Primary leiomyosarcoma of kidney with metastasis to contralateral kidney. Case report

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Abstract: *Introduction:* Renal leiomyosarcoma is a rare entity in the world and its understanding is based on reports and various cases; however, the prognosis is bleak for its malignant potential and an average survival of 18 months. *Clinical case:* A 54-year-old woman with a clinical picture of 6 months of pain in the right flank and a tomographic image of a bilateral renal lesion underwent right radical nephrectomy and left conservative surgery. The definitive histopathological study reported right primary renal leiomyosarcoma with left metastasis. *Conclusion:* Renal leiomyosarcoma is an entity of low incidence and high mortality; however, our case represents the minority of patients with contralateral kidney metastasis reported in the literature.

Keywords: renal leiomyosarcoma, metastasis, radical nephrectomy, renal neoplasia, tomographic findings

Introduction

Primary renal leiomyosarcoma is a rare tumor in the adult population, in the western hemisphere, which represents 0.1% [1] of all malignant tumors of the kidney and in the eastern, 0.6%–2.7% [2].

The most common effects appear in women among 50 years old or above. The prognosis is bleak, for example, the average survival is 18 months, with a survival rate of 29%–36% at 5 years. Metastases occur most frequently in specific zones, for example, the liver, bones, and lungs [3].

This is one of the few cases reported in the literature with metastatic activity related to the contralateral kidney. The focus is on the clinical picture, diagnostic imaging, immunohistochemical characteristics, surgical treatment, and follow-up at 6 months [4].

Clinical Case

A 59-year-old woman with oppressive pain in the right flank of 6 months of evolution, who came to our unit with abdominal tomography. Initially, the diagnosis of bilateral renal angiomyolipoma was concluded by the tomography. The findings indicate a 10-cm right kidney tumor with heterogeneous and irregular borders around it. A left renal tumor was also observed in the upper pole and a 1-cm paracaval lymph node, without evidence of other lesions *(Fig. 1)*.

Based on the tomographic findings, she suffered midline laparotomy and a right radical nephrectomy plus retroperitoneal lymphadenectomy was performed on her. A sample was taken and sent to a transoperative study to confirm the tumor etiology. For this reason, a left partial nephrectomy was performed and also a transoperative study that revealed negative surgical margins *(Figs 2 and 3)*. The patient had a good evolution and was discharged 5 days after the surgery.

The definitive histopathological study of the right and the left tumors reported cell necrosis, capsule and renal fat infiltrations, and metastasis in five lymph nodes. The immunohistochemical analysis was positive for actin, vimentin, calponin, and desmin and negative for cytokeratin AE2/AE3, CD68, BCL2, and WT1 *(Figs 4–7)*. With these findings, the diagnosis of high-grade primary

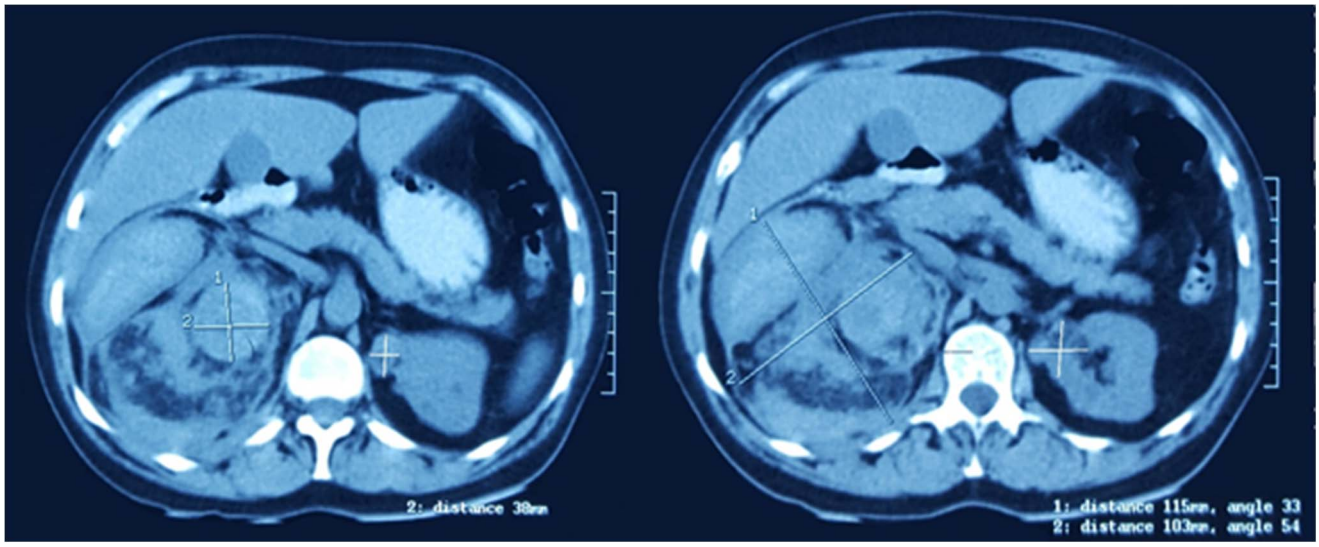

Fig. 1. Tomographic findings of the right primary tumor and contralateral metastasis

