CASE REPORT

Phaeochromocytoma presenting as an acute coronary syndrome

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SUMMARY

A 44-year-old Caucasian man presented to the emergency department in acute cardiogenic shock, with pulmonary oedema, secondary to an acute myocardial infarction and in a hyperosmolar hyperglycaemic state. The previous day he had undergone a colonoscopy, which revealed features of colitis, and was started on prednisolone. He had been previously diagnosed with type 2 diabetes, migraine and anxiety attacks. While awaiting a coronary angiogram he developed abdominal pain and a CT scan was performed and found a large right adrenal mass. Plasma-free metadrenaline levels were elevated. After 4 months, a right adrenalectomy was performed successfully. He made a good recovery with normalisation of his heart function and resolution of his diabetes. The diagnosis was delayed for years due to his episodic symptoms being attributed to other more common diagnoses. Although a rare diagnosis in itself, there are case reports of phaeochromocytoma initially presenting with an acute coronary syndrome.

BACKGROUND

The patient's diagnosis in this case was delayed because his symptoms were episodic, and other more common diagnoses were made to explain them. He had a colonoscopy the day before, which may have triggered a catecholamine surge from mechanical compression of the phaeochromocytoma, secondary to raised intra-abdominal pressure. Another possible precipitant was the administration of glucocorticoid following the procedure. This may have provoked an acute coronary syndrome, which led him on a path where the diagnosis was made incidentally.

CASE PRESENTATION

A 44-year-old Caucasian man presented to the emergency department at a district general hospital with sudden onset chest tightness over a few hours. This radiated to the interscapular region.

He had a history of type 2 diabetes mellitus and recurrent attacks of migraines. He was taking metformin 500 mg three times a day and sumatriptan 50 mg as required accordingly.

He had no family history of cardiac disease and was an ex-smoker. He was a nightclub owner and also worked as an information technology (IT) consultant.

On further history taking, a colonoscopy had been performed the previous day for a change in bowel habit with weight loss. He reported having diarrhoea (10 episodes per day). This showed active pancolitis with transverse colon sparing and

moderate proctocolitis. As this suggested the possibility of mild-to-moderate ulcerative colitis, he started taking prednisolone 40 mg shortly after the procedure, ~14 hours prior to his admission.

On arrival to the hospital, he was taken directly to the resuscitation bay and initial examination revealed a well-built muscular man who was distressed, tachypnoeic and cyanosed. His vital signs showed that he was hypotensive (81/54 mm Hg), tachycardic (150 bpm), hypoxic (oxygen saturations were 76% on 15 L O₂) and febrile (38.7°C). He was using accessory muscles of respiration and had bilateral widespread crepitations throughout the chest on auscultation. The lower limb peripheral pulses were palpable.

INVESTIGATIONS

The electrocardiogram and chest x-ray were abnormal (figures 1 and 2). Arterial blood gas analysis revealed a pH of 7.1 and lactate of 17 mmol/L with a low partial pressure arterial oxygen (paO₂) and a low partial pressure arterial carbon dioxide (paCO₂) indicating type 1 respiratory failure with severe metabolic acidosis.

His troponin was over 10 000 µg/L (<14). Other blood tests were: white cell count—25.2×10⁹/L (3.5–11), haemoglobin—153 g/L (130–150), platelets—455×10⁹/L (150–450), Na—135 mmol/L (135–145), K—3.7 mmol/L (3.5–5.0), urea—6.5 mmol/L (2.5–6.7), creatinine—148 µmol/L (70–150) (estimated glomerular filtration rate—45 mL/min/1.73m² (>60)), C reactive protein—3 mg/L (<5), glucose—46 mmol/L (3.5–5.5), serum ketones—0.4 mmol/L (<0.1), and amylase—45 U/dL (<180).

DIFFERENTIAL DIAGNOSIS

- ▶ Acute cardiogenic pulmonary oedema (related to a presumed non-ST segment elevation myocardial infarction (NSTEMI)).
- ▶ Acute pneumonia with severe sepsis and type 1 respiratory failure.
- ▶ Acute aortic dissection.

