Case Report

Organ preservation in leiomyosarcoma bladder: Case report and review of literature

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Abstract Leiomyosarcomas (LMSs) account for <0.1% of all bladder malignancies. Due to the infrequent occurrence of these tumors, established guidelines for management are lacking. Conventionally, radical extirpative surgery has been advocated. We present our experience with organ preservation in a young male presenting with LMS bladder. A brief review of literature supporting organ preservation in selected cases has also been presented.

Keywords: Carcinoma, leiomyosarcoma, partial cystectomy, radical cystectomy

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Received: 14.07.2017, **Accepted:** 18.09.2017

INTRODUCTION

Bladder leiomyosarcoma (LMS) is a rare malignancy, frequently presenting as high-grade advanced disease. Radical cystectomy with wide margins has been advocated in the management of bladder LMS. Partial cystectomy has historically been deemed as palliative surgery with poorer results in comparison to radical surgery.[1,2] We report a case of LMS bladder managed with partial cystectomy and no adjuvant therapy who remains disease free on follow-up of 48 months.

CASE REPORT

A 28-year-old male presented to us with lower abdominal pain and irritative lower urinary tract symptoms. Physical examination was unremarkable. Routine blood and urine investigations were normal. A CT scan of the abdomen revealed a 6.7 cm × 6.9 cm × 7.2 cm heterogeneously enhancing mass arising from the dome and anterior wall of the bladder. The rest of the bladder appeared normal, with no gross pelvic lymphadenopathy [Figure 1]. Cystoscopic examination showed a large mass occupying virtually the entire lumen of the bladder; however, the area of attachment of the tumor to the bladder wall, at the dome, was relatively small [Figure 2]. A cold cup biopsy of the lesion revealed a low-grade spindle cell tumor. Random biopsies of the apparently uninvolved bladder mucosa were also taken and were reported to be normal.

We proceeded with partial cystectomy. The tumor and an adequate margin of attached bladder were excised [Figure 3]. On histopathologic examination, the tumor was composed of bundles of interlacing fascicles of spindle-shaped cells with increased nucleocytoplasmic ratio and moderate amounts of eosinophilic cytoplasm. Mitotic activity (1–2/10HPF) including occasional

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/UA.UA_109_17

How to cite this article: Menon AR, Puthalath RT, Suresh N, Hegde S. Organ preservation in leiomyosarcoma bladder: Case report and review of literature. Urol Ann 2018;10:233-6.

atypical mitotic figures was seen. The tumor extended into the perivesical adipose tissue in foci [Figure 4]. A panel of immunohistochemical markers were done and the tumor cells were positive for Desmin, Vimentin and Ki 67 (8%–10%) and negative for ALK-1, H-Caldesmon, SMA, S100-P, CD34, MYOD1 and Pan cytokeratin, suggesting a diagnosis of low-grade LMS bladder. In consultation with medical oncology, he was counselled regarding the possible benefits of adjuvant chemoradiation. He declined further therapy and opted for surveillance, which included cystoscopy and imaging every 6 months. At the time of this report, he is 48 months postpartial cystectomy with no evidence of recurrence or metastasis.

DISCUSSION

The vast majority of the tumors arising from the bladder are of urothelial origin. Nonurothelial tumors of the bladder comprise less than 5% of all bladder malignancies. These include mesenchymal tumors, among which LMS are the most common, accounting for 0.1% of all primary bladder tumors.[1]

The proposed risk factors for the development of LMS within the bladder include the presence of retinoblastoma gene mutation, cyclophosphamide exposure, and pelvic radiation therapy, however, the vast majority of cases, as in our patient, do not have any identifiable predisposing causes. Although the peak incidence has been reported to be in the seventh to ninth decade of life,[1-3] patients with the afore-mentioned risk factors present at a younger age, in the second to third decades.[4]

