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Severe unexplained heart failure in a young person

Ciężka, szybko postępująca i niespodziewana ostra niewydolność serca u młodej osoby — opis przypadku

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Abstract

Acute heart failure in previously healthy young patients is a diagnostic challenge. We describe a case of severe acute heart failure in a 38 year-old patient complicated by acute renal failure and incidental abdominal mass.

Key words: heart failure, phaechromocytoma

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INTRODUCTION

Phaeochromocytomas are tumours derived from neuroectodermal tissue which can secrete large amounts of catecholamines. It has an incidence of 0.01% in the general population, and is most commonly encountered in the fourth and fifth decades of life, although it can present at any age. Phaeochromocytomas are characterised by a highly heterogeneous clinical symptomatology that mimics several other diseases, making it difficult to recognise [1]. The signs and symptoms of phaeochromocytoma are mostly due to hypercatecholaminaemia: hypertension, with paroxysms of adrenergic stimulation, causing palpitations, headaches, sweating, pallor, tremors and anxiety. Cardiogenic pulmonary oedema is also common in cases of phaeochromocytoma [2], sharing features similar to that of catecholamine-induced myocarditis, an important differential diagnosis in our case. Noncardiogenic pulmonary oedema, on the other hand, is an extremely rare manifestation, with only six cases reported in the literature between 1966 and 1999 [3].

CASE REPORT

A previously healthy 38 year-old Chinese man presented to us with acute abdominal pain radiating to the right flank and a three-day history of vomiting, associated with central chest tightness and dyspnoea. He was apyrexial with tachycardia, tachypnoea and hypertensive with a blood pressure of 164/127 mm Hg. Clinical examination was consistent with congestive cardiac failure. He also had a positive right renal punch. Chest radiography showed cardiomegaly with pulmonary congestion. An electrocardiogram showed sinus tachycardia.

He required mechanical ventilation for respiratory distress. A transthoracic echocardiogram revealed markedly impaired left ventricular systolic function, with left ventricular ejection fraction (LVEF) of 10–15%. A diagnosis of cardiogenic shock of uncertain etiology was made and pulmonary capillary wedge pressure was measured at 21 mm H_2O. He also developed acute anuric renal failure requiring dialysis, while ultrasonography showed right hydronephrosis with right renal outflow tract obstruction. A computed tomography (CT) scan of his abdomen showed a large retroperitoneal mass measuring 6 × 5 × 6 cm located between the medial aspect of the right kidney and the inferior vena cava. CT-guided biopsy of the mass was inconclusive. Urinary catecholamines could not be collected as he remained anuric.

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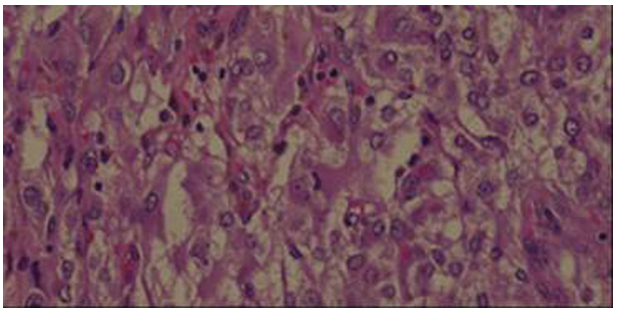

Figure 1. H&E section of pheochromocytoma. Tumour cells are seen in syncytial nests, in so-called 'zellballen pattern'

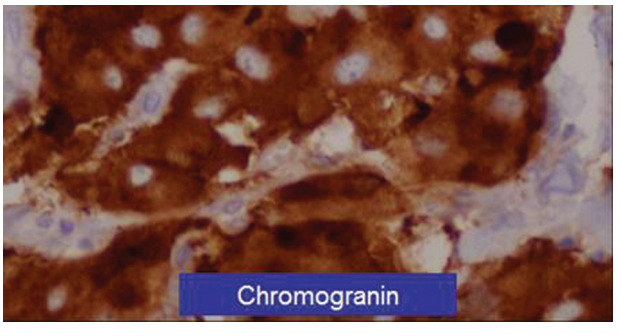

Figure 2. Immunohistological appearance of pheochromocytoma [chromogranin stain (brown), original magnification × 400]

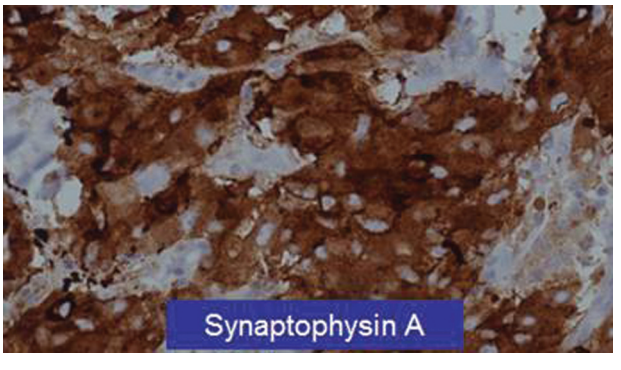

Figure 3. Pheochromocytoma staining positive for synaptophysin A

He improved haemodynamically and was started on a beta-blocker prior to surgery. At laparotomy, a firm mass with a diameter of 8 cm and a necrotic centre located between the inferior vena cava and the right kidney within Gerota's fascia was excised. Histology revealed features of phaeochromocytoma of adrenal origin (Figs. 1–3). He made significant clinical improvement and was discharged from hospital a week later. Echocardiography performed prior to discharge documented an improved LVEF of 40–45%.

DISCUSSION

Our patient was relatively young and had no risk factors for coronary artery disease, therefore we considered the rarer causes of cardiogenic pulmonary oedema. The symptoms of right flank pain probably resulted from the pressure effects of the mass, as well as abdominal angina with nausea and vomiting secondary to gastrointestinal ischaemic changes [4].

For patients with phaeochromocytoma presenting with severe acute heart failure, the prognosis is usually very poor. This is mainly due to the delay in obtaining a definitive diagnosis. If diagnosed correctly, and treated appropriately, surgical resection is successful in 90% of patients; however, the disease is fatal if not detected and treated promptly [5]. Thus, in patients presenting with acute heart failure without obvious cause, a diagnosis of phaeochromocytoma should be considered and urinary concentrations of 4-hydroxy-3-methoxymandelic acid and computed tomography should be undertaken as soon as possible.

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