Rapidly Progressing Malignant Epithelioid Renal Angiomyolipoma: A Case Report

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INTRODUCTION

Typical angiomyolipoma (AML) is a benign neoplasm, which consists of a mixture of smooth muscle, fat, abnormal blood vessels, and perivascular epithelioid cells, the latter of which have been described to be present in varying proportions.[1] Recent reports have described renal masses comprised almost entirely of perivascular epithelioid cells and have designated them as "monotypic epithelioid angiomyolipomas (EAMLs)" or "renal epithelioid oxyphilic neoplasms",[2,3] which are potentially malignant.[4] It has also been reported EAML sometimes exhibits aggressive growth, rupture, metastasis, or local recurrence.[5-7] Here, we report a case of rapidly progressing EAML.

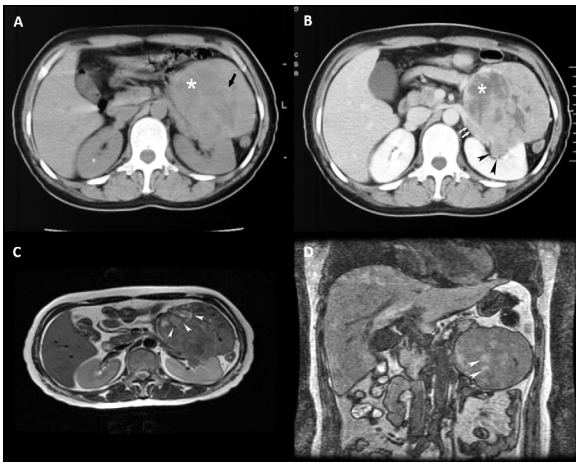

Figure 1. A 47-year-old woman with malignant epithelioid angiomyolipoma.
A) Precontrast-axial computed tomography image showing an 11 cm sized isoattenuating solid mass in the left kidney interpolar area. Note the subtle low-attenuated lesion (asterisk) in the central portion of the mass suggesting hemorrhage or necrosis; **B)** Contrast-enhanced axial computed tomography image showing a moderately enhancing solid mass in the left kidney interpolar area. The mass involved renal pelvis (arrowheads) and contained a hypoattenuating area suggesting hemorrhage or necrosis (asterisk). The image also depicted a hypoattenuated lesion in the left renal vein suggestive of renal vein thrombosis; **C)** Unenhanced axial T2-weighted (4000/110) image shows an isointense mass (relative to liver) in the interpolar area of the left kidney. Foci of hyperintensity (arrowheads) were also observed in the central portion on T2-weighted images possibly representing necrosis or hemorrhage; **D)** Out of phase coronal T1-weighted (450/10) precontrast image showing an isointense solid mass (relative to liver) in the left kidney. Focal hyperintense lesions without signal drop (arrowheads), possibly due to hemorrhage, were also noted.

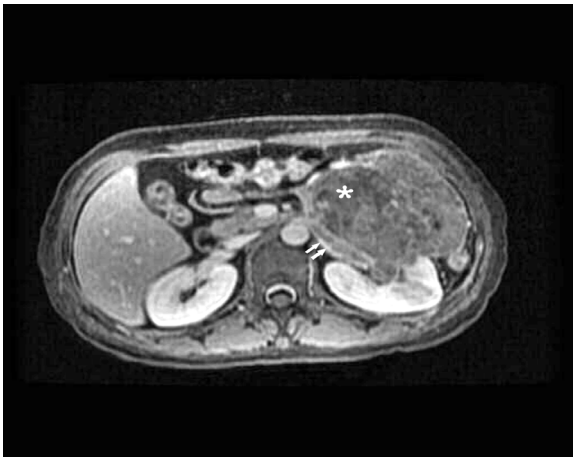

Figure 2. T1-weighted (450/10) postcontrast image showing a moderately enhancing solid mass in the left kidney interpolar area. The mass contained a non-enhancing hypointense area in its central portion suggestive of hemorrhage or necrosis (asterisk), and a non-enhancing filling defect was observed in the left renal vein suggestive of renal vein thrombosis (arrowheads).

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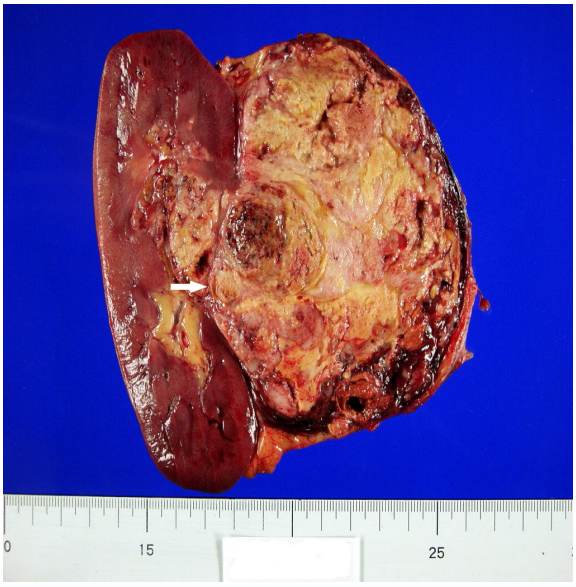

Figure 3. Photograph shows the solid renal mass with yellowish necrosis and reddish hemorrhage. Note the mass invaded the renal sinus (arrow).

