International Journal of Research and Review
[Indexed, Peer-Reviewed Journal]
Volume 12; Issue: 6; June 2025
Website: www.ijrrjournal.com
Case Report *E-ISSN: 2349-9788; P-ISSN: 2454-2237*

Cytohistomorphological Features of Primary Renal Leiomyosarcoma: A Rare Case Report

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DOI: https://doi.org/10.52403/ijrr.20250635

ABSTRACT

Primary sarcomas of the kidney are extremely rare tumor in adults accounting for about 1% of all tumours. The most common type is leiomyosarcoma (40%) followed by pleomorphic sarcoma, hemangiopericytoma, fibrosarcoma and unclassified sarcoma. Primary renal leiomyosarcoma is thought to originate from the smooth muscle cells of the renal pelvis, renal capsule or the renal vessels.

A 51-year-old female presented with abdominal pain and swelling in left flank. On systemic examination abdomen was soft, non-tender with a palpable mass of size 8*6 cm in left flank. CECT revealed a lobulated exophytic enhancing soft tissue mass lesion in left lumbar region abutting the lower pole of left kidney, left renal vessels and left pelviureteric junction provisional diagnosis of renal cell carcinoma. FNA smears showed clusters, bundles and fascicles of plump round to oval to spindle shaped cells with focal storiform pattern. Diagnosis of a retroperitoneal sarcomatoid lesion was made with differential diagnosis of Leiomyosarcoma, synovial sarcoma and sarcomatoid carcinoma. Histopathological examination and immunohistochemistry confirmed the diagnosis of leiomyosarcoma of kidney.

Radiological investigations are not helpful in differentiating leiomyosarcomas from other renal carcinomas. Cytological smears show predominantly atypical spindle cells. Histopathology and immunohistochemistry are mainstay for diagnosis.

Keywords: Kidney, cytology, histopathology, sarcoma, immunohistochemistry

INTRODUCTION

Primary sarcomas of kidney are rare tumors, accounting for approximately 1 - 3% of all primary renal tumors, with leiomyosarcoma being the most common subtype(40-60%) of renal sarcomas[1,2] followed by pleomorphic sarcoma, hemangiopercytoma, fibrosarcoma and unclassified sarcoma. Renal leiomyosarcomas have no specific symptoms or radiological findings to differentiate it from other renal malignancies[3], hence Fine-needle aspiration cytology (FNAC) and histopathological examination are vital tools in diagnosing PRL. Here we present a case of PRL managed at our hospital for further understanding of the disease.

CASE REPORT

A 51-year-old female presented with abdominal pain and swelling in left flank. On systemic examination abdomen was soft, non-tender with a palpable mass of size 8*6 cm in left flank. CECT revealed a lobulated exophytic enhancing soft tissue mass lesion in left lumbar region abutting the lower pole of left kidney, left renal vessels and left pelviureteric junction with a provisional diagnosis of renal cell carcinoma. (Fig.1)

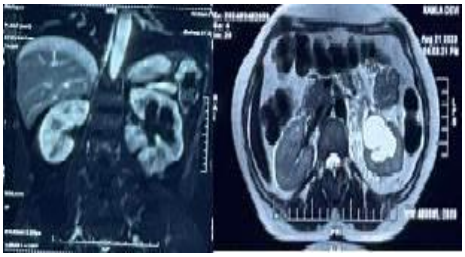

Fig 1. CECT showing a soft tissue mass lesion in left lumbar region abutting the lower pole of left kidney

An ultrasound guided FNAC was performed. Smears examined showed spindle shaped cells with blunt ends present in clusters, bundles and fascicles with a myxofibrillary stroma. (Fig.2,3) Anisonucleosis was moderate to focal marked. In addition, one field showed plump, round to oval cells and focally storiform pattern (Fig.4,5). A careful search showed an occasional cluster of benign cuboidal epithelial cells possibly of renal tubular origin? (Fig 6)

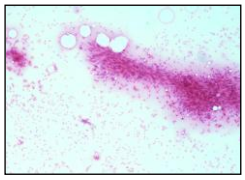

Fig.2.10x H&E. Fascicles and bundles of spindle cells.

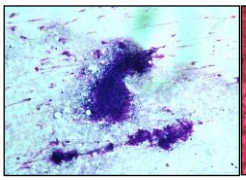

Fig 3. 10x, Giemsa. Smear showing myofibrillary stroma.

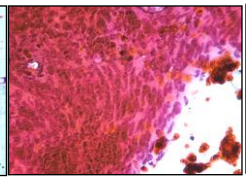

Fig 4. 40x H&E, round to oval plump cells.

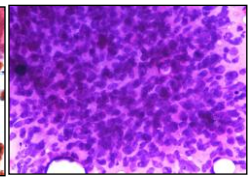

Fig 5. 40x, Giemsa Spindle cells showing pleomorphic nuclei.

