROUND

Phaeochromocytoma is a tumour, consisting of chromaffin cells, that usually originates from the adrenal medulla or, rarely, from an extra-adrenal site. The hallmark of this condition is excessive catecholamine release. It has been estimated to occur in 0.05–0.2% of patients with diastolic hypertension.¹

It is a deceptive condition with a very wide range of clinical symptoms and signs, ranging from an incidental asymptomatic mass to catastrophic life-threatening events such as seizures, intracranial bleeding or cardiogenic shock.¹ The clinical picture of a phaeochromocytoma is directly related to the episodes of circulatory catecholamine surge. These include paroxysms of severe headaches, sweating, anxiety, tremulousness, chest and/or abdomen pain, nausea and vomiting, as well as generalised weakness and labile blood pressure.¹ Cardiac features of phaeochromocytoma include different types of arrhythmias, cardiomyopathy and congestive heart failure, as well as characteristics of acute coronary syndrome.^{1 2}

The mechanism of cardiac injury secondary to catecholamines is not well understood, but

exposure to high catecholamine levels has been shown to disrupt permeability of myocardial cells with secondary increases in intracellular calcium levels, which has a direct toxic effect on cardiomyocytes. Also, free oxygen radical formation and high demand on oxygen have been implicated in myocardial injury in individuals with phaeochromocytoma. In addition, high catecholamine levels can cause myocardial ischaemia due to severe vasospasm and/or endothelial dysfunction.³ In experimental animal models, histopathological analysis of cardiomyocytes exposed to high levels of catecholamines showed evidence of cardiomyocyte necrosis with contraction band formation, cellular degeneration, myocarditis with inflammatory mononuclear cells infiltrates, areas of myocardial loss with fibrosis, smooth muscle hypertrophy within the myocardium and evidence of non-obstructive coronary atherosclerosis.³ Catecholamine-induced vasoconstriction can lead to ischaemic injury with multiorgan involvement that results in lactic acidosis and elevations of cardiac enzymes among other biomarkers.¹

Cardiac imaging can frequently show evidence of left ventricular hypertrophy secondary to hypertension. It can also show apical ballooning and hypokinesia in case of stress-induced cardiomyopathy, a frequent association. Otherwise, there may be non-specific regional or generalised left ventricular wall motion abnormalities.^{1 3}

Diagnosis of phaeochromocytoma is challenging as patients can present with a wide spectrum of symptoms and signs of a paroxysmal nature. A high degree of clinical suspicion is needed to

Figure 1 Patient's ECG on presentation to the emergency department, with sinus tachycardia and ST segment elevation in inferior leads.

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Figure 2 (A) Images of coronary angiogram showing normal right coronary artery (RCA) and mild atheroma in left anterior descending artery (LAD) with no evidence of obstructive coronary artery disease. (B) Images of coronary angiogram showing normal right coronary artery (RCA) and mild atheroma in left anterior descending artery (LAD) with no evidence of obstructive coronary artery disease.

recommend the appropriate investigations and to rule out phaeochromocytoma. Different biochemical markers are used to screen for phaeochromocytoma. However, plasma, and 24 h urinary metanephrine and normetanephrine, are the most sensitive initial tests for the diagnosis of phaeochromocytoma.¹

In this report, we describe a case of phaeochromocytoma presenting with ST segment elevation myocardial infarction (STEMI) in the absence of significant coronary artery disease on coronary angiography.

CASE PRESENTATION

A 59-year-old woman presented to our centre, with sudden onset chest pain and shortness of breath. On route to the emergency department, a 12-lead ECG showed ST elevation in her inferior leads (Figure 1). She was a heavy smoker with newly diagnosed non-insulin dependent diabetes, with no history of illicit drug abuse and an unremarkable family history. Clinical examination was remarkable for sinus tachycardia, with a heart rate of 104/min, respiratory rate 20/min, blood pressure 160/89 and normal heart sounds with no clinical features of heart failure.

As per standard protocol, the patient underwent an urgent coronary angiogram, which showed normal right coronary artery (RCA) and mild atheroma in her left anterior descending artery (figure 2A, B). Left ventriculogram showed reduced motion of the inferior wall and normal apical motion. Blood results showed a high sensitivity troponin level of 450 ng/L (normal range: 0–14 ng/L), and echocardiogram revealed hypokinesia of the inferior ventricular wall, consistent with myocardial infarction related to the RCA.

