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A rare case of spontaneous rupture of epithelioid angiomyolipoma

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ABSTRACT

A male patient, 40 years of age, arrived at our Institute with diffuse abdominal tenderness, right flank pain, hematuria and early stage of hemorrhagic shock with anemia and initial hypotension. The immediate clinical history revealed no significant previous trauma, only subsequently was reported inconstant pain in the right flank for 4–5 days with pallor and asthenia, signs and symptoms that the patient had not investigated. Abdominal CT scan with angiographic evaluation was performed showing right kidney mass and perirenal fluid collection by blood component. Immediate nephrectomy was performed and histopathological and further immunohistochemical study, revealed the epithelioid variant of angiomyolipoma.

Introduction

Epithelioid Angiomyolipoma (EAML) is a variant of Angiomyolipoma (AML) that consists of at least 80% of epithelioid cells. Although it is already in itself a rare and potentially malignant variant of AML, a spontaneous retroperitoneal rupture appears to be at the same time a unique and a challenging complication to manage since no reference has emerged from literature review. We would like to report the first case, to our knowledge, of spontaneous breakage of EAML and its management.

Case presentation

A 40 years-old male patient presented to our observation with diffuse abdominal pain and tenderness on palpation, right flank pain, hematuria and early stage of hemorrhagic shock with anemia and initial hypotension with blood pressure stabilized at 110/70 mmHg. Laboratory parameters showed hemoglobin 8,1 g/dl and white blood cell count was of 16,3 10^3/mm^3, others values did not show any particular pathologic findings. His past medical history revealed no significant previous trauma, only subsequently was referred an inconstant pain in the right flank for 4–5 days with pallor and asthenia, signs and symptoms not previously investigated. No other relevant pathologies emerged from his medical history. Immediate toracoabdominal Computed Tomography (CT) with angiographic assessment (Fig. 1) was performed showing a right kidney mass of about 13 cm of diameter with heterogeneous enhancement and perirenal fluid collection by blood component, compatible with renal and perirenal hematoma, no renal pedicle lesions were detected. CT also revealed pericardial and bilateral pleural effusion, perihepatic and perisplenic fluid flap. Patient started the hemotransfusion and underwent right nephrectomy with transperitoneal approach that allowed a better bleeding control, a quick and easy-removal of the kidney consenting at the same time to exclude injuries to other organs. The postoperative course did not revealed any complications, pleural effusion was managed with thoracic drainage while pericardial effusion resolved spontaneously. The patient was discharged 18 days after admission without consequences. Gross pathology examination reported the presence of a 8 × 6 cm exophytic renal mass, only partially surrounded by a pseudocapsule, invading the renal sinus and whom surface appeared ulcerated probably due to a spontaneous outbreak (Fig. 2). The microscopic pathology (Fig. 3 A) revealed mature fat cells, thick-walled vessels and widespread spindle and epithelioid cells with granular cytoplasm. Not rare were mitotic figures and necrosis (30% of the tumor volume). Further Immunohistochemical (Fig. 3 B) study showed tumor cells positive for Human Melanoma Black-45 (HMB45), S100 protein, Muscle-specific actin (MSA), CD34, CD31, negative for Epithelial Membrane Antigen (EMA) and Ki67 index 2–3%. The patient was then referred to the Oncology Unit for specific follow-up.

