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Controlateral and mesenteric metastasis of kidney leiomyosarcoma sixteen years after the surgery: A case report

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ABSTRACT

Introduction: Primary renal leiomyosarcoma is an extremely rare malignant tumour. It is more common in females than in males and in the right kidney. The diagnosis is difficult. The prognosis is poor because of frequent metastasis and recurrence. Case Report: A 41-year-old woman who had undergone left radical nephrectomy for a kidney tumour 16 years previously presented to our institution with right flank pain. On examination, the left sided mass was a 20 cm encapsulated firm tumor with necrotic and hemorrhagic foci compressing the residual kidney. Histological examination showed a high cellular proliferation of large spindle cells mixed with necrosis and calcification. Nuclei were irregular, hyperchromatic, 4 mitoses/10hpf were counted. Renal capsule and renal vein were tumor free. Tumors cells were positive for smooth muscle actin (AML), neuron specific enolase (NSE), with negative expression for, anti S100, cytokeratin, Epithelial membrane Antigen (EMA), HMB45 and CD34. Histopathological examination concluded to a low grade leiomyosarcoma without capsular effraction neither lymph node invasion measuring 20x13x10 cm. No postoperative treatment was indicated and the patient was lost to follow up for 16 years. Imaging finding (MRI) revealed a solid heterogenic mass at the lower pole of the right kidney. Besides, three mesenteric masses were noticed. A controlateral and mesenteric metastasis of the left kidney leiomyosarcoma was suspected. In order to confirm the diagnosis, an ultrasound guided biopsy of the right kidney lesion was performed. Histopathological findings concluded to a low grade leiomyosarcoma. The patient did not undergo surgery and was referred to a medical oncology center and chemotherapy is in progress. Conclusion: Leiomyosarcoma of the kidney is a rare aggressive tumour. Only few cases are reported in literature. Clinical features are non-specific making generally a delayed diagnosis. The treatment is based on surgery. The prognosis is still poor.

Keywords: Metastasis, Renal leiomyosarcoma, Recurrence, Surgery, Sarcoma

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INTRODUCTION

Primary renal leiomyosarcoma is an extremely rare malignant tumour that arises from the smooth muscle [1], from either intrarenal blood vessels or muscle fibers of the renal pelvis constituting less than 1% of all renal malignancies. It is more common in females than in males and in the right kidney. It is generally highly aggressive tumour with frequent metastasis and recurrence. Metastases are most likely to be found in lungs, liver, bone, and skeletal muscle [2]. In our cases, the controlateral and mesenteric metastasis of right kidney leiomyosarcoma was detected sixteen years after the first operation.

CASE REPORT

A 41-year-old woman presented to our hospital with a right flank pain. Her medical past history showed that she had undergone left radical nephrectomy for a kidney tumour 16 years ago. Histopathological examination concluded to a low grade leiomyosarcoma without capsular effraction neither lymph node invasion measuring 20x13x10 cm. No postoperative treatment was indicated and the patient was lost to follow up for 16 years.

She consulted again for left flank pain, described as a sensation of left abdominal heaviness, associated with chronic fatigue. Physical examination showed nothing abnormal but a palpable right flank mass. Besides, her vital signs were stable and no other abnormalities were found. Biological exams were normal: in this particular case, no renal failure was noticed.

The CT scan and abdominal magnetic resonance imaging (MRI) revealed a solid heterogenic mass approximately 88x74x64 mm at the lower pole of the right kidney. No thrombus in renal vein or vena cava was detected. Besides, MRI showed also the presence three mesenteric masses (Figure 1 and 2). There were located respectively at the left mesenteric edge (104x84x58 mm), the left paracolic gutter (38x36x25 mm) and a third one at the left side of the rectum (17x14x11 mm). The thoracic computed tomography and whole body bone scintigraphy did not reveal any other metastatic foci.

Based on these findings the diagnosis of a controlateral and mesenteric metastasis of the left kidney leiomyosarcoma was suspected. In order to confirm the diagnosis, an ultrasound guided biopsy of the right kidney lesion was performed. The biopsy only focused on the tumor with no renal parenchyma included. It showed a fusocellular proliferation with increased cellularity. Fascicules of long spindled eosinophilic cells showing a whirling, or haphazard architecture with no zone of necrosis. There were no epitheloid cells nor marked pleomorphism Irregular shaped and hyperchromatic nuclei were observed. We counted 5-6mitoses/hpf. Histopathological findings concluded to a low grade leiomyosarcoma (Figure 3). The immunohistochemical profile was diffusely positive with smooth muscle actin (Figure 4). The patient did not undergo surgery, she was referred to a medical oncology center and chemotherapy is in progress based on Doxorubicin in combination with Ifosfamide.

