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Case report

Renal epithelioid angiomyolipoma: 2 Cases report

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Abstract
Introduction: The 2004 World Health Organization Classification of Renal Neoplasms defined epithelioid angiomyolipoma as a potentially malignant mesenchymal neoplasm, characterized by a proliferation of predominantly epithelioid cells with approximately one third of patients experiencing metastases and one half of them having a history of tuberous sclerosis complex.
Observations: We report two cases of renal epithelioid angiomyolipoma diagnosed at our institution in order to analyze their clinical behaviour and histopathological features, and insist on diagnostic pitfalls.
Conclusion: Renal tumours with certain unusual features should be investigated immunohistochemically to exclude the possibility of epithelioid angiomyolipoma. These tumours are more likely to have an aggressive behaviour when they show more morphologic features predicting malignancy.

Introduction

Classic renal angiomyolipoma (AML) is a benign mesenchymal tumour containing fat, smooth-muscle cells and thick-walled blood-vessels. The epithelioid morphologic variant is defined by the 2004 World Health Organization (WHO) Classification of Renal Neoplasms as a potentially malignant neoplasm, characterized by a proliferation of predominantly epithelioid cells with approximately one third of patients experiencing metastases [1]. Epithelioid-AML (EAML) is included in the family of perivascular epitheloid cell tumour (PECT) which is associated genetically with tuberous scle-

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rosis complex (TSC) [2]. We herein present two cases of renal EAML.

Case reports

First case

A 28-year-old-woman having a history of TSC and bilateral dull lumbar pain was hospitalized with haematuria and acute left lumbar pain. Physical examination revealed bilateral lumbar masses. The woman then presented internal bleeding and acute severe anaemia (5 g/dl) resistant to reanimation and transfusion. Computed tomography (CT) showed bilateral renal heterogeneous masses. The biggest mass was involving nearly the whole left kidney (Fig. 1a). It was complicated by an intra tumoral and retroperitoneal haemorrhage. A radical left nephrectomy was performed immediately. Macroscopically, the tumour was yellowish non-encapsulated (Fig. 1b) measuring 16 cm × 9 cm × 7 cm and comprising areas of haemorrhage with a ruptured subcapsular haematoma. Histopathologically, the tumour was composed of epithelioid cells in sheet pattern (>80% of tumour size). These cells had often enlarged pleomorphic nuclei and abundant eosinophilic cytoplasm. Scattered multinucleated cells and mitotic figures were frequently seen (Fig. 1c and d). A perinephric fat infiltration was identified but neither necrosis nor renal vein invasion were seen. Immunohistochemically, Human Melanoma Black 45 (HMB45) was diffusely immuno-positive. The diagnosis of EAML was made. After a 4-month follow-up, no metastases were detected. A right renal AML embolization was planned subsequently.

Second case

A 56-year-old-man, known carrying a right renal AML diagnosed on ultrasonography 12 years ago, underwent evaluation for recent right dull lumbar pain. Physical examination revealed a right lumbar mass. A control ultrasonography showed an increased-size mid-polar hyperechogenous mass concomitant with the genesis of a new low-polar heterogeneous mass both involving the right kidney. CT concluded to a suspicious low-polar tumour (Fig. 2a). No evidence of TSC or tumours in other locations was found. A radical right nephrectomy was performed. Macroscopically, two encapsulated tumours were identified. The first was brown measuring 7 cm × 6 cm × 5 cm, while the second was yellowish measuring 5 cm × 5 cm × 4.5 cm (Fig. 2b). Histopathologically, the first tumour was a classic AML whereas the second showed similar histologic findings (Fig. 2c) as the first case's tumour except necro-

