Previously clinically “silent” adrenal phaeochromocytoma presenting as hypovolemic shock with paradoxical hypertension.

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Case Presentation
A 58-year-old male patient presented to the Emergency Department with a 6-8 hour history of left upper quadrant pain associated with general malaise, dizziness, nausea and vomiting. There were no other gastrointestinal or genitourinary symptoms; in particular the patient denied any evidence of gastrointestinal bleeding. He had a past medical history of hypertension, which was being treated with lisinopril.

On examination, the patient appeared unwell. He was fully alert but anxious with a Glasgow Coma Score (GCS) of 15. He was tachypnoeic (respiratory rate 25/min) with normal oxygen saturations (98%, FiO2 0.21) and temperature (36°C). He was sweaty with a tachycardia (heart rate 140 bpm) and prolonged capillary refill time (>5 seconds). The patient was also very hypertensive (BP 220/120 mmHg), but the rest of the cardiovascular and respiratory examination was normal. Abdominal examination demonstrated tenderness with guarding over the left upper quadrant and normal bowel sounds. There were no palpable masses.

Blood tests showed a leucocytosis (27.54 x 10⁶) with a neutrophilia (24.97 x 10⁶), normal haemoglobin (14.9g/dL) and platelet count (445,000/L). The urea and creatinine were raised (9.6mmol/L and 160mmol/L respectively). He also had an elevated alkaline phosphatase (249mmol/L), alanine aspartase (41nmol/L) and C-reactive protein (9nmol/L). He was also hyperglycaemic (random glucose 12.6mmol/L).

A chest radiograph showed an elevated left hemi-diaphragm. A computed tomography (CT) scan of the abdomen revealed a left peri-renal collection consistent with a haematoma and left adrenal tumour mass measuring 12cm in diameter (see Figure 1). Also of note was a large left pleural effusion with spontaneous rupture and retroperitoneal haemorrhage of a phaeochromocytoma.

Following blood pressure control and a normal synacthen test, the patient underwent an elective open left adrenalectomy via a trans-abdominal incision. Histology confirmed a malignant pheochromocytoma with central haemorrhagic necrosis and fibrinous changes (Figure 2). There was no evidence of tumour spread beyond the surgical margins or vascular system. The patient remained normotensive post-operatively, and made an uneventful recovery being discharged 10 days later.

Discussion
Phaeochromocytomas, first described by Frankel in 1886 [1], are rare catecholamine-secreting neuroendocrine tumours derived from chromaffin cells². The prevalence is 0.6% in patients with hypertension and up to 25% of patients with these tumours are normotensive or asymptomatic [2]. Sporadic forms of the tumour usually present in individuals aged between 40-50 years whereas hereditary forms are often diagnosed a decade earlier [2]. Formerly known as the 10% tumour, recent evidence would suggest this is no longer true. For instance, around 24% of phaeochromocytomas are thought to be hereditary following genetic testing, even in the absence of a prior history of familial disorders such as von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia (MEN) type 2, neurofibromatosis type 1 (NF1) and familial paragangliomas [2]. Bilateral adrenal tumours exceed 10% in MEN type 2 and VHL syndrome [2]. Furthermore, up to 20% of phaeochromocytomas may be extra-adrenal, 33% of which present as malignant disease as opposed to around 5% in adrenal tumours [2].

Clinically, patients may present with paroxysmal or sustained hypertension and sympathetic overdrive, including: headache, blurred vision, palpitations and diaphoresis. Patients presenting with spontaneous rupture and retroperitoneal

acid (HVA) and 5-hydroxyindacetic acid (5-HIAA) were normal. A repeat intravenous contrast CT thorax, abdomen and pelvis performed 3 days later now showed a small left pleural effusion with a large 10 x 9.6cm mass in the left suprarenal/upper pole left kidney consistent and left sided retroperitoneal collection. These findings were consistent with a spontaneous rupture and subsequent retroperitoneal

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Page 150

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haemorrhage are rare with only 25 reported cases [1,3]. These patients may present with an acute abdomen and back pain in association with hyper or hypotension, and occasionally hypovolemic shock [3]. In our case, the patient had a clinically silent phaeochromocytoma and presented as an acute abdomen with a hypertensive crisis but paradoxically had clinical features of hypovolemic shock.

Although the mechanism of rupture is largely unknown, the high intracapsular pressure may cause the haemorrhage and necrosis noted within the tumour [4]. Contrast CT is the imaging modality of choice in the emergency situation as it is readily available and has a similar sensitivity and specificity to MRI [3]. The mortality rate for emergency surgery following retroperitoneal rupture is 34% versus less than 1% following a period of preoperative optimisation and elective surgery [2,3]. However, intraperitoneal rupture is an absolute indication for emergency laparotomy [3].

The principles of management are good preoperative control of haemodynamic instability followed by elective adrenalectomy [2,3]. Phenoxybenzamine is preferable in controlling blood pressure and preventing a hypertensive crisis because it is a non-competitive $\alpha$-blocker whilst $\beta$-blockers are needed to control the tachycardia associated with high circulating catecholamine levels and $\alpha$-blockade [1]. Labetalol is a non cardio-selective $\beta$-blocker and selective $\alpha$-blocker that has been shown to be effective in controlling the hypertension as well as the tachycardia, although it has also been associated with paradoxical episodes of hypertension thought to be secondary to incomplete $\alpha$-blockade, making its use in the preoperative management in patients with phaeochromocytoma controversial [2]. Calcium channel blockers either alone or in combination with $\beta$-blockers have also become an acceptable form of management [1]. Laparoscopic elective adrenalectomy is the preferred option although it does not appear to have been attempted in patients with spontaneous tumour rupture [2,3]. Five-year survival for a malignant tumour is around 50% with a recurrence rate of about 14% for sporadic adrenal tumours [2]. Familial tumours on the other hand have recurrence rates as high as 33% [2]. Hence, all patients should be followed up annually for at least 10 years [2].

Conclusion

Spontaneous rupture of a phaeochromocytoma is an important differential to consider in an acute abdomen with a retroperitoneal mass. Although a rare diagnosis, elective management reduces mortality significantly.

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