CASE REPORT

Adrenaline rush: an unusual presentation of phaeochromocytoma

Benjamin Lindsey,¹ Mark David Eisner,² Hannah Katherine Mitchell,¹ Gerald Clesham¹¹Department of Cardiology, The Essex Cardiothoracic Centre (CTC), Essex, UK²Department of General Medicine, Basildon and Thurrock University Hospital, Essex, UK**Correspondence to**
Dr Mark David Eisner,
eisner1@gmail.com

Accepted 14 August 2015

SUMMARY

A 44-year-old man presented to the accident and emergency department with chest pain and shortness of breath. Admission ECG revealed ischaemic changes. He had markedly elevated troponin T and a severely impaired left ventricular ejection fraction with regional motion wall abnormalities. He was initially treated in intensive care for acute myocardial infarction. When his renal function improved, an angiogram was performed, which showed unobstructed coronary arteries. He was later found to have a phaeochromocytoma. This case illustrates a rare diagnosis presenting with common symptoms that could easily have been missed. On admission to hospital, patients can easily be labelled with a diagnosis and put on a treatment pathway, such as acute coronary syndrome. It is important for clinicians to keep an open mind and be prepared to review the diagnosis if the history does not fit.

BACKGROUND

On admission to hospital, patients can easily be labelled with a diagnosis and put on a treatment pathway, such as for acute coronary syndrome. It is important for clinicians to avoid diagnostic closure, keep an open mind and be prepared to review the diagnosis, as rare diseases may mimic more common presentations. Otherwise, the underlying diagnosis might easily be missed.

CASE PRESENTATION

A 44-year-old man presented to the emergency department with sudden onset of chest pain and vomiting. On examination, he was in respiratory distress, his oxygen saturation was 76% despite 15 L/min of oxygen and he had a blood pressure of 160/110 mm Hg. Admission chest radiograph showed pulmonary oedema.

He was under investigation by gastroenterology for long-standing diarrhoea. On the day prior to admission, he had undergone a colonoscopy, which showed ulcerative colitis. For this, he was started on a course of oral prednisolone.

He had recently been diagnosed with type 2 diabetes mellitus and had been started on metformin. It was noted that his blood glucose had been consistently high and difficult to control. He was an active man, with a normal body mass index. He had been treated by his general practitioner for anxiety and had noticed a resting tremor over the previous year. He was a non-smoker and did not abuse alcohol or other substances. There was no family history of note and no further cardiovascular risk factors were identified.

Admission ECG showed ischaemic changes (figure 1) and troponin T was elevated >10 000 ng/L. Echocardiogram showed a severely impaired left ventricular ejection fraction (25%) with regional motion wall abnormalities.

The patient was admitted to intensive care, where he was treated for acute coronary syndrome and pulmonary oedema before being transferred to a tertiary cardiology unit for further evaluation. Coronary angiography was initially not possible due to significant impairment of his renal function. On arrival to the ward, he reported abdominal pain. A CT of the abdomen without contrast was arranged to rule out a complication secondary to the colonoscopy. This revealed a right-sided mass, closely related to the adrenals. This was further evaluated with MRI (figure 2). A 12 cm heterogeneous mass was seen above the patient's right kidney arising from the right adrenal gland. A 24 h urine collection showed elevated levels of free nor-epinephrine and epinephrine, confirming a diagnosis of a phaeochromocytoma. When his renal function stabilised, a coronary angiogram was performed, which showed normal coronary arteries. An echocardiogram a month after admission showed that his left ventricular ejection fraction remained impaired at 40%.

Treatment for acute coronary syndrome was stopped and he was started on β -blockade. He was transferred to the endocrinology ward and started on α -blockade under the supervision of the endocrinology consultant, and referred for surgical excision of the tumour.

INVESTIGATIONS

Admission blood results—creatinine 533 μ mol/L, glucose 46 mmol/L, highly sensitive troponin T >10 000 ng/L and ketones 2.4 mmol/L.

ECG—dynamic ST depression V3–V6, ST elevation in V2, I, aVL (figure 1).

Echocardiogram 48 h after admission showed normal left ventricular cavity size with severely impaired systolic function and estimated left

Figure 1 Admission ECG.

To cite: Lindsey B, Eisner MD, Mitchell HK, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-211472

Figure 2 MRI showing large adrenal mass.

ventricular ejection fraction of 25–30%. Regional motion wall abnormalities were seen. The right ventricular cavity size was normal, with good systolic function. There was mild tricuspid regurgitation.

