

Report of primary leiomyosarcoma of renal pelvis and literature review

Abhidha Malik¹, Ritesh Kumar¹, Abhishek Shankar¹, Sunil Chumber², Sameer Bakhshi³, Seema Kaushal⁴, Bala Thirunavukkarasu⁴

¹Department of Radiation Oncology, All India Institute of Medical Sciences, New Delhi 110029, India.

²Department of Surgery, All India Institute of Medical Sciences, New Delhi 110029, India.

³Department of Medical Oncology, All India Institute of Medical Sciences, New Delhi 110029, India.

⁴Department of Pathology, All India Institute of Medical Sciences, New Delhi 110029, India.

Correspondence to: Dr. Abhidha Malik, Department of Radiation Oncology, All India Institute of Medical Sciences, New Delhi 110029, India.
E-mail: dr.abhidha@gmail.com

How to cite this article: Malik A, Kumar R, Shankar A, Chumber S, Bakhshi S, Kaushal S, Thirunavukkarasu B. Report of primary leiomyosarcoma of renal pelvis and literature review. *J Cancer Metastasis Treat* 2017;3:111-5.

ABSTRACT

Article history:

Received: 05-02-2017

Accepted: 24-03-2017

Published: 30-06-2017

Key words:

Primary,
leiomyosarcoma,
renal pelvis

Primary sarcomas of kidney are exceptionally rare tumors, accounting for only 1-2% of all malignant tumors of kidney. Leiomyosarcoma (LMS) is the most common histological subtype among all renal sarcomas. The authors describe here a case of primary leiomyosarcoma of renal pelvis in a 50-year-old lady, presenting with flank pain. Based on triple phase cardio-electroencephalographic covariance tracing abdomen, presumptive diagnosis of renal cell carcinoma/renal sarcoma/neurogenic tumor was made and patient underwent radical nephrectomy. Microscopy reflected spindle cell tumor which showed strong positivity for desmin and smooth muscle actin with negative epithelial markers, thereby confirming the diagnosis of renal LMS. Owing to aggressive nature and low survival rates of LMS patient received adjuvant treatment in form of chemotherapy and radiotherapy. Patient is doing well 1 year post treatment.

INTRODUCTION

Primary sarcomas of kidney constitute 1-2% of all malignant tumors of kidney.^[1] LMS is most common histological subtype, accounting 60-70% of all sarcomas of kidney, commonly seen in females in 4th-6th decades.^[2] Radical nephrectomy remains the treatment of choice.^[3] The role of adjuvant treatment remains debatable due to paucity of data on treatment of this rare neoplasm. Herein, we report

a case of a 50-year-old lady with leiomyosarcoma originating in renal pelvis along with relevant review of literature.

CASE REPORT

A 50-year-old postmenopausal lady, known case of hypothyroidism on T Thyroxin for 3 years, presented with diffuse dull aching paroxysmal pain in left flank of 2 months duration. On physical examination PORT



This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: service@oaepublish.com

Quick Response Code:



scar, due to laproscopic cholecystectomy done 2 years back was visualized. No mass palpable. Triple phase contrast-enhanced computed tomography (CECT) scan of whole abdomen revealed [Figure 1], well defined retroperitoneal mass in left para-aortic location with anterior displacement of left renal vein. Neurogenic origin/mesenchymal tumor. Patient planned for wide local excision of the mass, intra-operatively the mass was adherent to left renal vein and feeder vein, therefore decision taken to perform left nephrectomy along with adrenalectomy. Postoperatively, gross examination of the specimen reveals greyish white tumor measuring 8 cm × 4 cm × 2 cm, infiltration of the renal pelvis but not infiltrating into renal parenchyma. Renal sinuses, resected end of ureter and adrenal gland all were free of tumor. Microscopically tumor comprises of oval to spindle cells with moderate amount of cytoplasm and eosinophilic pleomorphic nuclei, tumor cells are arranged in interlacing fascicles and at places show cytoplasmic clearing, mitotic activity of 7-10/hpf, less than 50% necrosis, FNCLC grade 2. Immunopositive for desmin and smooth muscle actin (SMA), whereas immunonegative for Pan CK, CD-34, EMA, Bcl2, S-100, MIC-2. Overall features were suggestive of leiomyosarcoma [Figure 2]. Post operative CECT of chest, abdomen and pelvis were within normal limits. Patient received 6 cycles of adjuvant chemotherapy with single agent injection adriamycin 25 mg/m² day 1-3 followed by post operative radiotherapy to tumor

bed to a dose of 50 Gy/25#/5 weeks. Patient is doing well 1 year post treatment.

