



URINARY BLADDER LEIOMYOSARCOMA --- A RARE CAUSE OF HEMATURIA

Urology

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KEYWORDS

INTRODUCTION:

Cases of bladder leiomyosarcoma represent 0.1% of all nonurothelial tumours. Nonurothelial neoplasms of the bladder account for <5% of all bladder tumours. Urinary bladder leiomyosarcoma is a very rare entity with few cases reported in the literature. Given its scarcity, little is known about its etiology and consensus lacking on therapeutic management. Here we present the case of a 36year old female patient diagnosed with bladder leiomyosarcoma and then treated with partial cystectomy of the bladder and bladder reconstruction with bilateral pelvic lymph node dissection..

Case Presentation:

A 36 year old lady presented with complaints of hematuria for the past 2months. She had been operated previously in 2014 for ovarian cyst. A 8.21cm * 3.96cm hypoechoic heterogenous space occupying lesion, without any obvious internal vascularity, was noted in the anterior wall of the urinary bladder by means of USG of lower abdomen. CECT whole abdomen and pelvis (Image-1) confirmed the above findings and size of the large well circumscribed lobulated polypoidal lesion was 6.4cm (CC)* 4.3cm(Tr) * 4.4cm(AP). Heterogenous enhancement and mild thickening of the bladder wall was also seen. There was no upper urinary tract obstruction.No perivesical extension was noted. Associated findings were mildly bulky and cystic left ovary (4.2cm * 2.4cm).



Image-1- Computed tomography of the pelvis with intravenous contrast showing the mass on the right anterior part of the bladder.

The patient underwent cystoscopy and complete transurethral resection of the bladder tumour. Approximately 7*8cm tumour involving right dome and right superolateral border was noted along with 3*4cm base with punctate calcifications. Histopathology of the tumour bits revealed high grade spindle cell sarcoma, marked nuclear atypia, atypical mitosis and coagulative tumour necrosis. Mitotic count was 9-10/10HPF. Diagnosis of high grade leiomyosarcoma was made.(Image-2)

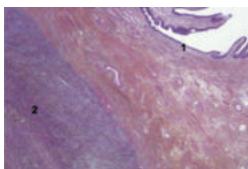


Image 2: histologic appearance of myxoid leiomyosarcoma of the

bladder hematoxylin eosin stain *25 ; 1- normal urothelium 2- massive invasion of the muscularis propria

The definitive procedure was done on Postoperative day 5 which comprised of partial cystectomy and bladder reconstruction and bilateral pelvic lymph node dissection(Image 3,4) . Multiple bilateral pelvic lymph nodes were seen, more likely to be reactive. No perivesical spread could be found. 4*4cm tumour base was excised with wide margins. The bladder was reconstructed over Foley's catheter in situ by vicryl 3-0. Drain was placed, wound closed with loop PDS and skin stapled.

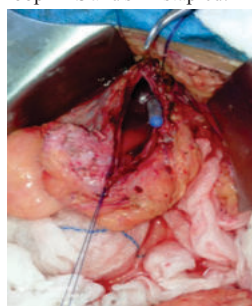


Image 3- Intra-op picture

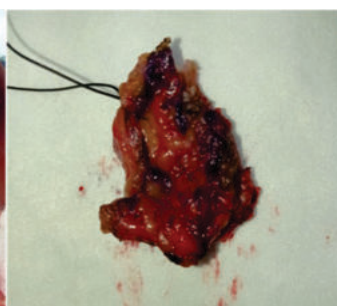


Image 4- resected specimen

Excised specimen consisted of partial cystectomy specimen which had focal area of residual leiomyosarcoma present. Immunohistochemistry performed on the block shows the lesional cells are positive for desmin, h caldesmon and SMA and negative for Cytokeratin . Lymph nodes sent for histopathology examination were found to be reactive (3 reactive right obturator and 4 reactive left pelvic) .

The postoperative period was uneventful . Patient was followed up at regular intervals and she experienced complete resolution of her urologic complaints .

DISCUSSION :

Only about 100 cases of bladder leiomyosarcoma have been reported in the literature, which represent 0.1% of all nonurothelial tumours¹ Sarcoma is the most common mesenchymal malignancy of the bladder and leiomyosarcoma is the most common bladder sarcoma in adults² Gross hematuria is present in more than 80% cases. There is strong evidence that the incidence of leiomyosarcoma is increased in patients receiving pelvic radiotherapy or systemic chemotherapy such as cyclophosphamide.³ Tobacco appears to not increase its incidence. There is no general consensus on treatment but it is considered to be highly aggressive that requires aggressive surgical resection with wide margins.⁴ Other favourable prognostic factors are tumour diameter of less than 5cm, low histologic grade, paratesticular or bladder tumour site. The Memorial Sloan Kettering Cancer Centre sarcoma staging was able to predict survival in such patients and suggested that urologic sarcomas behave similar to soft tissue sarcomas arising elsewhere in the body.

The standard surgical technique is en bloc removal of the bladder and the prostate and seminal vesicles in men, and uterus, cervix and vaginal cuff in women, along with bilateral pelvic lymph node dissection. It may result in low rates of positive tumour margins and local tumour recurrences. The urachus and the perivesical fat enveloping the upper hemisphere of the bladder should be ideally completely resected. Patients with advanced disease may benefit from neoadjuvant chemotherapy which would ultimately allow a radical procedure.⁵ Minimally invasive procedures like TURBT and adjuvant chemoradiation is an alternative for patients refusing a radical treatment. Partial cystectomy should only be considered as a palliative procedure and is possible only if the tumour is limited to certain locations of the bladder.

Bladder leiomyosarcomas may have 5 year disease specific survival rates of 84% (better than sarcomas in other organs). The overall recurrence rate is about 16% and most recurrences are seen in the pelvis. Local recurrences should be treated with systemic chemotherapy and/or pelvic external radiotherapy.

Bladder leiomyosarcomas were once thought to have had an extremely grim prognosis, however if an adequate surgical approach is sought for initially, patients may achieve significant long term survival⁶

CONCLUSION:

Leiomyosarcomas of the bladder have always been considered as a highly aggressive disease with poor prognosis. We present the case of a high grade bladder leiomyosarcoma primarily treated with minimally invasive surgery and then finally with partial cystectomy and bladder reconstruction and bilateral pelvic lymph node dissection and harbouring negative resection margins in the final pathology. Recent studies suggest that proper tumour free margin are essential for long term survival and lower rates of local recurrence.

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