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CASE REPORT

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Tako-tsubo cardiomyopathy precipitated by pheochromocytoma crisis

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Abstract

We report the case of a 69 year-old Chinese lady with pheochromocytoma who developed chest pain and acute ST-segment elevation in the anterior leads on electrocardiography. She was given intravenous phenoxybenzamine for alpha-blockade before undergoing urgent coronary angiography. This revealed minor coronary artery disease. Left ventriculogram demonstrated akinesis in the apex with hypercontraction of the basal segments. The Tako-tsubo cardiomyopathy eventually resolved with surgical removal of the pheochromocytoma. This case highlights the challenging management of suspected acute myocardial infarction in pheochromocytoma crisis and also reinforces the notion that Tako-tsubo cardiomyopathy is likely precipitated by catecholamine excess. (Cardiol J 2011; 18, 5: 564–567)

Key words: pheochromocytoma, Tako-tsubo cardiomyopathy, ST-segment elevation

Introduction

Pheochromocytomas are rare catecholamine-producing neuroendocrine tumors with myriad non-specific presenting symptoms. Typical presentations include headaches, palpitations, sweatiness, pallor and paroxysmal hypertension. Rare presentations have been reported, such as acute abdominal pain, septic shock-like syndrome, hyperthermia, pulmonary oedema and myocardial ischemia [1–3].

We present an unusual case of pheochromocytoma crisis precipitating Tako-tsubo cardiomyopathy.

Case report

A 69 year-old Chinese lady, with a known history of hypertension and hypercholesterolemia for five years, presented with intermittent epigastric pain for four days. This was associated with one episode of non-bilious vomiting, one month duration of loss of appetite and loss of weight. She had also had intermittent palpitations over the previous three years.

A thorough physical examination revealed normal blood pressure of 128/80 mm Hg, normal pulse rate of 74 beats per minute, dual heart sounds with no cardiac murmur, clear lung fields. Her abdomen was soft, non-tender to palpation and there was no organomegaly.

Electrocardiography at rest was normal. Initial blood investigations revealed mild anemia with hemoglobin of 10.1 mg/dL, normal renal and liver function with a normal serum amylase level.

An ultrasound scan of her abdomen revealed normal liver and kidneys, but an incidental finding of a right adrenal mass. Computed tomographic scan of her abdomen confirmed the presence of a large 8.2 × 5.8 × 8.5 cm heterogenously enhancing, vascular right adrenal mass (Fig. 1). A diagnosis of pheochromocytoma was made after 24 hour urinary collection revealed an elevated level of noradrenaline at 4,636 nmol/day, raised normetanephrines level at 54,943 nmol/day and elevated vanillyl mandelic acid level at 58 μmol/day.

On the fourth day of hospitalization, the patient complained of acute central chest pain associated

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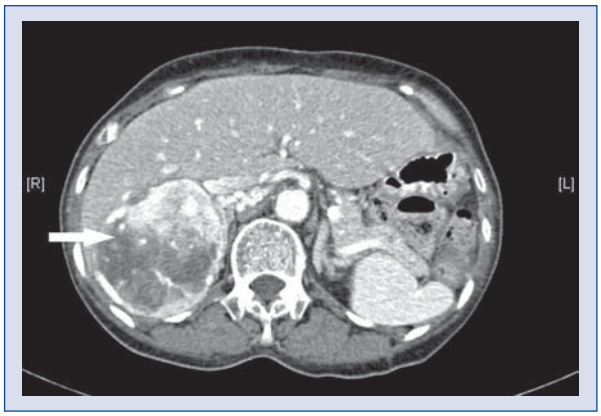

Figure 1. Computed tomographic scan of the abdomen showing a large right adrenal mass (indicated by arrow).

with diaphoresis and dyspnoea. Her blood pressure recordings increased to a maximum of 190/110 mm Hg. Electrocardiography revealed ST-segment elevation of 2 to 3 mm in leads V2, V3 and V4 but no reciprocal ST-segment depression in the inferior leads (Fig. 2). She had an elevated serum troponin I of 2.83 μg/L and raised creatine kinase-MB of 7.8 μg/L. The initial impression was that of a possible acute anterior ST-segment elevation myocardial infarction.

The patient was given intravenous phenoxybenzamine 10 mg for alpha-blockade before undergoing urgent coronary angiography. This revealed minor coronary artery disease. Left ventriculogram showed apical akinesia, inferior wall hypokinesia and basal hyperkinesia with overall left ventricular ejection fraction of 40% (Fig. 3). The final diagnosis was that of Tako-tsubo cardiomyopathy precipitated by pheochromocytoma crisis.