DISCUSSION

LMS are malignant neoplasm of smooth muscle origin. They are most commonly found in uterus, stomach, small intestine and retroperitoneum.^[4] LMS of renal origin are very rare and constitute only 0.12% of all malignant renal neoplasms.^[5] LMS of kidney was first described by Berry in 1919 but till date they have been reported only as case reports or as component

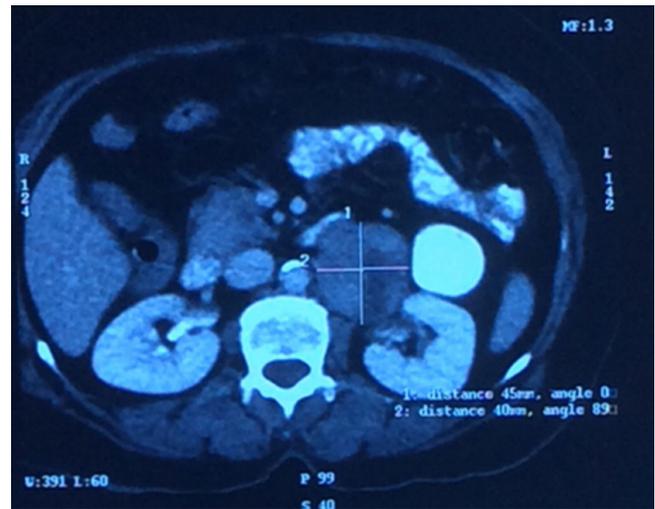


Figure 1: Contrast-enhanced computed tomography abdomen showing well defined retroperitoneal mass in left para-aortic location

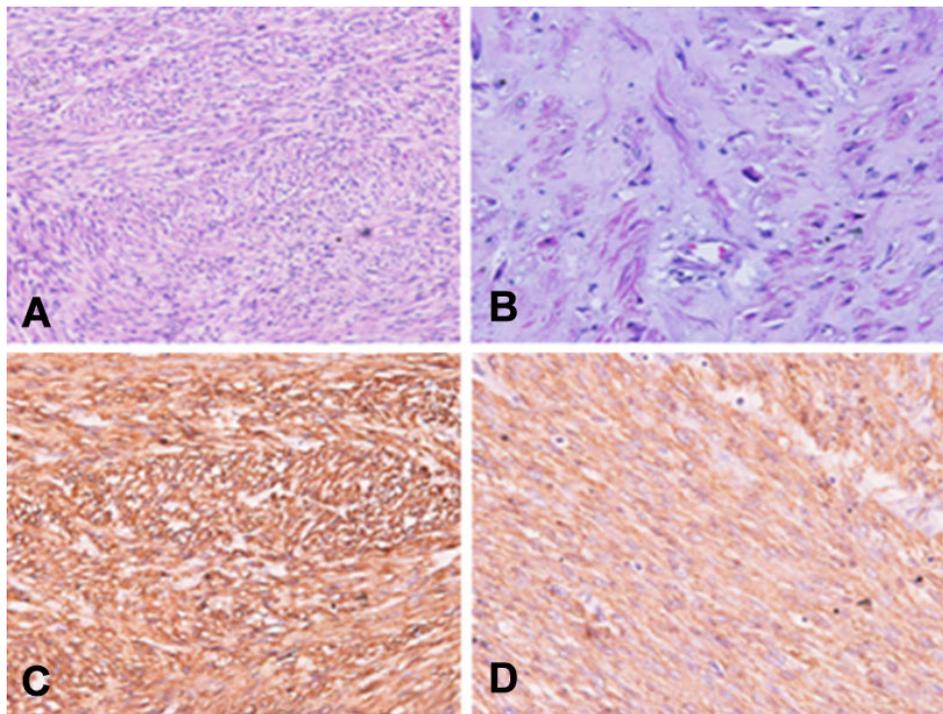


Figure 2: (A and B) Images comprising of oval to spindle tumor cells arranged in interlacing fascicles (A: 4x, B: 20x, HE); (C and D) images of desmin and smooth muscle actin immunopositivity, respectively (20x, HE)

Table 1: Summarizes clinic-radiological features, treatment and follow-up of previously reported cases of renal leiomyosarcoma in English literature in past 12 years

