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CASE REPORT

Prostatic High-Grade Stromal Sarcoma—A Rare Encounter

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Abstract Prostatic stromal sarcoma is a rare neoplasm, and we present a case of high-grade stromal sarcoma in a 50-year-old male with initial clinical presentation of difficulty in micturition. After initial evaluation by core needle biopsy, diagnosis was established on radical cystoprostatectomy specimen based on morphology and immunohistochemistry.

Keywords High-grade prostatic stromal sarcoma · Spindle cell neoplasm · Cystoprostatectomy

Introduction

Primary prostatic stromal sarcoma is a rare neoplasm with less than 25 cases reported in the literature to the best of our knowledge [1–4].The origin of this tumour is postulated to be from the prostatic stromal cells which are the hormone-responsive mesenchymal cells. The exact histopathogenesis is unknown. It could be attributed to an epithelial mesenchymal transition process or a primary stem cell defect [5, 6]. According to the World Health Organization (WHO) classification, there are two broad categories, namely STUMP (stromal tumour of uncertain malignant potential) and stromal sarcoma which are further classified as low grade and high grade [7]. We present a case of high-grade stromal sarcoma of the prostate because of its rarity and the importance in diagnosis due to its aggressive course.

Case Findings

A 50-year-old male presented with complaints of difficulty in micturition since 2 months. Despite urethroplasty, his symptoms did not subside. The patient had a 20-year history of smoking tobacco and consumption of alcohol. There was no other relevant history and general physical examination was unremarkable. Per rectal examination revealed a prostatic mass. His haematological and biochemical tests were unremarkable. Urine examination and culture revealed gram-negative bacilli. Positron emission tomography (PET) CT whole body scan depicted grossly enlarged prostate with mild heterogenous FDG uptake suggestive of carcinoma prostate. Additionally, enlarged hypermetabolic iliac and inguinal lymph nodes identified were indicative of metastases.

The patient underwent open biopsy from prostatic lesion which was opined as mesenchymal neoplasm. Subsequently, radical cystoprostatectomy with resection of the bulbar urethra and bilateral pelvic lymph node dissection was performed. On gross examination of cystoprostatectomy specimen, bladder mucosa was unremarkable. Prostate exhibited a large globular mass measuring 9 × 9 × 7 cm; cut surface was grey white, firm with whorled and lobulated appearance, and few soft yellowish areas. (Fig. 1a, b).

Histopathologic examination depicted diffuse proliferation of spindle cells with vesicular nuclei, moderate amount of eosinophilic cytoplasm admixed with numerous

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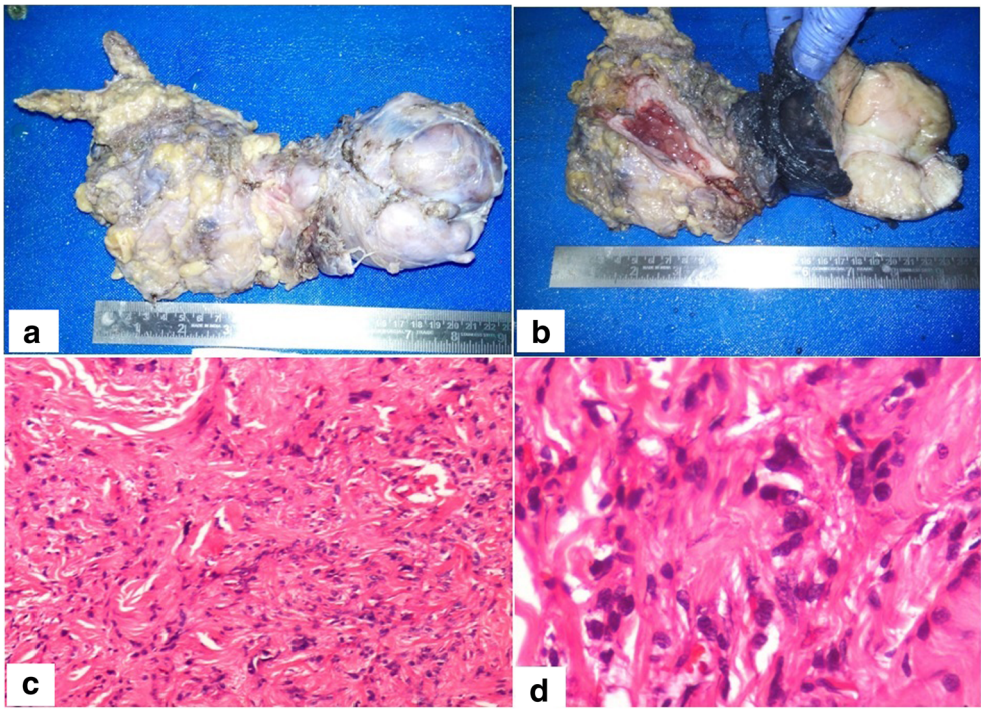

Fig. 1 **a**, **b** Gross: cystoprostatectomy specimen, cut section exhibiting unremarkable bladder. Prostate demonstrated a large globular mass measuring 9 × 9 × 7 cm; cut surface was greywhite, firm with whorled and lobulated appearance, and few soft yellowish areas. **c**,**d** Photomicrographs: diffuse proliferation of spindle cells admixed with numerous bizarre forms.**c** H&E, ×100. **d** H&E, ×400

multinucleated and bizarre forms and presence of mitotic figures. (Fig. 1c, d). Immunohistochemistry was positive for vimentin, CD34, progesterone receptor and focally for smooth muscle actin. Pancytokeratin, AMACR, h-caldesmon, myogenin, DOG-1, and CD117 were negative (Fig. 2). The expression of INI-1 was intact. A final diagnosis of high-grade stromal cell sarcoma of prostate was made.

