



## Primary Renal Leiomyosarcoma: A Case Report

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### Introduction

Primary sarcomas of kidney are rare tumors, which comprise 0.8% of all renal tumors. Leiomyosarcoma account for 50-60% of kidney sarcomas; followed by liposarcomas in 10-15% of cases<sup>1</sup>. They usually occur in the right kidney<sup>2</sup>. Mesenchymal tumors of the kidney lack natural barriers and hence are able to expand and present as a large abdominal lump. Sarcomas typically possess a pseudocapsule, which is often infiltrated by the tumor<sup>3</sup>. The documented biological behaviour of renal leiomyosarcoma is unpredictable which may be due to paucity of extensive literature. However, the disease is known to be associated with a poor prognosis and a high metastatic potential<sup>4</sup>.

There are only a limited number of case reports and two case series on primary renal leiomyosarcoma. We present the case below to improve understanding of clinical presentation, radiographic findings, and pathologic features of this rare and aggressive tumor.

### Case Report

A 50 year old lady was admitted with complaints of pain and lump in left loin for the past 4 months. The lump had been steadily increasing in size and was accompanied with dull aching pain. She also gave history of anorexia for the past one year. There were no additional symptoms and no hematuria at the time of admission.

### Physical Examination

On per abdominal examination, abdominal distention was observed and a lump was palpable in the left hypochondrium extending into left lumbar region and left epigastrium measuring about 13 cm below the costal margin and 8 cm medial to the mid clavicular line. There was no tenderness elicited. Her blood pressure was 100/70 mm of Hg and pulse rate 84/min.

Examination of all other systems were found to be unremarkable.

Urine routine and microscopic examination was within normal limits and was negative for malignant cells.

Her complete blood count, kidney function tests and liver function tests were all within normal limits.

CECT Abdomen: Well defined heterogeneous mass lesion arising from upper pole of left kidney measuring 10.2 x 18.8 x 13.2 cm with significant post contrast enhancement was observed. In non enhancing areas signs of necrosis were seen. Lesion was pushing pancreas and splenic vessels anteriorly with loss of fat planes. The mass was pushing the coeliac trunk inferiorly and laterally, extending towards the right side abutting the posterior abdominal wall with loss of fat planes at places. Left suprarenal gland was not visualized separately. A provisional diagnosis of Renal cell carcinoma was suggested.

In view of clinic-radiological impression of renal cell carcinoma, radical nephrectomy along with splenectomy was done. On gross examination, a specimen of tumor with kidney and spleen together measured 23x18x14.5 cm. Tumor was present on the upper pole of the left kidney measuring 18.5x15x14.5cm. The outer surface of the tumor was bosselated, partially encapsulated. On cut surface tumor was solid, grey white and soft to firm with whorled areas. Few areas of hemorrhage, cystic degeneration and necrosis were identified. No renal hilum, ureter or vessels were identifiable grossly. No adrenal gland was separately identified.

Spleen measured 10x 8x 3.5 cm, outer surface was unremarkable.

On cut section spleen appeared homogenous.



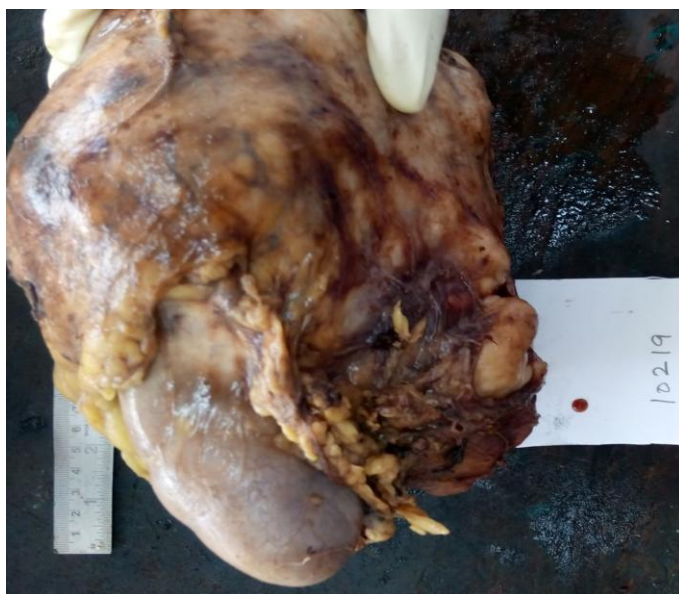
**Fig 1 & 2:** Radical nephrectomy specimen with spleen. Only a part of the kidney was identified. Tumor was present on the upper pole of the left kidney. The outer surface of the tumor was bosselated, partially encapsulated.



