Epithelioid PEComa (epithelioid angiomyolipoma) of the kidney: a rare tumor subtype for patients presenting with an enhancing renal mass

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Epithelioid angiomyolipomas, or perivascular epithelioid cell tumors (epithelioid PEComas) of the kidney, are histologically related to renal angiomyolipomas (AMLs). However, in contrast to typical AMLs, this rare tumor can exhibit an aggressive clinical course with approximately 50% of reported cases demonstrating disease progression. In this report, we present a case of a 24-year-old female with a history of stone disease who was incidentally found to have a 9.0 cm right renal mass that was difficult to characterize radiographically preoperatively. The patient underwent a right radical nephrectomy, and pathology revealed a renal epithelioid PEComa.

Key Words: renal mass, angiomyolipoma, epithelioid PEComa

Introduction

Epithelioid angiomyolipomas, or perivascular epithelioid cell tumors (epithelioid PEComas) of the kidney, are histologically related to renal angiomyolipomas (AMLs). However, in contrast to typical AMLs, this rare tumor can exhibit an aggressive clinical course with approximately 50% of reported cases demonstrating disease progression. Herein, we present a unique case of an epithelioid PEComa in a young female that was difficult to characterize radiographically.

Case report

A 24-year-old healthy, non-smoking female was found to have a 9.0 cm right renal mass at the time of evaluation for renal colic. A magnetic resonance imaging (MRI) of the abdomen was performed to characterize this lesion, demonstrating an enhancing

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central renal mass without the presence of any fat in the mass, Figure 1. As such, the mass was difficult to differentiate radiographically from a fat-poor angiomyolipoma or retroperitoneal leiomyosarcoma. Evaluation for metastatic disease was negative. The patient underwent an open right radical nephrectomy with regional lymphadenectomy. Final pathology revealed a 9.5 cm epithelioid PEComa (epithelioid AML) with negative margins and lymph nodes (stage II, pT2N0M0), and no evidence of necrosis, Figure 2.

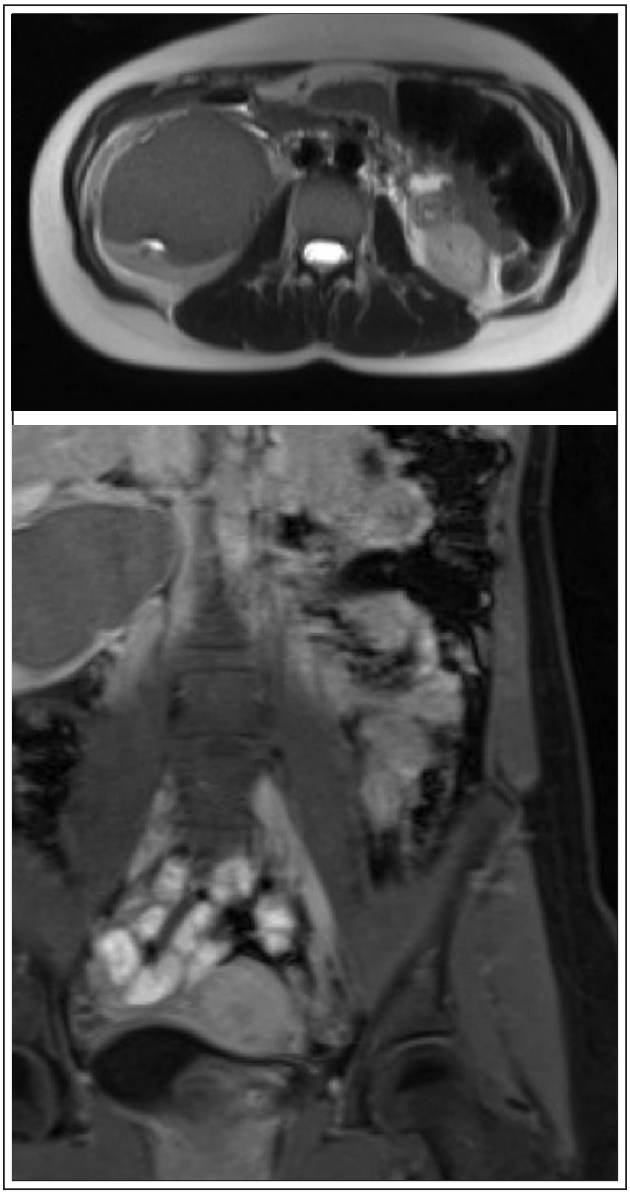

Figure 1. Representative MRI images of the enhancing, homogenous, centrally located right renal mass.

