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LEIOMYOSARCOMA OF THE KIDNEY: A VERY RARE CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Background: Renal leiomyosarcoma is a very rare malignancy. It presents with non specific clinical and radiological signs. Literature regarding this tumour has been limited to case reports. **Material and Methods:** To highlight its clinicopathologic features, we report a case of 74-year-old male with renal leiomyosarcoma. **Results:** The man was presented with a left flank pain. Primary diagnosis was based on endoscopic biopsy and immunochemical analysis. The patient wasn't amanable to surgery and he was given two lines of chemotherapy with no response, he was died after 26 months of follow up. **Conclusion:** Renal leiomyosarcoma has usually a dismal prognosis.Chemotherapy is a feasible option with the clinicians but its results are not well established.

KEYWORDS: Renal leiomyosarcoma, clinical presentation, imaging findings, treatment modalities, prognosis.

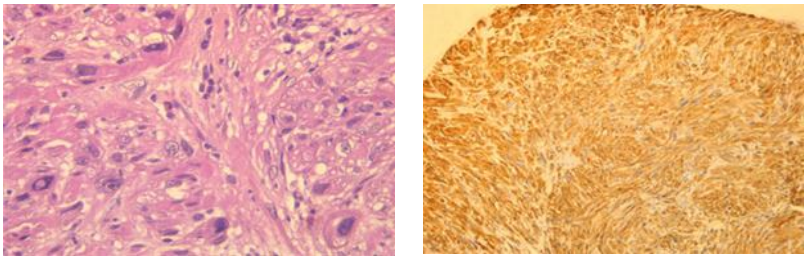
INTRODUCTION

Renal leiomyosarcoma (LMS) is a rare entity, comprising from 0.5 to 1% of all renal malignancies[1], while accounting for the majority of renal sarcomas (50 to 60%).[2] Commonly, it arise from the renal capsule, renal vein or the renal pelvis.[1] There is a paucity of data regarding the pathologic features and outcomes of patients with these tumors and usually it is a highly agressive disease with a dismal outcome.[2]

We report a case of primary renal LMS treated in 2013 in the medical oncology department of Fattouma Bourguiba university hospital, Monastir (Tunisia) to improve understanding of clinical presentation, imaging findings and treatment modalities.

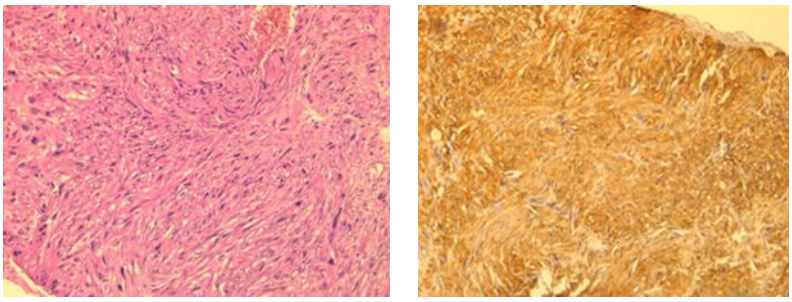
RESULTS

A 74-year-old male complained of oppressive pain in the left flank of 1 month of evolution. No abnormality was detected on his physical examination.The abdominal tomography showed 10 cm upper left polar renal tumor with heterogeneous and irregular borders around it, significantly enhanced after injection of contrast agent, encompassing the left iliac artery, compressing the renal vessels and closely related to the psoas muscle with no regional lymph nodes involvement. The metastatic workup was completed by bone scintigraphy revealing no metastases. Ureterorenoscopy with biopsy revealed the lesion to be a LMS.Immunostaining for Desmin was strongly positive in the tumour cells and for smooth muscle actin and weakly positive in epithelial membrane antigen (EMA)(**Fig 1-2**). The cells were, however, negative for anti-cytokeratin antibodies (anti CK) and PS100.The tumor was juged inoperable and hence the patient was given chemotherapy (CH) based on six courses of adriamycin in monotherapy with a radiological progression of the renal mass at evaluation but with no metastasis (**Fig. 3**). Then, the patient received 3 courses second line CH with Gemcitabin in monotherapy. Unfortunately, any response was developped and the patient was died after 26 months of follow up.



A B

Figure 1 : Fascicles of spindle cells with pleomorphic nuclei and eosinophilic cytoplasm (A: Hematoxylin Eosin x100, B: Hematoxylin Eosin x 400)



A B

Figure 2 : Diffuse immunostaining of tumor cells for Smooth Muscle Actin (A) and Desmin (B) (x 100)

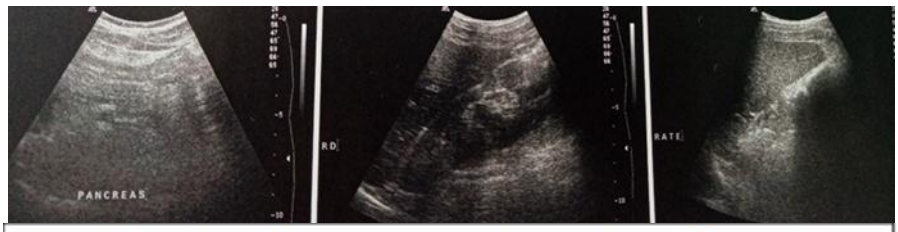


Figure 3 : Abdominal ultrasonography showing solid mass lesion of 16 cm in the left kidney after first line CH (progressive disease)

