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Case Report
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A rare case of Leiomyosarcoma of the Prostate presenting with Recurrent Hematuria and Retention of Urine

Md Shafiquzzaman[1], Md Habibur Rahman[2], Tohid Mohammad Saiful Hossain[3], Md Shamim Hossain[4], Saumitra Chakravarty[5], Md Serajul Islam[6]

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Abstract

Leiomyosarcoma of the prostate is an extremely rare aggressive tumor accounting for only 0.1-0.2% of all malignant prostatic tumors. About two hundred cases have been reported worldwide in the literature so far. There are no specific clinical features and imaging characteristics that can detect the condition, rather it is a histopathological and immunohistochemical diagnosis. So, a high degree of skepticism is needed in patients with hematuria, obstructive lower urinary tract symptoms with normal serum prostate specific antigen to detect the condition in time. Though there is no certain treatment modality for this condition, a multidisciplinary approach and aggressive management may benefit the patient. Here we report a case presented with recurrent episodes of hematuria, retention of urine and a history of transurethral resection of prostate multiple times which revealed benign prostatic hyperplasia. We have performed radical cystoprostatectomy of this patient which revealed Leiomyosarcoma of the Prostate. The patient was referred to clinical oncology for further management. But unfortunately, the patient died before receiving any systemic therapy.

Introduction

Carcinoma Prostate is the second most frequent carcinoma and histologically, adenocarcinoma is the commonest type. Whereas sarcomas of the prostatic stroma are extremely rare. In general terms, soft tissue sarcomas arise from tissues of mesodermal origin. They comprise 1% of all cancers and 0.1 to 0.2% of all prostate cancers. Leiomyosarcoma and rhabdomyosarcoma are the common subtypes of prostatic sarcomas. Of them, leiomyosarcoma accounts for 25-52% of all prostatic sarcomas. Leiomyosarcoma of prostate are common in adults and rhabdomyosarcoma of prostate are common in children.[1,2,3] Leiomyosarcoma of the prostate was first described by Sambert in 1853. A thorough search in literatures published in English shows fewer than 200 case reports regarding leiomyosarcoma of the prostate globally till now. Patients usually present with nonspecific lower urinary tract symptoms and upon further evaluation by physical examination and imaging investigation, most often misdiagnosed and treated as benign prostatic hyperplasia. As this is an aggressive tumor, patient returns with similar sort of symptoms. There is no specific treatment recommendation regarding the management of prostatic leiomyosarcoma due to its rarity. The disease has a poor prognosis due to its aggressive nature with a median survival of 17 months and 5-year survival rates of 26%.[4,5] Herein, we report

1. Resident, Department of Urology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh
2. Professor, Department of Urology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh
3. Professor, Department of Urology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh
4. Assistant Professor, Department of Urology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh
5. Assistant Professor, Department of Pathology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh
6. Medical Officer, Department of Urology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh
Correspondence: Dr Md Shafiquzzaman, Resident, Department of Urology, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh, Email: shafiq.zaman777@gmail.com

a case of Leiomyosarcoma of the Prostate in a 48 years old man, diagnosed after radical surgery who had history of repeated TURP and presented with worsening symptoms afterwards.

Case Presentation

A 48 years old gentle man with no comorbidities presented to the local hospital with the complaints of acute retention of urine in July,2020. Before that, he was suffering from poor flow of urine, frequency, urgency and occasional burning micturition but seek no medical attention. His International Prostatic Symptom Score (IPSS) was 27 of which storage 10 and voiding 17. Then he was catheterized and advised to take necessary medications. After one week, upon removal of catheter, he again developed urinary retention and recatheterisation was done and advised for surgical intervention. But he denied to do surgery and catheter was removed after one week. This time he was able to void with poor flow. During Coronavirus epidemic, he took conventional medicine for one year.

In later part of 2021, when situation didn't improve, he underwent transurethral resection of prostate (TURP) in a local clinic. The histopathology revealed benign pathology. He was symptomless for 4-5months. After that, he developed urinary tract infection (UTI) and microscopic hematuria several times and treated conservatively.

Then he underwent TURP for the second time in August, 2022 for similar sort of symptoms. The histopathology revealed acute and chronic prostatitis with prostatic abscess.

After being relatively better for several months, his lower urinary tract symptoms again worsened associated with gross hematuria. This time he developed clot retention and was treated with trichannel catheterization with irrigation in emergency department elsewhere. After removal of catheter, he was better for one month with medication.

