Stromal sarcoma of the prostate

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HENRY M, BRITTON C, COCO C, DADHANIA VS, SLOBODOV G. Stromal sarcoma of the prostate. *Can J Urol* 2019;26(1):9683-9685.

Primary spindle cell sarcoma of the prostate is exceedingly rare and accounts for 0.1% of all prostatic cancers. Typically, the disease course is rapid and portends a short and dismal prognosis. We report a case of prostatic stromal sarcoma (PSS) which likely lay dormant for several years after a transurethral resection of the prostate. This case is unique in that this cancer did not display the rapid growth of PSS in prior reports. Our patient received a cystoprostatectomy and an ileal conduit. This article describes a rare presentation of a rare tumor and a brief review of the literature.

Key Words: prostatic stromal sarcoma

Introduction

Prostate cancer is the leading cancer among men in the United States. The most common form of said cancer is adenocarcinoma. Variants account for upwards of 10% of all prostate cancers. Prostatic sarcomas (PSs) make up 0.1%-0.2% of all malignant tumors of the prostate. Less than 100 cases of primary prostatic spindle sarcomas are documented in the literature to date.[1] These tumors are comprised of malignant stroma with increased cellularity, mitotic figures, and pleomorphism, Figure 1. The histological features and immunophenotype of this patient's cancer are most consistent with a stromal sarcoma, likely of prostatic origin. Of note, tumor size, grade, and histological subtype of prostate sarcoma have no significant influence on actuarial survival.[2] Stromal sarcomas can exhibit local infiltration, as in this case, and also have the capacity for distant metastasis. We describe a rare case of prostatic stromal sarcoma (PSS) with indolent features, in addition to a review of the literature regarding these tumors.

Accepted for publication January 2019

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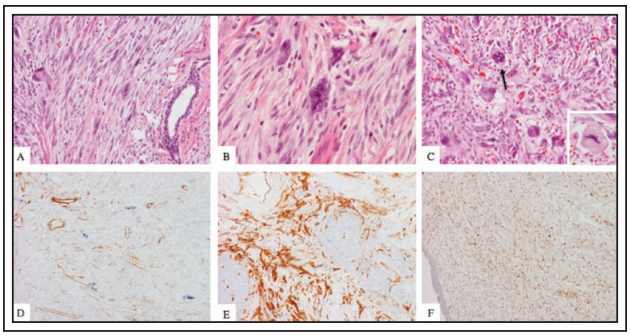

Figure 1. Stromal sarcoma is hypercellular and composed of spindle cells with infiltrative growth between prostate glands (**A**). Nuclear atypia and multinucleated giant cells (**B**). Mitotic figures are numerous (**C**). Immunoreactivity for smooth muscle actin (**D**), CD34 (**E**), and progesterone receptor (**F**).

Case report

A 31-year-old Caucasian male presented with acute urinary retention. Voiding was managed with a suprapubic tube over the past year. At age 21, he was diagnosed with chronic prostatitis, which manifested with dysuria and painful ejaculations. This was treated with antibiotics and NSAIDs for several years. Eight years later at age 29, he developed urinary retention requiring suprapubic tube placement and eventual transurethral resection of the prostate (TURP). The TURP specimen was positive for desmoid tumor of the prostate. Two years later he had a recurrence of acute urinary retention requiring the placement of a suprapubic tube under anesthesia. A transrectal ultrasound (TRUS) and prostatic biopsy were taken at that time, and the biopsy confirmed the presence of a desmoid tumor.

An MRI revealed an irregular, heterogenous lesion within the central zone of the prostate extending into the bladder neck and encroaching rectal margin with concern for extraprostatic extension, Figure 2 and Figure 3. Subsequently, the patient was treated with a radical cystoprostatectomy and an ileal conduit for urinary diversion. Pelvic lymphadenectomy was performed at that time and all nodes were negative. Cystoprostatectomy was chosen as it offered the most extensive resection of the cancer, which aside from an early diagnosis, is the most curative option.[2] Final histopathology revealed a diagnosis of spindle cell sarcoma, consistent with primary prostatic stromal sarcoma extending into the inked surgical margins of the membranous urethra and multiple prostate margins, 0/14 nodes were positive. Patient has remained cancer free with no evidence of disease for 3.5 years postoperatively, followed by serial PET scans via a medical oncologist.

