Recurrent episodes of hematuria: A rare presentation of leiomyosarcoma of prostate

ABSTRACT
Leiomyosarcoma of the prostate is an extremely rare entity. Sarcomas account for about 1% of all malignant tumors and less than 5% of them arise from the genitourinary tract. Majority of patients present with urinary obstructive symptoms. The outcome is generally poor. Surgery with or without radiotherapy / chemotherapy forms the mainstay of treatment for patients with operable tumors. We report a patient presenting with recurrent episodes of hematuria.

KEY WORDS: Leiomyosarcoma, presentation prostate

INTRODUCTION
Leiomyosarcoma of the prostate is an extremely rare entity. Sarcomas account for about 1% of all malignant tumors and less than 5% of them arise from the genitourinary tract. It is known to have a poor outcome. We report a gentleman presenting with repeated episodes of hematuria diagnosed to have leiomyosarcoma of the prostate.

CASE REPORT
A 67-year-old gentleman presented with history of repeated episodes of hematuria and poor urinary stream. He was diagnosed to have benign prostatic hyperplasia (BPH) two years ago, for which he had undergone transurethral resection of prostate (TURP) tumor, the slides of which were not available to us for review. A rectal examination revealed hard prostatomegaly (grade 4) with obliteration of the median sulcus and enlarged right lobe abutting the rectum but not fixed to it. This mass was found to infiltrate the periprostatic tissue, reaching up to the pelvic sidewall. Prostate-specific antigen (PSA) at presentation was normal. An ultrasonogram of the abdomen revealed a heterogeneous hypoechoic mass arising from the prostate gland infiltrating the bladder base and projecting in to the bladder lumen. The urinary bladder contained blood clots with thickened walls. MRI of the pelvis showed a 7.5 x 4.3 cm tumor involving the right lobe of prostate, disrupting the capsule with extension into the periprostatic fat, neurovascular bundle and bladder base [Figure 1]. Cystoscopy showed a large fleshy growth in the prostatic urethra and bladder lumen along with multiple clots. The right lobe of prostate was enlarged, especially at the apex. Biopsy from the prostatic mass showed a spindle cell sarcoma destroying prostate with an occasional spared benign prostatic gland entrapped in the tumor [Figure 2]. The tumor was composed of integrating fascicles. The spindle-shaped cells revealed elongated blunt-ended cigar-shaped hyperchromatic nuclei and eosinophilic fibrillary cytoplasm characteristic of leiomyosarcoma [Figure 3]. Nuclear pleomorphism, raised mitotic activity and areas of necrosis confirmed the malignancy. Metastatic work-up did not reveal any evidence of distant metastasis. The hematological profile was within normal limits. In view of the disease infiltrating into the surrounding structures and the expected morbidity, surgery was ruled out. He received two cycles of neoadjuvant chemotherapy using Ifosfamide (500 mg/m²) and Epirubicin (60 mg/m²), followed by external beam radiotherapy to the prostate gland with adequate margins using 3D-CRT technique delivering a dose of 6,000 cGy/30#45days. He completed the radiotherapy treatment without any breaks with...
Sarcomas account for 1% of all malignant tumors and less than 5% of them arise from the genitourinary tract, accounting for 1-2% of all malignant genitourinary tumors. In a large series of histologically proven cases of soft tissue sarcoma admitted to the Memorial Sloan-Kettering Cancer Center (MSKCC), Russo et al reported that 2.7% had tumors arising from the urinary tract and male genital tract (urological sarcoma). In this series, the commonest site of origin of adult urological sarcoma was paratesticular, followed by the prostate, seminal vesicles, urinary bladder and kidney; and the commonest histological type was leiomyosarcoma, followed by rhabdomyosarcoma and liposarcoma.

Sarcomas and related proliferative lesions of specialized prostatic stroma are rare. Lesions have been classified into prostatic stromal tumor of uncertain malignant potential (P-STUMP); and prostatic stromal sarcoma based on the degree of stromal cellularity, presence of mitotic figures, necrosis and stromal overgrowth. Cytogenetic analysis of primary leiomyosarcoma of the prostate reveals clonal chromosomal rearrangement involving Chromosomes 2, 3, 9, 11 and 19.

Our patient presented with hematuria, which is a relatively rare presenting symptom described in literature. Most authors have reported dysuria and outlet obstruction as the presenting symptoms. In a series by Chen et al, dysuria was the first symptom for all patients with prostate sarcoma. Mondaini et al reported three cases of prostate sarcoma presenting with stranguria, symptoms of bladder outlet obstruction. None of the patients had hematuria. Cambronero et al reported an exophytic tumoral mass in the perineum as a rare presentation. Cuesta et al reported leiomyosarcoma detected in a patient having BPH who underwent TURP. Digital rectal examination generally reveals a prostatic mass, but needle biopsy is required for confirmation of diagnosis.

Surgery, radiotherapy and chemotherapy have been used in the management of leiomyosarcoma of prostate, but there are no standard recommendations. For operable tumors, the primary treatment is surgery, followed by chemotherapy and/or radiotherapy. Early diagnosis and complete surgical resection offer patients the best chance for survival.

The long-term survival rate and prognosis of patients with prostate sarcoma is poor. In a series by Russo et al, the prognostic variables which were found to be of significance on univariate analysis included tumor diameter less than 5 cm, low histological grade, paratesticular or bladder tumor site and complete surgical resection.

In a series of 14 patients reported by Chen et al, all patients underwent surgery, chemotherapy and/or radiotherapy. Eleven died of the disease, two of whom died approximately 12 months after diagnosis. Two patients were long-term survivors. Wang et al reported a series of seven patients with prostatic sarcoma. Two patients underwent radical cystoprostatectomy and survived for 13 and 21 months respectively after surgery. Two patients underwent total pelvic exenteration, followed by chemotherapy and radiotherapy and remained alive for 15 months after surgery. The other three received chemotherapy and radiotherapy and died approximately seven months after surgery.
Ahlering et al described 11 patients with leiomyosarcoma (seven with primary bladder and four with prostate tumors). Those having nonbulky disease underwent surgical resection and were observed if the margins and nodes were negative. Patients with positive surgical margins or nodes were treated adjunctively with external beam radiotherapy, 4,500-5,000 cGy and chemotherapy. Patients with bulky disease were treated with preoperative chemotherapy with or without radiotherapy, followed by an exenterative operation. Of the 11 patients, nine were without evidence of the disease, with a mean follow-up of 61 months (range 35 to 96 months). Camuzzi et al have reported a patient successfully treated with transperineal radon seed implantation and external irradiation.

Various chemotherapeutic regimes have been attempted for the treatment of leiomyosarcoma of prostate. Kuroda et al reported a case of leiomyosarcoma of the prostate accompanied by multiple hepatocellular carcinomas that was given combined chemotherapy (CVVADIC: cyclophosphamide, vincristine, adriamycin and DTIC). He died one year and two months after the initial diagnosis due to hepatic failure. During autopsy, it was revealed that the histology of the liver tumors was hepatocellular carcinoma and though the leiomyosarcoma of the prostate had invaded the wall of the bladder and the rectum, there was no obvious distant metastasis. Mondaini et al have used doxorubicin in one of the patients.

Patients with primary prostatic sarcomas usually present obstructive symptoms. The reported outcome of patients with nonmetastatic primary soft tissue sarcoma of the prostate gland is poor. Relatively good outcome in terms of disease and symptom control can be achieved with a combined modality approach comprising surgery, radiation therapy and chemotherapy.

REFERENCES