CASE REPORT

Epithelioid angiomyolipoma associated with a classic angiomyolipoma

Seiichi Kato, MD,[1] Kensaku Seike, MD,[1] Takako Masue, MD,[2]
Naoki Yamamoto, MD,[3] Shinichi Maeda, MD[1]

[1]Department of Urology, Toyota Memorial Hospital, Toyota, Japan
[2]Department of Urology, Gifu University Hospital, Gifu, Japan
[3]Department of Primary Care, Shinsei Hospital, Nagano, Japan

KATO S, SEIKE K, MASUE T, YAMAMOTO N, MAEDA S. Epithelioid angiomyolipoma associated with a classic angiomyolipoma. The Canadian Journal of Urology. 2009;16(5):4857-4859.

We report a case of renal epithelioid angiomyolipoma that arose in association with a classic angiomyolipoma in a 54-year-old Japanese man without tuberous sclerosis. Histologically, the tumor was composed of polygonal cells exhibiting diffuse hemorrhage, multifocal necroses, and vascular invasion. Immunohistochemical staining was positive for melanoma specific antibody and focally positive for smooth muscle actin. On the basis of a review of the literature, we suggest that large epithelioid angiomyolipoma may have malignant potential and therefore requires close follow up.

Key Words: epithelioid angiomyolipoma, malignant potential

Introduction

Several anecdotal cases of epithelioid angiomyolipoma (EAML), an uncommon variant considered potentially malignant, have been reported; all cases were fatal.[1,2] We describe a case of EAML that arose in association with a classic angiomyolipoma (AML).

Accepted for publication May 2009

Address correspondence to Dr. Naoki Yamamoto, Department of Primary Care, Shinsei Hospital, 851 Obuse, Obuse-chou, Kamitakaigun, Nagano, Japan 381-0295

Patient presentation

A 54-year-old man without tuberous sclerosis consulted us with a 1 week history of fever and diarrhea. A computed tomographic (CT) study confirmed a well circumscribed, 15 cm tumor arising from the left kidney, Figure 1. Marked enhancement of the tumor parenchyma was noted, with a non enhancing, patchy internal area, Figure 1. A fat component was noted in the tumor on CT. There was also a small tumor with the same appearance in the right kidney, Figure 1. A selective left renal angiogram showed a bizarre vascular pattern, characterized in particular by

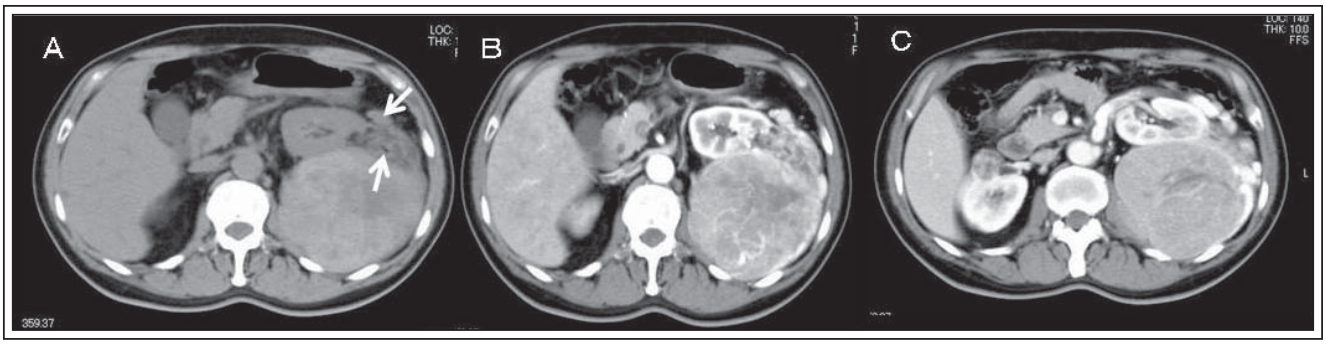

Figure 1. (A) The left side renal tumor was pedunculated. The CT image shows a space-occupying lesion of mixed attenuation interspersed with areas of fat attenuation in a stalk of tumor (arrow). (B) Marked enhancement of the tumor parenchyma is noted, with a non enhancing, patchy internal area. (C) A small tumor with the same appearance is seen in the right kidney.

multiple saccular aneurysms throughout the tumor, Figure 2. With a provisional diagnosis of AML, the feeding arteries were embolized with ethanol, followed by placement of platinum microcoils, Figure 2. However, the patient's fever continued, and the tumor did not decrease despite embolization; therefore, left nephrectomy was performed.

Histology

In part, a variable admixture of mature adipose tissue characterized the histologic appearance of the tumor with tangles of tortuous thick walled blood vessels, and sheets of smooth muscle, features typical of AML. However, in most of the tumor, tumor cells exhibited variation in size and shape, nuclear hyperchromatism, mitoses, and bizarre giant cells. The epithelioid cells stained positive for melanoma specific antibody (HMB-45), focally positive for smooth muscle actin (SMA), and negative for CD10, which confirmed that the tumor was EAML, Figure 3.