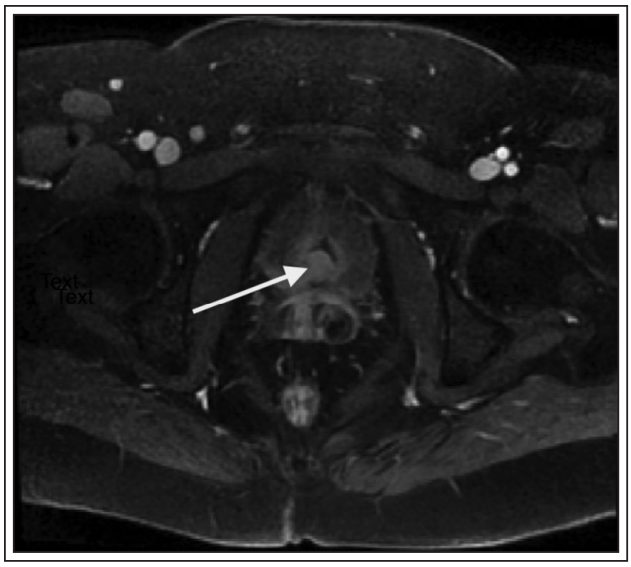

Figure 2. T2 MRI, transverse cut of prostate demonstrating primary tumor.

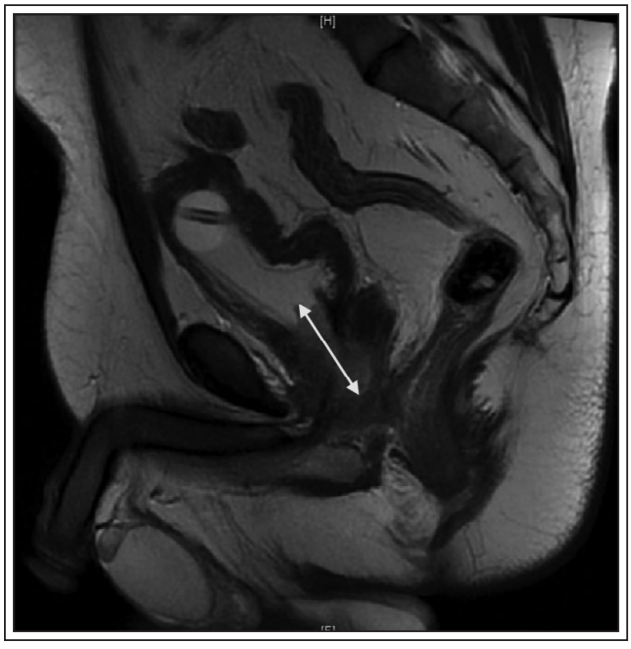

Figure 3. T2 MRI, sagittal cut of prostate demonstrating stromal sarcoma.

Discussion

PSs are exceptionally rare malignant tumors of the prostate gland.[3] While leiomyosarcoma is the most common variant seen in the adult population, PSS are much rarer, comprising less than 30 published cases.[4] PSS together with stromal tumor of uncertain malignant potential (STUMP) are histologically defined by the World Health Organization as specialized stromal tumors of the prostate.[5] In a clinicopathologic study of 50 cases of specialized stromal tumors of the prostate, the average age of diagnosis was 58 years, with subjects ranging from 27-83 years. Patients most commonly present with obstructive urinary symptoms.[5] Transrectal ultrasound-guided biopsy or transurethral resection most commonly provide the diagnosis. The most significant long term prognostic factors were negative surgical margins and absence of metastatic disease at presentation. Tumor size and grade, and histological subtype of the primary tumor have no significant influence on survival.[2] Average life expectancy is less than 1 year.[1] Typically, PSA levels are lower in patients with PSS compared to those with prostatic adenocarcinoma.[3] Due to the rarity and lack of long term follow up, the prognosis of these tumors is largely unclear.

Since the patient presented with lower urinary tract symptoms (LUTS) nearly a decade prior to the resection of his primary spindle cell sarcoma, it likely followed an indolent course. Although the first LUTS may have been a result of a different issue, we know for certain that this tumor lay dormant for a minimum of 2 years after the first biopsy. This is unique to these tumors, for they usually follow a rapid and aggressive course.[1] A number of factors may contribute to the indolent nature of this case. First, our patient's age was younger than average. This could indicate that he was diagnosed earlier, had a different variant of PSS, or was healthier due to his younger age. Also, his tumor of 1.4 cm x 1.4 cm was smaller than the average PSS size of > 4 cm diameter,[3] potentially signifying an idle course. □

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