Case Report

Prostatic sarcoma: A case report with review of literature

Parveen Rana, Amrita Duhan, M. K. Garg[1], Vijay Punia[2]

Departments of Pathology, [1]Surgery, Bhagat Phool Singh Government Medical College for Women, Khanpur Kalan, Sonepat, [2]Medical Officer, Haryana Civil Medical Services, Haryana, India

ABSTRACT

Nonepithelial neoplasms of prostate are relatively infrequent and include a broad array of entities including both benign and highly aggressive tumors. Because of their rarity and limited understanding, when encountered, they may pose a diagnostic challenge, due to histological overlap between them or their rarity. The ancillary studies including immunohistochemistry (IHC) have often limited utility and the main criteria for diagnosis lies on morphology findings by hematoxylin and eosin (H and E) staining. We present here this rare entity which was not suspected either clinically or radiologically, was diagnosed on routine H and E staining after excision biopsy and confirmed with use of limited IHC panel.

Key words: Differential diagnosis, morphology, prostate, sarcoma

INTRODUCTION

Nonepithelial neoplasms of prostate are rare representing only <1% of all prostatic tumors. They include both benign and malignant tumors specific to the prostate that is specialized stromal tumors and those arising from extraprostatic sites like smooth muscle tumor, myofibroblastic tumor, solitary fibrous tumor (SFT), and gastrointestinal tumor (GIST).[1,2] The majority of tumors are characterized by spindle cell pattern with significant overlap in morphological pattern. For this, appropriate use of immunohistochemistry (IHC) and molecular studies are necessary for accurate diagnosis, prognosis, or prediction therapy. The diagnosis of nonepithelial neoplasms always raises the question whether the tumor is primary or secondary. The serum prostate specific antigen (PSA) level has its own limitation, because in the tumors of nonacinar origin, it is typically not elevated, unless there is focus of concomitant adenocarcinoma.[1]

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The literature defining these entities clinically and pathologically is very limited. Some aspects are still evolving requiring further studies. An accurate distinction is warranted because of significant differences in their therapeutic implications.

CASE REPORT

An old male aged 75 years was admitted in our hospital with chief complaints of retention and dribbling of urine along with burning micturition and occasional hematuria. Digital rectal examination revealed a massive growth anterior to the rectum with obliteration of median sulcus. On ultrasonography, prostate appeared large in size and lobulated in appearance with median lobe indenting bladder base; measuring 73 cm × 65 cm × 60 cm and weighing 151 g. No free fluid was seen in pelvis. On chest X-ray no active intrathoracic lesion was detected. PSA was 1.1 ng/ml (normal = 0-4 ng/ml). An impression of benign prostatic hyperplasia was made. Subsequently an open biopsy of the prostate was performed. Peroperative, both lobes of prostate were found to be moderately enlarged. Biopsy was received in multiple grey white to grey brown globular soft tissue pieces measuring together 7 cm × 6.5 cm × 5 cm. Cut-section was uniform, solid, and glistening with presence of myxoid change [Figure 1]. Microscopy showed a diffusely infiltrating spindle cell tumor with areas of myxoid degeneration, moderate

Address for correspondence: Dr. Amrita Duhan, H. No. 143, Sector 4, Rohtak - 124 001, Haryana, India. E-mail: amrita.duhan@gmail.com

to marked nuclear pleomorphism, and high mitotic activity (12/10 high power field). No epithelial element was noted [Figure 2]. Immunohistochemically, the tumor cells showed positivity for vimentin only while staining was negative for PSA, smooth muscle actin, CD34, CD68, myoglobin, and cytokeratin [Figure 3]. Therefore, a diagnosis of primary sarcoma of prostate (undifferentiated) was offered. However, the patient was not compliant for further treatment and was lost to follow-up.

DISCUSSION

The malignant mesenchymal lesions of the prostate have been classified into sarcomas of specialized prostatic stroma and other sarcomas equivalent to their soft tissue counterparts. The latter group accounts for only 0.1-0.2% of all primary prostatic tumors.[3] World Health Organization (WHO) histological classification of prostatic sarcomas is shown in Table 1.[4] Rhabdomyosarcoma (RMS) is the most frequent mesenchymal tumor within the prostate in childhood, whereas leiomyosarcoma (LMS) is the most common in adults.[5]

Majority (75-80%) of prostate cancers develop in the peripheral zone, 10-20% in the transitional zone, and remaining 5-10% in the central zone.[6] Patient age is generally not helpful as sarcomas occur over a wide age range, except for RMS and occasionally inflammatory myofibroblastic tumor (IMT) which should be considered in the differential diagnosis of patients under the age of 20 years.[7] The tumors are bulky, and patients present mainly with nonspecific symptoms such as obstructive urinary symptoms, hematuria, rectal fullness, and abnormal digital rectal examination findings. Serum PSA level is also not elevated because of their nonacinar origin. Imaging does not help much, it is neither specific for the type of lesion nor definitely depicts its malignant potential.[1,7] In our case, the patient was elderly with diffuse enlargement of prostate and clinically as well as radiologically misdiagnosed as benign nodular hyperplasia.