Discussion

Prostatic stromal sarcomas are uncommon and comprise 0.7% of prostatic tumours [4]. The present case highlights the histopathological features favouring a high-grade stromal sarcoma. Gross findings in sarcomas of the prostate are variable in comparison to other mesenchymal lesions. It can manifest as a

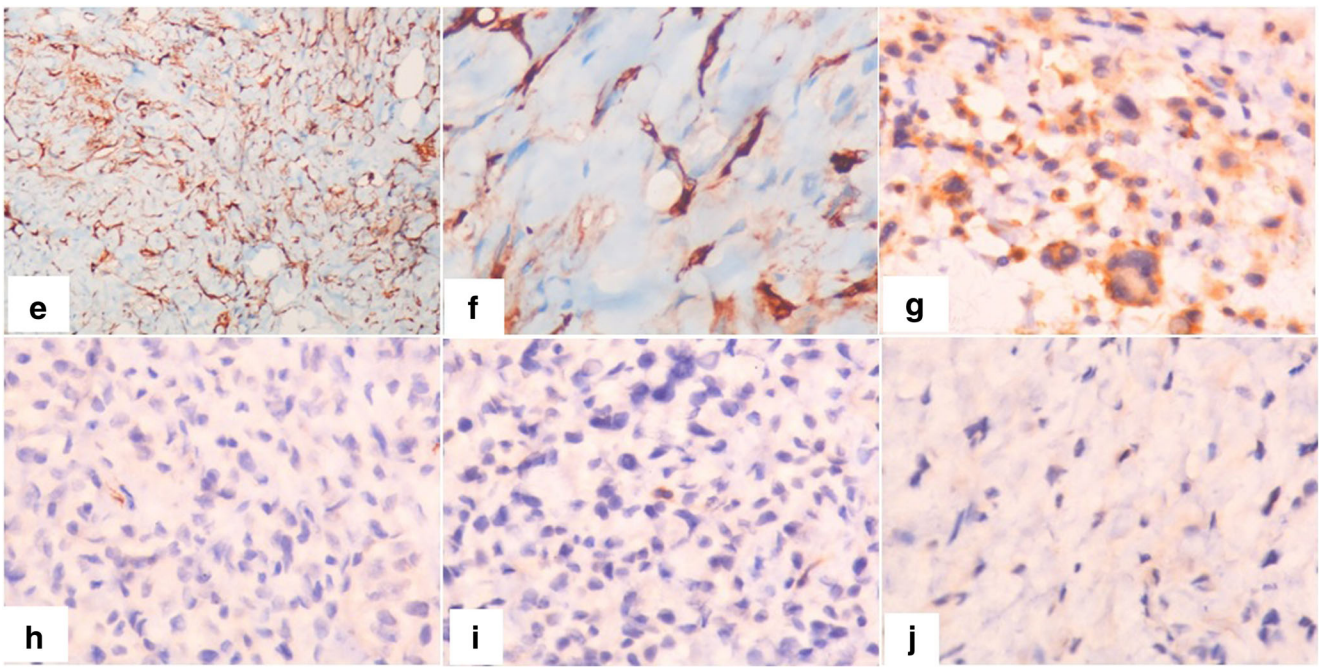

Fig. 2 **e**–**j** Photomicrographs:immunohistochemistry, **e**, **f** CD34 positive in tumour cells, ×100, ×400. **g** Smooth muscle actin positive in tumour cells, ×400. **h** Caldesmon negative, ×400. **i** DOG-1 negative, ×400. **j** S100 negative, ×400

well-circumscribed lesion in contrast to inflammatory myofibroblastic tumour which may present with infiltrative margins.

Histopathological examination is the mainstay in the diagnosis as there are several benign and malignant tumours with overlapping histopathologic features posing a diagnostic challenge. Accurate typing is essential as therapeutic and prognostic options differ. The diagnostic possibilities are many which include STUMP, inflammatory myofibroblastic tumour, solitary fibrous tumour, synovial sarcoma, leiomyosarcoma, Gastro-Intestinal Stromal Tumour (GIST), and sarcoma of myofibroblastic origin [5, 8, 9]. STUMP is a close mimic of benign prostatic hyperplasia but has a propensity to recur and generally seen in the younger population. Histologically, spindle cells with mild increase in cellularity and degenerative atypia are seen. It may be associated with low-or high-grade stromal sarcoma as demonstrated in 14% of stromal tumours of the prostate [10]. However in our patient, an associated STUMP was not identified. Low-grade sarcomas are characterised by a diffuse proliferation of spindle cells with minimal mitotic activity and absence of nuclear atypia [8]. High-grade stromal sarcomas exhibit marked cellularity associated with nuclear atypia and notable mitotic activity with or without necrosis as seen in our patient.

Close attention to morphological details in conjunction with IHC is the key to making the right diagnosis. Immunohistochemical studies in the present case supported the diagnosis of stromal sarcoma; however, it is reported that the role of IHC and other ancillary tests are of limited utility [3, 11]. There are currently no approved molecular tests, yet studies have indicated that overexpression of p53, p16, and MED12 gene mutation imply a poor prognosis. On the contrary, 17q duplication and increased PRUNE 2 protein expression have shown a favourable outcome [6].

Surgical intervention, chemotherapy and radiotherapy are the treatment modalities based on the initial presentation and stage [4, 12–14].The 5-year mean survival is 44% and is dictated by stage and metastasis. As there are no biological parameters such as elevated PSA to diagnose or monitor therapy status, prostatic sarcomas usually manifest at advanced disease process usually with obstructive symptoms and are associated with poor prognosis [6]. Absence of metastases and clear surgical margins has a positive bearing on long-term survival [12].

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