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CASE REPORT

Renal epithelioid angiomyolipoma mimicking urothelial carcinoma of the upper urinary tract

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Summary *Epithelioid angiomyolipoma is a rare mesenchymal tumor arising mainly in the kidney that can potentially behave aggressively. Epithelioid angiomyolipoma can often resemble sarcomatoid renal cell carcinoma, high grade renal carcinoma or sarcoma. Its similarity to renal cell carcinoma has been emphasized in most of the cases reported in literature. With the purpose of contributing to the awareness of this similarity, a 32-year-old female patient with renal epitelioid angiomyolipoma in the left kidney which radiologically mimicked urothelial cell carcinoma of the upper urinary tract is presented.*

KEY WORDS: Renal; Epithelioid angiomyolipoma; Treatment.

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INTRODUCTION

Angiomyolipomas (AML) are benign tumours of the kidney and are composed of blood vessels, smooth muscle cells and mature fat cells. They comprise 2-6.4% of all renal tumors. Angiomyolipomas are among the most common benign lesions of the kidney (1, 2). These tumors may be formed either as a part of the *tuberous sclerosis complex* (TSC) or as an isolated renal lesion (3). In 50% of patients with TSC, AMLs tend to be multifocal and bilateral involvment may occur (3). Angiomyolipomas are most frequently seen in the kidneys and less commonly found in extra-renal sites such as the retroperitoneum and liver (4). *Epithelioid angiomyolipoma* (EAML) is a variant of AML. Although it is histologically a benign tumor, it may show clinically aggressive behavior and may mimick renal cell carcinoma in imaging studies. Most reports in literature regarding EAML are related to its radiologic and histologic similarity to renal cell carcinoma. Presented in this paper is a case of EAML radiologically mimicking urothelial cell carcinoma of the upper urinary tract.

CASE REPORT

A 32-year-old female patient who referred with the complaint of left flank pain for a nine month period was hospitalized in our clinic. The physical examination findings were normal. Urine test displayed the presence of many erythrocytes and blood chemistry was normal. At urinary system ultrasonography, a solid mass lesion of 76 x 49 mm in size was observed at the mid pole of the left kidney. The right kidney was found to be normal. The upper-lower abdominal phase contrast-enhanced computed tomography (CT) and abdominal dynamic magnetic resonance (MR) images that the patient had prior to coming to our hospital were studied. The abdominal dynamic MR imaging showed a mass lesion 8 x 4 x 6.5 cm in size localized at the mid pole of the renal pelvis which extended towards the exterior and contained hemorrhagic foci. In the post-contrast sections, the lesion showed hemorrhagic foci of minimal heterogenous contrast which decreased in number as the lesion extended towards the renal parenchyma (Figure 1). The urine cytology was benign. With diagnostic flexible ureteroscopy, a tumoral lesion filling the left renal pelvis and calyces was observed. Urethelial carcinoma was suspected and radical nephro-ureterectomy was performed with removal of the cuff from the bladder. At histopathological examination, tumoral structure including thick-walled vascular structures, wide necrotic and hemorrhagic areas are observed adjacent to the kidney tissue. The tumoral structure consisted of round-oval nucleoled spindle-shaped cells, some multinucleated, some ganglion-like appearance, showing palisading areas and a few mitotic figures. Tumoral cells were immunohistochemically HMB45 positive, focally CD68 positive, Vimentin positive and nonreactive with S-100, SMA, MSA, EMA, PANCK, DESMIN, CD34, CD10, NSE, melan-1, factor XIIIa, c-kit. The tumor was histopathologically reported as an epithelioid angiomyolipoma (Figure 2A-B). Seventeen months postoperatively, abdominal MR imaging confirmed that there was no local recurrence or far metastasis in the patient.

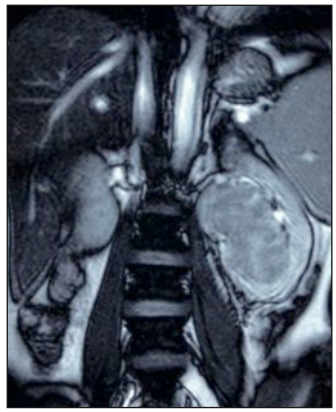

Figure 1. *Abdominal MRI appearance of the left kidney mass lesion.*

No conflict of interest declared.