The presenting symptoms are attributable to the mechanical effect of the mass within the bladder causing storage or voiding symptoms based on the location of the mass within the bladder. Hematuria develops when the mass ulcerates and has been reported to be the most common symptom in most series.[2] The lesion is usually first detected on imaging studies and subsequently confirmed on cystoscopy and biopsy. Microscopically, infiltrative interlacing fascicles of spindle cells are noted. Vimentin, muscle-specific actin, and desmin are usually positive on immunohistochemistry. Epithelial markers and ALK-1 are usually negative. Bladder leiomyoma in contrast to LMS are noninfiltrative smooth muscle tumors, lacking mitotic activity, cytologic atypia, and necrosis.[5] Other spindle cell lesions including, sarcomatoid carcinoma, inflammatory myofibroblastic tumor, and postoperative spindle cell nodule need to be distinguished from LMS. This is usually accomplished by histology and immunochemistry.[6]

LMS has been subdivided into low grade and high grade based on mitotic count (≥5/10 hpf) and nuclear atypia. In a series of 15 cases, no disease recurrences were reported during a mean follow-up of 62 months when the tumor had one or fewer mitotic figures.[7] The

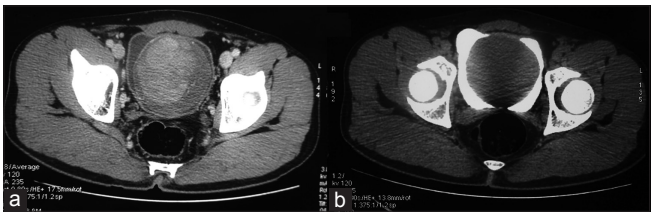

Figure 1: CECT Abdomen: a) Heterogeneously enhancing tumour in bladder. b) Delayed images showing site of attachment of the tumour to the dome to the bladder

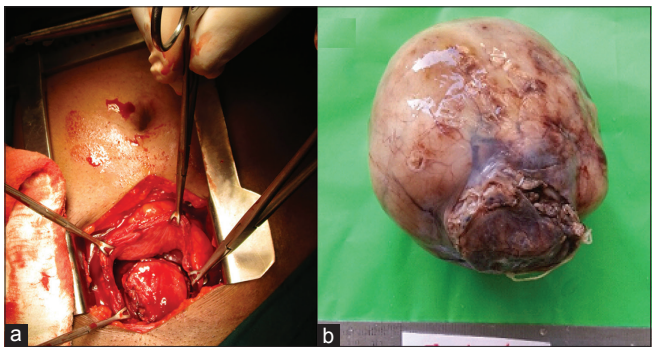

Figure 3: a) Intraoperative image: Tumour excised with adequate margin of normal bladder. b) Gross specimen of tumour

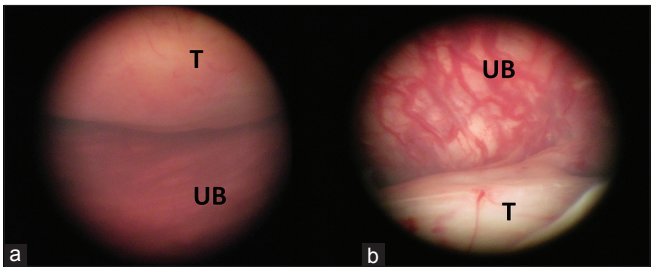

Figure 2: Cystoscopy Image: a) Tumour (T) seen inside the bladder (UB). b) Narrow area of attachment of Tumour (T) to bladder (UB)

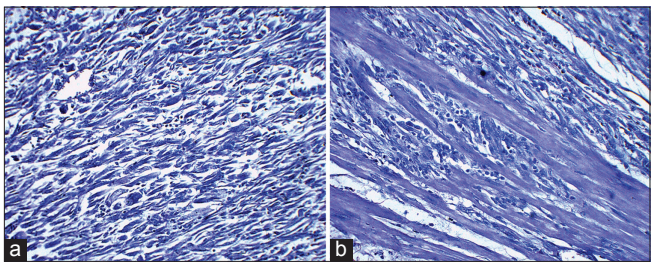

Figure 4: Photomicrograph: a) Tumour cells in interlacing fascicles with mild nuclear atypia. b) Tumour cells invading the muscular layer