Table 1: WHO histological classification of prostatic sarcomas

Prostatic stromal tumors	Mesenchymal tumors
Stromal tumors of uncertain malignant potential	Leiomyosarcoma
Stromal sarcoma	Rhabdomyosarcoma
Metastatic tumors	Chondrosarcoma
	Angiosarcoma
	Malignant fibrous histiocytoma
	Malignant peripheral nerve sheath tumor
	Hemangioma
	Chondroma
	Leiomyoma
	Granular cell tumor
	Hemangioepricytoma
	Solitary fibrous tumor

WHO: World health organization

Figure 1: Gross photograph showing external and cut surface of excised prostatic biopsy

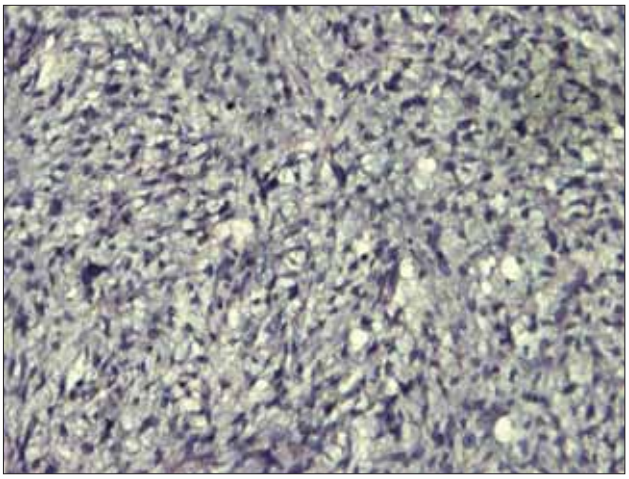

Figure 2: Photomicrograph showing spindle cell tumor with moderate to marked pleomorphism H and E, ×200

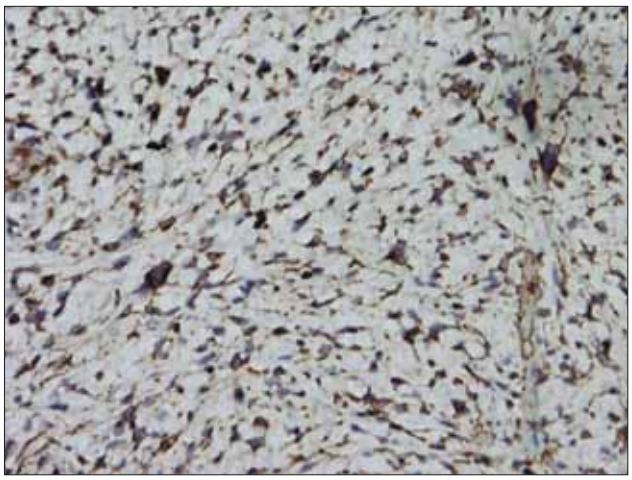

Figure 3: Tumor cells showing vimentin positivity (×400)

On gross inspection, there is wide variation in the lesional size irrespective of its benign or malignant behavior. Infiltrative borders are also not specific for malignancy as some prostatic sarcomas can have a circumscribed growth pattern while lesions with a typically benign course, such as IMT, may appear infiltrative.[7] The neoplasms with diffuse growth are often misdiagnosed and confused with diffuse benign prostate diseases such as nodular hyperplasia, nonspecific chronic inflammation, malakoplakia, and tuberculosis.[8]

Microscopically, there is overlapping of morphological features. In RMS, more than 80% are embryonal subtype making it difficult to differentiate from small cell carcinoma of prostate and lymphoma. Diagnosis is confirmed by using antibodies to muscle antigens including MyoD1 and myogenin specific for RMS. LMS consists of interlacing fascicles of spindle cells with blunt ended nuclei. Nuclear atypia, tumor necrosis, and mitosis are variable in extent. It should be differentiated from GIST of rectum with the help of c-kit and CD 34 immunostaining.[5] Stromal tumor of uncertain malignant potential (STUMP) and prostatic stromal sarcoma (PSS) of specialized prostatic tissue express progesterone receptor (PR), supporting their derivation from the hormonally responsive prostate mesenchyme.[9] Other sarcomas of prostate reported in literature include fibrosarcoma, malignant fibrous histiocytoma, angiosarcoma, and osteosarcoma, etc.[5]

The histologic subtype of prostate sarcoma appears to have significant prognostic significance amongst various tumor-related factors. The overall survival for adults with non-RMS histologies is poor with a median survival of only 2 years. The prognosis of pediatric patients with RMS is much better with a median survival of over 10 years. However, the presence of metastasis is a poor prognostic factor. In terms of treatment-related factors, surgery alone should not be the first choice as it is considered inadequate. Patients who received chemotherapy or chemoradiation adjuvant to surgery have a better survival rate.[10]

Prostate sarcomas are highly aggressive, with limited therapeutic options. Knowledge of the various lesions occurring at this site along with study of morphologic features is the most important tools in the differential diagnosis and making an early and proper diagnosis.

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