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IP Journal of Diagnostic Pathology and Oncology

Journal homepage: https://www.jdpo.org/

Case Series

Primary leiomyosarcoma of prostate: Report of 3 cases

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ARTICLE INFO

Article history:
Received 15-04-2022
Accepted 25-04-2022
Available online 19-05-2022

Keywords:
Histopathology
immunohistochemistry
leiomyosarcoma
prostate sarcoma

ABSTRACT

Prostate sarcomas originate from the mesenchymal tissues including smooth muscle, fibromuscular stroma, paraganglia, nerves, and blood vessels. They account for less than 0.1% of all prostate tumors and often present with obstructive symptoms. Leiomyosarcoma is the most common sarcoma involving the prostate in adults affecting men between the ages of 40 and 78 years. Patients with leiomyosarcoma of the prostate commonly have a poor prognosis and their life expectancy depends on the stage of the disease at the initial diagnosis. Histopathological examination is essential for definitive diagnosis and can be performed at an early stage using guided transrectal prostate biopsy. Since prostate specific antigen (PSA) levels are generally normal, digital rectal examinations are extremely important. Tumor cells commonly express vimentin, smooth muscle actin and desmin, and up to 25% express cytokeratins. We report 3 cases of prostatic leiomyosarcoma with clinical, radiological, histopathological features and immunohistochemistry.

1. Introduction

Prostate leiomyosarcoma is a rare primary malignant neoplasm of prostatic smooth muscle that accounts for less than 0.1% of all prostate malignancies and typically follows an aggressive clinical course.[1] Less than 200 cases have been reported in the literature globally. Its risk may be related to prostatitis, perineal trauma, previous prostate biopsy and radiation.[2]

2. Case Reports

2.1. Case 1

A 70 years old male patient presented with lower urinary tract obstructive symptoms and hematuria for 3 months. Serum PSA levels was 4 ng/ml. The CT scan report showed a prostatic mass measuring 10x9.5x9 cm (Figure 1). The transrectal ultrasound (TRUS) biopsy was done one month back which revealed high grade prostatic leiomyosarcoma. We received a radical cystoprostatectomy specimen (Figure 2). Grossly, the specimen weighed 750 grams and measured 13x12x8 cm. Urinary bladder measured 7x6x5 cm and prostate measured 10x9x7cm. The entire prostate was replaced by a nodular grey white fleshy mass measuring 12x10x8 cm seen reaching upto the bladder. The cut surface was fleshy and firm and showed areas of necrosis and hemorrhages. Microscopic examination showed tumor cells arranged in interlacing fascicles and bundles (Figure 3a). Individual tumor cells were spindle shaped cells with enlarged hyperchromatic and pleomorphic nuclei (Figure 3b,c) with evidence of necrosis (Figure 3d) and multinucleated tumor giant cells. 10-12 mitotic figures were seen per 10 hpf. The immunohistochemical (IHC) profile demonstrated positive reactivity for SMA (Figure 4), Desmin, Vimentin while CD117 and S100 were negative. Based on histology and IHC findings, the diagnosis of

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https://doi.org/10.18231/j.jdpo.2022.034

prostatic high grade leiomyosarcoma was confirmed. This patient on pre-operative workup didn't have any metastasis. Post-operatively for 10 months, the patient was doing fine and was started on radiotherapy.

2.2. Case 2

A 35 years old male patient visited a urologist in September 2021 for sense of incomplete evacuation, nocturia and lower urinary tract symptoms for 15 days. Serum PSA was 0.82 ng/dl. MRI prostate was done which revealed 6.4x5.5x5.1 cm prostate and 8.3x7x9.3 cm exophytic lesion on left lateral aspect and involving the base of the bladder, laterally displacing bilateral seminal vesicles, distal sigmoid colon and rectum. Features suggestive of neoplastic etiology. Right and left lobe of prostate TRUS biopsy was done and sent for histopathological examination. Microscopy showed a tumor arranged in interlacing fascicles (Figure 5a). Individual tumor cells were spindle cells with large elongated nuclei, moderate amount of eosinophilic cytoplasm. Tumor was infiltrating the extraprostatic tissue (Figure 5b). Tumor showed moderate cellularity and focal areas of necrosis. Immunohistochemistry (IHC) was done which showed positivity for SMA, Ki-67 (15-20%) and negativity for desmin, CD34, CD 117 and S100. Based on histology and IHC findings, the diagnosis of prostatic low grade leiomyosarcoma was confirmed. Patient didn't come for follow up and unfortunately he expired within a year.

