A case of giant, isolated renal angiomyolipoma in an elderly female

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Introduction

Renal angiomyolipoma is an uncommon, benign neoplasm. Although it is seen in association with tuberous sclerosis, the majority (80%) occur in isolation. Isolated tumours predominantly occur in middle aged females, and mostly arise from the right kidney [1]. Larger tumours are known to be associated with a significant risk of rupture (traumatic or spontaneous) resulting in haemorrhagic complications [2]. We report a case of a giant, uncomplicated, isolated angiomyolipoma arising from the left kidney in an elderly female.

Case report

A 76-year old female presented with a history of progressive abdominal distention of 4 months duration without any other symptoms. She had no history of tuberous sclerosis or other medical illness. Examination showed generalised abdominal distention with a large, ballotable mass in the left flank. Ultrasonography and CT suggested a large left renal angiomyolipoma with remnants of left kidney (Figure 1). She underwent a left radical nephrectomy and a tumour weighing 4.5 kg was excised. Histology confirmed the diagnosis of renal angiomyolipoma.

Discussion

Angiomyolipomas are composed of 3 tissue elements: mature adipose tissue, smooth muscle cells, and thick-walled blood vessels. These can develop in 2 forms. The isolated form which is the commonest (80%), occurs sporadically, and the remaining 20% are associated with tuberous sclerosis. The isolated type is often solitary; presents at a mean age of 43 years; is commoner in women and 80% arise from the right kidney [2]. In this case, the patient was a 76-year old female where the tumour involved the left kidney.

The size can vary from a few millimeters to being larger than 20 cm. The majority (60%) are asymptomatic. However, about 82% to 94% of angiomyolipomas equal to or greater than 4 cm in diameter are symptomatic, and the main presenting symptoms are related to intratumoural or retroperitoneal haemorrhage [3]. Typical angiomyolipomas can be diagnosed without histological confirmation by a combination of ultrasound and CT in up to 95% of cases. On ultrasonography they cannot be confidently differentiated from renal cell carcinoma. However, the characteristic detection of fat within the lesion by CT is diagnostic [4].

Figure 1. CT images showing the angiomyolipoma.

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The management depends on factors such as tumour size, risk of haemorrhage, and whether the lesion is sporadic or associated with tuberous sclerosis. Symptomatic, large tumours require surgical intervention. Smaller (<4 cm), asymptomatic tumours can be managed conservatively with close monitoring using newer imaging techniques (CT, MRI). Radical nephrectomy should be the procedure of choice if there is any suspicion of malignancy [5].

References

Platypnoea-orthodeoxia syndrome due to right to left interatrial shunting despite normal intracardiac pressures

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Introduction

Platypnoea-orthodeoxia syndrome (POS) is a rare condition in which hypoxaemia and breathlessness occur when upright and resolve when prone. POS has been reported in association with intracardiac and intrapulmonary shunting [1]. This case describes a patient who presented with hypoxaemia and platypnoea and investigations showed an interatrial defect with right to left shunt, without pulmonary hypertension.

Case report

A 45-year old man was admitted with worsening shortness of breath. He was breathless on exertion and felt more so when seated and was relieved when lying flat. Three months prior to admission he underwent a tube thoracostomy for a suspected pneumothorax after falling from a height. When breathing room air his oxygen saturation was 90% in the supine position and 82% in the erect position. During the Valsalva manoeuvre it dropped to 78% with little or no improvement with inspired oxygen. He looked plethoric, and his haemoglobin was 17.7 g/dl with a haematocrit of 55%. Examination of the abdomen was normal. Arterial blood gas showed significant hypoxaemia (po2 68 mmHg). Lung functions, chest CT and CT pulmonary angiogram were normal. 99 mTc pulmonary perfusion scan ruled out pulmonary embolism, but high tracer uptake in the systemic side suggested a right to left shunt. Trans-thoracic echocardiography failed to reveal an intra-cardiac shunt. Trans oesophageal echocardiography showed an atrial septal defect and the contrast crossing confirmed a right to left shunt. Estimated tricuspid pressure gradient was only 28 mmHg. While awaiting right heart catheterisation he developed an ischaemic stroke involving the right parietal lobe. He developed acute hydrocephalus and underwent ventriculo-peritoneal shunt insertion. Thrombophilic screening, including prothrombin gene mutation, was negative. He recovered fully from the stroke, without any residual neurological deficit. A subsequent cardiac-catheter study showed normal pulmonary venous drainage. There was no pulmonary arteriovenous fistula. Mean pulmonary artery pressure was 12 mmHg. He underwent a device closure of the atrial septal defect. After the procedure platypnoea and orthodeoxia resolved.

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