Discussion

While closely related to classic renal AMLs, renal epithelioid PEComas are rare tumors which are radiographically difficult to differentiate from fat-poor AMLs. In the largest review of renal PEComas, the primary tumor is usually > 4 cm. Furthermore, PEComas display a more aggressive phenotype with a 47% rate of disease progression. In a recent review, the mean age at presentation was 40.7 years (range 14-68 years), and the most common metastatic sites are the liver, lung, and peritoneum/mesentery.[1] Specific clinicopathologic factors associated with disease progression include associated tuberous sclerosis complex and/or concurrent angiomyolipoma, necrosis, tumor size > 7 cm, extrarenal extension

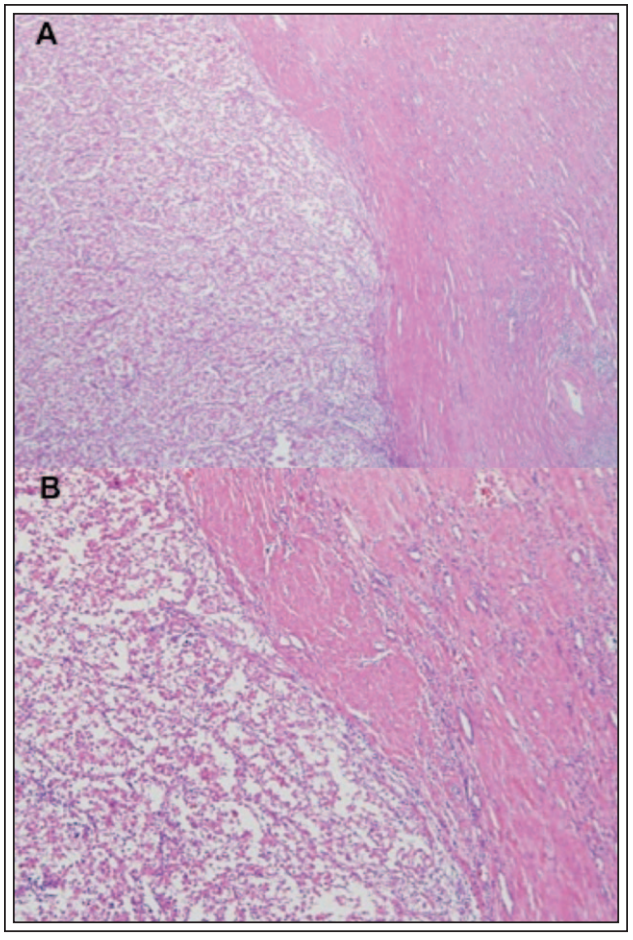

Figure 2. a) Epithelioid PEComa (epithelioid AML) with adjacent renal parenchyma (4X); **b)** Higher magnification of epithelioid PEComa (epithelioid AML) (10X).

and/or renal vein involvement, and carcinoma-like growth pattern.[1] Tumors with less than two adverse prognostic features (our patient) are considered low risk for disease progression (15% rate). Intermediate (2 or 3 parameters) and high risk (> 4 parameters) tumors have a 64% and 80% rate of disease progression, respectively.

Conclusions

Herein, we presented an interesting case of a 24-year-old with a large right renal mass, pathology from which revealed a 9.5 cm epithelioid PEComa. Epithelioid PEComas can display a more aggressive phenotype compared to its closely related classic renal AML counterpart. Specific clinicopathologic factors are associated with disease progression and can be used to guide clinical and radiographic follow up. □

References

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