No.	Ref.	Year	Age (year)	Gender	Clinical symptoms	Side	CT diag	Size (cm ²)	Sx	Adj Rx	FU	LF	DF	Site of DF
1	[17]	2004	54	Male	Abd discomfort	Left	RCC	8 × 7.5	N	No	6 months	No	No	
2	[18]	2005	52	Female	Incidental	Left	-	2	PN	No	2 years	No	No	
3	[19]	2006	48	Female	Flank mass	Right	RCC	12.5 × 10.5	RN	No	Unknown	-	-	
4	[20]	2006	44	Male	Hematuria	Left	RCC	5 × 4	RN	No	3 years	No	No	
5	[21]	2007	42	Female	Incidental	Left	RCC	5 × 3	PN	No	Unknown	-	-	
6	[1]	2007	55	Female	Abd pain	Right	RCC	4 × 2	NSS	No	15 months	No	No	
7	[8]	2007	60	Male	Urinary frequency	Right	RCC	10 × 8	N	No	6 months	No	No	
8	[15]	2007	55	Male	Flank pain	Left	RCC	3 × 2.5	RN	Yes	6.5 years	No	No	
9	[6]	2009	42	Female	Flank pain	Right	RCC	15	RN	No	7 months	No	No	
10	[22]	2009	71	Male	Flank mass	Left	RCC	13 × 6.5	RN	Yes	7 months	No	No	
11	[23]	2009	65	Female	Flank mass	Right	RCC	15 × 11	RN	No	1 year	No	No	
12	[24]	2010	55	Female	Flank pain	Left	RCC	20 × 16	RN	No	Unknown	-	--	
13	[25]	2011	57	Female	Flank mass	Left	-	25 × 23	RN	No	3 years	No	No	
14	[26]	2011	65	Female	Flank pain	Right	-	13 × 11	RN	No	15 months	No	No	
15	[27]	2012	74	Female	Abd pain	Left	RCC	8 × 6	RN	No	1 month	No	No	
16	[28]	2013	70	Male	Flank pain	Right	RCC	4	LN	No	2 months	No	Yes	Bone
17	[29]	2013	69	Female	Flank mass	Right	RCC	18 × 15	RN	No	5 years	No	No	
18	[30]	2013	20	Male	Hematuria	Left	RCC	7 × 6	LN	No	2 months	No	Yes	Lung
19	[31]	2014	65	Female	Flank mass	Right	RCC	18 × 8	N	No	2 months	No	No	
20	[14]	2015	50	Female	Flank pain	Left	-	10 × 6	RN	No	1 year	No	No	
21	[32]	2015	39	Male	Flank mass	Left	RCC	16 × 14	RN	No	1 year	No	No	

Diag: diagnosis; Adj: adjuvant; Sx: surgery; Rx: treatment; FU: follow up; LF: local failure; DF: distant failure; Abd: abdomen; CT: computed tomography; RCC: renal cell carcinoma; N: nephrectomy; PN: partial nephrectomy; RN: radical nephrectomy; LN: laparoscopic nephrectomy; NSS: nephron sparing surgery

of larger series of renal sarcoma literature [Table 1].^[6] Histogenesis of renal LMS is believed to be from renal capsule or smooth muscle fibers in renal pelvis or from the renal vessels.^[7]

The renal LMS has preponderance in women, with women being twice more commonly affected than men and majority of patients presenting in 4th-6th decades of life.^[8] The cause of female preponderance is not fully known, but studies suggest that some malignancies are associated with genes located on X chromosome that escape X inactivation.^[9] Renal LMS is found to occur equally on both sides and also bilaterally, however the etiology remains obscure.^[10]

The patients of LMS usually presents with flank pain, hematuria and abdominal mass, thus mimicking renal cell carcinoma (RCC). It's difficult to differentiate between LMS and RCC. Ultrasound, tomography or magnetic resonance neither is able to differentiate between the two and diagnosis is usually made postoperatively.^[11] Because renal sarcoma is a rare condition; no effective treatment has yet been established. It is believed that surgery offers the best chance of cure therefore radical nephrectomy is treatment of choice, but recently partial nephrectomy has been shown to give good results in these patients.^[12]

Histologically, leiomyosarcoma has to be differentiated from sarcomatoid RCC, leiomyoma and angiomyolipoma. Leiomyosarcoma can be differentiated from leiomyoma by presence of mitosis and necrosis in malignant tumor although cellular pleomorphism can be seen in both. Renal angiomyolipoma shows fascicles of smooth muscle cells admixed with mature fat and thick walled blood vessels. Sarcomatoid RCC forms the closest differential diagnosis of renal LMS.^[13]

