Case Report

Extrarenal retroperitoneal angiomyolipoma

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ABSTRACT

Angiomyolipoma of the retroperitoneum is extremely rare with only 7 cases reported to date in the literature. Spontaneous hemorrhage and shock is the most common presentation that requires emergency intervention. We report a case of large extrarenal retroperitoneal angiomyolipoma that illustrates a different form of presentation for such a rare tumor mimicking a large locally advanced renal parenchymal tumor. The diagnosis was made by histopathology after performing radical nephrectomy. Although rare, angiomyolipoma of the perinephric fat may have variable presentations and should be considered in the differential diagnosis of large renal tumors particularly in view of possible kidney sparing management.

Keywords: Angiomyolipoma, retroperitoneal neoplasms, kidney.

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Angiomyolipomas are ubiquitous tumors that arise most commonly in the kidneys and generally behave as benign tumors, although occasional malignant ones have been reported. Extrarenal angiomyolipomas are rare and may arise in the liver, uterus or retroperitoneum. Other unusual locations such as head, vagina, penis, abdominal wall, spermatic cord and colon were also reported.[1] Retroperitoneal angiomyolipomas are exceedingly rare and represent a small group of the extrarenal angiomyolipomas of urological interest due to their close relationship with the kidney and adrenal gland.[2] To date, there are only 7 reported cases of extrarenal retroperitoneal angiomyolipoma in the literature, most of them presented with hemorrhage and shock and their management generally led to nephrectomy. Herein, we report a case of angiomyolipoma of the perinephric fat that had a different clinical presentation and radiological appearance.

Case Report. *Patient One.* A 72-year-old man presented to his local hospital with symptoms of right-sided abdominal swelling associated with mild flank pain for 8-months. There was no fever, hematuria or lower urinary tract symptoms. Intravenous urography (IVU) and abdominal ultrasound performed at the referral hospital showed severely dilated and compressed upper and middle right kidney calyces by a large soft tissue mass containing areas of calcifications and normal left kidney (**Figure 1**). Family history of tuberous sclerosis was negative. Abdominal examination revealed a large palpable, non-tender and irregular mass with restricted mobility occupying the right side of the abdomen, extending down to right iliac fossa, and crossing the midline. Serum creatinine and electrolytes were normal, and white cell count was slightly increased at 12.7 x 10^9/l. (normal 4-11). Midstream urinalysis was unremarkable with

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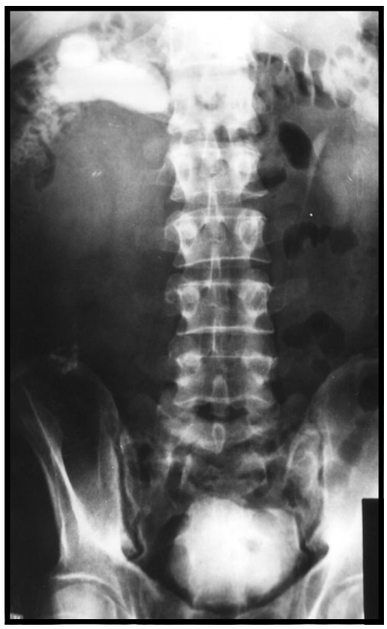

Figure 1 - Intravenous urography showing severely dilated and compressed right kidney by a large soft tissue mass containing areas of calcifications.

negative growth on culture. Repeat abdominal ultrasound showed a large solid mass with mixed echogenicity involving most of the right kidney with patent renal vein and inferior vena cava. Computerized tomography (CT) of the chest and abdomen showed a large solid tumor mass with heterogeneous densities (attenuation coefficient was 36.7 Hounsfield units) arising from the mid and lower portions of the right kidney. The mass did enhance with contrast medium, with extensions outside the Gerota's fascia and suspicious direct involvement of inferior vena cava at L4-L5 level **(Figure 2a & 2b)**. Right renal vein and inferior vena cava were patent and there was no evidence of distant metastases. Renal angiographic studies or CT-guided percutaneous biopsy of the tumor were not performed, as the radiological interpretation was consistent with locally advanced renal parenchymal malignant tumor. Right radical nephrectomy was performed, convalescence was uneventful and the patient was discharged from the hospital 5 days postoperatively. The entire specimen weighted 2900 gm and the golden yellow tumor mass measured approximately 19 x 15 x 13 cm. The kidney was flattened and deformed by the tumor mass, which was adherent to the kidney but separate from it. Microscopic examination revealed nests of epitheliod cells with markedly pleomorphic nuclei, nuclear chromatin, and abundant eosinophilic cytoplasm with rare mitotic figures. Other areas showed thickened blood vessels and mature fat cells **(Figure 3)**. The kidney showed no angiomyolipomatous lesions but arteriolonephrosclerosis and hydronephrosis secondary to chronic inflammation and the adrenal gland was unremarkable. These findings were consistent with angiomyolipoma of the perinephric space. Follow up CT of the abdomen 3 and 6 months after surgery revealed no evidence of residual or recurrent tumor.