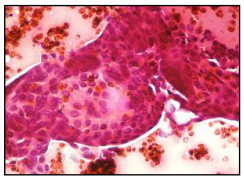

Fig 6. 40x H&E. Benign epithelial cells.? renal tubular origin

A diagnosis of a retroperitoneal sarcomatoid lesion was made with possible differentials that were considered were sarcomatoid renal cell carcinoma, gastrointestinal stromal tumor, hemangiopericytoma, pleomorphic sarcoma, leiomyosarcoma and synovial sarcoma. Subsequently a radical nephrectomy specimen was received. Grossly, a grey white mass was seen at the lower pole of kidney pushing into the renal pelvis. (Fig.7) Multiple section examined showed tumor infiltrating the perinephric fat and peripelvic fat. (Fig.8) The tumor was well demarcated from the normal renal parenchyma and separated by a thin fibrous capsule. (Fig.9) The tumor was composed of atypical spindle cells present in bundles and fascicles. (Fig 10.)

Fig.7. A gross image showing tumor in the lower pole of kidney and abutting the renal pelvis

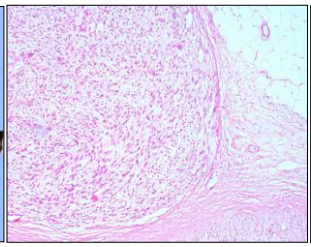

Fig 8. 10X, H&E. Tumor reaching up to the perinephric fat. stroma.

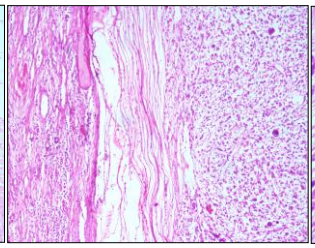

Fig 9.10x H&E. Tumor with renal capsule and normal renal tissue

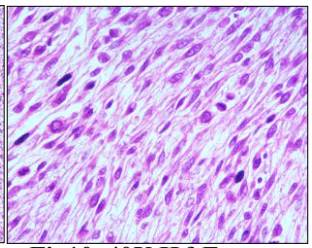

Fig10. 40X H&E Spindle cells arranged in fascicles and bundles. Mitotic activity present

Immunohistochemistry in our case showed two positive myogenic marker, h-caldesmon & SMA and was also CD99 positive. (Fig.11,12) PanCK, CK7 were negative thus ruling out SRCC. (fig.13) CD68, TLE-1 was negative ruling out possibility of SRCC, SS, pleomorphic sarcoma. (fig 14) Thus a final diagnosis of leiomyosarcoma of kidney was made.

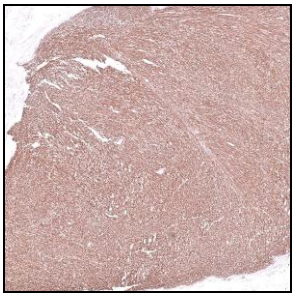

Fig 11. H-caldesmon, diffuse and strong positive.

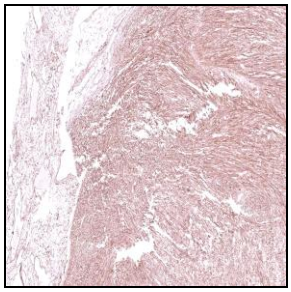

Fig 12. CD99, positive.

