Phaeochromocytoma presenting as Takotsubo cardiomyopathy

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We report a case of a 72-year-old woman who presented with ST-elevation myocardial infarction (STEMI). However, coronary angiography showed unobstructed arteries while echocardiography (ECHO) showed severe left ventricular (LV) apical hypokinesia with ejection fraction (EF) of 25–30%. Seven months later she presented with a transient ischaemic attack and a repeat ECHO showed a normal EF. A few months later, she was diagnosed with breast cancer and as part of staging procedure, an incidental left adrenal mass was identified. This was biochemically confirmed as phaeochromocytoma (PY) and she underwent laparoscopic adrenalectomy.

PY is a rare catecholamine secreting tumour arising from adrenomedullary chromaffin cells. Excessive catecholamine-induced stimulation can present as transient, reversible cardiomyopathy similar to Takotsubo cardiomyopathy and cerebrovascular events. The diagnosis of PY is often delayed but it is important to recognize PY as a cause of reversible cardiomyopathy. Early intervention is essential to improve mortality from cardiovascular and cerebrovascular complications.

KEYWORDS: Acute myocardial infarction, transient ischemic attack, Takotsubo cardiomyopathy, phaeochromocytoma

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Case presentation

A 72-year-old woman presented with typical cardiac chest pain lasting an hour. She had intermittent history of palpitations, headache, dizziness, sweatiness and weight loss over the last 18 months. Her past medical history included hypertension and type 2 diabetes. She had never smoked and consumed alcohol occasionally. Her mother had ischaemic heart disease. She denied any recent stressful life events. Her medications included ramipril, 5 mg once daily, and metformin, 500 mg twice daily.

On examination in the emergency department, she was afebrile, oxygen saturations at 99% on air, respiratory rate was 18 breaths per minute, pulse was 64 beats per minute and blood pressure was 158/85 mmHg. Systemic examination revealed normal cardiorespiratory and abdominal findings.

Blood results were haemoglobin of 130g/L, glycated haemoglobin (HbA1c) of 60 mmol/mol, mean corpuscular volume of 90 fL, C-reactive protein of <5 mg/L, creatinine of 85 μmol/L, urea of 6 mmol/L and troponin of 1.372 μg/L. Chest X-ray showed no evidence of pulmonary congestion or effusion. Electrocardiography (ECG) showed ST segment elevation myocardial infarction (STEMI), deep T wave inversion and QT interval prolongation. She underwent coronary intervention in a tertiary cardiac unit.

Diagnosis

Common causes of life-threatening cardiovascular chest pain include acute coronary syndrome (ACS), acute myocarditis, aortic dissection and acute pulmonary embolism. The most common cause is ACS due to plaque rupture and vessel occlusion. In presence of angiographically normal coronary arteries, one has to consider coronary vasospasm, vasculitis, myocarditis or type 2 myocardial infarction (supply vs demand).

Initial management and prognosis

Since our patient had typical symptoms, diagnostic ECG changes with very high cardiac markers, ACS was the most likely diagnosis. Coronary angiography, however, showed unobstructed normal arteries. Echocardiography (ECHO) showed apical hypokinesia, preserved basal segments and severe left ventricular (LV) systolic dysfunction with estimated ejection fraction (EF) of 25–30%. She was discharged home on secondary prevention with aspirin, bisoprolol, rosuvastatin with plans for local follow-up and repeat ECHO in 3 months to reassess LV function.

Case progression and outcome

After 7 months, she experienced a transient ischaemic attack with expressive dysphasia. A 12-lead ECG showed complete resolution of her previous ECG changes. Magnetic resonance imaging (MRI) of the brain and carotid Dopplers were normal. ECHO was repeated and showed complete resolution of previous LV wall changes and function, hence ticagrelor and eplerenone were stopped.
Unfortunately, 4 months later, she was diagnosed with intraductal breast cancer. A staging computed tomography (CT) of the abdomen and pelvis identified a left adrenal mass and this prompted an endocrinology consult. Twenty-four hour urinary metadrenaline were significantly elevated and pheochromocytoma (PY) was diagnosed. Preoperatively, her blood pressure was controlled with phenoxycbenzamine followed by beta blockers. She underwent laparoscopic adrenalectomy and, postoperatively, phenoxycbenzamine was stopped. Her HbA1c was back within normal range at 39 mmol/mol and blood pressure stable on bisoprolol. She awaits wire-guided wide local excision of breast lesion.

**Discussion**

PY is a rare catecholamine secreting tumour arising from adrenomedullary chromaffin cells which typically manifests with sustained or paroxysmal clinical features including hypertension, palpitations, tachycardia, headache, and diaphoresis. Excessive catecholamine-induced stimulation of cardiac myocytes can present as transient, reversible Takotsubo cardiomyopathy (TC). Patients with PY may show typical cardiovascular, cerebrovascular, and renal effects of prolonged hypertension, such as congestive heart failure with pulmonary oedema, cardiogenic shock and cardiac arrhythmias. TC, also known as apical ballooning syndrome or broken-heart syndrome, is a recognised but uncommon occurrence with PY presents as ACS characterised by severe left ventricular (LV) dysfunction that typically recovers spontaneously within days or weeks. It accounts for 2.0% of ST-segment elevation infarct syndrome, is a recognised but uncommon occurrence with PY. Hypertension was the commonest presenting feature and only 4% of patients had the classic triad of headaches, excessive catecholamine production, and cerebrovascular and renal effects of prolonged hypertension, such as congestive heart failure with pulmonary oedema, cardiogenic shock and cardiac arrhythmias.

**Learning points**

- For the cardiologist, transient typical LV apical akinesis or dyskinesis on ECHO, with absence of significant stenosis on coronary angiography should prompt consideration of Takotsubo cardiomyopathy. In selected patients, this should lead to an active search for a PY including measurement of blood or urinary catecholamines as all the classical clinical features of PY may not be present at the initial presentation.
- For the endocrinologist, the diagnosis of a PY should lead to cardiac imaging (ECHO or MRI) in order to detect a potential silent cardiomyopathy. This will shorten the delay in introducing heart failure-specific drugs as well as improve perioperative management.
- For the stroke/transient ischaemic attack clinic specialists, in patients with suggestive history and typical ECHO findings, PY should be considered as a risk factor. Since our patient presented with various systemic symptoms to different specialists, recognition of this rare but potentially curable condition ought to be considered and collaborative working is crucial.

**References**


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