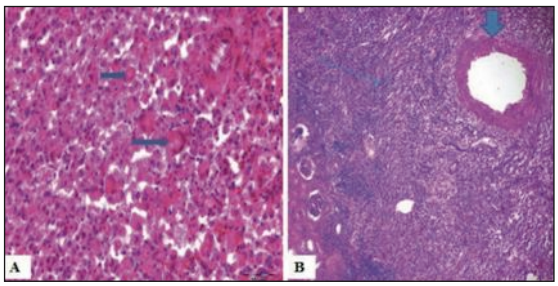

Figure 2.
A. *(HEX400) Atypical cells with prominent nucleoli, exhibiting multinucleated or ganglion-like appearance.*
B. *(HEX100) Tumoral formation including palisades areas (thin arrow) and thick-walled vascular structures (thick arrow).*

Discussion

Angiomyolipoma is a mesenchymal tumor composed of dysmorphic blood vessels, fat tissue, and smooth muscle tissue in varying proportions. Only 1% of renal angiomyolipomas show only epithelioid morphology (5).

Epithelioid angiomyolipoma is a rare mesenchymal tumor recognized in recent years and first reported by *Mai et al.* (6) in 1996. For many years, the tumor was misclassified as an AML. In 2004, the *International Agency for Research on Cancer* (IARC) of the World Health Organization classified EAML as an entity different from typical or classical AML and described it as a mesenchymal tumor with malignant potential. The tumor is primarily composed of epithelioid cells, whereas in some cases, it may show similarity to typical AML (7). Although the growth pattern of EAML may be similar to that of AML, EAML may also display an invasive growth pattern where the tumor tissue shows hemorrhage, necrosis, and degeneration. It sometimes causes lymph metastasis. However, true cystic lesions and peripheral vascular and renal sinus invasion are rarely seen (8). The epithelioid morphology combined with cytologic atypia may render diagnosis difficult and lead to inaccurate diagnoses such as metastatic melanoma or renal cell carcinoma. Immunohistochemistry plays the key role in differential diagnosis (5). The tumor cells of EAML stain negative for the S-100 protein and epithelial markers and stain positive for variable expressions of smooth muscle markers (smooth muscle actin, muscle specific actin) and melanocytic markers (HMB-45 and/or melanA) (9). In our case, the immunohistochemical staining was positive for HMB-45, CD-68, and vimentin and negative for desmin and cytokeratin.

It is difficult to differentiate malignant EAML from other solid renal tumors such as oncocytoma, renal cell carcinoma and sarcomatous lesions with only imaging studies. CT or MR imaging is frequently used to detect the fat foci which are characteristic of the tumor. However, the diagnosis of EAML is difficult because abnormal blood vessels and mature fat cells are also present in typical AML, but not apparent in EAML. The specific image characteristics of EAML have not been described in literature until recently (10). Most EAML reports in literature are related to its radiologic and histologic similarity to renal cell carcinoma. In our case, due to the suspicion of urothelial carcinoma on the abdominal MR images, diagnostic flexible ureteroscopy was performed as a first step. In ureteroscopy, a tumor completely filling the renal pelvis and calyces was observed. According to the prediagnosis of urethelial carcinoma of the upper urinary tract, the cuff was removed from the bladder and nephro-ureterectomy was performed. The reported histopathological diagnosis of the tumor was EAML. The patient was not given any adjuvant therapy. At post-operative 17 months, abdominal MR images of the patient confirmed that there was no local recurrence or any far metastasis. In conclusion, EAML is a rare tumor that can mimic malignant or benign tumors and has unpredictable behaviour. It should be kept in mind that this potentially malignant tumor may radiologically and histologically be confused with renal cell carcinoma and sarcomatous lesions and that it may radiologically mimic urethelial carcinoma of the upper urinary tract.

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