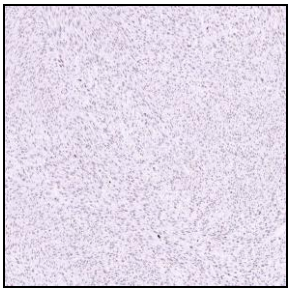

Fig 13. PANCK-Negative

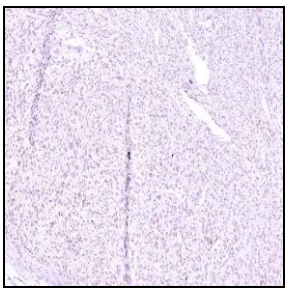

Fig 14. TLE1 Negative

DISCUSSION

Primary renal leiomyosarcomas are rare tumors. The incidence of the primary renal leiomyosarcoma increases with the age of the patient. This tumor is found to be more common in females than in males[4]. It usually has an insidious clinical presentation with late-stage symptoms of abdominal pain, vomiting, weight loss and a palpable mass[5]. As the tumor increases in size, it produces symptoms such as lumbar pain and haematuria. Radiological investigations are not of much help in differentiating leiomyosarcomas from other renal cell carcinomas[3]. FNAC of renal sarcomas reveal mainly spindle cells giving varied possible differentials of leiomyosarcoma, SRCC, SS, hemangiopericytoma, fibrosarcoma, Pleomorphic histiocytic lesion and gastrointestinal stromal tumor[5-10]. This case showed fascicles and bundles of round to oval to spindle cells. Absence of malignant epithelial component excluded the diagnosis of SRCC and SS. Similarly, the absence of endothelial cells and arborizing capillaries with tumour aggregates excluded hemangiopericytoma. Pleomorphic fibrohistiocytic lesion is characterised by marked pleomorphism which was not seen our case. Another possible differential of fibrosarcoma shows spindly cells with uniform, elongated nuclei with tapering ends, small inconspicuous nucleoli and scanty, wispy cytoplasm associated with myxoid material unlike leiomyosarcoma. (Table.1) Histological examination of leiomyosarcoma, show a smooth muscle tumor with a fasciculated architecture, made up of spindle-shaped cells arranged in fascicles and bundles. The cells have sharp-ended, non-tapered nuclei with eosinophilic cytoplasm. Indicators of malignancy such as necrosis, nuclear pleomorphism with mitotic figures are present as was seen in our case also. (Fig.8-10). Histologically, it is extremely difficult to differentiate a renal leiomyosarcoma from a renal sarcomatoid carcinoma. Both tumors have similar clinical, radiographic and pathological features[11]. Only the absence of an epithelial component on morphological examination and the absence of cytokeratin expression on immunohistochemical examination can rule out a sarcomatoid carcinoma of the kidney. Primary monophasic synovial sarcoma of the kidney also shows monophasic spindle cells. The spindle cells are plump with irregular cell borders. They tend to grow in sheets and usually have trapped renal tubules in the form of cysts. However, these tumours show positivity for Bcl-2[7]. (Table 1). PML show positive immunostaining for smooth muscle actin, h-caldesmone, desmin and vimentin with negative staining for epithelial markers, s-100, cd117, TLE1[11].

	Sarcomatoid RCC	Synovial sarcoma	Fibrosarcoma	GIST	Renal Leiomyosarcoma(our case)
Age	54-63yrs M:F -2:1	Median age-35 yrs M:F-1.2:1	Median age-45yrs M:F-1.3:1	60-65yrs M:F-1:1	40-75yrs, female preponderance
Incidence	8% of RCC's	1% of malignant renal tumors	1% of adult sarcomas	<1% of extraintestinal sarcomas	40% of renal sarcomas
Radiological findings	Mass lesions within kidney	Round to oval lobulated mass	Hyperechoic mass lesion	Indefinite findings	Mass lesion abutting kidney
Cytology	Spindle cells and a malignant epithelial component	biphasic pattern of spindle-shaped and epithelioid cells. Small clusters of monotonous cells with oval to spindle-shaped cytoplasm, branching tumor tissue fragments, and scant background mucin and cohesive epithelial cells.	Cellular, showing spindly cells with uniform, elongated nuclei; small, inconspicuous nucleoli; and scanty, wispy cytoplasm associated with myxoid material. No significant nuclear pleomorphism or mitoses.	bland spindle to epithelioid cells arranged in fascicles.	Fascicles and bundles of round to oval cells to spindle cells
HPE	Foci of typical RCC, Storiform pattern of spindle cells	hyperchromatic and cellular spindle cell proliferation with staghorn vessels.	Highly cellular fibroblastic proliferation in herringbone pattern	Bland spindle cells with eosinophillic cytoplasm, elongated nuclei, inconspicuous nucleoli, artifactual vacuoles	Spindle cells in fascicles and bundles
IHC	CK- positive SMA- pos/neg Desmin, h-cal- neg	TLE1-80-90% + CK-Variable(depending on the epithelial component) EMA: 29 - 90%, variable BCL2-79-100% B-Catenin- 30 - 73% cd99- (91%), CD56(100%), calretinin (56 - 71%) Neg stains- CD34, desmin h-caldesmon, myogenin, MyoD1, FLI1, WT1	Reticulin + fibers surrounding each cell Phosphotungstic acid-hematoxylin + vimentin, type 1 collagen, p53 + High Ki67 Neg-s-100, keratin	c-kit , Dog1 positive	H-caldesmon, SMA, desmin positive Pan-CK- Negative

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Table 1. Differentiating features of primary sarcomas of kidney.

Renal leiomyosarcomas is thought to have possible origin from renal capsule, renal pelvis or renal vessels[12]. This case showed tumor extending into the renal pelvis and capsule was intact, indicating possible origin from renal pelvis. No vascular involvement was seen in this case.

CONCLUSION

Primary renal leiomyosarcoma is a rare but highly aggressive malignancy. FNAC provides preliminary cytological diagnosis, while histopathological examination, supported by immunohistochemistry, offers definitive diagnosis.

Abbreviations

FNAC- Fine needle aspiration cytology
PRL- Primary renal leiomyosarcoma
SRCC- Sarcomatoid renal cell carcinoma
SS- Synovial sarcoma

Declaration by Authors

Ethical Approval: Not required
Acknowledgement: None
Source of Funding: None
Conflict of Interest: No conflicts of interest declared.

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How to cite this article: Neelam Sood, Nidhi Varshney, Anamika Rawat. Cytohisto-morphological features of primary renal leiomyosarcoma: a rare case report. *International Journal of Research and Review*. 2025; 12(6): 301-305. DOI: *https://doi.org/10.52403/ijrr.20250635*

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