In February,2023, he once again developed features of UTI, hematuria, perineal pain and voiding LUTS and presented to BSMMU. He was evaluated with proper medical history, his previous medical records and physical examination. On digital rectal examination, prostate was found hugely enlarged in size, firm in consistency, palpable surface was smooth and upper limit couldn't be reached. He was getting treatment for UTI. But during treatment he again developed clot retention and was admitted in our emergency department and managed accordingly. He was shifted to indoor department and underwent necessary investigations. Urine R/M/E revealed both Pus cell and RBC was plenty/HPF and urine C/S revealed growth of E. coli which was sensitive to colistin sulphate and polymyxin B. Ultrasonography showed prostate size was 110cc with median lobe protrusion within urinary bladder. There was also 86cc hypoechoic mass within the prostate suggesting hematoma (Fig.1). S. Creatinine was 1.1mg/dl and S.PSA was 0.5ng/ml.

Then he underwent Urethrocystoscopy (Fig.2) and found organized blood clot within the urinary bladder and prostatic tissue that was resected. The tissues were sent for histopathology which revealed a benign neoplasm composed of smooth muscle arranged in fascicles which was Masson Trichrome stain positive suggestive of benign spindle cell neoplasm most probably stromal nodule or leiomyoma. Upon Immunohistochemistry the tumor cell was found positive for Smooth Muscle Actin (SMA), Epithelial Membrane Antigen (EMA) but negative for Pancytokeratin, CD 34, Progesterone Receptors (PR), Anaplastic Lymphoma Kinase (ALK) and Desmin suggesting leiomyomatous or stromal lesions.

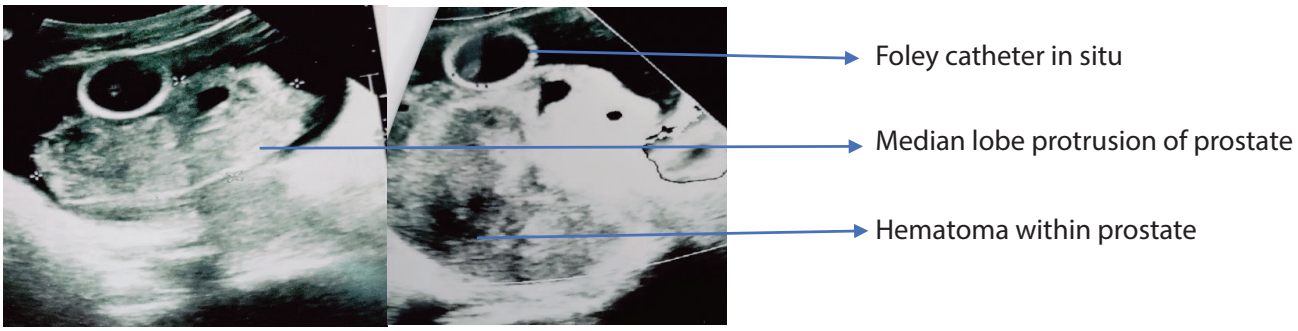

Fig. 1: *USG KUB with Prostate showing hugely enlarged prostate, size- 110cc with median lobe protrusion within urinary bladder. And also, 86cc hypoechoic mass within the prostate suggesting hematoma.*

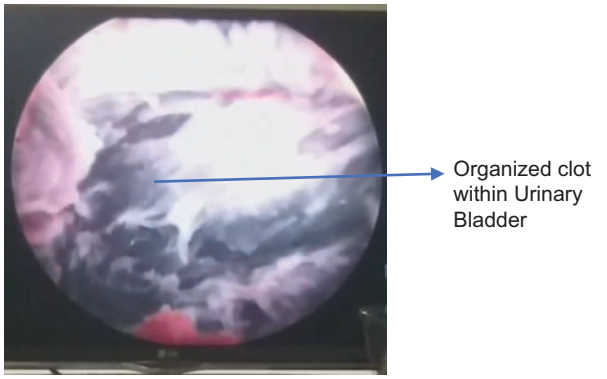

Fig.2: *Urethrocystoscopy showing organized blood clot within Urinary Bladder.*

He was advised to perform an Ultrasonography after one month. This time the ultrasonography (Fig.3) report revealed a urinary bladder mass of about 13.4 x 9.95cm. Prostate volume was 25cc.There was also upper tract change this time in the form of bilateral hydroureteronephrosis. His S.Creatinine was found elevated i.e. 2.58mg/dL. So, we advised Non contrast CT scan KUB (Fig.4) which revealed soft tissue density mass at the base of urinary bladder which could not be separated from the prostate compressing the rectum from anteriorly measuring about 7.4cm x 8cm x 10.8cm. There were also prostatic calcification and bilateral hydroureteronephrosis.

