

Phaeochromocytoma the Great Mimicker: A Case Report

Dear Editor,

Phaeochromocytomas are catecholamine producing neuroendocrine tumours that can manifest a variety of symptoms which mimic other diseases. They are also known for their “rule of 10” where 10% are extra-adrenal, of which 10% are extra-abdominal, 10% are malignant, 10% occurs in normotensive patients and 10% are hereditary.¹ Their typical presentations are headaches, palpitations, sweatiness, pallor and paroxysmal hypertension. However, it has been reported in literature that rare presentations of phaeochromocytomas include acute abdomen, septic shock-like syndrome, hyperthermia, pulmonary oedema and myocardial ischaemia.^{1,2}

The following case that presented to our hospital demonstrated a case of phaeochromocytoma mimicking an acute coronary syndrome.

Case Report

A 43-year-old Chinese gentleman, with known hypertension for the last 10 years, presented at the Accident and Emergency Department (A&E) of our hospital with non-vertiginous giddiness since early morning with vomiting and bitemporal headaches. He also complained of abdominal pain and difficulty in passing urine. He had experienced paroxysmal attacks of palpitations for the last 4 months. It was associated with diaphoresis and shortness of breath.

At the A&E, he was noted to be febrile with low-grade temperature of 37.9 degrees Celsius and tachycardic at 119 beats per min with an elevated blood pressure (BP) of 185/119 mm Hg. The apex beat was not displaced. The first and second heart sounds were normal. There were no cardiac murmurs. The lungs were clear. There was no focal neurological deficit. He did not have features of neurofibromatosis. The ECG (Fig. 1) showed diffuse ST

segment depressions of 3 mm. His cardiac biomarkers were elevated with creatine kinase 778 U/L, creatine kinase MB 21.22 ng/mL and troponin-T 1.620 ng/mL. At the A&E, he was given a stat dose of nifedipine 5 mg.

His BP was very labile. It decreased to 142/97 mm Hg with nifedipine but rebounded up to 250/190 mm Hg. Intravenous nitroglycerin was started but his BP then plummeted to systolic 55 mm Hg only. Nitroglycerin was stopped and he had to be fluid resuscitated with 1 L of normal saline. His BP improved to 90/50 mm Hg and he was admitted to the intensive care unit (ICU) with a working diagnosis of acute non-ST elevation myocardial infarction (NSTEMI). He was treated with aspirin and subcutaneous low molecular weight heparin.

In the ICU, his BP rebounded up to 175/100 mm Hg after he was started on intravenous dopamine, which was then stopped. His BP continued to swing from 90/60 mm Hg to 290/160 mm Hg and heart rate increased to 149 bpm. His abdomen was guarded and a mass was palpable. The possibility of a catecholamine hypertensive crisis was suspected in view of the labile BP and the abdominal mass. Intravenous sodium nitroprusside was started and an urgent computed tomography (CT) scan of his abdomen was arranged. CT abdomen showed a large right adrenal mass (8.0 cm by 7.0 cm) with possible rupture as evidenced by extravasation of contrast.

Oral phenoxybenzamine 10 mg tds was started and intravenous nitroprusside was continued overnight until the phenoxybenzamine took effect. Beta-blockade with propranolol 20 mg tds was added 1 week later. Meanwhile, his 24-hour urinary catecholamines were collected and despatched. They were highly elevated (Table 1).

Subsequent repeat ECGs in the ICU showed resolution of the ST depressions (Fig. 2). A transthoracic 2D



Fig. 1. Marked ST segment depressions on lead V2 to V6, I, II, III and AVF suggestive of myocardial ischaemia.



Fig. 2. Normalisation of the ST segments.

Table 1. Urinary Catecholamines on 2 Consecutive Days

	1 st set	2 nd set	Normal range
24h urine tests			
VMA (vanillylmandelic acid)	938.8 umol/day	475 umol/day	0-34.3
Metanephrine	337542 nmol/day	171572 nmol/day	400-1600
Normetanephrine	26347 nmol/day	15257 nmol/day	600-1900
Dopamine	1044 nmol/day	866.7 nmol/day	424-2612
Adrenaline	66504 nmol/day	16264.8 nmol/day	0-109
Nor adrenaline	16168 nmol/day	3828.1 nmol/day	89-473

Table 2. Urinary Catecholamines Postadrenalectomy

	1 week postoperative	10 days postoperative	Normal range
24h urine tests			
VMA (vanillylmandelic acid)	21.1 umol/day	–	0-34.3
Metanephrine	929 nmol/day	895 nmol/day	400-1600
Normetanephrine	2066 nmol/day	2155 nmol/day	600-1900
Dopamine	1092 nmol/day	937 nmol/day	424-2612
Adrenaline	<18 nmol/day	<18 nmol/day	0-109
Nor Adrenaline	182 nmol/day	228 nmol/day	89-473

echocardiogram done on Day 2 of admission, showed normal left ventricular size but moderately impaired left ventricular systolic function with a left ventricular ejection fraction of 35%. There were hypokinesis of the basal to midventricular segments but normal apical contractility. On Day 15 of admission after his BP was stabilised, a coronary angiography was performed to assess his coronary arteries. The coronary angiogram showed normal coronary arteries (Figs. 3 and 4). A repeat transthoracic 2D echocardiography post-angiogram revealed the left ventricular ejection fraction had normalised to 55%

Three months later, the patient underwent a right total adrenalectomy. It was uneventful and he recovered well. The histology of the excision showed phaeochromocytoma with extensive necrosis. His urinary catecholamines returned to normal values postoperatively (Table 2). A repeat 2D echocardiogram showed normal left ventricular systolic function with no segmental wall motion seen. He was followed up for next 2 years with no evidence of recurrence of phaeochromocytoma. However, his BP remained elevated requiring nifedipine LA 30 mg bd and atenolol 50 mg om for long-term control.