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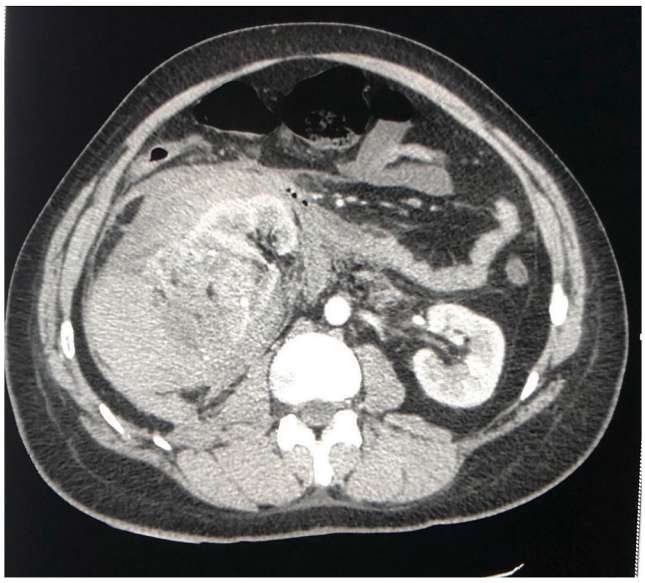

Fig. 1. CT image showing a right kidney mass with heterogeneous enhancement andperirenal fluid collection by blood component, compatible with renal and perirenal hematoma.

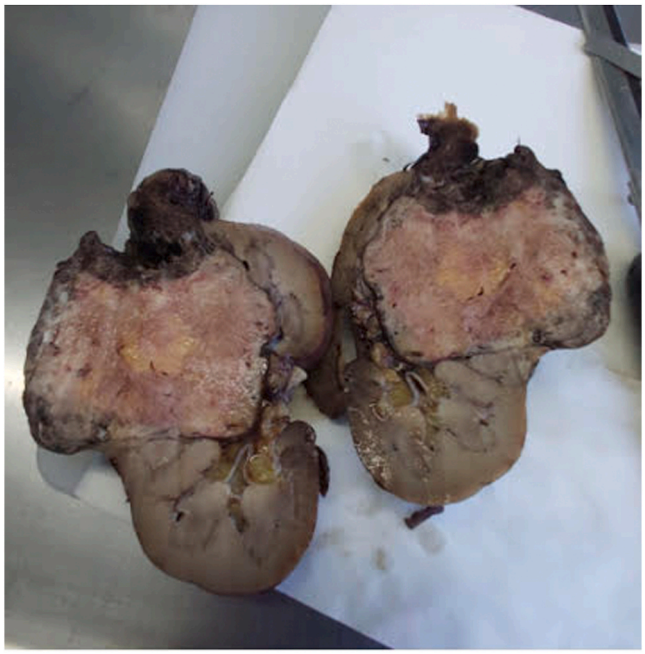

Fig. 2. Section of the kidney showing a 8 × 6 cm exophytic renal mass, only partially surrounded by a pseudocapsule, invading the renal sinus and whom surface appeared ulcerated probably due to a spontaneous outbreak. The upper edge of the lesion appears almost totally damaged as a consequence of a spontaneous rupture.

Discussion

AML is a benign mesenchymal tumor and belongs to a family of so-called perivascular epithelioid cell tumors (PEComas). EAML is a very rare variant of AML that occurs in 1% of cases and consists of at least 80% epithelioid cells.[1] According to the literature, main adverse prognostic factors are necrosis, nuclear atypia in ≥70% of the epithelioid cells, mitotic count ≥2 per 10 high power field, and presence of atypical mitotic figures, although some authors included also tuberous sclerosis (TS), extrarenal extension and/or the involvement of the renal vein,