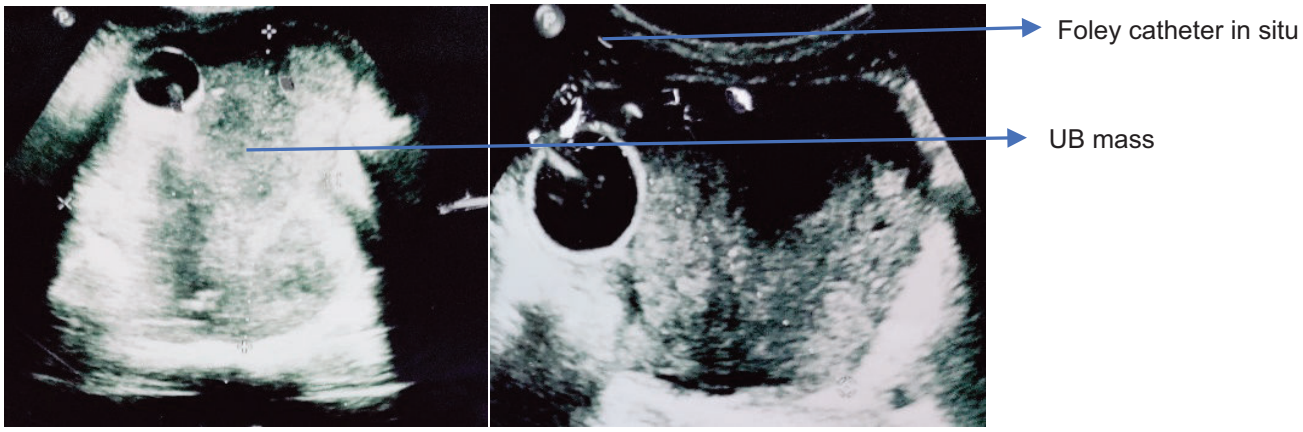

Fig. 3: *USG showing large lobulated mixed echogenic mass measuring about 13.4cm x 9.95cm in the UB. There is also foley catheter in situ. Prostate size-25cc. MCC-113ml*

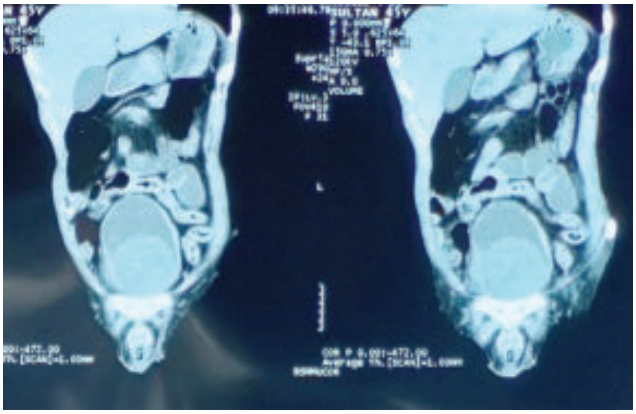

Fig.4: *Non contrast CT KUB showing soft tissue density mass at the base of urinary bladder which could not be separated from the prostate compressing the rectum from anteriorly measuring about 7.4cm x 8cm x 10.8cm*

His renal function was deteriorating and S. Creatinine was found to be as high as 6.8mg/dL at one stage and he was anemic, Hb% was 7.8g/dL and his S. Albumin was found 2.5g/dL. Then, multidisciplinary approach was taken involving faculties of nephrology, clinical oncology, radiation oncology, pathology, radiology and imaging and critical care medicine. The patient was optimized as per recommendation for radical surgery. His S. Creatinine was reduced to 1.99mg/dl, Hb% was increased to 13.1g/dl and S. Albumin was 3.17g/dl before the surgery. Then the patient underwent Radical Cystoprostatectomy with bilateral cutaneous ureterostomy. A soft friable mass was found during operation which arose from the prostate and occupied whole urinary bladder but it was separated from the urinary bladder wall. The mass was measured about 8 x 6 x 7 cm. (Fig.4) The specimen was sent for histopathological examination.