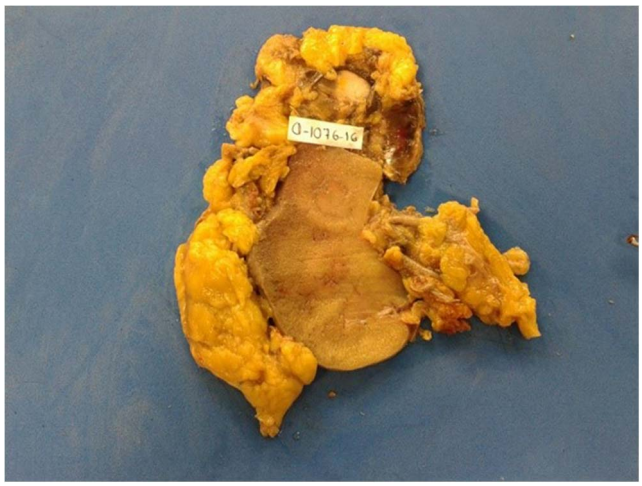

Fig. 2. Surgical sample of the primary tumor

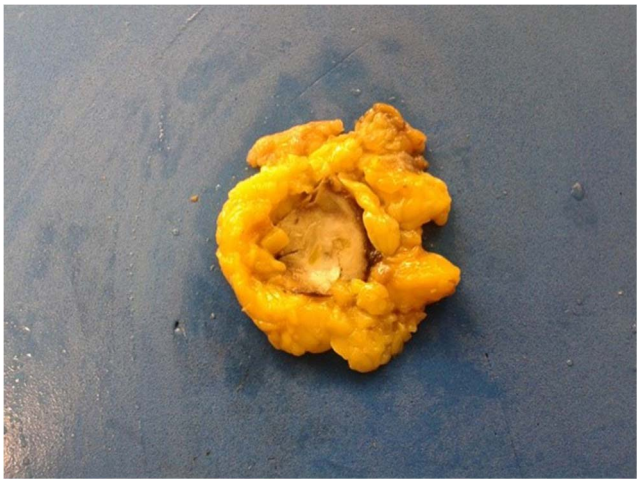

Fig. 3. Product of the contralateral metastasectomy of the kidney

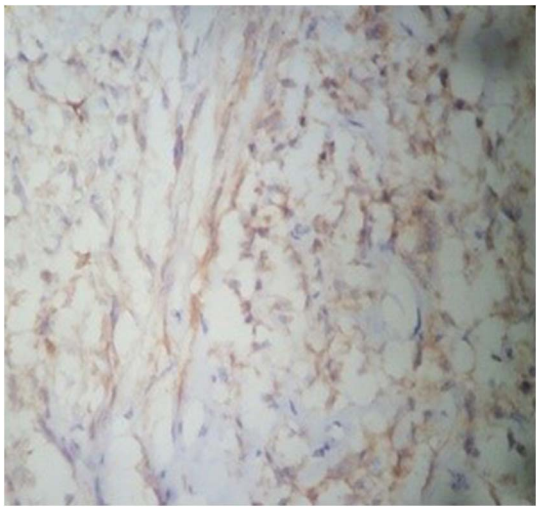

Fig. 4. Positive histological sections at S100

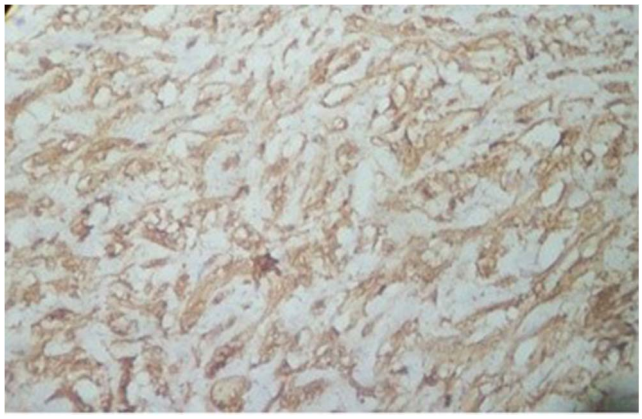

Fig. 5. Positive histological sections to actin

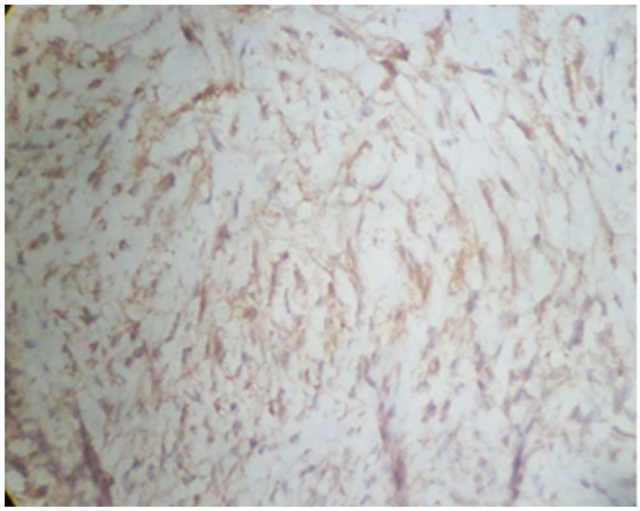

Fig. 6. Positive histological sections to vimentin

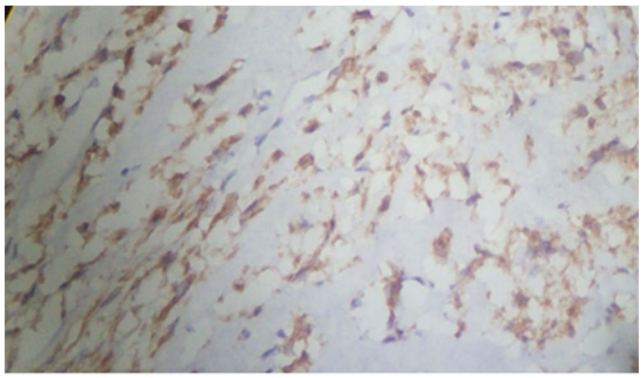

Fig. 7. Positive histological sections to calponin

renal leiomyosarcoma with metastasis to the left kidney was concluded.

Discussion

Histological types of renal sarcoma include leiomyosarcoma, liposarcoma, fibrosarcoma, rhabdomyosarcoma, and malignant fibrous histiocytoma. Leiomyosarcoma is the most common type, representing 50%; however, it is still a rare neoplasm originating in the intrarenal blood vessels or smooth muscle fibers of the renal pelvis [5]. The cause of the predominance in women is unknown; however, some studies suggest association with the X chromosome [6].

The symptoms are indistinct with respect to the carcinoma; however, in the literature, it is reported as the incidental finding of abdominal tumor, abdominal pain, lumbar pain, and hematuria and in isolated cases as fever [7]. In our patient, the form of presentation was an abdominal tumor associated with pain in the right flank, which motivated her to seek medical assistance.

Tumors, such as leiomyosarcoma or leiomyoma, contain a dense group of fusiform muscle cells and a variable amount of fibrous connective tissue, the latter produce a low-intensity sequence in T1, in magnetic resonance [8]. In computed tomography, areas of fibrous connective tissue produce a pattern of low reinforcement in the early phase and a greater reinforcement in the late phase. This form of behavior in imaging studies is characteristic of non-epithelial tumors, but it is not exclusive of these neoplasms [8].