The patient's postoperative course was uneventful, and he was discharged without fever.

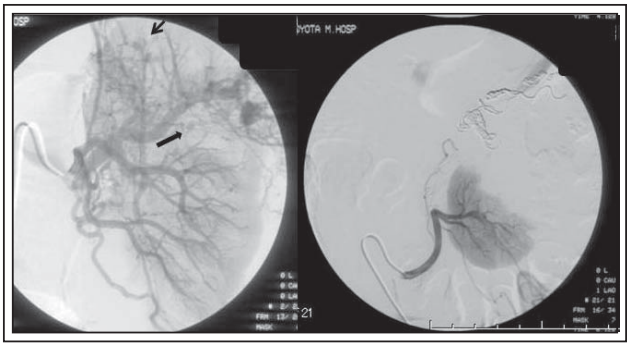

Figure 2. (A) A selective left renal angiogram shows a tumor with a bizarre vascular pattern, characterized in particular by multiple saccular aneurysms throughout the tumor (arrow). (B) Feeding arteries were embolized with ethanol and platinum coils.

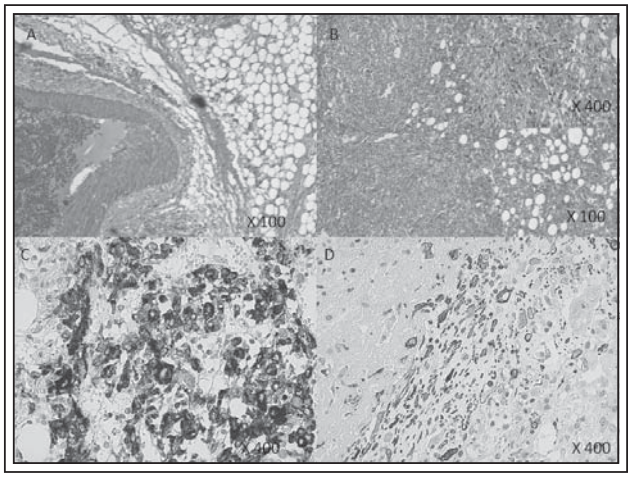

Figure 3. (A) Perivascular epithelioid cells arranged in solid nests or cords with well defined cytoplasmic borders and abundant cytoplasm that varies from eosinophilic and granular to clear, indicating classical angiomyolipoma. (B) Moderate to severe nuclei pleomorphism and hyperchromatism with bizarre multinucleated giant cells and mitoses indicating epithelioid angiomyolipoma. (C) Positive staining of the epithelioid cells for melanoma specific antibody (HMB-45). (D) Focally positive staining for smooth muscle actin (SMA).

Discussion

Since some AMLs exhibit regions of cellular atypia, the pathologic differential diagnosis can include several types of sarcoma. Positive immunoreactivity for HMB-45 is characteristic of AML or EAML. Additionally, it is important to recognize that EAML can develop adjacent to conventional AML or even within it.[3]

EAMLs often resemble renal cell carcinomas radiographically. The presence of fat is quite specific for making the diagnosis of AML. However, approximately 5% of AML cases present with a homogeneous pattern due to a minimal fat component.[4] Complete absence of adipose tissue would suggest renal cell carcinoma rather than EAML.[4] Since EAML is rarely reported, its characteristic imaging features are unclear, and accurate preoperative diagnosis is still difficult.

The present case did not respond to embolization, although transcatheter arterial embolization has usually been used for the treatment of symptomatic AML.[5] To date, there is no absolute protocol of treatment for EAML owing to its rarity. Treatment options for EAML usually include surgery;[1,2,3,6] however, incomplete surgical resection of EAML can result in aggressive recurrence. There is insufficient evidence for the use of additional postoperative therapies, such as irradiation and chemotherapy.[1,2,4] Further clinical investigations are necessary. □

References

1. Lowe BA, Brewer J, Houghton DC et al. Malignant transformation of angiomyolipoma. *J Urol* 1992;147(5):1356-1358.
2. Kragel PJ, Toker C. Infiltrating recurrent renal angiomyolipoma with fatal outcome. *J Urol* 1985;133(1):90-91.
3. Gupta C, Malani AK, Gupta V, Singh J et al. Metastatic retroperitoneal epithelioid angiomyolipoma. *J Clin Pathol* 2007;60(4):428-431.
4. Bonzanini M, Pea M, Martignoni G et al. Preoperative diagnosis of renal angiomyolipoma: fine needle aspiration cytology and immunocytochemical characterization. *Pathology* 1994;26(2): 170-175.
5. Han YM, Kim JK, Roh BS et al. Renal angiomyolipoma: selective arterial embolization-effectiveness and changes in angiomyogenic components in long-term follow-up. *Radiology* 1997;204(1):65-70.
6. Park HK, Zhang S, Wong MK et al. Clinical presentation of epithelioid angiomyolipoma. *Int J Urol* 2007;14(1):21-25.