DISCUSSION

LMS is still a rare malignant disease originating in the intrarenal blood vessels or smooth muscle fibers of the renal pelvis.[3] These tumors occur mainly in elderly between the fourth to eighth decades of life whereas cases in children have also been reported.[4] Male to female ratio is 1:2.[5] The cause of the predominance in women is unknown; however, some studies suggest association with the X chromosome.[3] A preference for right side is noted and also bilaterally.[4,6] Although, since the renal vein is longer on the left side, in cases of LMS arising from the renal vein, a greater proportion of left sided lesions might be expected.[7]

There was an increasing frequency of LMS in immunocompromised patients and an association with Epstein Barr virus, penis carcinoma, renal calculus, xeroderma pigmentosa and tuberous sclerosis.[6]

They generally have an insidious clinical presentation[2] and symptoms usually indicate locally advanced disease.[8] Associated signs are indistinct with respect to the carcinoma including flank pain as our patient, abdominal mass, hematuria and loss of weight and appetite. Emergency presentations with spontaneous rupture and severe peri-renal hemorrhage were seldom reported.[9]

Imaging findings are non specific and quite comparable to other renal tumors. At computed Tomography (CT), renal LMS manifests as a well-defined solid mass with heterogeneous and delayed enhancement of the fibrous

stroma. Rarely, as a multilocular cystic mass with peripheral enhancement. Central necrotic areas is also seen in large tumors and sometimes signs of locoregional invasion such our case. At magnetic resonance imaging (MRI), LMS appears as a soft-tissue mass with low-signal-intensity areas on T1- and T2-weighted images showed delayed enhancement, this form of behavior in imaging studies is characteristic of non-epithelial tumors, but it is not exclusive of these neoplasms.[3,10] So, neither ultrasonography, tomography or MRI are able to differentiate between LMS and renal cell carcinomas.Therefore, the diagnosis is usually made postoperatively.[5] Even, ureterorenoscopy with biopsy may fail to obtain an adequate tissue specimen.[1] In the presented case, fine needle aspiration and immunohistochemical study were performed with good results.

Histologically, LMS show characteristics of smooth muscle tumor with alternating fascicles of fusiform cells. The cells have blunt ended, nontapering nuclei and eosinophilic cytoplasm. Indicators of malignancy are necrosis, nuclear pleomorphism and more than rare mitotic figures.[11] Immunohistochemically, they show reactivity for vimentin, actin, smooth muscle myosin, desmin, H-caldesmon and basal lamina components including laminin and type IV collagen.[6] Andrea T et al. Indicates that LMS have tendency toward higher-grade lesions and tumor behavior correlated is with grade.[7] Renal LMS may be confused with renal carcinoma with sarcomatoid differentiation since both contain a large number of fusiform cells and the differential diagnosis for LMS of kidney includes leiomyoma,fibrosarcoma, urothelial carcinoma and angiomyolipoma.[5,12] Renal LMS usually have an aggressive biological behavior with poor prognosis and 5-year survival rates of 29–36%.[8,13] Their lethality arises from the early haematogenous dissemination, most of the patients develop distant metastases at the time of detection and the lungs were the primary site of spread.[13] These patients are not amenable to surgery.[14] Miller et al. Noted in their series of 27 renal LMSs a distant metastases in 90% of patients after an average follow-up of 2.8 years.[15] Renal LMS should be treated as sarcoma using multimodality therapy so as to optimize survival and reduce tumor recurrence.[9] Radical or enlarged nephrectomy with wide margins is the treatment of choice but it is uncertain whether it's sufficient or radical nephroureterectomy is superior.[1] The major prognostic factor is total surgical resection.[8] Although the role of lymphadenectomy remains controversial.[8] Interestingly, conservative surgical treatment by partial nephrectomy has been attempted in exceptional cases of localized, small, polar renal nodules, low grade tumor with a unique kidney or bilateral LMSs.[3,4,9] Triple therapy including surgery, CH and radiotherapy (RT) has been advocated because of the aggressiveness of these disease but results are not well established.[7] Adjuvant CH through Adriamycin, Ifosfamide, Dacarbazine and RT at a dose of 44 Gy to the renal fossa and adjacent lymphatic area is justified by Vasquez Ciriaco et al. especially, when having any of the following high risk factors such as higher than 5-cm tumor, high histologic grade, retroperitoneum, and presence of necrosi.[3] Neoadjuvant CH has been prescribed by some authors but more studies are needed to demonstrate its effectiveness in 5-year survival.[3,4] The possibility of treatment with tyriosine kinase inhibitors such as sunitinib has been reported in phase II trial.[16] In a study of 34 metastatic LMS (with 29 uterine location), 53% had some response to Gemcitabine and Docetaxel; yet the median survival was still only 17.9 months.Accordingly, CH for metastatic LMS is not successful.[15]

CONCLUSION

In conclusion, renal LMS is a very rare tumor that clinically and radiographically mimics more common renal malignancies with high metastatic potential. It presents a diagnostic challenge.The extensive surgical excision of the tumor is the only way for the exact histopathological diagnosis and the cornerstone of the treatment but LMS should be treated aggressively to achieve better results.[12]

We reported a rare case of primary inoperable LMS arising from the renal pelvis in an old male which differential diagnosis is only possible by histopathological analyses based on percutaneous endoscopic biopsy. Because of the surgical irresectability, this unfortunate patient was only treated by CH with unfavorable results. Paucity of cases and absence of long-term follow up with controlled randomized studies is hampering definitive treatment protocols.

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