Phaeochromocytoma—"the great mimic": an unusual presentation

Louisa Mitchell, Fionn Bellis

We report the case of a patient who presented to the emergency with the common symptoms of chest pain and dyspnoea and who was subsequently found to have the rare diagnosis of a phaeochromocytoma. We highlight the need to maintain a high index of suspicion of the various differential diagnoses in any case presentation and the importance of trusting clinical intuition. We comment on the benefit of the use of emergency ultrasound.

A 62-year-old man presented to the emergency department with central chest pain. Past medical history included a non-ST elevation myocardial infarction (NSTEMI), hypercholesterolaemia, diabetes and hypertension.

At presentation, he had an atypical dull chest ache. He was hypertensive, with a systolic blood pressure of 210 mm Hg, and tachypnoeic, but felt well in himself and was insistent on being discharged. An electrocardiogram (ECG) showed sinus tachycardia and partial right bundle branch block. Blood gas analysis showed metabolic acidemia and relative hypoxia. A working diagnosis of a pulmonary embolus was made.

A contrast computed tomography (CT) pulmonary angiogram showed no evidence of pulmonary embolus but immediately afterwards the patient deteriorated catastrophically with flash pulmonary oedema. He became increasingly tachycardic, with unrecordable blood pressure and oxygen saturations. ECG monitoring revealed dynamic changes of an NSTEMI.

The patient was intubated and ventilated and an epinephrine (adrenaline) infusion commenced to maintain mean arterial pressure. Hydrocortisone was administered to cover the possibility of a CT contrast reaction. Urgent echocardiography showed only globally impaired left ventricular function. There was a mild neutrophilia but other blood tests including C reactive protein and creatinine kinase were normal.

The patient was transferred to the intensive care unit (ICU) with a working diagnosis of a NSTEMI, leading to acute left ventricular failure and respiratory failure. On arrival at the ICU he again became hypertensive, yet central venous pressure was normal. Inotropes were discontinued and a nitrate infusion was commenced.

Such rapid change in the patient's clinical condition led to reconsideration of the working diagnosis. A consultant

intensivist performed an informal abdominal ultrasound finding an indistinct loculated mass above the right kidney, raising the possibility of an adrenal tumour. Transfer for definitive imaging was delayed as the patient remained cardiovascularly unstable. Questioning the patient's wife revealed no previous symptoms of phaeochromocytoma other than persistent poorly controlled hypertension.

The patient remained clinically unstable in the ICU. He developed renal failure requiring haemofiltration. His metabolic acidosis persisted and his blood pressure became labile.

Recommencing his routine atenolol on day 3 resulted in the blood pressure becoming still more labile with dramatic swings from 60 mm Hg systolic to 300 mm Hg and back every few minutes. The clinical picture was now consistent with a diagnosis of a phaeochromocytoma secreting paroxysmal surges of epinephrine, exacerbated by β -blockade. Ultrasound was performed in the ICU by a consultant radiologist, showing an adrenal mass consistent with phaeochromocytoma. The patient was initially treated with phentolamine and vasopressin, and samples were sent to an off-site specialist laboratory for catecholamine concentrations.

Cardiovascular stability was finally achieved with a combination of atenolol, phenoxybenzamine, phentolamine and magnesium. A tracheostomy was performed, and on weaning from sedation the patient's level of consciousness improved.

Urine analysis results returned with epinephrine: creatinine ratios 10 $\times$ normal, confirming the diagnosis. There was debate about the need for further imaging, but it was felt necessary to identify any secondary deposits in a potential malignancy before surgery. The patient underwent contrast CT scanning of the abdomen showing a 7 cm complex mass arising from the right adrenal gland, with no evidence of direct or indirect metastasis (fig 1).

Nineteen days after admission the patient underwent laparotomy and right adrenalectomy. Surgery was uncomplicated with blood pressure remaining reasonably stable. His post-operative recovery was slow but unremarkable with no major neurological sequelae. Haemofiltration was discontinued and he was transferred to his local hospital for rehabilitation after 27 days.

DISCUSSION

While "common things are common", an open mind to possible differential diagnoses is essential in unusual cases. In this case, suspicion of an adrenal tumour first arose with the flash onset of pulmonary oedema with intravenous contrast. The epinephrine surge from the underlying phaeochromocytoma had in retrospect been initiated by an NSTEMI then exacerbated by the intravenous contrast. Non-radiologist led ultrasound supported further reconsideration of the differential diagnosis.

Our diagnosis was confirmed in this patient with definitive ultrasound. However, adrenal masses or "incidentalomas" on radiological investigation are common, being found in 2.1% of all postmortem examinations, only 1–4% of which are phaeochromocytoma.¹ Only 0.1% of cases of hypertension are caused by phaeochromocytoma, so it should not be diagnosed on the basis of an adrenal mass found in a hypertensive patient.² This means formal imaging studies for suspected phaeochromocytoma are only really justified after the diagnosis has been established biochemically.³ Epinephrine: creatinine values as high as our patients' have a sensitivity of 91% and a specificity of 75%.²

This is an unusual, but not unknown, presentation of phaeochromocytoma.

Figure 1 Phaeochromocytoma (arrow) arising from the right adrenal gland.

Most patients present between the age of 40 and 50 years with 3 years of gradually worsening symptoms before diagnosis.³ While ideally patients with proven pheochromocytoma "should be managed by an experienced team of endocrinologists, endocrine surgeons, and anaesthetists",⁴ this was clearly not possible with an acute presentation of a rare condition to a small district general hospital.

This case emphasises the fact that clinical intuition should be trusted and followed up early in the management of an unusual case. It demonstrates the potential value of non-radiologist led investigative ultrasound and serves as a

reminder for those working in emergency medicine of the importance of keeping a high index of suspicion for rarer conditions.

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REFERENCES

- 1 **Alderazi Y**, Yeh MW, Robinson BG, *et al*. Pheochromocytoma: current concepts. *Med J Aust* 2005;**183**:201-4.
- 2 **Zendron L**, Fehrenbach J, Taverna C, *et al*. Pitfalls in the diagnosis of pheochromocytoma. *BMJ* 2004;**328**:629-30.
- 3 **Lenders JWM**, Eisenhofer G, Mannelli M, *et al*. Pheochromocytoma. *Lancet* 2005;**366**:665-75.
- 4 **Prys-Roberts C**. Pheochromocytoma – recent progress in its management. *Br J Anaesth* 2000;**85**:44-57.

IMAGES IN EMERGENCY MEDICINE

How innocuous it is! Or is it?

Ajit Singh Kashyap, Kuldip Parkash Anand, Surekha Kashyap, Kulamani Sahoo

A 28-year-old man was hit on the face during a gunfight. Apart from some local discomfort he had no complaints, or visual or neurovascular deficits. The wound of entry was below the medial canthus of the right eye (fig 1), but there was no wound exit. A cranial computed tomographic (CT) scan revealed injury to the right orbit floor and right maxillary antrum, but no bullet or metal fragment. Plain x ray of the neck (anteroposterior and right lateral) revealed a bullet pointing upwards and lying in soft tissue on the right side (figs 2 and 3). The bullet was removed uneventfully from the carotid sheath lying between the internal jugular vein and the carotid artery.

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Figure 1 Entry wound. Informed consent was obtained for publication of this figure.

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Figure 2 Anteroposterior radiograph showing position of bullet.

Figure 3 Right lateral radiograph showing position of bullet.