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Prostatic stromal sarcoma- A rare tumour with aggressive behaviour

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Abstract
Prostatic stromal sarcomas are extremely rare, accounting for less than 0.1 % of malignancies of prostate. They arise from mesenchymal cells and exhibit aggressive behaviour. We report a case of 55-year-old man with presented dysuria, haematuria and pain abdomen with normal PSA findings. Diagnosis of this tumour poses a challenge due to its rarity and nonspecific clinical symptoms, hence final diagnosis is made with correlation between clinical, radiological, histopathological and immunohistochemistry findings.

Keywords: Prostate specific antigen (PSA), prostatic stromal sarcomas, transurethral prostatic resection (TURP)

Introduction

Prostatic stromal sarcoma (PSS) are very rare tumours of prostate, representing less than 0.1–0.2 % of primary malignancies. These tumours arise from mesenchymal stromal cells and include stromal tumours of uncertain malignant potential (STUMP), stromal sarcomas, and undifferentiated pleomorphic sarcomas [1]. Majority of the cases are characterized by spindle cell pattern. Thus, for definitive diagnosis immunohistochemistry (IHC) and molecular studies are necessary. In the tumours of nonacinar origin the serum prostate specific antigen (PSA) level has its own limitation, because, it is typically not elevated, unless there is focus of concomitant adenocarcinoma [2]. They have a poorer prognosis in comparison with epithelial tumours [3]. We herein report a case of 55yr old male presented with dysuria, haematuria and pain abdomen.

Case report

A 55years old presented with dysuria, haematuria and pain abdomen.

On investigations PSA was 0.590ng/mL. Ultrasound abdomen showed large well defined lobulated fluid density lesion noted in pelvis of size approximately 8.9x6.5x6cms with thickened irregular walls and multiple septae. Small lesion seen adjacent to it on left side of pelvis. Impression of? Neoplastic aetiology /? Abscess was given. Cystoscopy showed distorted position of ureter and distorted lobes of prostate, possibility of? Ruptured abscess or? Sarcoma was given. CECT showed fairly well defined heterogeneously hypodense lesion near completely involving the prostatic parenchyma and bilateral seminal vesicles with enhancement pattern, central necro-cystic areas, locoregional and lymph node extension likely mitotic aetiology. Followed by radiological investigations transurethral prostatic resection (TURP) was performed and specimen sent for histopathological examination. Histopathological examination of TURP chips showed extensive areas of necrosis with few foci displaying highly pleomorphic round to spindle shaped tumour cells arranged in diffuse sheets frequent atypical mitosis, bizarre cells and tumour giant cells were noted (Figure 1 & Figure 2). On IHC Vimentin (Figure 3) and CD34 were positive.

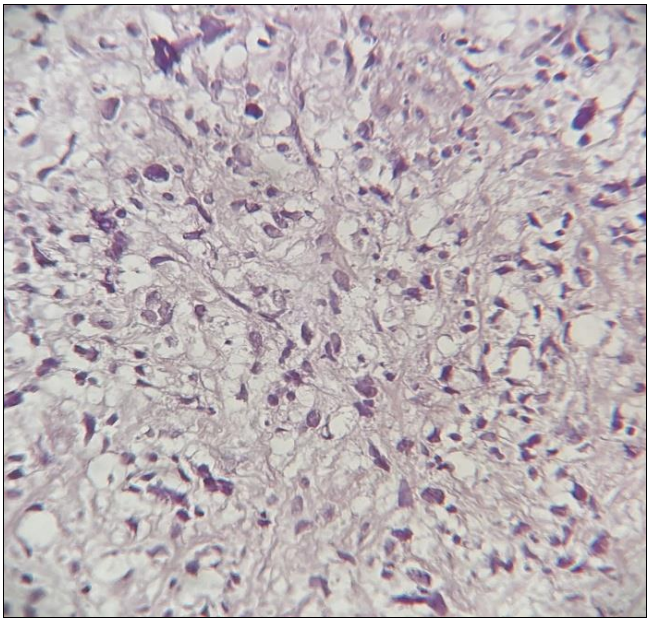

Fig 1: (H&E, 40X): Tumors cells which are round to oval, spindle shaped along with bizarre cells