TREATMENT

He was given treatment for cardiogenic pulmonary oedema and pneumonia, and was admitted to intensive care for non-invasive ventilation with continuous positive airway pressure (CPAP). An echocardiogram showed normal left ventricular size with global hypokinesia and an ejection fraction of 20–25%. Once stabilised, his heart rate reverted to sinus rhythm without conversion to atrial fibrillation. He was weaned off inotropic support and

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Figure 1 ECG (at initial clinical presentation) showing an on-ST segment elevation myocardial infarction (NSTEMI) with atrial flutter with 2:1 block.

Figure 2 Chest X-ray (at admission) showing acute pulmonary oedema.

transferred to a neighbouring cardiology centre for further investigation and treatment.

While in the cardiology ward he developed abdominal pain and vomiting. An abdominal CT scan was performed which revealed a right adrenal mass. This was confirmed with greater clarity on MRI of the abdomen (figure 3). This scan showed a heterogeneous right adrenal mass with cystic and solid components, and an area of haemorrhage.

As a screening test, 24-hour urinary catecholamines were performed and were found to be elevated: free norepinephrines 1166 nmol/24 hours (100–560), and free epinephrines 1422 nmol/24 hours (2–110).

As per the US Endocrine Society Clinical Practice guidelines, plasma free metadrenaline levels were sent for analysis and found to be above 10 000 nmol/L (<0.50) and plasma normetadrenaline levels above 40 000 pmol/L (120–1180) confirming the diagnosis.¹

At this point the history was revisited. The patient had a 5-year history of recurrent episodes of headache, flushing and palpitations. His general practitioner (GP) had treated him for migraine, anxiety and panic attacks. His blood pressure had never been checked during these episodes. His symptoms were sudden paroxysms associated with an impending feeling of doom and some of these occurred during sexual intercourse, causing psychological distress. He had attended a rapid access chest pain clinic twice and on each occasion had been investigated, reassured and discharged.

Figure 3 MRI of the right adrenal phaeochromocytoma.

OUTCOME AND FOLLOW-UP

After the diagnosis was made, a coronary angiogram was performed employing non-ionic contrast medium, which showed normal coronary arteries. A repeat echocardiogram showed that his ejection fraction had improved to 40–45%. His care was transferred to the endocrinology team for the necessary pre-operative preparation, which included α blockade. This was started cautiously given his recent myocardial infarction. He was then transferred to a high-dependency unit at a specialist centre, where a metaiodobenzylguanidine (MIBG) scan demonstrated isolated uptake in the right adrenal gland with no uptake elsewhere. Thereafter he was discharged home. He was later readmitted for a preoperative assessment, during which an echocardiogram showed a further improvement of his ejection fraction to 55–60%. The multidisciplinary team wanted to give further time for his cardiac function to improve prior to surgery. Four months after his initial presentation, an open laparotomy and a right adrenalectomy was performed with no complications. He made a good recovery. Histology later confirmed a completely excised phaeochromocytoma with no adverse features. His oral glucose tolerance test (OGTT) post-surgery showed only impaired glucose tolerance. His insulin was weaned off and he was discharged with no diabetes medication.

DISCUSSION

Phaeochromocytoma is a rare neuroendocrine tumour that originates from chromaffin cells of the adrenal medulla, leading to increased catecholamine production. It presents most often in those aged 40–50 years and 90% are sporadic. In 25% of cases, diagnosis is made incidentally on CT scan. To aid in diagnosis and to rule out metastasis, an MIBG scan is helpful. This has a sensitivity of 84% and a specificity of 73% when used in detecting catecholamine-secreting tumours.²

Diagnosis can be challenging due to the episodic rather than sustained secretion of hormones from the tumour. This results in 50% of patients having only episodic hypertension, leading to false-negative investigation results. In some cases, the detection rate is improved by screening patients when they are symptomatic. The condition also mimics other disorders, as illustrated in this patient's misdiagnoses of migraines and panic attacks. This led to a delay of several years in his diagnosis.