CASE REPORT

A 47-year-old woman was referred for the evaluation of acute left abdominal pain. The patient had no history of tuberous sclerosis, and had no abnormal findings during a routine hematological workup or by urine analysis. A more thorough physical examination revealed a large palpable mass in the left abdomen. Precontrast computed tomography (CT) scan showed an isoattenuated mass (versus the liver) with a hypoattenuated portion in the interpolar area of the right kidney (**Figure 1A**). Postcontrast CT depicted a well demarcated, heterogeneously enhancing, necrotic mass with renal vein thrombosis (**Figure 1B**). The mass was of isosignal intensity versus the liver on T2-weighted images (**Figure 1C**), and no detectable signal drop was observed on out-of phase chemical shift magnetic resonance (MR) images (**Figure 1D**). Dynamic enhanced MR images with fat suppression revealed heterogeneous enhancement and a necrotic portion (**Figure 2**). Based on these findings, renal cell carcinoma with renal vein thrombosis was diagnosed preoperatively. Resultantly, left nephrectomy and retroperitoneal lymph node dissection were performed.

On gross examination, the tumor was a gray, solid, expansible, 10.7 × 10 × 7.5 cm sized mass (**Figure 3**) with necrosis and hemorrhage. In addition, adhesion between the tumor and perirenal fat and renal vein thrombosis were observed.

Permanent pathologic evaluation determined the left renal tumor to be EAML comprised mainly of epithelioid cells. Tumor cells were cytologically malignant and exhibited marked pleomorphism and atypical mitotic figures. Typical AML was observed in less than 5% of the mass. No fat component was seen in the typical AML portion, and more than 80% of the mass was necrotic. Immunohistochemical findings were positive for anti-gp100 (HMB)-45, vimentin, α-smooth muscle actin, and CD-10, and negative for cytokeratin. One month after surgery, the patient presented with an abnormal liver function test (increased alkaline phosphatase, aspartate aminotransferase, and alanine transaminase), and a postcontrast CT scan and ultrasonography guided biopsies demonstrated the presence of multiple liver metastases, which were histopathologically confirmed as EAML.

DISCUSSION

AML arises mainly in the kidney and usually follows a benign course. AML can usually be diagnosed based on the radiologic detection of fat in the mass. However, it can sometimes be difficult to detect fat in a renal mass radiologically,[8] because intratumoral hemorrhage obscures fat, little fat is present[9] or because the mass represents renal cell carcinoma or potentially malignant EAML.[10] These latter possibilities are of clinical importance because they have substantially more severe clinical courses.

EAML is a recently recognized variant of AML characterized by the presence of epithelioid cells[2,3] and its diagnosis is difficult as mature adipose tissue is not evident in this tumor. In fact, some cases of EAML have been misdiagnosed as renal cell carcinoma.[11] However, HMB45 is positive in sarcomatoid renal cell carcinoma, renal sarcoma, and in EAML.[12] In our case, HMB45 was positive which is a specific indicator of renal EAML. Although several authors have discussed the appearance of EAML on CT scans, a correct preoperative diagnosis can only be achieved by pathologic examination. On CT scans these tumors appear as solid renal masses with or without a necrotic center and with accompanying metastases to a lymph node or another abdominal organ.[10,13] VanderBrink and colleagues reported a case of EAML with an enhancing element,[14] but to the best of our knowledge no report has been issued on the MRI findings or out-of-phase chemical shift MRI characteristics of EAML.

In a previous study, EAML demonstrated invasion, recurrence, and metastasis, but poor prognoses were rare.[5-7,14] The renal mass in our patient showed peripheral enhancement with central low attenuation, suggesting necrosis, and there was no evidence of fat. These find-

ings are not consistent with the typical imaging findings of AML or AML with minimal fat and suggest the possibility of malignancy. In addition, our patient showed renal vein thrombosis on CT and MR images, which raised level of suspicion for renal cell carcinoma.

CONCLUSIONS

The radiologic differentiation of EAML and renal cell carcinoma appears difficult, but whenever a solid renal mass with a necrotic portion and no fat component is encountered, even if accompanied by renal vein thrombosis, the possibility of malignancy should be considered and EAML included in the differential diagnosis.

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CONFLICT OF INTEREST

None declared.

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