A rare initial clinical presentation of pheochromocytoma

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DESCRIPTION

A 44-year-old man with treated hypertension presented to the emergency department after experiencing acute right flank pain. He described measuring hypertension, followed by malaise sensation, sweating and rapidly progressive right flank pain. On arrival, the patient was conscious, normotensive, with sinus tachycardia, alternating with supraventricular tachycardia and non-sustained ventricular tachycardia. He was diaphoretic with pale clammy skin and cold extremities. Laboratory studies showed lactic acidosis, hyperglycaemia, with a haemoglobin drop (from 139 to 110 g/L within an hour). A focused assessment with sonography for trauma (FAST) ultrasound showed a large hypoechogenic right pararenal mass (figure 1). An abdominal CT scan showed an acute haemorrhage originating from a right suprarenal hypervascularised mass compatible with a ruptured pheochromocytoma (figure 2). Blood pressure became extremely labile with severe hypertension and moments of hypotension.

The patient was admitted to the intensive care unit and treated with aggressive antihypertensive therapy. He developed multiple organ failure with severe stress cardiomyopathy (30% left ventricular ejection fraction) and type 2 Non-ST-Elevation Myocardial Infarction, acute renal insufficiency, low hepatic dysfunction and abdominal compartment syndrome. Plasma normetanephrines and metanephrines came back highly positive. After stabilisation, right adrenalectomy was performed. Histopathology of the right suprarenal mass confirmed an 8 cm ruptured pheochromocytoma, with important intratumorous haemorrhage. The clinical outcome thereafter was favourable, with normalisation of left ventricular ejection fraction.

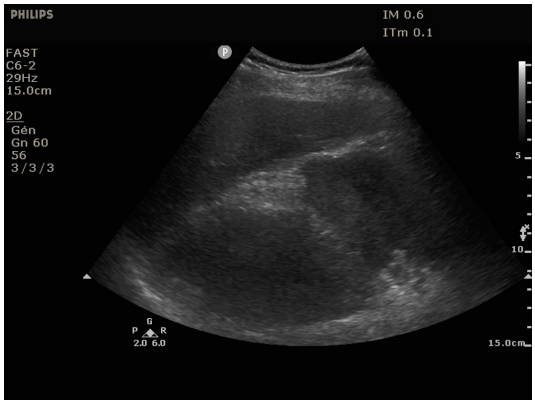

Figure 1 Hypoechogenic mass on focused assessment with sonography for trauma (FAST) ultrasound.

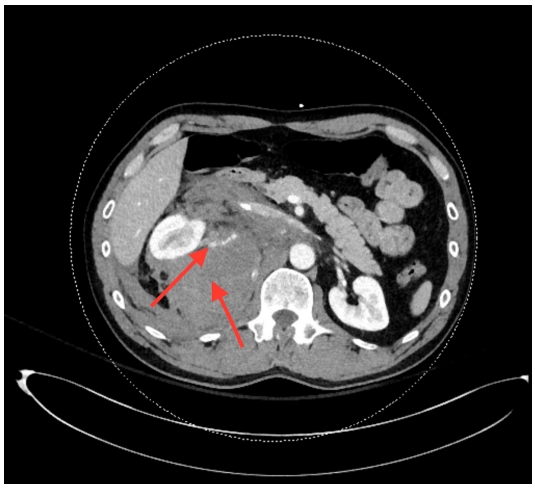

Figure 2 Active low intensity haemorrhage (left arrow), perirenal and retroperitoneal haematoma (right arrow).

In this young patient with a large tumour, a germline mutation in one of the genes responsible for hereditary pheochromocytomas and/or paragangliomas was searched for. The analysis included variants in 21 different genes and came back negative.

Pheochromocytoma is a rare disease with variable clinical presentation, affecting less than 0.2% of patients suffering from hypertension.[1] Pheochromocytoma rupture with haemorrhage is a very rare initial clinical presentation of pheochromocytoma, with less than 60 cases described in the literature. If not rapidly recognised, it can result in the patient's death, most commonly from hypovolemia due to haemorrhage, heart failure from catecholamine cardiomyopathy or respiratory disorder from pulmonary oedema.[2 3] Initial correct diagnosis is made in only 30% of cases.[2] Most cases are diagnosed with CT. In this case, we show the use of FAST ultrasound in locating the pararenal

Learning points

- Pheochromocytoma is a very rare disease with an extremely variable clinical presentation.
- Pheochromocytoma rupture is a rare clinical presentation, but can be life-threatening if not rapidly recognised, due to hypovolemia, catecholamine cardiomyopathy or respiratory disorder.
- Focused assessment with sonography for trauma (FAST) ultrasound can be used in the emergency setting, although CT scan is the preferred imaging modality for diagnosis.

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haemorrhage, which guided ordering emergent CT, confirming the rare diagnosis.

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