Phaeochromocytoma is treated by surgical resection, which is a high-risk procedure. The preoperative medical therapy is

aimed at volume expansion and reducing the blood pressure to prevent a hypertensive crisis on the operating table. The most commonly used drug is phenoxybenzamine, a non-selective α blocker. It is usually started 7–10 days preoperatively, followed by a β blocker. However, a longer duration of preoperative α -adrenergic blockade is indicated in patients with recent myocardial infarction or catecholamine cardiomyopathy, as in this case.³ Other options include selective α -1 blockers or calcium channel blockers.

On further reflection, the cause of this patient's presentation was likely to be multifactorial. The near lethal paroxysm could have been precipitated by the colonoscopy performed the previous day, which may have caused an increased intra-abdominal pressure during the procedure.^{4 5} This mimics the abdominal compression test used previously to clinically assess a suspected pheochromocytoma in a patient with secondary hypertension.

Another precipitant could have been the prednisolone he received, after the colonoscopy result had revealed colitis. Corticosteroids are implicated as a precipitant in the US Endocrine Society Clinical Practice guidelines.¹ Several case reports have noted this phenomenon. The mechanism behind this is thought to be the induction of catecholamine synthesis and the inhibition of vasodilatation in the heart and peripheral vasculature.^{6 7}

Intense splanchnic vasoconstriction could explain the patient's colitis, which may have been ischaemic in nature. The acute coronary syndrome (type II NSTEMI) and the global hypokinesia could have been due to coronary vasospasm and catecholamine cardiomyopathy, which resolved after the tumour removal.

Patient's perspective

- ▶ After years of playing rugby, training and participating in fitness activities, I noticed that intense exercise such as squats would cause me to have heart palpitations and blackouts.
- ▶ I did the natural thing and reduced my intensity. However, this then seeped through into other activities such as snowboarding, Moto-X and then sex—activities I perceived to be low-to-mid intensity.
- ▶ Very soon it became apparent that my tolerance to heat was low, summers became unbearable and my body temperature would sky rocket in any situation other than complete calm.
- ▶ I went to two GP surgeries over a period of 6–7 years with these symptoms and even cases of unprovoked palpitations. On each occasion I was told it was my diet, age or health, which did not seem correct given my sporting history, and the only test I was referred for was a 24-hour ECG. This revealed nothing as I had work and did not do anything strenuous at the time and so further incidents just kept occurring.
- ▶ Shortly after, my condition revealed other illnesses such as diabetes and then ulcerative colitis. When given steroids for my colitis, the first day I found myself with severe palpitations, vomiting and pains in my shoulders and as a last resort called the medics. I was taken into hospital with heart failure, renal failure, chest issues, pneumonia and I actually died for a brief period of time.
- ▶ It was here that the hospital did further investigations after realising it was something else. This was when the tumour was diagnosed and found to be one of the biggest of its kind.

Here, catecholamine excess in the circulation is likely to have affected multiple systems, manifesting as gastrointestinal and cardiac pathology. Although a rare diagnosis in itself, there are multiple case reports in the literature of pheochromocytoma presenting as an acute coronary syndrome.^{8–10}

Learning points

- ▶ Pheochromocytoma can be life-threatening, as illustrated by this patient presenting perimortem with an acute coronary syndrome.
- ▶ Diagnosis may be delayed by the fact that the symptoms of pheochromocytoma are often episodic and can be attributed to more common diagnoses.
- ▶ Raised intra-abdominal pressure and administration of corticosteroids are important precipitants of a pheochromocytoma crisis.

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Contributors TI was involved in working up this patient in detail, presenting this case in their team meeting and writing up the manuscript. PF, managed this patient in the ward, presented him in the medical grand rounds and was involved in his follow-up care. He provided the idea to write up this case and collected all the necessary details towards publication. He edited the final manuscript. AC-K managed this patient and was involved in the clinical management and in the discussion of the clinical management. RK was the endocrine consultant fully involved in the clinical management of this patient and in the follow-up care. He supervised the case presentation and the write-up of this case report.

Competing interests None declared.

Patient consent Obtained.

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