Retroperitoneal haemangiopericytoma

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DESCRIPTION
A 55-year-old woman, diagnosed with a renal mass by abdominal ultrasonography (USG) in an external medical centre, presented to our hospital. The medical history was unremarkable except for diabetes mellitus and hypertension. The haemogram and urine tests were normal. The only pathology revealed in routine biochemical tests was increased glucose level (164%mg/dL). The repeat USG performed in our hospital revealed a solid hypoechoic mass lesion approximately 46×46×cm in diameter closely adjacent to the right kidney. At Doppler ultrasonography, the tumour demonstrated intense vascularisation. An abdominal CT scan was performed to characterise the lesion. The CT scan revealed a 42×39×mm mass lesion with regular contours adjacent to the right kidney. In the arterial phase, the tumour demonstrated intense enhancement and in the delayed phase the images showed washout (figure 1). The patient underwent surgery and after histopathological examination retroperitoneal haemangiopericytoma was diagnosed and no relapse or distant organ metastasis was detected throughout the 2-year follow-up.

Haemangiopericytoma is a rare mesenchymal tumour with high vascularity; it arises from capillary pericytes and accounts for less than 1% of all vascular tumours. It can be considered as a soft tissue sarcoma with very low grade malignancy potential. These tumours often occur in adulthood particularly between 50 years to 60 years of age; they can occur anywhere where there are capillary vessels, but are most frequently localised in the musculoskeletal system.1 2 Aetopathogenesis is not known completely but past traumas, steroid treatment, hypoglycaemia and high renin levels have been indicated in the aetiology.2

Haemangiopericytomas tend to grow slowly. Patients are often asymptomatic, and when the tumour is first detected they have already reached a large size. As it grows, the tumour may compress adjacent organs and cause pain. Besides this, the risk of malignancy increases in large tumours.1 Mitotic activity, haemorrhage and presence of thrombosis are also considered in favour of malignancy besides tumour size. Hypoglycaemia, hypertension and gynaecomastia can be seen in cases in which paraneoplastic syndrome develops. The definitive diagnosis of haemangiopericytoma is made histopathologically.2

On radiological imaging, CT scan shows a well demarcated solid mass. Due to its hypervascularity, it demonstrates intense enhancement on contrasted images.3 The hypodense areas inside the mass belong to necrotic or haemorrhagic components. The probability of malignancy increases if haemorrhagic and necrotic components are present. Calcifications are rather rare. On MRI, the necrotic, haemorrhagic or calcific components in the internal structure cause inhomogeneity. A pseudocapsule in the form of a hypointense rim has been reported in some cases.2 1

The prognosis varies, and the main determinant is the histological pattern. Radiological differential diagnoses include liposarcoma, malignant fibrous histiocytoma and leiomyosarcoma. It can be distinguished from liposarcomas with its hypervascularity and intense contrast enhancement, and from malignant fibrous histiocytomas with its well defined borders, but these findings are non-specific.1 3

Learning points
► Haemangiopericytoma is a rare mesenchymal tumour, arising from capillary pericytes.
► Past trauma, steroid treatment, hypoglycaemia and high renin levels have been indicated in the aetiology.
► On radiological imaging, CT scans show a well demarcated, hypervascular solid mass lesion with hypodense areas inside the mass belonging to necrotic or haemorrhagic components.

Contributors

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