2.3. Case 3

A 53 years old male patient came for urology consultation in august 2021 in view of painless hematuria with dysuria associated with passage of blood clots and tissue for 15 days. Serum PSA was 3.60 ng/ml. The CT scan showed 4.5x4x3 cm soft tissue lesion involving the right lateral wall of bladder with loss of fat planes with prostate volume of 40cc. Histopathology of prostate chips done one and half years back suggested the possibility of leiomyoma. Prostate mass biopsy was done and sent for histopathological examination. Microscopical examination revealed a tumor composed of interlacing fascicles of smooth muscle cells which have elongated cigar shaped nuclei and moderate amount of eosinophilic cytoplasm (Figure 6a). Few areas of the tumor show (10%) nuclear pleomorphism (Figure 6b) and increased cellularity. There are occasional areas of necrosis seen (Figure 6c). Mitotic figures ware variable, maximum was 1-2/hpf. On immunohistochemistry, desmin showed positivity and Ki-67 showed variable positivity from 2-10% (Figure 6d). Myogenin, S100 and P63 was negative. Based on findings, the diagnosis was offered as smooth muscle tumor of uncertain malignant potential (STUMP). Same patient came after two months with same complaints. The MRI suggestive of mass arising from the prostate and neck of the bladder measuring 6x5.9 cm. Transurethral resection of bladder tumor (TURBT) was sent for histopathological examination. Microscopy revealed similar histomorphological findings as previous with few areas showing atypical large pleomorphic nuclei with prominent nucleoli (Figure 6b) and focal areas of necrosis (Figure 6c). Based on microscopy and IHC findings, the diagnosis of low grade leiomyosarcoma was confirmed. Four months postoperatively patient was doing fine with no metastasis.

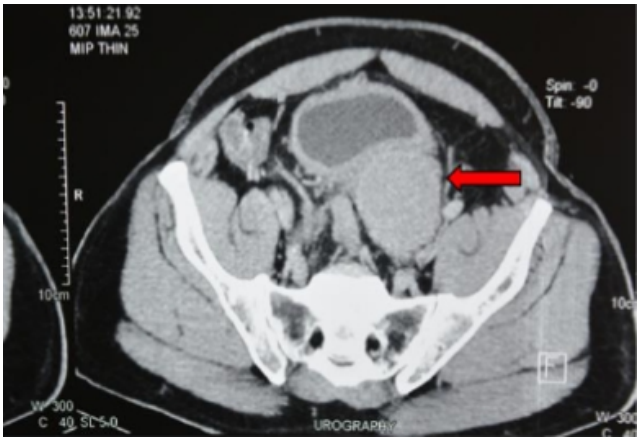

Fig. 1: Showing prostatic mass

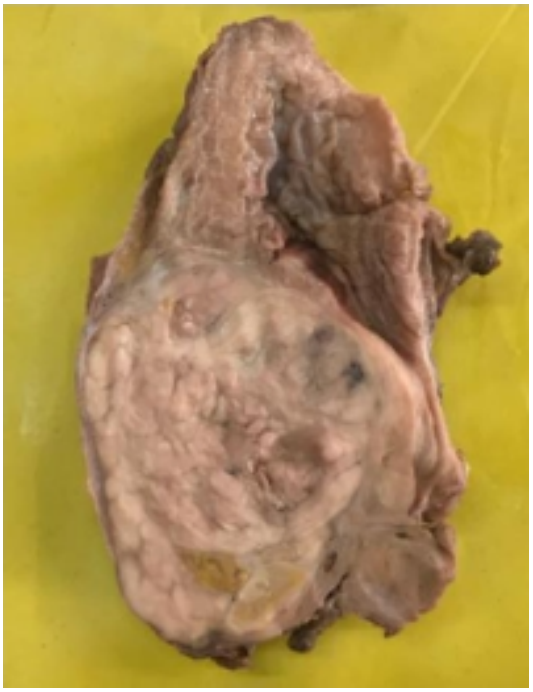

Fig. 2: Gross specimen of prostatic leiomyosarcoma. Cut surface showed grey white, firm, fleshy mass with areas of haemorrhages and necrosis, reaching upto the bladder