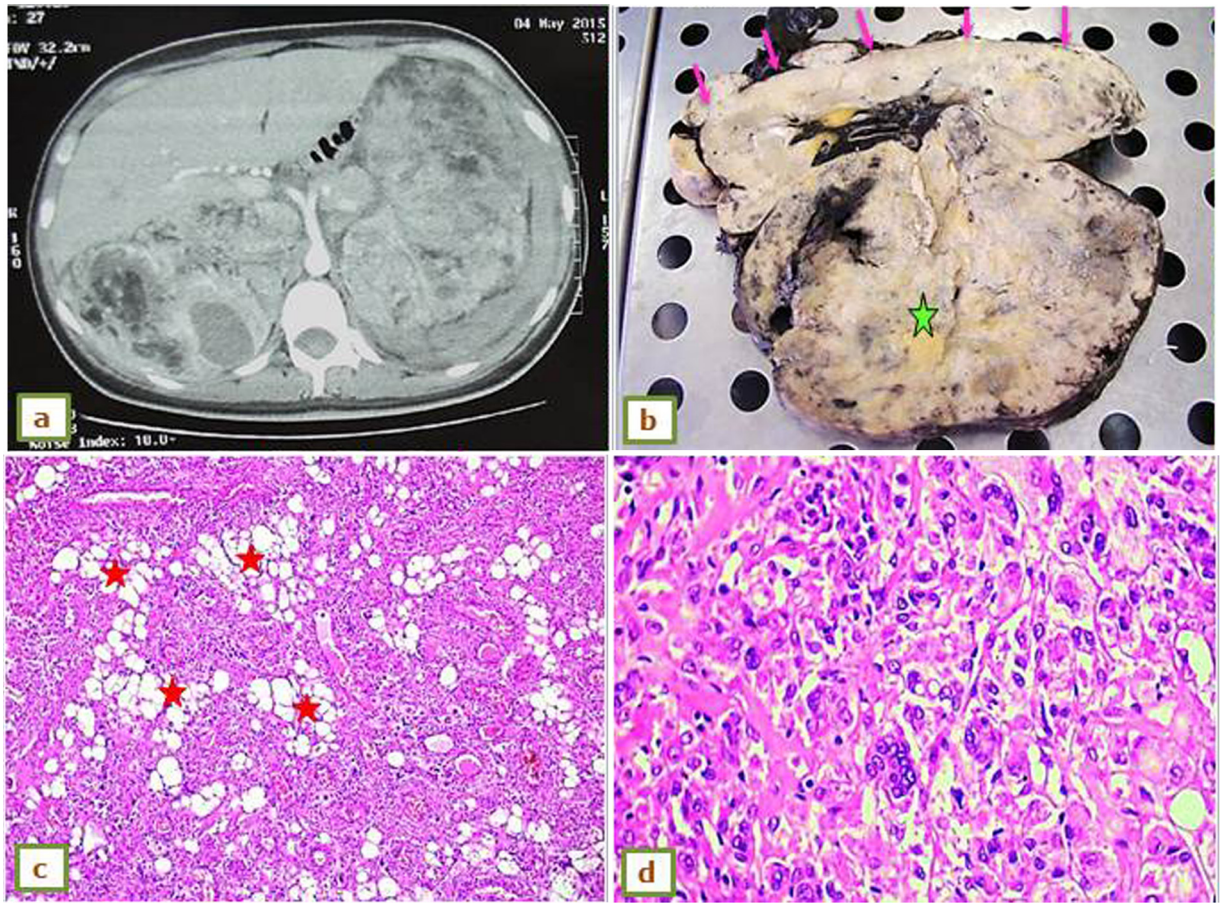

Figure 1 (case 1) (a) CT scan: multiple heterogeneous masses of both kidneys with an intra-tumoral haemorrhage in the left kidney extending to the retro peritoneum (b) Gross appearance in EAML: a lobulated white yellowish tumour (green star) with areas of haemorrhage. The tumour pushes to the periphery the normal renal parenchyma (pink arrows). (c) Histologic features of EAML: mature fat (red stars) and smooth muscle cells (haematoxylin and eosin, original magnification 20×). (d) Histologic features of EAML: Epithelioid cells with abundant eosinophilic cytoplasm (haematoxylin and eosin, original magnification 40×).