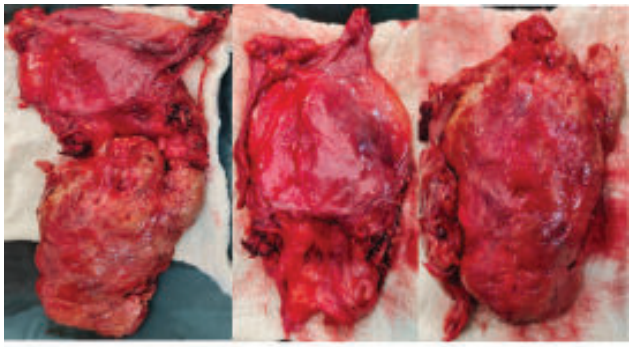

Fig.5: (A) Radical cysto-prostatectomy specimen, (B) Resected Urinary Bladder, (C) Resected prostate having friable soft tissue mass of about 8 x 6 x 7 cm.

Gross examination revealed prostatic specimen with tumor sized 16x8x7cm and urinary bladder measuring 10x8.5x6.5cm. On microscopic examination, the specimen of prostate showed a malignant neoplasm composed of pleomorphic spindle cells arranged in intersecting fascicles with areas of necrosis and frequent mitosis (Fig.5). The histopathological findings confirmed the presence of Grade-II Prostatic Leiomyosarcoma according to French Federation of Cancer Centre Sarcoma Group (FNCLCC) grading. The urinary bladder was found to be free of tumor. No immunohistochemistry of the resected specimen was done.

Follow up:

We discharged the patient with necessary medication advices and referred to medical oncology department for further management. The patient came after three months of surgery with moderate anemia, hyponatremia and raised serum creatinine. His anemia and hyponatremia were corrected. But he couldn't start chemotherapy. The patient died in October,2023; four months after the surgery and twenty-one months after seeking first medical attention.