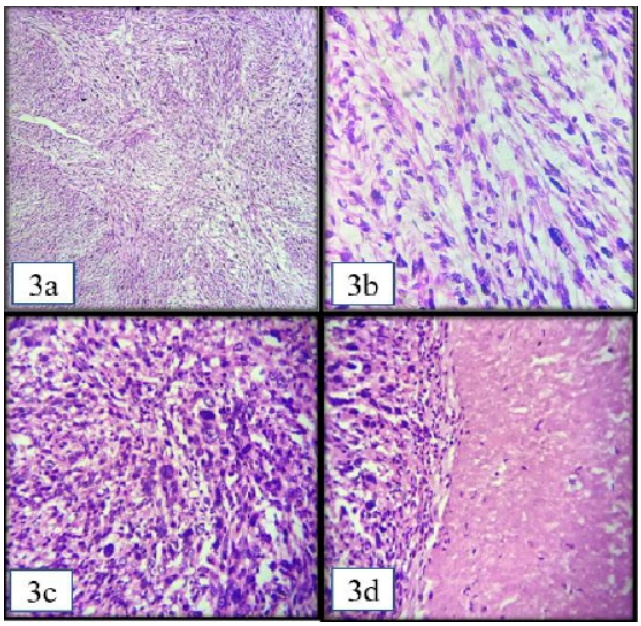

Fig. 3: Photomicrograph of prostatic leiomyosarcoma showing; **a:** Tumor cells arranged in interlacing fascicles and bundles.(H&E, 10x); **b:** Spindle shaped cells with nuclear pleomorphism.(H&E, 40x); **c:** Enlarged hyperchromatic and pleomorphic nuclei. (H&E,40x); **d:** Areas of necrosis. (H&E, 40x).

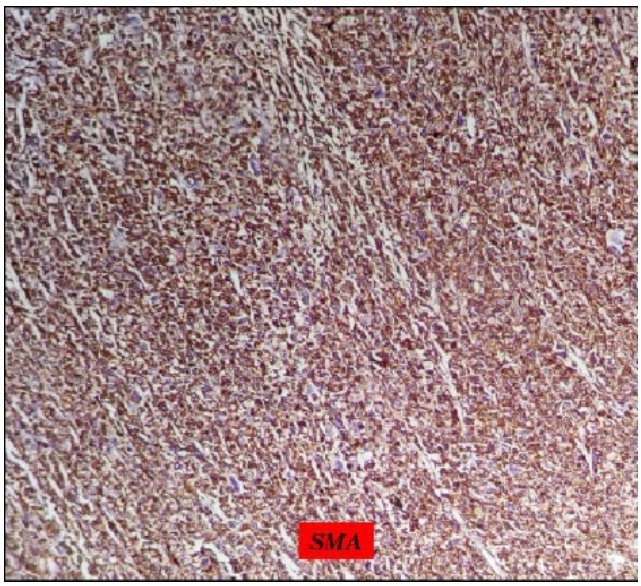

Fig. 4: Immunohistochemical profile result SMA positivity

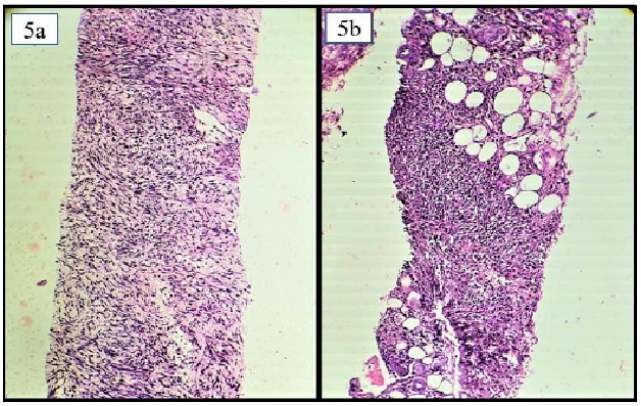

Fig. 5: a: Tumor cells arranged in interlacing fascicles. (H&E, 10X); **b:** Tumor infiltrating the extraprostatic tissue. (H&E, 10X).