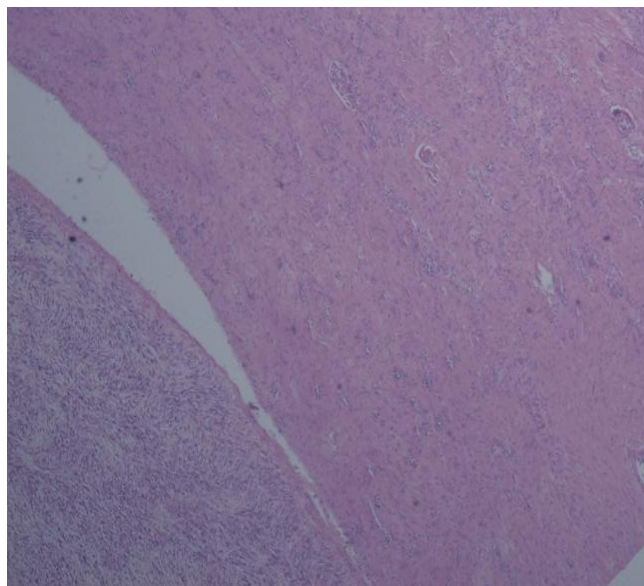
**Fig 3:** On cut surface tumor was grey white, firm with whorled areas. Few areas of hemorrhage, cystic degeneration and necrosis were identified. No renal hilum, ureter or vessels were identified on gross.

### On Histopathological Examination

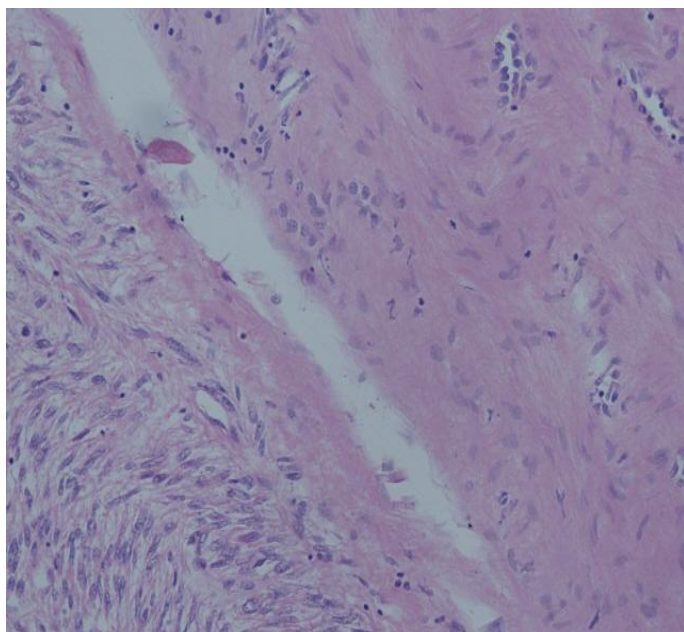
Histologically, the tumor was composed of alternating fascicles of spindle cells with eosinophilic cytoplasm and blunt ended, non tapering nuclei. Though focal necrosis (< 50% of specimen) was found, the tumor predominantly had myxoid areas. Mean mitosis were 7/10 per high power field. Minimal inflammation and



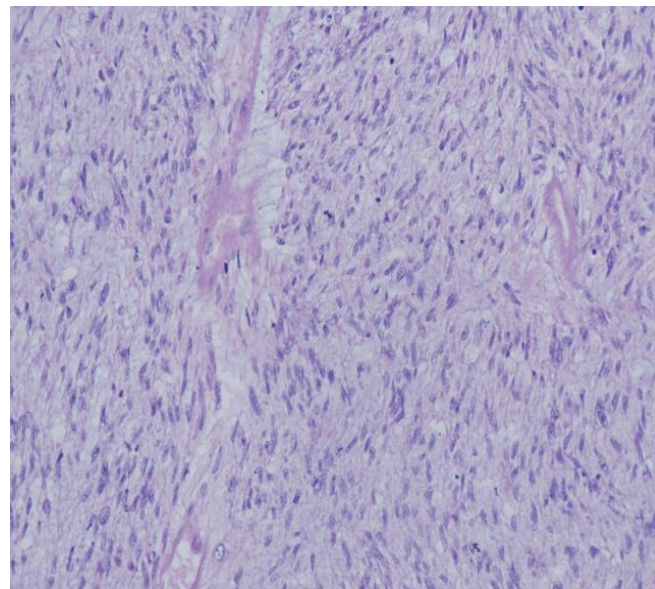
lymphovascular invasion was observed. No epithelioid cells were seen, however focal areas of high- gradepleomorphism were found. The tumor was limited to the kidney and probably arose from the renal vessels. Immunohistochemically the tumor was SMA, Vimentin and Bcl-2 positive and was negative for cytokeratin and final diagnosis of leiomyosarcoma was made.