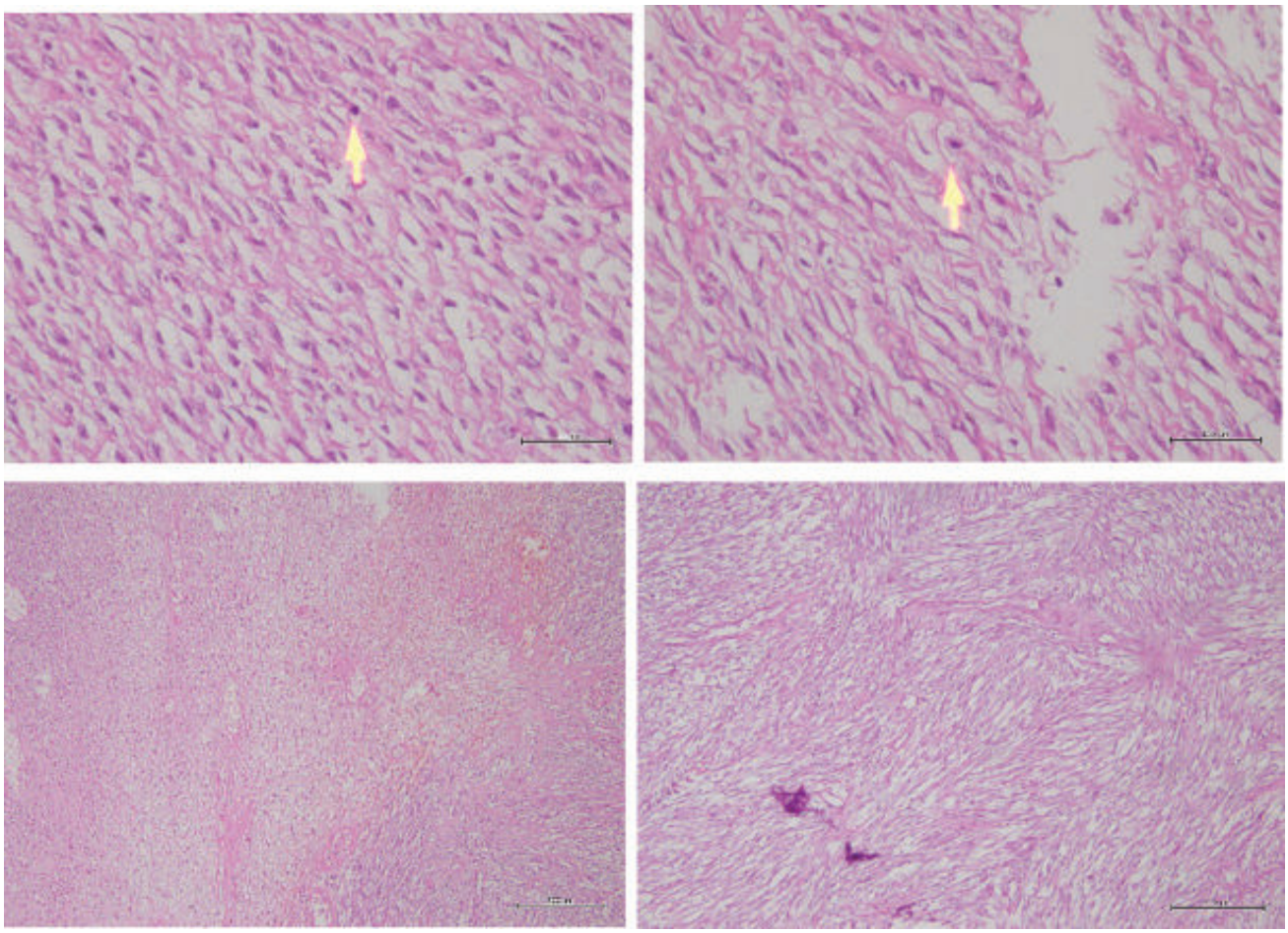

Fig.6: *Microscopic appearance of the specimen showing nuclear pleomorphism with increased nuclear cytoplasmic ratio with mitoses and areas of necrosis.*

Discussion:

Adenocarcinoma is the most common prostatic malignancy accounting for more than 95%. Many unusual tumors of almost similar clinical features but of variable prognosis have been reported within prostate. These unusual tumors may originate from prostatic epithelium, stroma or ectopic cells within the prostate.[6]

Sarcoma of the prostate is a rare genitourinary malignancy. It comes from the interstitial tissue of the prostate. It is classified pathologically into- rhabdomyosarcoma, leiomyosarcoma, fibrosarcoma and spindle cell sarcoma. Under microscope, prostatic leiomyosarcoma exhibits moderate to severe abnormal spindle cells. To distinguish the low grade malignant smooth muscle sarcoma and leiomyoma, the degree of cell atypia, lesion site, degree of mitosis, presence of necrosis and rate of infiltration can be used. More atypia or mitosis in leiomyosarcoma cells denote higher degree of malignancy. Leiomyosarcoma lacks normal glands unlike stromal sarcoma.[7]

Prostatic leiomyosarcoma is an aggressive rare neoplasm. The usual age range is 41 to 78 years. Due to non-specific presentation, it is difficult to detect the disease early. As a result, most of the patients present at an advanced stage with metastasis to lung, liver etc. The most common presentation is urinary obstruction that may be associated with perineal pain, recurrent hematuria, strangury, burning on ejaculation and constitutional symptoms. Recurrent hematuria most often related directly to prostatic leiomyosarcoma due to hypervascularity. As the disease is non-epithelial in origin, serum PSA is typically within normal limits or may be raised a little. The disease requires histopathological diagnosis. For this, TRUS guided needle biopsy or trans urethral resection of prostate is needed. Histopathological examination and FNCLCC grading system are used for definitive diagnosis. The tumor may of variable sizes ranging from 3 to 21cm. The tumor shows marked necrosis, hypercellularity, nuclear pleomorphism and positive immunohistochemical stains like smooth muscle actin, desmin, vimentin, calponin and CD34.[8,9]

CT and MRI scans can provide information about local and distant spread of the disease. It is most commonly metastasized to lung, liver, bone and rarely to brain.

There is no established treatment strategy for prostatic leiomyosarcoma. The treatment options include a multimodal approach of combined surgery, radiation and chemotherapy. Surgery remains the mainstay of treatment in resectable tumors with the options of radical cystoprostatectomy, radical retropubic prostatectomy, suprapubic prostatectomy and pelvic exenteration. Surgery can be preceded or followed by chemotherapy and/or radiotherapy. Neoadjuvant therapy can help in reducing tumor burden and facilitate tumor resection.[10]

The prognosis of leiomyosarcoma of prostate is very poor. If the patient is fit and the tumor is localized, multimodal treatment including radical surgery and chemotherapy may improve patient survivality. The median survival age is 17months. The survival rates of 1, 3 and 5year are 68%, 34% and 26% respectively. Absence of metastasis and negative surgical margin are the only factors for long term survival[11].

Conclusion:

As prostatic leiomyosarcoma is a very rare and aggressive neoplasm with poor prognosis, a high degree of suspicion is needed to diagnose it at early stage. Still there is no consensus on its appropriate management. Multimodal regimens including radical surgery, chemotherapy and radiotherapy may benefit the patient survival. There should be a global multicenter trial on management of prostatic leiomyosarcoma to establish an appropriate treatment algorithm.

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