Cystic epithelioid angiomyolipoma mimicking a cystic renal cell carcinoma: A case report

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ABSTRACT

Here we report a case of epithelioid angiomyolipoma with cystic changes in a 42-year-old woman. Preoperative imaging studies revealed a 9.8 cm cystic tumor arising from the lower pole of the right kidney, with multiple enhancing solid components and septation diagnosed as a cystic renal cell carcinoma. The patient underwent laparoscopic radical nephrectomy. Histopathology revealed a lipid-poor angiomyolipoma with epithelioid features.

1. Introduction

Angiomyolipoma (AML) is a common benign renal tumor composed of varying amounts of three components: immature blood vessels, smooth muscles, and adipose tissue. The presence of fat tissue in AMLs is a helpful finding on ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI), especially for classic AML. However, lipid-poor AMLs are devoid of this classic fat feature, which leads to difficulties in differentiating them from renal cell carcinoma. Epithelioid AML may show malignant features during the follow-up period. Here we report a case of cystic epithelioid angiomyolipoma mimicking cystic renal cell carcinoma on preoperative imaging.

2. Case presentation

A forty-two-year-old woman, with no past medical illness, presented to the urology clinic of our institute because of incidental findings of a large right renal lower pole exophytic cystic lesion with a solid component on ultrasound, indicative of cystic renal cell carcinoma (RCC) (Fig. 1). Her primary complaint at presentation was mild right flank pain. Her serum creatinine level was 66 μmol/L, and her hemoglobin level was 12.5 gm/dl. Computed tomography (CT) scan was performed, and revealed an 8.4 x 8 x 9.8 cm complex cystic mass lesion arising from the lower pole of the right kidney with multiple enhancing solid components and septation with cystic renal cell carcinoma, with no invasion of the collecting system. No extension beyond the perirenal fascia was observed. The mass abutted the adjacent 2nd and 3rd part of the duodenum, with no obvious invasion. A single patent renal artery and vein was observed. The IVC was also patent. Imaging of the left kidney was unremarkable, with no suspicious solid lesions (Fig. 2).

The diagnosis of cystic RCC was explained to the patient, and she was counseled to undergo right laparoscopic radical nephrectomy in view of the large renal mass T3. The patient tolerated laparoscopic right radical nephrectomy, which was uneventful, and her postoperative creatinine and hemoglobin levels were 82 mg/dl and 11.9 gm/dl, respectively. The patient had a smooth post-operative course.

The histopathology report revealed that the tumor was 8.5 cm in the greatest dimension at the lower pole of the right kidney, and consisted of a mixture of myoid cells, dysmorphic blood vessels, and a minor component of mature adipose tissue. Most myoid cells exhibit epithelioid differentiation in the form of sheets of large polygonal cells with eosinophilic cytoplasm and mildly atypical nuclei with prominent nucleoli. Scattered mitotic figures were also observed. Immunohistochemical staining revealed that the tumor cells were positive for HMB45 and melan-A. Desmin was negative. CKAE1/AE3 and GATA3 tests were negative. The overall morphology and immunoprofile were consistent with a diagnosis of angiomylipoma. The presence of epithelioid features in this case might indicate a potentially aggressive behavior in the form of recurrence or metastasis (Fig. 3). At follow-up, the vascular and
3. Discussion

Renal AML is a solid renal tumor commonly encountered in clinical practice. AML is a solid tumor composed of variable numbers of immature blood vessels, smooth muscle cells, and mature adipose tissue, and is classified as a type of perivascular epithelioid cell tumor, according to the World Health Organization classification 2002.

AMLs are usually discovered incidentally and generally have no abnormal symptoms or signs on medical examinations, although they may present as hematuria, pain, and mass palpation. In some cases, retroperitoneal bleeding can occur when the tumor size is more than 4 cm. AML can be easily diagnosed using imaging methods, such as ultrasonography, computed tomography, and magnetic resonance imaging.
imaging. However, recent research has shown that some AMLs contain too little fat to be detected on unenhanced CT. This makes it difficult to distinguish between AMLs with cystic lesions and renal cell carcinoma. In the present case, it was difficult to diagnose AML because of the cyst-like formation and the lack of fat tissue on preoperative imaging. Moreover, the tumor had the potential for malignancy due to the enhanced septa and the presence of masses within the cystic lesion. Thus, the patient underwent radical nephrectomy for a large tumor. The final histopathological diagnosis was cystic epithelioid angiomyolipoma, with a mixture of myoid cells, dysmorphic blood vessels, and a minor component of adipose tissue.

The term “fat poor angiomyolipoma” was first reported in 1997, with this phenomenon accounting for approximately 5% of all AMLs. Herein, we report a large complex cystic right lower renal mass that presented as a complex renal mass discovered incidentally. Radiological investigation was highly suggestive of malignancy, and histopathological examination was required to confirm the diagnosis due to imaging findings indicative of a malignant tumor such as RCC. However, if the imaging modalities indicate the possibility of benign nature, this will guide the decision toward nephron-sparing surgeries.

4. Conclusions

Here, we report a case of epithelioid AML with cyst-like changes, which was difficult to differentiate from cystic renal cell carcinoma using preoperative imaging alone. We believe that further characterization of this variant and further study of its malignant potential could reduce the need for invasive management.

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