Discussion. Angiomyolipomas are mixed mesenchymal tumors, usually regarded as choristomas (a distorted arrangement of mature tissues occurring at a normal location) that contain a mixture of smooth muscle, thick-walled blood vessels and mature fat. They are uncommon tumors

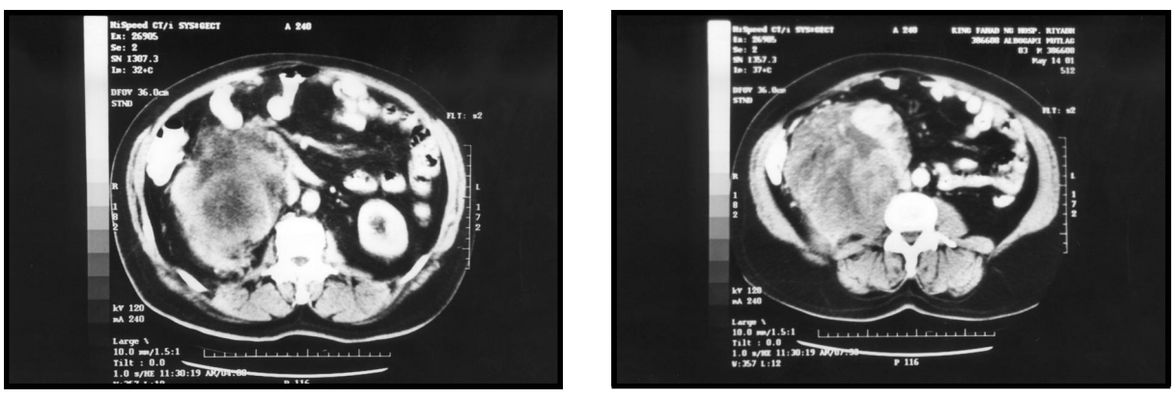

Figure 2 - Computerized tomography of the abdomen showing **(a)** large heterogenous tumor involving the right kidney. **(b)** Suspicious direct involvement of inferior vena cava.

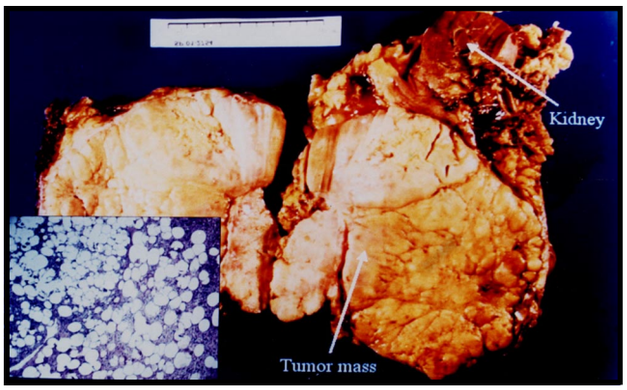

Figure 3 - Gross and microscopic (reduced from x400) appearance of extrarenal retroperitoneal angiomyolipoma with immature smooth muscle and fat.

that frequently occur in the kidneys, and in this situation they are often multiple and bilateral. Forty percent of the patients with angiomyolipoma will also have features of tuberous sclerosis, and as many as 80% of the patients with tuberous sclerosis will have angiomyolipoma.[3] Extrarenal angiomyolipomas are considered rare pathological entities; the most common site is the liver. Bleeding, pain or symptoms secondary to tumor compression to adjacent organs, or both, are the most common presenting symptoms of angiomyolipoma at extrarenal sites.[1] Retroperitoneal angiomyolipomas represent a small group of extrarenal angiomyolipomas; they have a tendency to bleed extensively like their renal counterpart, due to richness of the vascular supply and the intrinsic structure of their blood vessels being thick-walled and deficient of elastic fibrillae.[4] Of the 7 reported cases, 5 of them presented with hemorrhage and shock that required blood transfusion and emergency intervention. Management included surgical exploration to control hemorrhage and generally led to nephrectomy. Recently, Murphy et al[5] reported a case with severe hemorrhage that was successfully managed non-operatively by percutaneous selective embolization with salvage of the kidney. The presence of a fat density lesion on abdominal CT usually makes the diagnosis of angiomyolipoma rather easy as the attenuation values of the fat can clearly be identified. Moreover, the existence of aneurysm-like vascular lesions outside the renal outline with normal arterial branching inside the kidney on angiography should raise the possibility of extrarenal angiomyolipoma or renal angiomyolipoma with exophytic growth. In patients with theses radiological findings conservative approach should be considered. The radiological appearance of the tumor mass in our case is unusual for angiomyolipoma and diagnosis was only made after radical nephrectomy. The absence of fat densities as well as the tumor mass enhancement after contrast injection made our interpretation as locally advanced renal tumor. There was no suspicion that the tumor can be primarily arising from the retroperitoneum rather than from the kidney itself, and accordingly renal angiographic studies or percutaneous biopsy of the tumor were not performed. Percutaneous needle biopsy of the tumor mass in similar cases can provide further information and suggest the benign etiology with possible kidney salvage. Due to its closeness to the kidney and the frequent presence of intratumoral and extratumoral hemorrhage and hematoma, extrarenal retroperitoneal angiomyolipoma usually lead to difficulties in the interpretation of the radiological findings and must be differentiated from liposarcoma of the retroperitoneum. While the former generally arise from perinephric fat, liposarcomas usually arise outside of Gerota's fascia.[6] Since angiography for renal tumors is not a routine investigation and is generally performed in cases with severe hemorrhage or for tumor embolization, extrarenal angiomyolipomas of the perinephric fat should be persuasively considered in the differential diagnosis of large renal parenchymal tumors, retroperitoneal masses and in cases of spontaneous retroperitoneal hemorrhage, especially in view of a possible renal sparing treatment.

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