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Case report

Prostatic Leiomyosarcoma: Case Report

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Leiomyosarcoma of prostate is one of the rarest malignancies which is encountered in oncology practice. Symptomatically patients present with obstructive urinary complaints with no specific tumour marker or PSA abnormal value. Due to the limited number of cases till now no specific guidelines for treatment, diagnosis is available. Mostly treated on experience based on other prostate malignancy and sarcoma of other sites. Here we are reporting a case of primary prostatic leiomyosarcoma of a 17-year-old male and challenges regarding his diagnosis and treatment.

Introduction

Background

One of the rarest malignancies in the prostate is sarcoma [1]. They account for not more than 0.1% of all prostatic malignancies. Out of different sarcomas, leiomyosarcoma histology contributes 20-52% of cases. Generally, the initial presentation of a patient is with urinary complaints, mostly secondary to obstructive cause. Till now no specific serum marker is available for screening. PSA levels are almost within normal limits in patients of leiomyosarcoma. As of no treatment guideline available for prostatic sarcoma due to rarity of tumor [2]. Patients are treated with protocol based on other prostatic malignancies and leiomyosarcoma of different sites. We are here reporting our experience of primary prostatic sarcoma in 17 yr. old male.

Clinical case presentation

A 17 yr. an old man presented in the urology outpatient department with a complaint of urinary retention for the last 3-month duration associated with a complaint of pain in the abdomen. On presentation, patient ECOG performance status was 1 and other physical examinations were within normal limits. Patient underwent CPE + Foley catheterization. Ultrasound whole abdomen pelvis showed enlarged prostatic mass. MRI pelvis showed soft tissue mass measuring 7x6x9 cm in the prostate infiltrating the left lateral wall of the urinary bladder (Figure 1).

Figure 1. CEMRI Pelvis Showing Tumour in Sagittal and Coronal Section.

Mass was T1 isointense and iso hypo intense on T2 and showed diffusion restriction. Multiple enlarged pelvic lymph nodes and bone metastasis in left femur, sacral ala present. Level of serum LDH and PSA was 1796.56u/l and 0.15ng/dl respectively. Patient underwent a USG guided biopsy of prostatic mass. Biopsy showed tumour cells arranged in diffuse sheet, nest and also exhibit perivascular arrangement with high N: C ratio. Diagnosed it as a poorly differentiated malignant

tumor.

IHC was done, showed VIMENTIN positive, and negative PANCK, CD45, S100, SMA, CK7, CK20, CD117, OCT4, CD34, UROPLAKIN, PSA, PAX8. On the basis of IHC prostatic undifferentiated sarcoma or prostatic leiomyosarcoma was considered as differential. In view of high LDH and multiple pelvic lymph nodes, bone metastasis RHABDOMYOSARCOMA was ruled out by repeating IHC. Patient was then treated on the basis of sarcoma histology. Chemotherapy (LIPODOX +IFOSFAMIDE +MESNA), 4-week regimen was started. Patient is on our follow-up after 3 cycles. planned for response assessment after 4 cycles of chemotherapy completed.

Discussion

Prostatic stromal sarcoma are very rare malignancies, not accounting for even more than 1/1000 prostatic malignancies. Leiomyosarcoma contributes to more than 25% of these cases. so maximum experience regarding treatment of these tumors are from case reports and case series. One thing which clearly came out in this limited literature is the aggressive nature of these tumours and some survival advantage secondary to surgery. When it comes to histology, LMS has a variety of histology. One of the major challenges posed in LMS is early stage diagnosis. Mostly due to obstructive urinary symptoms often misdiagnosed as LUTI or BPH. This seriously hampers chances of early diagnosis. Most of the patients present as a locally advanced prostatic mass with obstructive LUTS. 25-40% cases come with metastatic disease to lung and liver most commonly [3]. Our patient presented with complaints of difficulties in urination resistant to conservative and medical management. After USG pelvis suspected pelvic mass and later confirmed via CEMRI pelvis, the patient was planned for USG guided prostatic biopsy. By considering age of child and rarity of adenocarcinoma prostate in this age group, LMS prostrate was in our differential secondary to Rhabdomyosarcoma in view of high pelvic lymph nodal status.

As expected in most malignancies pathological diagnosis is an easy way to confirm the diagnosis as compared to imaging and clinical finding. Light microscopy in our case tumor revealed spindle cells with hyperchromatic nuclei showed tumour cells arranged in diffuse sheet, nest and also exhibit perivascular arrangement with high N: C ratio and increased mitotic activity, which is suggestive but not specific to LMS. In IHC vimentin came to be positive with negative PANCK, CD45, S100, SMA, CK7, CK20, CD117, OCT4, CD34, UROPLAKIN, PSA, PAX8. This was an exception to previous case reports as most of them have SMA, CD44, CALPONIN, DESMIN positivity.

MRI pelvis showed soft tissue mass measuring 7x6x9 cm in the prostate infiltrating the left lateral wall of the urinary bladder. Mass was T1 isointense and iso to hypo intense on T2 and showed diffusion restriction. Multiple enlarged pelvic lymph nodes and bone metastasis in left femur, sacral ala present. Due to the exophytic nature of tumour in our patient as expected not much diagnostic challenge was posed if we compared to non-exophytic growth LMS. Presence of metastasis in our patient confirmed the aggressive nature of this disease. Brain CEMRI was not done as not indicated due to low chances of brain metastasis. FDG PET CT is a potentially useful modality for staging of locally advanced disease and useful for response evaluation [4].

As already discussed, management principles are based on our knowledge about treating prostate cancer and LMS elsewhere. disease within the prostate is usually treated with surgical options [5]. Surgically unfit patients can be taken up for EBRT. Surgical options include Radical retropubic prostatectomy, radical cystoprostatectomy, suprapubic prostatectomy, and pelvic exenteration. Bulky disease can be offered preoperative chemotherapy with or without EBRT, followed by an attempt for surgical resection.

Metastatic patients are treated with palliative chemotherapy or best supportive care. The commonly used regimens are either Adriamycin with ifosfamide or gemcitabine with docetaxel [6]. In our patient we gave ifosfamide + doxorubicin. Follow-up after 3 cycles showed improvement in

patient symptoms.

In conclusion, even though LMS is not so common, treating oncologists should keep it as one differential diagnosis when it comes to dealing with prostatic carcinoma. clinical and radiological nonspecific presentation poses diagnostic challenges. These tumors should be treated more aggressively. There is a need for a cancer registry of these tumors on national and global level to know more about these tumors' natural history, biological behavior, prognosis and most importantly treatment.

Acknowledgments

Statement of Transparency and Principals:

• Author declares no conflict of interest

• Study was approved by Research Ethic Committee of author affiliated Institute.

• Study's data is available upon a reasonable request.

• All authors have contributed to implementation of this research.

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