The distinctive histological characteristics are the presence of fusiform cells with abundant necrosis, nuclear polymorphism, and high mitotic activity [9]. The immunohistochemical analysis expresses positivity to actin, calponin, desmin, and h-caldesmin [10]. The differential diagnosis of this neoplasm includes renal carcinoma with sarcomatoid differentiation, leiomyoma, and classic carcinoma, which are distinguishable only in the histopathological study.

The treatment of choice is a total radical nephrectomy [11]. Deyrup et al. [12] reported the nephron-sparing surgery in a patient with a 4-cm tumor as an option. In our patient, we chose for a conservative surgery in the left kidney because it is a bilateral tumor, with a negative intraoperative pathological margin of 1 and 0.5 cm. Considering the general aggressiveness of sarcomas and their high capacity for local and distant recurrences, treatment with adjuvant chemotherapy through mesna, adriamycin, ifosfamide, and dacarbazine and radiation therapy at a dose of 44 Gy to the renal fossa and adjacent lymphatic area is justified [13]. Especially, when having any of the following high risk factors, such as higher than 5-cm tumor, high histologic grade, retroperitoneum, and presence of necrosis. In the case of neoadjuvant therapy with chemotherapy or radiotherapy, more studies are needed to demonstrate its effectiveness in 5-year survival.

Conclusions

Renal leiomyosarcoma occurs in less than 0.1%, its clinical evolution is similar to other renal neoplasms, although its tomographic image may suggest the diagnosis in some cases. Its biological behavior is aggressive with an overall survival of 27%–36% at 5 years. Our case is one of the few reported with contralateral renal metastasis, treated with radical surgery for the primary tumor and conservative surgery in the metastasis, with survival until this time of 6 months.

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