A second set of troponin showed an increase of up to 945 ng/L, again consistent with myocardial infarction; and follow-up ECG showed resolution of ST segment elevation with new development of deep T-wave inversion on inferior leads (figure 3). As the coronary angiogram did not fully explain the chest pain, tachycardia, the significant serial rise in troponin and the transient ST elevation, work up for an alternative diagnosis was performed. Chest X-ray was normal; D-dimer was 3.0 µg/mL (normal <0.5 µg/mL). Subsequently, we ordered a CT pulmonary angiogram (figure 4). This was negative for pulmonary embolus, but showed a large left-sided incidental adrenal mass of 4.9 cm×4.8 cm.

Urinary catecholamines and plasma metanephrine showed very high levels (plasma normetanephrine was 12.9 nmol/L

Figure 3 ECG the next day showing resolution of ST elevation with development of deep T-wave inversion in inferior leads and V6.

Figure 4 CT image showing left heterogeneous adrenal mass suggestive of pheochromocytoma.

with normal reference range of less than 1.29), consistent with a diagnosis of pheochromocytoma.

OUTCOME AND FOLLOW-UP

The patient was started on dual antiplatelet therapy for 6 months, and then aspirin for life, atorvastatin, isosorbide mononitrate, β blockers and α antagonist. She was referred for adrenalectomy, which was achieved successfully 6 months after presentation. There were no further symptomatic episodes suggestive of ischaemia and ECG returned to normal with no ischaemic changes.

DISCUSSION

This is an unusual case of pheochromocytoma presenting acutely as STEMI. Coronary angiogram showed no culprit coronary artery occlusion to explain this patient's symptoms, ST elevation on ECG, the rise in troponin or the echocardiographic finding of hypokinesia at the RCA territory. This is consistent with intense coronary spasm with the surge of catecholamines, which probably caused a significant limitation of blood flow along the RCA. This has been reported and suggested previously with cases of pheochromocytoma and STEMI,^{2 4–6} while several other cases mentioned association of pheochromocytoma with non-STEMI acute coronary syndrome.^{7–9} In each of those cases, there were symptoms suggestive of myocardial ischaemia, ECG changes and a rise in cardiac enzymes. Frequently, the imaging studies were consistent with the picture of ischaemic injury rather than stress-induced cardiomyopathy. One of the limitations of our report is the lack of documented evidence of coronary artery spasms, but this is related to the transient nature of these spasms. In cases other than of procedure-induced coronary spasms, they are rarely captured on coronary angiogram.

An alternative explanation is direct toxicity to the myocardium with the surge of catecholamine, but this is usually reported in cases of stress-induced cardiomyopathy,^{3 10} and usually comes with left ventricular angiographic, ECG and echocardiographic features different from those seen in this case.¹⁰

The consequences of this rare cardiac presentation of pheochromocytoma should raise the discussion of when to screen for pheochromocytoma in patients with primary coronary spasm

resulting in acute coronary syndrome. Given the incidence and rarity of both conditions, it may still be early to recommend blind screening for pheochromocytoma, but at least looking for more clinical features of this rare disorder is reasonable.

The other question is whether there is enough evidence to suggest screening for pheochromocytoma in cases with chest pain, abnormal ECG, high cardiac enzyme levels, normal angiogram and normal or minimally abnormal left ventricular or apical wall motion on echocardiogram. A normal angiogram and high troponin are increasingly encountered in the cardiac catheterisation laboratory, especially with the use of high sensitivity assays. Up to 21% of one cohort of patients had true myocardial infarction on cardiac MR, as shown by a recent meta-analysis.¹¹

Learning points

- ▶ Pheochromocytoma is a rare condition with various cardiovascular manifestations.
- ▶ Pheochromocytoma can present as an acute coronary-like syndrome.
- ▶ Normal coronary artery angiogram in the setting of acute cardiac ischaemic event-like scenarios should raise the suspicion of other conditions such as pheochromocytoma.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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