Diagnosis of sarcomatoid RCC can be made if typical RCC is seen somewhere in the tumor, so a thorough sampling of tumor is required to rule out any epithelial component of sarcomatoid RCC. Immunohistochemically tumor cells of leiomyosarcoma are positive for desmin, SMA calponin, H caldesmon and negative for CK, S-100 and HMB-45. The angiomyolipoma will show HMB-45 positivity while sarcomatoid variant of RCC will be CK positive.^[14]

To make diagnosis of primary renal sarcoma the following criteria should be met: (1) the patient must not have or have had sarcoma elsewhere to rule out metastasis; (2) gross must be compatible with origin in kidney rather than involvement due to retroperitoneal sarcoma; (3) sarcomatoid variant of RCC must be excluded.^[8]

Small size < 5 cm, low histological grade and renal limited disease are associated with more favorable outcomes. Histological grade of tumor is assigned based on mitotic count, necrosis and nuclear pleomorphism.^[8] The most important prognostic factor is tumor free resected margin. Large size and metastasis to adjacent organs at the time of diagnosis makes the prognosis poor.

Adjuvant treatment in form of chemotherapy and radiotherapy has been tried differently by different people. Sharma et al.^[15] prescribed chemotherapy with mesna, adriamycin, ifosphamide and dacarbazine regimen and sandwich radiotherapy with a dose of 44 Gy/22#/4.5 weeks to the renal bed and adjoining lymphatics. Beccia et al.^[16] prescribed vincristine (1.4 mg/m²) or cyclophosphamide (700 mg/m²) plus actinomycin D (0.04 mg/m²) to patients following surgery. As no randomized control trials have demonstrated their long term effects, treatment can be tailored individually. In our patient, we administered single agent doxorubicin (25 mg/m² day 1-3). Before administering treatment, we thoroughly explained our patient about potential benefits of adjuvant treatment, despite negative margin status and absence of metastasis. Our recommendations were based on high risk features like abdominal/retroperitoneal location, size greater than 5 cm, high grade histology and presence of necrosis.

To conclude, renal leiomyosarcoma is a rare tumor should be differentiated from sarcomatoid variant of renal cell carcinoma and angiomyolipoma. Radical nephrectomy is treatment of choice but in spite of successful resection it usually shows an unfavorable prognosis. Aggressive treatment with adjuvant chemotherapy and radiotherapy can offer better results. Paucity of cases and absence of long-term follow up with controlled randomized studies is hampering definitive treatment protocols.

Authors' contributions

Conceptualisation of this work: R. Kumar, A. Malik

Providing all necessary information:

B. Thirunavukkarasu, A. Shankar

Jointly supervising the work: S. Chumber, S. Bakhshi, S. Kaushal

Financial support and sponsorship

None.

Conflicts of interest

There are no conflicts of interest.

Patient consent

Patient consent was obtained as per institutional policy.

Ethics approval

The ethics approval was obtained from the institutional ethical committee for preparation and publication of this paper.

