# Primary Leiomyosarcoma of the Kidney: A Case Report

**Naveen Thimiri Mallikarjun***

*Department of Urology, Mahatma Gandhi Medical College, Puducherry, India*

***Corresponding Author****

Naveen Thimiri Mallikarjun
Department of Urology,
Mahatma Gandhi Medical College,
Puducherry, India
E-mail: tm.naveen1992@gmail.com



**Received:** July 14, 2023, Manuscript No. OCCRS-23-106248; **Editor assigned:** July 21, 2023, PreQC No. OCCRS-23-106248 (PQ); **Reviewed:** August 04, 2023, QC No. OCCRS-23-106248; **Revised:** January 03, 2025, Manuscript No. OCCRS-23-106248 (R); **Published:** January 10, 2025, DOI: 10.35248/2471-8556.25.11(1).003

## Abstract

Renal Leiomyosarcoma (LMS) is a rare and aggressive mesenchymal tumour. The tumour usually arises from the smooth muscle component of the kidney and has a high tendency for local recurrence. Herein, we report a case of primary leiomyosarcoma in a 55-year-old woman, presenting with upper abdominal pain.

**Keywords:** Renal leiomyosarcoma • Renal sarcoma • LMS • Mesenchymal tumour • Smooth muscle

## Introduction

Primary renal sarcoma is a rare entity and represents only 1% among all malignant renal tumors [1,2]. Leiomyosarcoma (LMS) is the most common variant among renal sarcomas and arises from the mesenchymal component of the kidney. Renal leiomyosarcoma tends to displace adjacent structures rather than invade and is characterized by rapid growth rate with hypovascular pattern [3]. Renal LMS shows a strong predilection for the female gender with peak incidence in the fifth decade of life [4]. Herein, we report a case of a 55-year-old woman who presented with non-specific upper abdominal pain for 6 months with significant loss of weight. Upon radiological evaluation, she was diagnosed with an exophytic large left renal mass without lymphadenopathy. A diagnosis of primary renal leiomyosarcoma arising from the renal capsule was established after histopathological and immunohistochemistry analysis.

## Case Report

A 55-year-old woman presented to our outpatient department with complaints of vague upper abdominal pain associated with history of significant loss of weight for 6 months. The pain was unrelated to diet. On physical examination, her vital signs were within normal limits. Laboratory data were also within normal limits. An abdominal ultrasound done revealed a heterogenous mass measuring 6.5 × 5 cm originating from the lower pole of left kidney (Figure 1). For further characterisation, a Contrast-Enhanced Computed Tomography (CECT) of abdomen was done which showed a large heterogeneous tumour (7 cm) at the lower pole of the left kidney. Lymphadenopathy and venous thrombosis were absent (Figure 2). High Resolution CT (HRCT) of the chest was normal. Based on the tentative diagnosis of renal cell carcinoma (cT2N0M0), we proceeded with open left partial nephrectomy. Intra-operative frozen section confirmed negative tumour margins. nephrectomy.

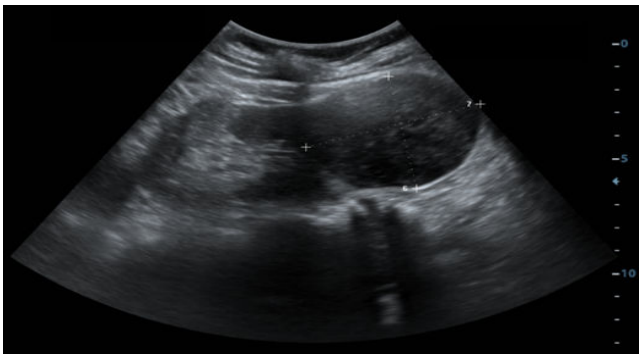

**Figure 1.** Ultrasound of abdomen showing a heterogenous lesion arising from the lower pole of left kidney.

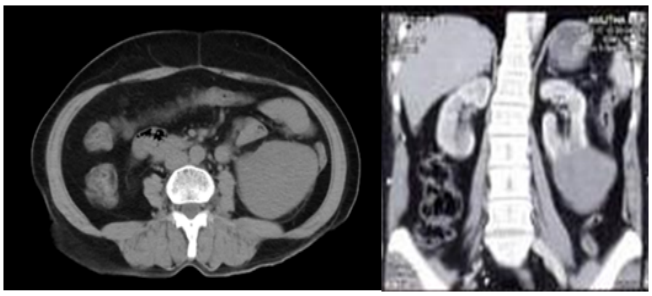

**Figure 2.** Computed tomography of abdomen showing exophytic tumour (7 × 6.5 cm) originating from the lower pole of the left kidney, breaching the perirenal fat and gerota's fascia.