Table 1: Summary of Bladder LMS articles published over the last ten years

Author (Reference)	Number of Patients	Procedure	Location	Grade	Adjuvant Therapy	Follow up Period	Recurrence	Metastasis
Spiess PE[4]	3/16	Partial cystectomy	NA	NA	1 Yes 2 No	NA	1 (Salvage Cystectomy)	2
Minagawa T[5]	1 (RB Mutation)	Partial cystectomy	Left Lateral Wall	NA	Nil	16	Yes – Managed by Radical Cystectomy	No
Labanaris AP[9]	1/7	Partial cystectomy	NA	Low	No	8	No	No
Gupta DK[10]	1	TURBT	Bladder Neck	Low (3-4/10 hpf)	Chemoradiation	12	Nil	Nil
Zhong D[11]	1	Partial cystectomy	Left Lateral Wall	NA	Nil	48 Months	Nil	Nil
Hamadalla NY[12]	1	Partial cystectomy + Right distal ureterectomy	Right Lateral Wall	High (22/10 hpf)	Chemoradiation	4	Nil	Nil
Xu Y[13]	1	Partial cystectomy	Left Lateral Wall	Low (MSKCC Stage 1)	Nil	72	Nil	Nil
Tsujita Y[14]	1	Partial cystectomy (+Ileal resection)	Bladder Diverticulum	High	Nil	1	Yes	Yes
Cumplido JD[15]	1	Partial cystectomy	NA	NA	Yes	12	No	No

Memorial Sloan–Kettering Cancer Center soft-tissue sarcoma staging system is used to stage LMS of the bladder based on tumor grade, size, invasion, and presence of metastasis. Rodríguez *et al*. reported poorly differentiated histology in 63.2% cases in a series of 183 LMS, with 50% cases presenting with locally advanced or metastatic disease.[1]

Due to the infrequent occurrence of these lesions, there are no established guidelines for management. Conventionally, aggressive surgical extirpation, which involved radical cystectomy with wide margins, was advocated for the treatment of bladder LMS.[2,8] Partial cystectomy was considered suboptimal therapy with a higher likelihood of recurrence.[9] The evidence for an organ conserving approach is limited primarily to case reports. A PubMed search for bladder LMS articles published over the last 10 years revealed only nine articles that had follow-up data available specifically for patients with bladder LMS who underwent partial cystectomy or transurethral resection. These are summarized in Table 1. There was no reported recurrence or metastasis when organ preservation was performed for low-grade LMS, confined to the bladder. In cases where recurrence or metastasis did develop, the tumors were either high-grade or had extravesical spread. In lesions close to the ureteric orifice or the bladder neck, where a partial cystectomy was technically unfeasible, a transurethral resection of the tumor was performed.[10]

Radiation, chemotherapy, or both have been administered either in the neoadjuvant setting in locally advanced or metastatic disease or as adjuvant therapy in tumors found to have poor pathologic features post radical cystectomy.[2] In the series by Rosser *et al*. of 10 patients with locally advanced disease treated with neoadjuvant chemotherapy, three had partial response and one had complete response, with all patients deemed to have resectable disease posttherapy. The benefit of either radiation or chemotherapy, has not been proven in randomized control trials. Our patient did not receive adjuvant therapy, however, he remains disease free 48 months after partial cystectomy.

In conclusion, LMS of the bladder is a rare malignancy with clinical behavior strongly dependant on histologic characteristics. Majority of LMS present with high-grade advanced disease requiring radical surgery. However, there is growing evidence that in low grade, early stage LMS organ preservation is a viable option.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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