The patient's condition stabilized in the intensive care unit with general supportive management, oral phenoxybenzamine 20 mg thrice daily and oral propranolol 20 mg thrice daily. She eventually underwent excision of the adrenal mass about one month into her hospitalization. Histology confirmed the adrenal mass to be a pheochromocytoma with extensive necrosis. The patient recovered uneventfully following surgery. Two-dimensional echocardiography performed before discharge revealed mild concentric left ventricular hypertrophy but otherwise normal-sized heart chambers, normal left ventricular ejection fraction, absence of segmental wall motion abnormality, and normal heart valves.

Discussion

Pheochromocytomas are rare tumours of chromaffin tissue derived from the embryonic neural crest. They produce their distant effects by secreting excessive amounts of catecholamines. They are known for their 'rule of 10' whereby 10% are extra-adrenal, of which 10% are extra-abdominal, 10% are malignant, 10% occur in normotensive patients and 10% are hereditary [1–3].

Cardiac manifestations of pheochromocytoma include cardiac arrhythmias, electrocardiographic abnormalities such as ST-segment and T wave changes, ventricular hypertrophy, acute coronary

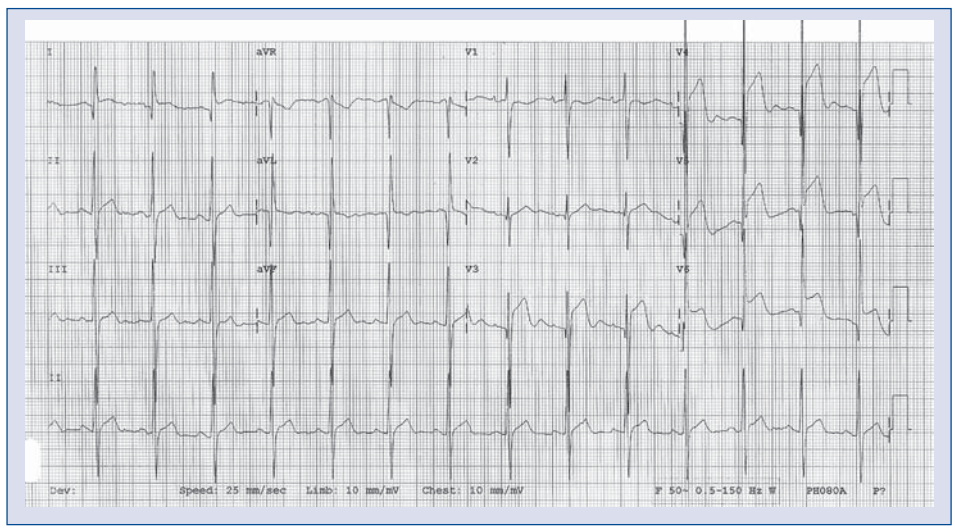

Figure 2. Electrocardiography revealed ST-segment elevation in leads V3 to V6 without reciprocal ST-segment depressions in leads II, III, aVF.