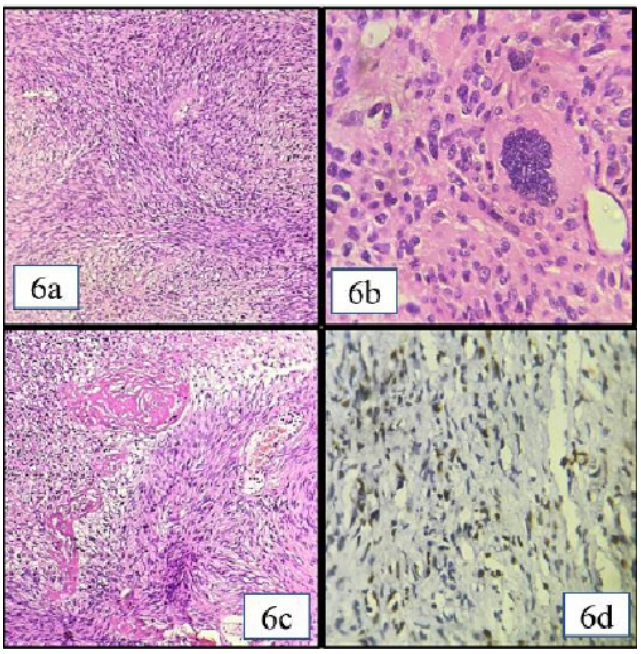

Fig. 6: Photomicrograph of prostatic leiomyosarcoma showing; **a:** Tumor cells arranged in interlacing fascicles. (H&E, 10X); **b:** Bizarre nuclei with nuclear pleomorphism. (H&E, 40X); **c:** Areas of necrosis. (H&E, 10X); **d:** Immunohistochemistry results Ki67 positivity (2-10%).

3. Discussion

Primary prostate leiomyosarcoma is the most common sarcoma affecting the prostate, although it is generally a rare tumor.[3] Leiomyosarcoma of prostate was described by Gaudin et al[4] in 1998. World Health Organization classifies this tumor as a distinct spindle cell neoplasm.[5,6] The etiology of primary prostate leiomyosarcoma is unknown or poorly understood.[3] Histological types can be divided into prostate leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma and spindle cell sarcoma. Benali et al[3] and

Heddat et al[7] in their study stated that the most frequent reason for consultation is a urinary obstructive syndrome including hematuria, burning with urination, perineal pain, urgency, acute urine retention and weight loss as observed in our case.[3,6–9] Patients typically present between the ages of 41 and 78 years (mean age 61 years).[1,10]

Serum PSA levels are typically normal due to their non-epithelial origin, making it highly difficult to differentiate these from benign prostatic diseases. The final diagnosis is usually accomplished via TRUS guided needle prostate biopsy, transurethral resection of prostate (TURP), and less frequently by open surgery.[5,9,11]

Abdominopelvic ultrasound would reveal hydronephrosis, thickening of the bladder wall or infiltration of the base of the bladder.[7,12] Transrectal prostate ultrasound is a useful technique to better assess the characteristics of the prostate. It can show heterogeneous hypoechogenic lesions in the prostate, an invasion of the capsule or an extension in the rectum, the pelvic sidewall, the seminal vesicle, or the ejaculatory duct.[7,12,13] Echo-guided transrectal biopsy tends to be the usual approach to obtain samples of the prostate lesion for the histological examination which would establish the diagnosis of leiomyosarcoma of the prostate.[7,12] Transurethral resection of the prostate for symptoms of the lower urinary tract or retention of urine may lead to the accidental diagnosis of leiomyosarcoma during the histological examination of the resection coupons.[7]

Tumors range in size from 3 to 21 cm and are highly infiltrative. Gross examination in Zazzara et al[1] study reveals an ill-defined mass with fleshy to firm consistency and a tan-pink appearance with focal areas of haemorrhage, necrosis and/or cystic degeneration. Heddat et al[7] stated that leiomyosarcoma can also present as an exophytic hypogastric mass of the prostate infiltrating the rectum, a perineal mass, or by metastases to the liver and lungs.[1,7]

Our case showed a nodular fleshy grey white mass seen reaching upto bladder.