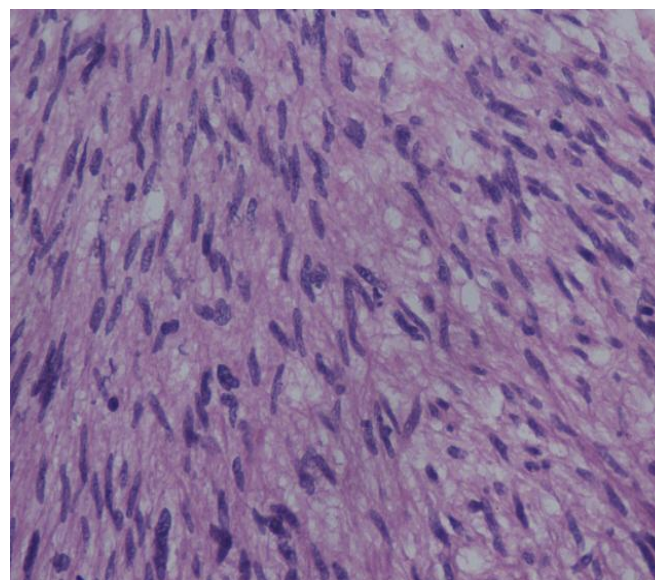
**Fig 4:** 4X: The tumor was encapsulated with clear demarcation from normal kidney.



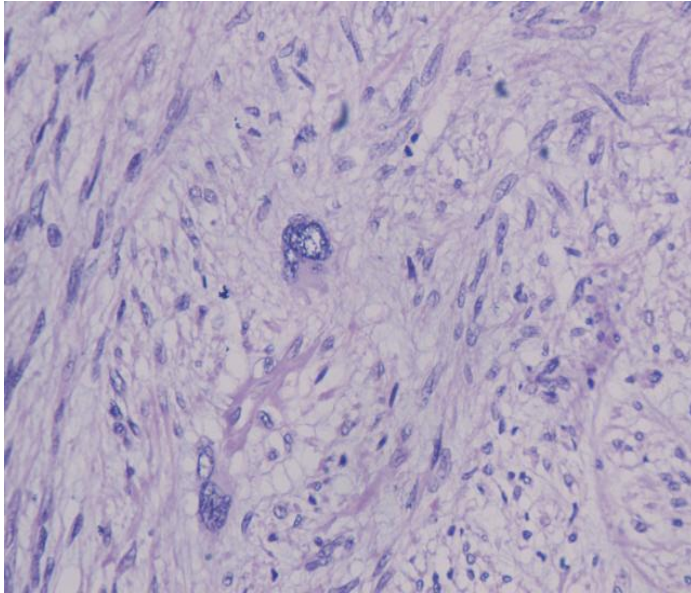
**Fig 5:** 20X: The tumor was limited to the kidney and probably arose from the renal vessels.



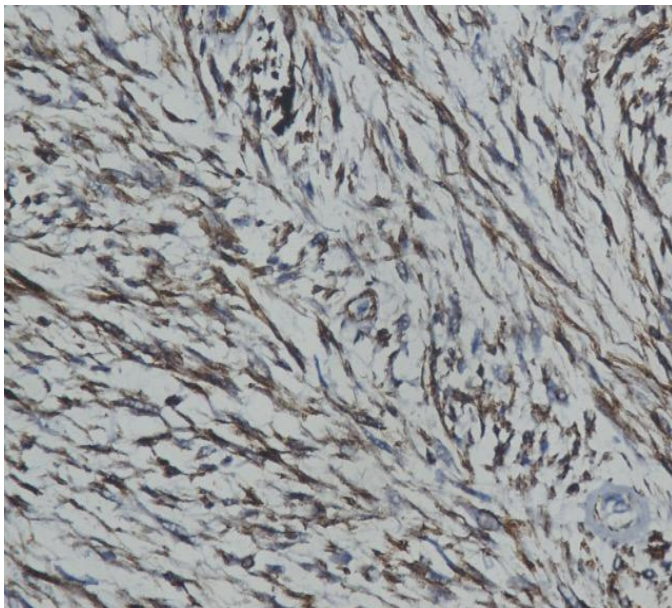
**Fig 6:** 10X: The tumor predominantly had myxoid areas. Mean mitosis were 7/10 per high power field.



**Fig 7:** 40X: Tumor was composed of alternating fascicles of spindle cells with eosinophilic cytoplasm and blunt ended, nontapering nuclei. Minimal inflammation and lymphovascular invasion was observed.



