Case report

When numbers do not add up!

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A 62-year-old gentleman was admitted to the surgical admissions unit in 2009 with a history of vomiting and abdominal pain on a background of abnormal renal (urea 13.6 mmol/l, creatinine 165 umol/l) and liver function tests (alanine transaminase 48 U/l, gamma glutamyl transaminase 100 U/l, alkaline phosphatase 156 u/l). Blood pressure was 170/120 mm Hg on admission and a 12-lead electrocardiogram (ECG) revealed sinus tachycardia and anterolateral ischaemia. Troponin I levels were elevated at 2.88 μg/l. A provisional diagnosis of acute cholecystitis and acute coronary syndrome was made by the admitting surgical team. Intravenous fluids and antibiotics were commenced followed by an improvement in patient’s condition.

Abdominal imaging revealed normal liver, gall bladder and a 5.4 × 5.6 cm mass close to the right kidney (Figure 1). A differential diagnosis of perinephric abscess and a solid/cystic malignancy was entertained and a computed tomography (CT)-guided biopsy was performed. The histopathology revealed large polyhedral tumour cells in sheets staining positively for chromogranin A (Figure 2) and synaptophysin, consistent with a phaeochromocytoma or a paraganglioma. An urgent urine 24 h fractionated metanephrines revealed marked elevations of urine metadrenaline and normetadrenaline (6.20 and 9 μmol/collection, respectively, normal range 0.3–1.7 μmol/collection, normetadrenaline 0.4–3.4 μmol/collection).

Further history revealed classical episodes of ‘panic attacks’ and a ‘sudden urge to run out of the bloody place’, each episode lasting for ~20 min. Interestingly, our patient had a history of achalasia cardia and would occasionally rub his abdomen after meals to ‘facilitate food passage into the stomach’. This would invariably provoke a classical episode of panic attack.
An urgent referral to the endocrine surgeon led to tumour removal after adequate alpha and beta blockade. Extensive fibrosis and adhesions led to the removal of 4.2-cm paraganglioma enbloc with the right kidney, a normal right-adrenal gland and perinephric fat (Figure 3). The patient made a good post-operative recovery with complete cessation of symptoms and return of urinary fractionated metanephrines to normal range.

Discussion

‘Catecholamine crisis’ is a rare life threatening endocrine emergency which can have a devastating course if unrecognized. The diagnosis hinges on correlating the varied clinical manifestations and maintaining a high index of clinical suspicion. The presence of multi-organ abnormalities, elevated troponin levels associated with ischaemic ECG changes, high-blood pressure on admission and the CT findings should have raised the suspicion of a ‘catecholamine crisis’ in our case.

A focussed history in such a setting may give vital clues to diagnosis. Particular triggers that provoke an ‘attack’ include anaesthetic agents, metoclopramide, opiates, radiographic contrast and even simple abdominal palpation. It was of great interest that our patient gave a history of classical symptoms after rubbing his abdomen, which is well recognized with an intra-abdominal phaeochromocytoma/paraganglioma. Percutaneous biopsy of such masses can be a recipe for disaster as serious complications including hypertensive crisis, severe haemorrhage, capsular disruption and even death have been reported. In addition, biopsy can also result in inflammation and adhesions, thus complicating resection, as in our case. Therefore, urgent confirmatory biochemical investigations are mandatory prior to biopsy of masses close to adrenal gland or kidneys in the appropriate clinical setting, to prevent devastating sequelae.

Conflict of interest: None declared.

References