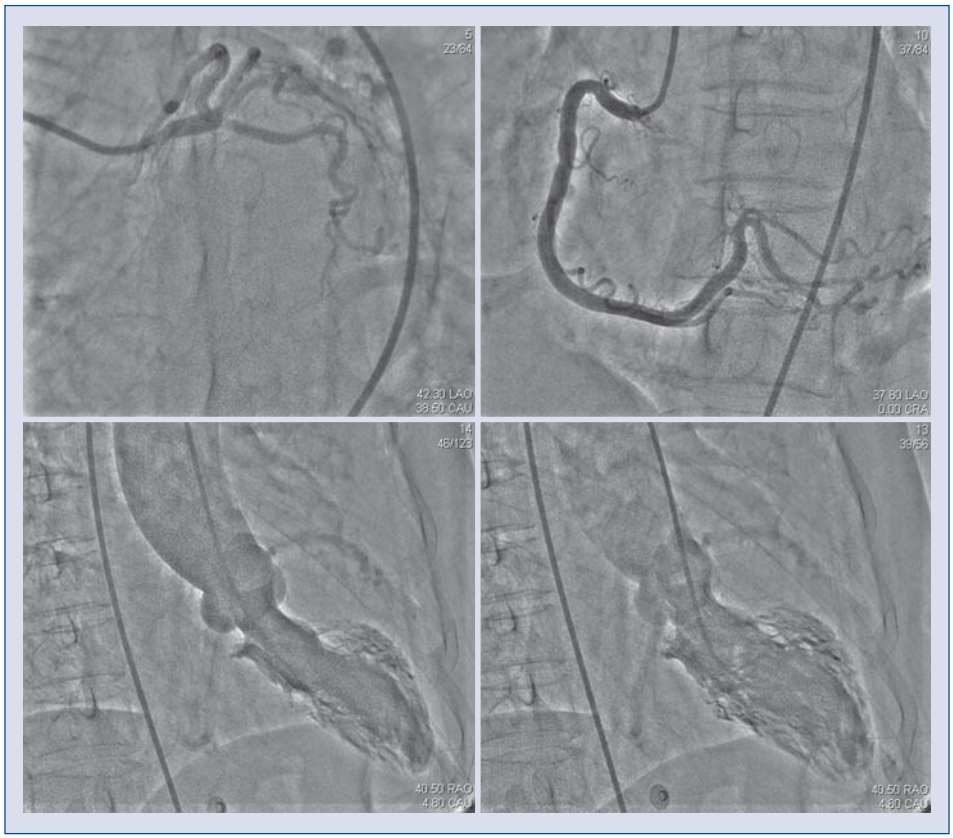

Figure 3. Invasive coronary angiography showed minor coronary artery disease and left ventriculography revealed apical akinesia with hyperkinesis of the basal segments of the heart.

syndromes, congestive heart failure secondary to myocarditis or cardiomyopathy, and cardiogenic shock [4–9]. Our patient developed acute chest pain and ST-segment elevation in the anterior leads on electrocardiography during her pheochromocytoma crisis. What was initially surmised to be an acute anterior ST-segment myocardial infarction eventually turned out to be Tako-tsubo cardiomyopathy.

Tako-tsubo cardiomyopathy is also referred to as transient left ventricular apical ballooning syndrome. It is characterized by transient left ventricular dysfunction causing apical ballooning with compensatory basal hyperkinesis, electrocardiographic changes that can mimic ST-segment elevation acute myocardial infarction, and minimal release of cardiac enzymes in the absence of obstructive coronary artery disease. It is a rare syndrome, typically seen in post-menopausal women, who present with a history of recent emotional or physical stress. With conservative treatment, the apical ballooning resolves spontaneously within an average of 18 days (range 9 to 53 days) [10].

Considerable evidence suggests that enhanced sympathetic activity may explain the transient myocardial dysfunction seen with Tako-tsubo cardiomyopathy. Plasma catecholamine and neuropeptide levels in patients with Tako-tsubo cardiomyopathy induced by acute emotional stress are markedly elevated compared to patients with myocardial infarction [11]. ^{123}I-metaiodobenzyl-guanidine imaging in patients with apical ballooning reveals impaired sympathetic innervation in the dysfunctional apex, despite normal perfusion [12]. Case reports of Tako-tsubo cardiomyopathy due to catecholamine-secreting tumors [13] and acute brain injury [14] further support the notion of sympathetically mediated myocardial stunning. This report further implicates enhanced sympathetic stimulation in the pathogenesis of this syndrome by demonstrating a causal link between catecholamine excess and the development of Tako-tsubo cardiomyopathy.

This interesting case also illustrates the challenging management of a patient with pheochromocytoma crisis who develops acute chest pain associated with ST-segment elevation on electrocardiography. Intravenous thrombolytic agents were contraindicated in this patient, in view of her severe hypertension and resultant high risk of

hemorrhagic stroke. Invasive coronary angiography would be both diagnostic and therapeutic, serving to confirm ST-segment elevation acute myocardial infarction and proceeding on to percutaneous coronary intervention if necessary. For this reason, invasive coronary angiography was carried out, albeit at significant risk to the patient. Alpha-blockade was achieved with intravenous phenoxybenzamine prior to the procedure in a bid to ameliorate the risk. Besides, Tako-tsubo cardiomyopathy cannot be definitively diagnosed via electrocardiography alone without invasive coronary angiography and left ventriculography.

Conclusions

Acute coronary syndromes can occur as a complication of pheochromocytoma crisis, but the possibility of Tako-tsubo cardiomyopathy should be considered, with diagnosis via invasive coronary angiography and left ventriculography.

Acknowledgements

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