**Fig 8:** 40X: No epithelioid cells were seen, however focal areas of high- grade pleomorphism were found.



**Fig 9:** 10X: Immunohistochemically the tumor was SMA (figure), Vimentin and Bcl-2 positive and was negative for cytokeratin

### Discussion

Renal leiomyosarcomas are observed to arise from the smooth muscle fibers of renal pelvis, renal capsule or renal vessels. Leiomyosarcoma originating from the renal vessels are most frequently documented<sup>5</sup>. These tumors usually present insidiously, with symptoms and signs occurring at late stages of the disease: which include abdominal pain, palpable mass, vomiting,

hematuria and also cachexic symptoms such as weight loss<sup>6</sup>. Neither ultrasonography, tomography or magnetic resonance are able to differentiate between leiomyosarcomas and renal cell carcinomas<sup>7</sup>.

Data from the American Joint Committee on Cancer (2010) has revealed that the prognosis of soft-tissue sarcomas was directly associated with the disease stage<sup>8</sup>. In addition to the tumor-node-metastasis (TNM) classification of renal sarcomas (Table I), the specific histological grade of the sarcoma is also used in tumor staging. However, the use of the TNM classification in the staging of sarcomas does not sufficiently predict the prognosis. Therefore, the histological grade is determined based upon the scoring system of the French Federation of Cancer Centers Sarcoma Group (Table II)<sup>9</sup>.

Surgical excision and the stage of the tumor are the most important predictors of prognosis for cases of renal sarcoma. Surgical resection is the only prognostic factor able to confer increased survival rates for patients with a primary tumor, or for those who present with a primary tumor and metastatic disease. An inability to perform surgical resection appears to be the most unfavorable prognostic variable for overall survival<sup>10</sup>.

The presence of metastasis at the time of diagnosis is another important predictor of prognosis, as the mean survival rate is usually shorter in those patients with metastatic disease at diagnosis<sup>11</sup>. A tumor grade is assigned according to the number of mitoses per high-power field, the degree of cellularity, the cellular and nuclear morphology, and the presence of necrosis. Discordance between pathologists with respect to tumor grading and histological subtype can be substantial<sup>12</sup>.

Increased rates of necrosis, poor differentiation, mitotic activity and increased histological grade are associated with a poor prognosis. A study by Deyrup et al revealed an association between the increasing histological grade of renal leiomyosarcomas and the survival rate<sup>13</sup>.

Higher histological grade was defined as a poor prognostic factor. Low-grade soft-tissue sarcomas exhibit limited metastatic potential, but also tend to recur locally. Therefore, surgical excision, which includes the removal of negative tissue margins measuring 1-2 cm or more in all directions, is the recommended treatment for patients with early-stage sarcomas<sup>14</sup>.

The involvement of regional lymph nodes, although extremely rare in cases of kidney sarcoma, is predictive of a poorer prognosis.

### Conclusions

Primary Renal Leiomyosarcoma is a rare diagnosis, but should always be considered in asymptomatic older females with massive renal lump, given its metastatic potential and grim prognosis. As a presumptive diagnosis of leiomyosarcoma on clinical and radiological grounds remains fairly difficult, histopathological analysis remains the mainstay for diagnosis and staging.

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**Appendix****Table I.**

Sarcoma stage according to the TNM system of the American Joint Committee on Cancer.

Stage Tumor characteristics

IA T1, N0, M0, G1 or GX: Tumor is  $\leq 5$  cm

IB T2, N0, M0, G1 or GX: Tumor is  $> 5$  cm

IIA T1, N0, M0, G2 or G3: Tumor is  $\leq 5$  cm

IIB T2, N0, M0, G2: Tumor is  $> 5$  cm

III T2, N0, M0, G3: Tumor is  $> 5$  cm; OR Any T,

N1, M0 or G: Tumor can be any size

IV Any T, N, M1 or G: Tumor can be any size

T, tumor; N, node; M, metastasis; G, grade; GX, the grade cannot be assessed.

**Table II.**

Grading system based upon the French Federation of Cancer Centers Sarcoma Group.

Tumor necrosis

Score 0, no necrosis

Score 1,  $< 50\%$  tumor necrosis

Score 2,  $\geq 50\%$  tumor necrosis

Tumor differentiation

Score 1, good

Score 2, intermediate

Score 3, poor

Mitotic count

Score 1, 0-9 mitoses per 10 HPF

Score 2, 10-19 mitoses per 10 HPF

Score 3,  $\geq 20$  mitoses per 10 HPF

Histological grade

Grade 1; total score 2,3

Grade 2; total score 4,5

Grade 3; total score 6,7,8