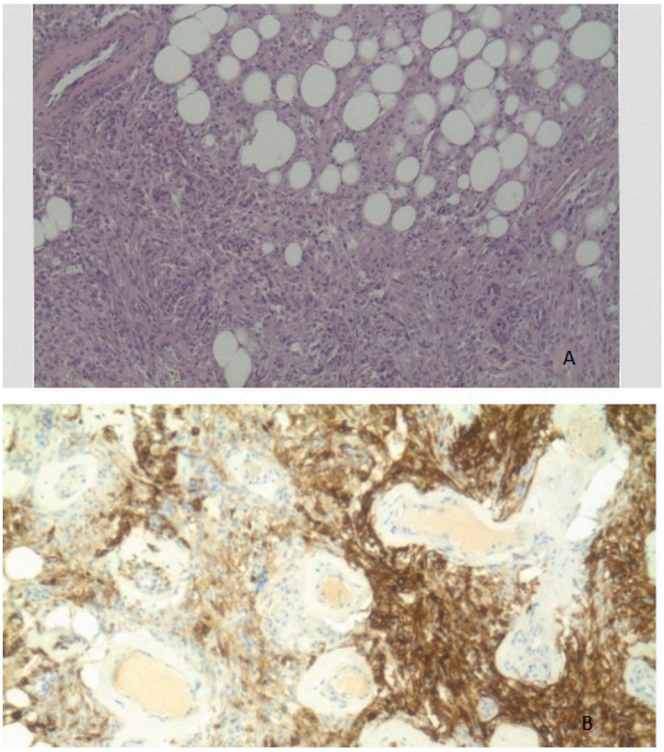

Fig. 3. Thi simage shows both the histopathological features of epithelioid angiomyolipoma (mature fat cells, thick-walled vessels and widespread spindle and epithelioid cells with abundant cytoplasm) (Fig. 3 A) and the immunohistochemical positivity for HMB45 (Fig. 3 B).

tumor size ≥7cm.[2] As with most renal masses, EAML is generally asymptomatic, its diagnosis is often incidental or for sudden onset of symptoms including pain and/or bleeding. The main risk factors for rupture and consequently for bleeding include tumor size, grade of angiogenic component, tumor growth pattern (≥50% exophytic growth), genetic abnormality and it can occurs in the retroperitoneum or into the collecting system which can be life threatening. EAML with diameter ≥ 4cm is more likely to develop rupture both for the increase of the proportion of angiogenic component and for the risk to develop aneurysms or vessels with incomplete vascular walls inside the lesion; TS patients with loss of TSC1 or TSC2 genes were more likely to suffer from aneurysm, leading to dysregulation of the normal angiogenic pathways and to rapid growth and spontaneous rupture of the tumor.[3] Due its potential malignant behavior, being able to even produce metastases, especially in the liver and the lung, surgical management is the reference treatment that contemplates nephrectomy or nephron sparing surgery according to the features of the renal mass. The preoperative diagnosis by imaging remains a challenge; the radiological appearance of most EAML has a tendency to be hyperattenuating on unenhanced CT images, with low intensities on T2-weighted images and could be a heterogeneously solid, homogeneously solid or multilocular cystic lesion with massive hemorrhage.[4] However other renal tumors have also shown similar patterns on CT scans focusing on the need of renal biopsy for the characterization of indeterminate renal masse for more precise treatment decision-making. The immunohistochemical stain appears to be determinant in the diagnosis of EAML that is usually positive for HMB-45 even if it can be co-expressed with SMA and S100; the negative EMA can rule out renal cell carcinoma while Ki67 index exceeding 10% may indicate the malignant behavior of the tumor.[5] Although EAML is well described in literature, to our knowledge, this is the first case of spontaneous rupture reported and we wanted to provide our experience of a complication that may seem complex to deal with. The EAML we observed, presented two of the main risk factors for spontaneous

bleeding referring to the size of the lesion and the amount of angiogenic component as reported in the pathology report; then the immunoractivity to the specific markers, confirmed the diagnosis. The evaluation of the CT images, of the vital signs and of the laboratory parameters made the nephrectomy necessary. Even if it is a challenging procedure, it reveals to be safe and effective to manage the bleeding, exclude other lesions and quickly reach the kidney and its vascular structures. No major complications emerged after the surgery and after the discharge the patient is following regular oncological follow-up.

Conclusions

EAML is a rare entity but severe adverse events can always occur. Bleeding can reveals to be life threatening therefore an early diagnosis and a prompt treatment can be decisive to prevent major complications. Due to its malignant potential and aggressive behavior, strict follow-up is mandatory.

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