On hemisection of the soft tissue mass along with attached fibrofatty tissue, a firm grey white well circumscribed tumour measuring 6.5 × 6 cm was found. Upon microscopic examination, the tumour was found to be composed of well-encapsulated spindle cells arranged in bundles with a degree of high cellularity and an interlacing fascicular pattern. Most tumour cells had moderate eosinophilic cytoplasm (Figure 3). No renal epithelial tumour was identified in the extensive sampling. Immunohistochemical analysis showed that the tumour cells were diffusely positive for smooth muscle actin. Based on these findings, a final diagnosis of primary renal leiomyosarcoma originating from the renal capsule was made. No adjuvant therapy was provided after surgery.

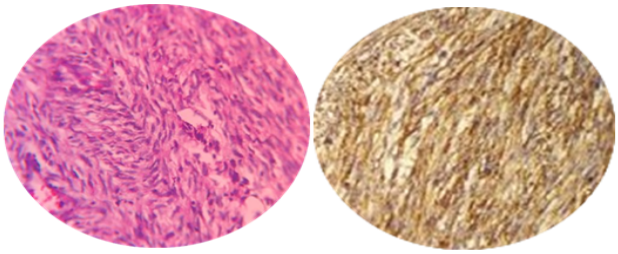

**Figure 3.** Histopathologic section showing spindle shaped tumour cells arranged in bundles with a high degree of cellularity and an interlacing fascicular pattern with moderate eosinophilic cytoplasm. IHC showing tumour cells with strong positivity for Smooth Muscle Actin (SMA).

## Discussion

Primary renal leiomyosarcoma represents <1% among all renal malignancies and has a poor prognosis [1,5]. Renal sarcomas are derived from mesenchymal component and hence are free of natural barriers that limit dissemination. These tumours are typically surrounded by a pseudocapsule that is often infiltrated with malignant cells. Suspicion of renal LMS should be based on certain specific findings which includes origin of mass from renal capsule or perisinous region of kidney, large size renal mass in the absence of lymphadenopathy and hypovascular pattern [3].

The most important prognostic factors are margin status and tumour grade. Low-grade sarcomas often have an indolent course whereas high-grade sarcomas often metastasize, with lungs being the primary site of spread [6]. There is no specific grading system for primary renal leiomyosarcoma, though the French Federation of Cancer Centers classification system is often used. According to this system, the tumour is given a score ranging from 1-3 depending on the degree of differentiation, a second score of 1-3 for the number of mitoses, and a third score of 0-2 for the degree of necrosis. A tumour with a total score of 2 or 3 is defined as a grade-I tumour, 4 or 5 as a grade-II tumour, and 6–8 as a grade-III tumour. Grade II and III tumours are referred to as high-grade tumours [3,7].

On microscopic analysis, leiomyosarcoma needs to be differentiated from sarcomatoid variant of renal cell carcinoma, leiomyoma, angiomyolipoma, osteogenic sarcoma, and malignant peripheral nerve sheath tumour. Renal leiomyosarcoma can be differentiated from leiomyoma by the presence of cellular pleomorphism, increased mitosis, and necrosis whereas sarcomatoid carcinoma lacks the uniform fascicular pattern typical of leiomyosarcomas. An extensive tumour sampling in renal leiomyosarcoma is required to rule out any epithelial component of sarcomatoid renal cell carcinoma. Renal angiomyolipoma has fascicles of smooth muscle cells with an admixture with mature fat. Leiomyosarcoma is immunoreactive for SMA, calponin, desmin, and h-caldesmon and nonreactive for CK, S-100, and HMB45. Sarcomatoid carcinoma is positive for CK [1,8]. Radical nephrectomy is the standard treatment for primary renal leiomyosarcoma [1,3]. There is no consensus on the role of neoadjuvant or adjuvant radio/chemotherapy in the management of renal LMS [9].

## Conclusion

To conclude, primary leiomyosarcoma of kidney is a rare entity and clinical presentations as well as radiology imaging, simulate other renal malignancies. Clinching the diagnosis holds relevance for its therapeutic and prognostic bearing. A thorough histopathological analysis and careful interpretation of immunohistochemical markers are necessary to arrive at the correct diagnosis for proper management.

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**Cite this article:** Mallikarjun NT. "Primary Leiomyosarcoma of the Kidney: A Case Report". Oncol Cancer Case Rep, 2025, 11(1), 1-2.