REFERENCES

- Demir A, Yazici CM, Eren F, Türkeri L. Case report: good prognosis in leiomyosarcoma of the kidney. *Int Urol Nephrol* 2007;39:7-10.
- Srinivas V, Sogani PC, Hajdu SI, Whitmore WF Jr. Sarcomas of the kidney. *J Urol* 1984;132:13-6.
- Kavantas N, Pavlopoulos PM, Karaitianos I, Agapitos E. Renal leiomyosarcoma: report of three cases and review of the literature. *Arch Ital Urol Androl* 1999;71:307-11.
- Weiss SW. Smooth muscle tumors of soft tissue. *Adv Anat Pathol* 2002;9:351-9.
- Kendal WS. The comparative survival of renal leiomyosarcomas. *Can J Urol* 2007;14:3435-42.
- Aiken W, Gibson T, Williams S, Gaskin D. Leiomyosarcoma of the kidney. *West Indian Med J* 2009;58:183-4.
- Niceta P, Lavengood RW, Fernandes M, Tozzo PJ. Leiomyosarcoma of kidney. Review of the literature. *Urology* 1974;3:270-7.
- Dhamne SA, Gadgil NM, Padmanabhan A. Leiomyosarcoma of the renal pelvis. *Indian J Pathol Microbiol* 2009;52:549-51.
- Brown CJ, Greally JM. A stain upon the silence: genes escaping X inactivation. *Trends Genet* 2003;19:432-8.
- Polianko NI. Leiomyosarcoma of both kidneys. *Arkh Patol* 1986;48:70-2. (in Russian)
- Kurugoglu S, Ogut G, Mihmanli I, Korman U, Durak H. Abdominal leiomyosarcomas: radiologic appearances at various locations. *Eur Radiol* 2002;12:2933-42.
- Lacquaniti S, Destito A, Candidi MO, Petrone D, Weir JM, Servello C, Pisanti F, Alcini E. Two atypical cases of renal leiomyosarcoma: clinical picture, diagnosis and therapy. *Arch Ital Urol Androl* 1998;70:199-201.
- Grignon DJ, Ayala AG, Ro JY, el-Naggari A, Papadopoulos NJ. Primary sarcomas of the kidney. A clinicopathologic and DNA flow cytometric study of 17 cases. *Cancer* 1990;65:1611-8.
- Srivastava P, Prasad R, Khanna G. Primary leiomyosarcoma of kidney: a rare case report. *Muller J Med Sci Res* 2015;6:157-9.
- Sharma D, Pradhan ES, Aryya NC, Shukla VK. Leiomyosarcoma of kidney: a case report with long term result after radiotherapy and chemotherapy. *Int Urol Nephrol* 2007;39:397-400.
- Beccia DJ, Elkort RJ, Krane RJ. Adjuvant chemotherapy in renal leiomyosarcoma. *Urology* 1979;13:652-4.
- Minami H, Ueki O, Tanaka T, Nishida H, Hashimoto T, Kawaguchi K. Case of leiomyosarcoma of the renal pelvis. *Int J Urol* 2004;11:122-4.
- Cocuzza M, Arap S, Lucon AM, Saldanha LB. Renal Leiomyosarcoma treated with partial nephrectomy. *Clinics (Sao Paulo)* 2005;60:345-6.
- Adhikari RC, Sayami G, Dali S, Shrestha HG. A case of leiomyosarcoma of the kidney. *J Inst Med* 2006;28:74-6.
- Kartsanis G, Douros K, Zolota V, Perimenis P. Case report: leiomyosarcoma of the renal pelvis. *Int Urol Nephrol* 2006;38:211-3.
- Chung YG, Kang SC, Yoon SM, Han JY, Seong DH. Leiomyosarcoma arising from the blind end of a bifid renal pelvis. *Yonsei Med J* 2007;48:557-60.
- Dubey A, Koul R. A rare case of renal sarcoma with the review of literature. *Int J Nephrol* 2009;6:1-5.
- Choudhury M, Singh SK, Pujani M, Pathania OP. A case of Leiomyosarcoma of kidney clinically and radiologically misdiagnosed as renal cell carcinoma. *Indian J Cancer* 2009;46:241-3.
- Venkatesh K, Lamba SM, Niveditha SR, Krishnagiri C, Babu

- S. Primary leiomyosarcoma of the kidney. *Patholog Res Int* 2010;2010:652398.
25. Azizun-Nisa, Hasan SH, Raza Y. Primary renal leiomyosarcoma. *J Coll Physicians Surg Pak* 2011;21:713-4.
26. Ellouze S, Abid N, Kossentini M, Gouiaa N, Charfi S, Mhiri N, Boudawara T. Leiomyosarcoma of the kidney. *Clin Genitourin Cancer* 2011;9:68-9.
27. Pong YH, Tsai VFS, Wang SM. Primary leiomyosarcoma of the kidney. *Formasan J Surg* 2012;45:124-6.
28. Valery JR, Tan W, Cortese C. Renal leiomyosarcoma: a diagnostic challenge. *Case Rep Oncol Med* 2013;2013:459282.
29. Beardo P, José Ledo M, Jose Luis RC. Renal leiomyosarcoma. *Rare Tumors* 2013;5:e42.
30. Cho EY, Yoon JH, Kim W. Leiomyosarcoma of the renal pelvis: report of a case and review of the literature. *OMICS J Radiol* 2013;3:154.
31. Babu S, Singhai A, Hussain N, Singh V. Renal leiomyosarcoma -- a rare entity. *J Case Rep* 2014;4:29-32.
32. Bavikar R, Deshmukh S. Primary leiomyosarcoma of kidney in a young male treated with partial nephrectomy -- a case report. *Sarcoma Res Int* 2015;2:1013.