Case Study of the Month

A Case of Renal Capsular Liposarcoma with Intracaval Fat Thrombus

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1. Case report

A 58-yr-old woman was referred to our hospital for hematuria and a painless, rapidly growing abdominal mass. Computed tomography showed a rounded mass, 23 × 19 cm in diameter, spread over the left retroperitoneal space. A thrombus was observed occupying the left renal vein until the vena cava. Total left nephrectomy with excision of the retroperitoneal mass was carried out. A cavotomy highlighted the intraluminal thrombus, which was easily detached with a forced Valsalva maneuver. The pathologic examination demonstrated a well-differentiated liposarcoma of the renal capsule with fat thrombus in the vena cava. At 24 mo follow-up, there was no evidence of recurrence.

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the patient remained asymptomatic, without evidence of recurrence on control with CT scan.

2. Discussion

Sarcomas of the genitourinary (GU) tract are most commonly reported to be leiomyosarcomas (47% renal, 50% bladder, 57% prostate, and 19% paratesticular sarcomas), followed by liposarcomas [1,2]. Liposarcoma accounts for at least 20% of all soft-tissue sarcoma in adults, with a peak incidence between 50 and 65 yr of age and a male predominance. It may occur anywhere in the body, although the most common sites are the thigh and the retroperitoneum [3]. It could be difficult to establish the exact origin of a tumor that usually derives from fat, from

Fig. 1 – Preoperative picture showing the abdominal mass.

Fig. 2 – Preoperative computed tomography scan (a,b) showing the presence of a mass originating from the inferior pole of the left kidney and angiography (c) showing a double left renal artery.
sinus, or from the renal parenchyma [4]. Primary tumors of the renal capsule are uncommon, and liposarcoma of the renal capsule is distinctly rare. There have been only 18 reports of liposarcoma arising from the renal capsule [5–7], and to the best of our knowledge, this is the first case of a concomitant fat thrombus in the vena cava that was well identified with the use of an ultrasonographic contrast agent and was easily removed through a minimum cavotomy with a forced Valsalva maneuver caused by the anesthesiologist.

Liposarcomas are classified into five histologic subtypes: well differentiated, dedifferentiated, myxoid, round cell, and pleomorphic [8]. The well-differentiated liposarcoma is a locally aggressive, nonmetastasizing, highly locally recurrent, malignant mesenchymal neoplasm composed of a mature adipocytic proliferation with significant variation in cell size and focal nuclear atypia. A well-differentiated liposarcoma usually presents as a deep-seated, painless, enlarging mass that can slowly grow for a long time to attain a very large size. Well-differentiated liposarcomas can be subdivided morphologically into four main subtypes: adipocytic (lipoma-like), sclerosing, inflammatory, and spindle cell [3]. Histologic grade, reflecting the extent of differentiation, remains the most important prognostic factor [8]. Common sites of metastases of renal sarcoma are the lung, the lymph nodes, and the liver [9].

The renal adipocytic liposarcoma has no metastatic potential unless it faces dedifferentiation; when it does, it may more correctly be named atypical lipoma or atypical lipomatous tumor [4].

Liposarcomas are often considered difficult to distinguish from angiomylipomas (AMLs) because both are large fat-containing lesions [9]. Because prognosis and often treatment differ for these two conditions, it is important to have an accurate diagnosis before any treatment or management is instituted. Three major imaging findings (defect in the renal parenchyma, vessels in the lesion, presence of additional AMLs) are crucial for making the distinction [10]. Evidence suggests that complete surgical resection with a negative margin can offer the best chance of cure in patients who present with primary disease.

The relatively poor prognosis of GU soft-tissue sarcomas may be explained by the higher proportion of high-grade
tumors, the proportion of patients who present with metastatic disease, the larger tumor size, and the anatomic site [1]. Cancer-related survival in patients with GU sarcoma is more unfavorable than in patients with sarcoma at all sites and is reported to be 56% at 5 yr. The prognosis of renal sarcoma is especially poor, with a reported 5-yr survival rate of 29% [1,9].

Primary surgical resection is the main treatment. The most important prognostic factors for survival are histologic subtype or grade and completeness of resection [8], as occurred in our case, despite the presence of a wide intracaval vein thrombus that was easily removed without complications.

In conclusion, after an accurate literature review, we can assert that the case presented is the first of a kidney liposarcoma associated with a fat intracaval thrombus that, after radical removal, does not seem to influence prognosis after 24 mo of follow-up.

Conflicts of interest: The authors have nothing to disclose.

References


EU-ACME question

Please visit www.eu-acme.org/europeanurology to answer the following EU-ACME question online (the EU-ACME credits will be attributed automatically).

Question:
Is a differential diagnosis between angiomyolipomas and liposarcomas important?

A. It is not important distinguish angiomyolipomas and liposarcomas because treatment and prognosis is the same for both.

B. A differential diagnosis between angiomyolipomas and liposarcomas is not able to be performed with imaging studies.

C. It is important because treatment and prognosis are different.

D. Angiomyolipomas and liposarcomas do not have similar characteristics.