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Oncology Leiomyosarcoma of prostate: Case report and literature review

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1. Introduction

Prostate sarcoma originates from the prostatic stroma and it is an extremely rare neoplasm that accounts for less than 0.1% of primary prostate malignancies; globally, less than 200 cases have been reported in the literature.¹ Leiomyosarcoma is the most common primary prostate sarcoma in adults and constitutes 38–52% of them.² It has an aggressive clinical course.³ Here we present a case of prostate leiomyosarcoma and a literature review regarding the clinical and pathological features, diagnostic modalities, therapeutic aspects and prognosis of this rare entity.

2. Case report

A 62-year old man performed an urological consultation for moderate lower urinary tract symptoms, in particular nocturia and frequency; macrohematuria wasn't detected. He was a heavy smoker, presented dyslipidemia and hypertension; he hadn't a family history of genitourinary cancer.

Serum PSA was 1.6 ng/ml. On digital rectal examination an apical node was palpated. On trans rectal ultrasonogram (TRUS) an apical non-homogeneous hypoecogeneous area was detected.

Furthermore, the patient performed a TRUS guided prostate biopsy whom histopathological examination revealed a suspect diagnosis of an atypical stromal tumor of uncertain malignant potential (STUMP).

As clinical staging, he performed a PET-CT scan revealed a significant uptake of the right prostate lobe, Chest/Abdomen CT scan and Bone scan revealed no pathological features, mpMRI prostate revealed a prostate volume of 94 cc, with a 12 mm suspect pathological node in the left prostate lobe (peripheral zone) and a 13 mm suspect pathological node in the right prostate lobe (anterior transitional zone) with no clinical lymph node involvement (Fig. 1a-b).

The patient underwent an extra-peritoneal robot assisted radical prostatectomy and extended lymphadenectomy.

Specimen histopathological examination revealed a high grade prostatic leiomyosarcoma, with no lymphnode pathological involvement and negative surgical margins; a complete resection was achieved. Immunohistochemical analysis showed no expression of cheratin AE1/AE3, PSA; proliferation rate of Ki67 was >20%; smooth muscle actin, desmin, vimentin and calponin were expressed (Fig. 2a–b).

Post-operative adjuvant therapy was performed with eight docetaxel cycles of 75 mg/m², weekly. After 6 months from surgery, the patient is alive and cancer free.

3. Discussion

Prostate sarcoma is a rare malignant genitourinary cancer. According to different histological types, it can be divided into prostate leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma and spindle cell sarcoma.⁴ 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 immunohistochemical profile of both prostatic STUMP and prostate stromal sarcoma demonstrate positive reactivity for CD34, which may aid in distinguishing

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Fig. 1. mpMRI prostate: a) suspect pathological node in the left prostate lobe (peripheral zone); b) suspect pathological node in the right prostate lobe (anterior transitional zone).



Fig. 2. Microscopic pathological features: a) Hematoxylin and eosin stain; b) Expression of desmin in immunohistochemical analysis.

them from other prostatic mesenchymal neoplasms such as rhabdomyosarcoma or leiomyosarcoma.

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.³

Patients range in age from 41 to 78 years at presentation with a mean age of 61 years. The lack of early specific symptoms results in more advanced disease at presentation. In fact, up to a third of patients have demonstrable metastases at presentation, usually to the lung, and sometimes to the liver as well.^{2,3}

Individuals most commonly present with signs and symptoms of urinary obstruction. Additional associated symptoms include perineal pain, hematuria, burning on ejaculation, constipation and weight loss. Serum PSA is typically within normal limits, attributable to the non epithelial origin of this disease. Diagnosis is made by TRUS guided needle biopsy or transurethral resection in most patients, and less commonly by open surgical procedures.^{2,3}

Tumors range in size from 3 to 21 cm and are highly infiltrative.³ Gross examination reveals an ill-defined mass with fleshy to firm consistency and a tan-pink appearance with focal areas of hemorrhage, necrosis and/or cystic degeneration. The majority of prostate leiomyosarcomas have a high grade appearance microscopically with areas of viable tumor consisting of hyper cellular, intersecting bundles of eosinophilic, spindle shaped cells exhibiting variable degrees of nuclear atypia and mitotic activity. Necrosis and cystic degeneration can be prominent. Tumor cells commonly express vimentin, smooth muscle actin and desmin, and up to 25% express cytokeratins^{3,5}. Progesterone receptor expression has also been reported in leiomyosarcomas.⁵

As clinical staging is necessary to perform a chest-abdominalpelvic CT scan and a bone scan under the circumstances that lung is the most common site of metastasis followed by liver and bone. $^{2,3} \ensuremath{\mathsf{C}}$

Multimodality treatment regimens including surgery, radiotherapy and chemotherapy are recommended.²

Overall prognosis for prostate leiomyosarcoma is poor, and 50%-75% of patients die of cancer within 2–5 years.^{2,3,5} 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.²

Patients with large tumors involving surronding structures should be considered for neoadjuvant RT with or without chemoterapy to improve the probability of a complete resection and minimize the extent of the resection in order to provide the best functional outcomes.

4. Conclusion

Leiomyosarcoma is the most common primary prostate sarcoma in adults and has an aggressive clinical course. Multimodality treatment regimens including surgery, radiotherapy and chemotherapy are recommended. Overall prognosis is poor and 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.

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