Prostatic stromal lesions that were not obvious sarcomas have been designated STUMP. The presence of necrosis, atypical mitotic figures, marked hypercellularity, and nuclear pleomorphism without degenerative features are features of sarcoma, rather than STUMP.The majority of prostate leiomyosarcomas have a high grade appearance microscopically with areas of viable tumor consisting of hypercellular, intersecting bundles of eosinophilic, spindle shaped cells exhibiting variable degrees of nuclear atypia and mitotic activity. Necrosis and cystic degeneration can be prominent.[1]

The immunohistochemical profile of both prostatic STUMP and prostate stromal sarcoma demonstrate positive reactivity for CD34, which may aid in distinguishing them from other prostatic mesenchymal neoplasms such as rhabdomyosarcoma or leiomyosarcoma.[1] In cases of prostatic leiomyosarcoma, tumor cells commonly express vimentin, smooth muscle actin and desmin, and up to 25% express cytokeratins.Progesterone receptor expression has also been reported in leiomyosarcomas.[1,14]

The pathological diagnosis, supplemented by an immunohistochemical study is essential for the classification of these tumors.[3]

Recommendations regarding the management of prostate sarcomas weren't sufficient and contradictory. Sen and associate evaluated the results of 13 patients treated at the Mayo Clinic between 1970 and 1985, 10 patients were treated for the cure: 3 with leiomyosarcoma, 2 with rhabdomyosarcoma and 5 with carcinosarcoma. No distinction was made regarding the histological subtypes. All 3 patients who were treated with partial cystectomy had recurrence.[3]

Therapeutic combinations include surgery, pre- or postoperative radiotherapy, and neoadjuvant or adjuvant chemotherapy. However, the use of adjuvant chemotherapy increased cystic necrosis without actual tumor response. Post-operative radiotherapy was reserved for residual disease.[3]

Radical retro-pubic prostatectomy is a good curative option for prostatic sarcoma.[1,9]

Overall prognosis for prostate leiomyosarcoma is poor, and 50% to 75% of patients die of cancer within 2 to 5 years. Prognosis is improved in patients with no evidence of distant metastases at initial presentation and in those with localized disease in whom complete resection can be achieved surgically with microscopically negative margins.[1,9]

As clinical staging is necessary to perform a chest-abdominal-pelvic CT scan and a bone scan under the circumstances that lung is the most common site of metastasis followed by liver and bone. Multimodality treatment regimens including surgery, radiotherapy and chemotherapy are recommended. Patients with large tumors involving surrounding structures should be considered for neoadjuvant radiotherapy with or without chemotherapy to improve the probability of complete resection and minimize the extent of the resection to provide the best functional outcomes.[1,9]

4. Conclusion

Adult prostatic leiomyosarcoma is a rare tumor, often metastatic at diagnosis, with normal PSA levels. CT and especially MRI play an important role in the assessment of extension and post-treatment follow-up, but only the Prostate biopsy supplemented by immunohistochemical analysis can confirm the diagnosis. Their therapeutic management is not currently codified, and their prognosis remains very poor and can only be improved with a multi-disciplinary approach and early diagnosis, making it possible to perform complete radical surgery, the only effective therapy. Histopathology and

immunohistochemistry have very important roles in the diagnosis.

5. Acknowledgement

None.

6. Source of Funding

None

7. Conflicts of Interest

None.

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Cite this article: Patil MA, Padmanabhan A, Gadgil NM. Primary leiomyosarcoma of prostate: Report of 3 cases. *IP J Diagn Pathol Oncol* 2022;7(2):140-144.