DISCUSSION

Primary sarcomas are rare malignant tumour. Its incidence ranges between 0.8 and 2.7% in adults [1–3].

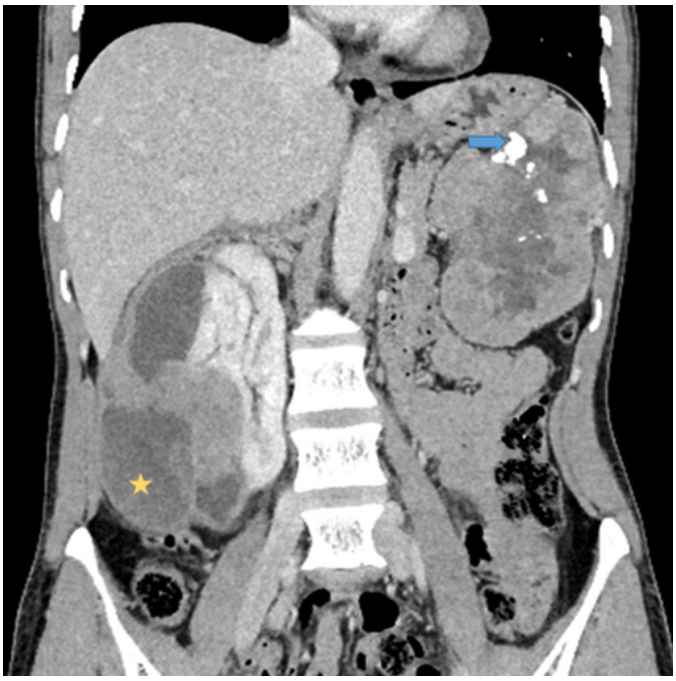

Figure 1: Contrast enhanced coronal CT scan shows two lesions: One mesenteric in the left upper abdomen and the other in peripheral cortex of right kidney with perinephric extension. They contain three components: solid component with moderate enhancement, large central necrosis (Yellow star) and coarse calcification (Blue arrow).

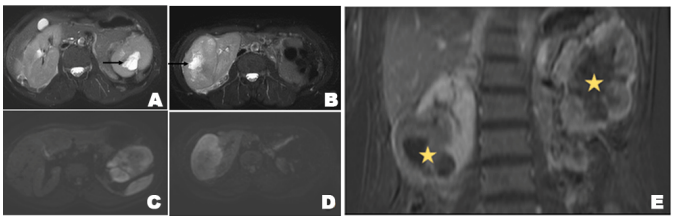

Figure 2(A–E): Axial T2-weighted image show the two lesions with 2 different signal intensity portions one peripheral solid component and another central necrotic (Black arrow) (A, B). Axial Diffusion-WI Hyperintense components/restricted diffusions are seen in the mass (C, D). Coronal reformatted gadolinium-enhanced 3DFSPGR reveals a heterogeneous enhancement of the tumor (Yellow star) (E).

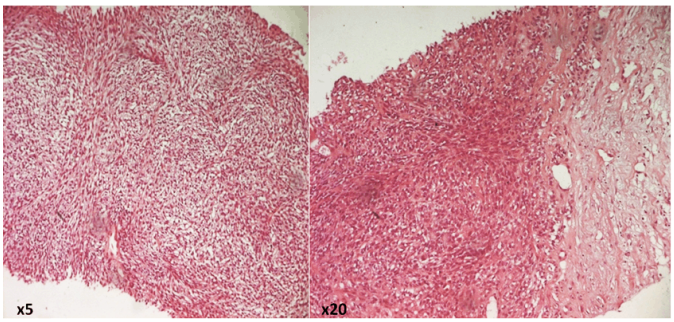

Figure 3: Low grade renal leiomyosarcoma: Histological examination showing fascicles of long spindle eosinophilic cells showing a whirling, or haphazard architecture. Nuclear pleomorphism is low to moderate. We counted 5-6mitoses/hpf. No renal tissue was observed.