Discussion

Phaeochromocytoma is a rare disorder with an incidence of less than 0.5% in patients with hypertension. It commonly

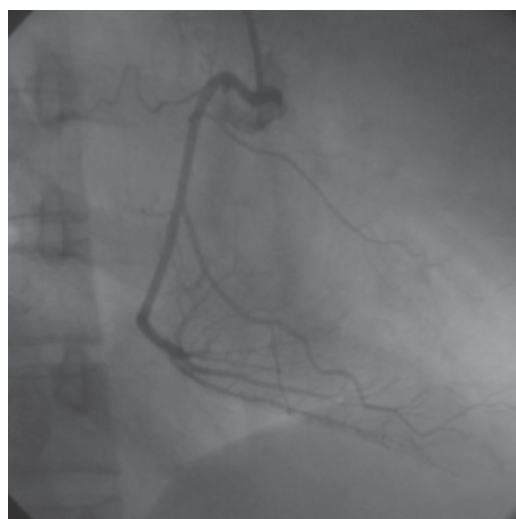


Fig. 3. Right coronary artery – normal.



Fig. 4. Left coronary artery with branches to left anterior descending artery and left circumflex artery – normal.

presents as persistently elevated BP, rather than paroxysmal swings of BP, (which occur in only 10% of pheochromocytoma). It was first reported locally in 1967 (Tan and Chee) and again in 1974 (Tan et al).³ No cardiac findings were reported then. In 1986, a case of a patient with neurofibromatosis and phaeochromocytoma were reported locally who presented with hypotension.⁴ That patient had T wave inversions and elevated cardiac enzymes. Our patient presented similarly with a picture of NSTEMI, except that his ECG showed ST depressions instead of T wave inversions, and paroxysm of BP swings. A coronary angiogram, however, was done for this patient. The findings of normal coronaries exclude ischaemic heart disease, and support the postulation that the mechanism of NSTEMI is likely due to coronary vasospasm from catecholamine surges. The sudden onset of abrupt swings in BP, after a 10-

year history of stable hypertension, may be precipitated by the rupture and necrosis of the phaeochromocytoma.

ECG abnormalities in patients with phaeochromocytoma were observed and reported since the 70s. Several cases of phaeochromocytomas presenting as acute coronary syndrome have been reported worldwide. Most recently, Sanchez-Recalde et al from Spain,⁵ Carron et al from Switzerland⁶ and Tournoux et al from France⁷ reported similar cases in their respective countries. ECG abnormalities reported include ST depression and T wave inversions.

Liao et al from Kaohsiung, Taiwan⁸ had reported in 2000 that 6 of their 25 patients with phaeochromocytoma had abnormal ECG changes. Among the 6 patients, 5 had symptoms of chest pain, which prompted coronary angiography to assess the coronary arteries. All 5 turned out with normal coronaries. Thus, 1/5 of patients with phaeochromocytoma had abnormal ECG changes and chest pain but normal coronaries.

Akashi et al⁹ described a form of cardiomyopathy called “Takotsubo cardiomyopathy” which they postulated to be due to exposure to high levels of catecholamines and phaeochromocytoma-related. The original Takotsubo was coined by Satoshi et al¹⁰ and was characterised by the left ventricle resembling a “octopus-fishing pot” formed by akinetic mid to apical walls of the left ventricle but hyperdynamic basal contraction. Satoshi attributed it to coronary vasospasm as his patients showed near normal catecholamines level. However, Sanchez-Recalde et al¹¹ reported the “inverted Takotsubo cardiomyopathy” which is more akin to this case report with respect to the pattern of hypokinesia and postulated causal event. They believed that the pathophysiology of Takotsubo cardiomyopathy (or stress-related cardiomyopathy) and phaeochromocytoma-related cardiomyopathy are similar and mediated by catecholamines.¹² Rarely, phaeochromocytoma can present as acute pulmonary oedema, as a result of sudden severe impairment of left ventricular systolic function.¹³

We postulate the mechanism of myocardial ischaemia is likely that of vasospasm from the catecholamine surge, leading to ST depressions and release of cardiac biomarkers. The presence of multiple segmental wall motions, and not just a global left ventricular hypokinesia, seen on transthoracic echocardiogram appeared to support this postulation. The vasospasm of the coronaries is reversible. Thus, with adequate alpha- and beta-blockade, resolution of ST depression on the patient's ECGs was noted to be associated with improvement of his left ventricular systolic function.

Conclusion

Phaeochromocytoma is rare and can present with a multitude of symptoms mimicking other disease. This case illustrates the need to consider the differential diagnosis of phaeochromocytoma in a patient presenting with an acute coronary syndrome, especially when it is associated with paroxysm of BP fluctuations.

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