CASE REPORT

Primary Low-Grade Fibromyxoid Sarcoma of Kidney—an Extremely Rare Entity

Rohit Bhattar¹ · S. P. Aggarwal¹ · S. S. Yadav¹ · V. Tomar¹

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Abstract Renal LGFMS is an extremely rare tumor. So far, only four cases have been reported in literature. LGFMS are indolent tumor with distinctive histopathological features with potential for late recurrences and metastasis. The diagnosis is made on histopathological examination and supported by immunohistochemical analysis. A 35-year-old young male was presented with flank pain and lump and diagnosed as low-grade fibromyxoid sarcoma after surgery on immunohistochemical analysis. This is the fifth case of renal LGFMS and we presented the review of literature. Radical surgery remains the primary treatment modality. Long follow-up is mandatory.

Keywords Kidney · Low grade · Recurrence · Sarcoma

Case Report

A 35-year-old male presented with left flank pain and lump for 3 months with no other associated symptoms. Patient gave past history of left sided open pyeloplasty in 2010. On examination, a mobile 9 × 8 cm, non-tender lump was present in the left lumbar area. The rest of the examination was normal.

Laboratory studies were unremarkable but ultrasound showed the presence of a solid, heterogeneous mass in lower pole of left hydronephrotic kidney. On contrast-enhanced computed tomography (CECT) of abdomen showed a 7 × 6 cm

non-enhancing, hyper dense lesion with calcifications in lower pole of the hydronephrotic left kidney. Subsequent magnetic resonance imaging (MRI) showed heterogeneous signal intensity mass lesion in lower calyces of the left kidney with gross hydronephrosis (Fig. 1a). No vascular invasion was identified. The contra-lateral kidney was normal. High-resolution computed tomography (HRCT) chest showed no evidence of any metastatic lesion. So, a working diagnosis of poorly functioning kidney with a renal mass was made. Urine cytology and cystoscopy of bladder were done (keeping urothelial tumor as a differential diagnosis), which were found to be normal.

Radical nephrectomy was planned. At surgery, the kidney was grossly hydronephrotic up to the pelvic brim. There was an endophytic tumor present in the lower pole. The specimen was removed en-bloc without violation of Gerota's fascia. On cut section, there was a 7 × 6 × 5 cm bosselated, pedunculated grayish brown solid mass.

On histopathological examination (HPE), grossly, a polypoidal lesion covered with metaplastic squamous epithelium was seen. The surface of polyp was ulcerated at some areas and was replaced by inflammatory granulation tissue. On microscopic examination, the tumor composed of spindle cells arranged in bundles, with abundant collagen and myxoid stroma with interspersed prominent vessels. There was mild to moderate anaplasia with occasional mitotic figures (Fig. 1b). Immunohistochemistry (IHC) was done with Flex Envision Detection System. The tumor cells were immunonegative for cytokeratin, EMA, SMA, desmin, CD 34, S-100, HMB 45, and MUC 4, ruling out rhabdosarcomas and melanomas. So, a final diagnosis of low-grade fibromyxoid sarcoma was made. Final staging was T2 deep N0 M0. Patient was counseled about the pathology and need for regular follow-up. Post-op recovery was good and at 18-month follow-up, no recurrence was seen and patient is doing well.

✉ Rohit Bhattar
bhattarrohit@gmail.com

¹ Department of Urology and Renal Transplantation, SMS Medical College, Jaipur, Rajasthan, India

Fig. 1 **a** T2 weighted SPAIR sequence MRI showing a heterogeneously hyperintense lower pole mass in left hydronephrotic kidney. No contrast enhancement was seen. **b** H&E stain high power view showing spindle cells with occasional mitotic figures and mild to moderate anaplasia

Discussion

Soft tissue sarcomas (STS) are a heterogeneous group that account for 1% of all malignant tumors, with the genitourinary tract accounting for 2.1% of STS [1]. There are about 133 adult cases and 33 pediatric cases of LGFMS reported in literature, so far. Renal LGFMS are extremely rare tumors, with only four cases (three in adults and one in pediatric age group) reported so far [2–6]. LGFMS are rare indolent tumors with deceptively benign histological appearance with a potential of late local recurrences and metastasis [7]. Radical surgery is the primary treatment of choice but there is a role of radiotherapy and chemotherapy in the cases of recurrences or metastases [8]. The estimated 5-year overall survival is over 90%, but very late local relapses and distant metastasis are reported, which underlines the need for a long-term follow-up. Negative surgical margins and tumor size are the most important prognostic factors for local recurrence and metastasis [8]. On microscopy, there are bland fibroblasts in a whorled or

linear arrangement, admixed with myxoid stroma. Tumor cells are small and have pale eosinophilic cytoplasm, round to ovoid nuclei and sparse mitotic figures with absent nuclear anaplasia and necrosis [3–6, 8]. The current role of IHC is to rule out other soft tissue sarcomas and melanomas.

The clinical presentation, pathology, and treatment outcomes of the four reported cases are discussed in Table 1.

Like all other previous reported cases, the tumor in our case was present in a male patient, whereas LGFMS in other sites have an equal gender distribution. We did not perform FNAC preoperatively and straight forward went for radical nephrectomy. The histology was suggestive of LGFMS and we performed IHC panel keeping other sarcomas also in differential. In our case, the immunohistochemical results were negative for all markers, i.e., cytokeratin, EMA, SMA, desmin, CD 34, S-100, HMB 45, and MUC 4, ruling out rhabdosarcomas and melanomas similar to Rubinstein et al., whereas Alevizopoulos et al. had vimentin, CD34, and c-kit (focally weak positivity) positive tumors and Ki-67 = 1%. Rubinstein

Table 1 Clinical and treatment outcome of previously reported cases

Authors	Sex	Age (years)	Side	Presentation	Investigation	Tumor size (cm)	Treatment	Outcome and follow-up
Silverman et al. (2000)	M	70	Bilateral	Back pain	FNAC	—	Right partial nephrectomy	No recurrence
Gonzalez et al. (2009)	M	28	Left	Abdominal pain and lump	FNAC	25 × 20 × 14	Radical nephrectomy	Recurrence and Died after 8 months
Alevizopoulos et al. (2012)	M	48	Left	Renal colic	Ureteroscopy and biopsy	4.3 × 4 × 4.6	Radical nephroureterectomy	Recurrence free at 7 months
Rubinstein et al. (2014)	M	6	Right	Abdominal lump	IHC, Fish	16.4 × 13.5 × 12.7	Radical nephrectomy	Recurrence free at 18 months
Current case	M	35	Left	Abdominal pain and lump	IHC	7 × 6 × 5	Radical nephrectomy	Recurrence free at 18 months

et al. had reported FISH positivity for ESWR1–CREB3L1 gene fusion in their case, which we did not perform as it was not available in our institute. We had a good surgical outcome with no recurrence seen at 18 months of follow-up.

Conclusion

Primary renal low-grade fibromyxoid sarcoma is a rare tumor with distinct histopathological features for diagnosis that are supplemented by immunohistochemistry. Radical surgery should be done to achieve negative surgical margins. Lifelong follow-up is required due to chances of late recurrence and metastasis.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Abbreviations *CECT*, contrast-enhanced computed tomography; *FNAC*, fine needle aspiration cytology; *H&E*, hematoxylin and eosin stain; *HPE*, histopathological examination; *IHC*, immunohistochemistry; *LGFMS*, low-grade

fibromyxoid sarcomas; *MRI*, magnetic resonance imaging, *STS*, soft tissue sarcomas

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