Follow-up echo showed an ejection fraction of 40–45%.

MRI of the abdomen showed a 12 cm heterogenous mass replacing the right adrenal gland with mass effect on adjacent organs.

A 24 hour urinary catecholamines were free norepinephrine 1166 nmol/24 h (<560 µmol) and free epinephrine 1422 nmol/24 h (<144 nmol).

DIFFERENTIAL DIAGNOSIS

- ▶ Non-ST elevation myocardial infarction
- ▶ Myocarditis
- ▶ Pheochromocytoma
- ▶ Adrenal adenocarcinoma.

TREATMENT

The patient was initially placed in intensive care, and treated for acute coronary syndrome and pulmonary oedema with continuous positive airway pressure, glyceryl trinitrate infusion, aspirin, clopidogrel and low-molecular-weight heparin.

After the diagnosis of pheochromocytoma was made, anticoagulation was stopped, α and β -blockade was started and the patient was referred for surgical excision of the tumour.

OUTCOME AND FOLLOW-UP

A pheochromocytoma was successfully resected and the patient made a good recovery.

DISCUSSION

Pheochromocytomas are rare, catecholamine secreting tumours originating from chromaffin tissue, usually arising from the adrenal medulla. They typically present with symptoms of increased sympathetic activity, ranging from a tremor to hypertensive crisis.¹

We present an unusual case of a patient with a longstanding history of adrenergic symptoms who acutely deteriorated, presenting with symptoms of an acute myocardial infarction.

He became unwell the day following a colonoscopy, raising the possibility that the catecholamine surge was triggered by the direct pressure from the colonoscope on the tumour. A second possibility could have been recent commencement of prednisolone therapy. Previous case reports have described similar presentations after the administration of exogenous glucocorticoids. In these cases, the catecholamine storm occurred between 5 and 36 h after steroid treatment, a timescale that fits with our case report. This association is given biological plausibility by *in vitro* studies, which show increased synthesis and secretion of catecholamines by pheochromocytoma cells exposed to glucocorticoids.²

Pheochromocytomas represent an uncommon disease entity; cases mimicking acute myocardial infarction represent less common presentations of the disease. However, a number of such cases have been described in the literature.^{3–6}

In the long term, it is well documented that patients with pheochromocytoma have increased cardiovascular morbidity. One study found that 13% of patients with pheochromocytoma had a cardiac event, compared to 1.1% in age-matched controls with essential hypertension.⁷ Prolonged exposure to high levels of circulating catecholamines can induce fibrosis in the myocardium, causing long-term cardiac changes.⁸ However, in the acute presentation, the mechanism of myocardial damage is less clear. A number of cases in the literature describe acute plaque rupture, which was not the underlying pathology in this case. Other cases describe catecholamine-induced vasospasm of the coronary vessels; these cases tended to have unobstructed coronaries on angiography.⁹

Takotsubo cardiomyopathy is well described in pheochromocytoma. Presentation often mimics myocardial infarction. It typically presents with acute left ventricular dysfunction with normal coronary arteries. This case differs from Takotsubo cardiomyopathy, as it lacks the characteristic findings on echocardiogram. Typically, there is apical ballooning of the left ventricle with only transient impairment of the ejection fraction. In the case we describe, there was global impairment of the left ventricle and only moderate improvement in the ejection fraction after 1 month.

This case illustrates the important point of maintaining a wide differential, particularly in younger patients presenting acutely unwell. Clinical pathways serve an important purpose, but clinicians should not blindly follow pathways and should be prepared to question the diagnosis when there are features in the history that do not fit.

Learning points

- ▶ There are advantages to having care pathways for common conditions such as acute coronary syndrome, however, clinicians should maintain an open mind and, when the clinical picture does not completely fit the diagnosis, a differential should be considered.
- ▶ History taking remains the most important tool in diagnosing uncommon diseases. A young man presenting in such a way without significant cardiac risk factors should prompt the clinician to widen the differential diagnosis.
- ▶ Pheochromocytoma can present in a variety of ways, and paroxysms of hypertension are not always present. The history of anxiety and tremors should have acted as pointers.

Contributors BL was involved in planning of paper, drafting of case report and discussion. MDE was responsible for planning of paper, drafting discussion, editing paper and revision of paper. HKM participated in editing of case report and discussion and assistance with planning of article. GC consultant responsible for patient and guidance with writing case report and overall supervision.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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