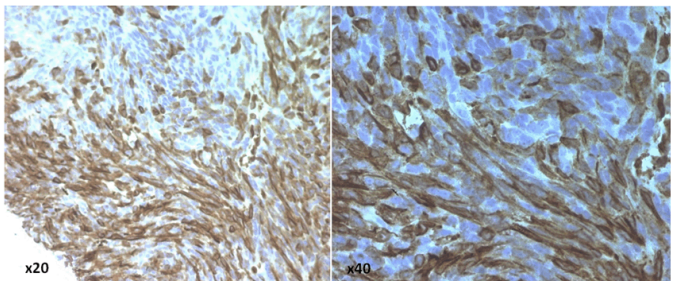

Figure 4: Spindled tumour cells showing positive cytoplasmic immunostaining for smooth muscle actin.

Although leiomyosarcoma is the most frequent type, it remains rare and is generally located in soft tissues (11%) or in the uterus in 30% of cases. It has been reported in the retroperitoneum, bones, blood vessels, thyroid gland or skin. Kidney is a rare location of leiomyosarcoma and its incidence does not exceed 0.1% of all invasive renal malignancies [4]. Mostly, it takes origin from the smooth muscle fibers of the renal vessels or the renal capsule.

It occurs preferentially in women than in men and more common in the right kidney. It is diagnosed in most cases in the fourth to sixth decades of life [2–5].

Clinical features are non-specific, patients most commonly presented with abdominal pain or palpable mass accompanied by hematuria. Other associated symptoms include weight loss and gastrointestinal symptoms [2–6]. In our cases, the patient presented with an isolated right flank pain.

Imaging investigations offers potential advantages for diagnosis and detection of metastatic locations. On computed tomography (CT) and MRI, Leiomyosarcoma appear as heterogeneous mass with irregular borders of varying density and signal intensities [1, 7–8]. However, leiomyosarcoma cannot be distinguished from renal cell carcinoma based on imaging.

Confirmation of the diagnosis is usually made by histopathological examination [6–8]. The microscopic features of leiomyosarcoma include the presence of classic spindle cells with eosinophilic cytoplasm hyperchromatic nuclei that show varying degrees of mitotic activity [8–9]. Cellular atypia, mitotic activity, and necrosis determine the degree of differentiation. In this case, our patient's pathologic specimen revealed fascicles of long spindle cells with the presence of 5 to 6 mitoses and the absence of necrosis. These characteristics are compatible with a low grade leiomyosarcoma. Immunochemical analysis is mandatory to precise the histological type and confirm the diagnosis since the difference between leiomyosarcoma and other histological types of renal tumours is based on its response to a panel of markers [1, 2, 5, 6–8]. Classically, it is mainly positive for smooth muscle actin, and it could also be positive to Calponin, Desmine and Vimentin with negative expression for KL1, Anti S100 or Cytokeratin [9–10]. In our case, the immunohistochemical profile was strongly positive with smooth muscle actin.

Concerning the therapeutic strategy, surgery seems to be the only curative treatment for primary renal leiomyosarcoma [1, 2, 6, 8, 11, 12]. Adjuvant chemotherapy or radiotherapy can be used in selected cases and the results are still controversial. Published studies show better local control of leiomyosarcoma but no survival benefit [1, 5, 11]. Our patient did not undergo surgery because of metastatic lesions. A palliative chemotherapy is in progress.

The prognosis of leiomyosarcoma is poor because of frequent metastasis and recurrence, although occasionally long-term survivors are reported in literature [13]. Our patient is still alive for 16 years after surgery which make this case particular.

Low histological grade, absence of lymph node metastasis, size less than 5 cm and radical surgery are associated with better prognosis. Usual secondary locations are lungs, liver and bone. Controlateral and mesenteric metastasis are rare [1–8].

CONCLUSION

The leiomyosarcoma of the kidney is a rare malignant tumour. The diagnosis remains difficult because of non-specific presentations. Radical surgery represents the only curative treatment. Adjuvant therapies can be used in selected cases. The prognosis is worse with frequent recurrence and metastasis.

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Author Contributions

Skander Zouari – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Mouna Ben Othmane – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Wissem Hmida – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Yesser Ben Cheikh – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Mehdi Jaidane – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ali Faouzi Mosbah – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.