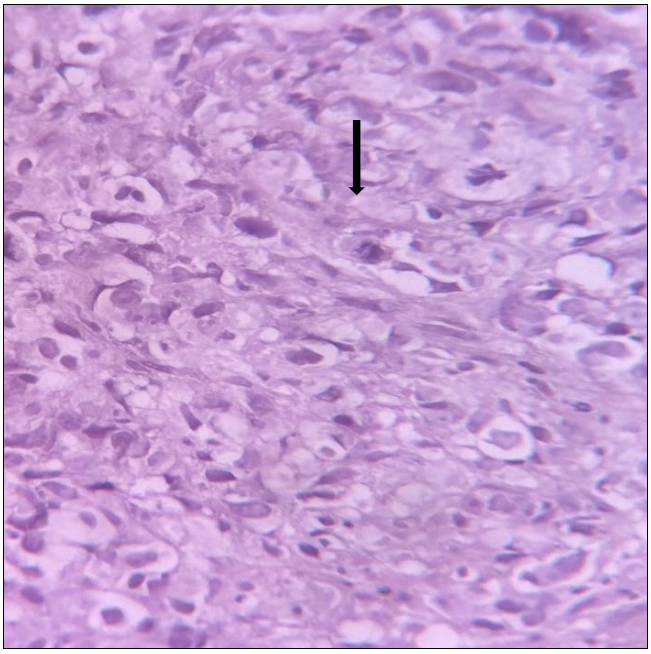

Fig 2: (H&E, 40X): Atypical mitotic figure noted amidst the tumor cells

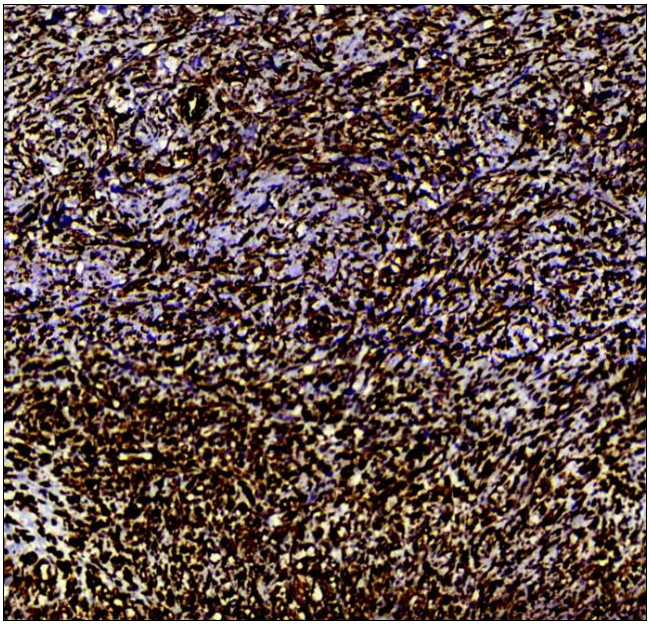

Fig 3: (H&E, 40X): Immunohistochemistry for Vimentin tumour cells show strong cytoplasmic staining

Discussion

Prostate sarcomas constitute less than 0.1% of all malignant tumours of prostate. They arise from the mesoderm in the reproductive tract [4]. Gaudin *et al*. in 1998 was first to classify prostatic sarcoma into PSPUMP (prostatic stromal proliferation of unknown malignant potential) and PSS (prostatic stromal sarcoma) based on the stromal cellularity and mitotic figures, stromal overgrowth and necrosis [5]. The first PSS case report was published in 1998, and only ~30 cases have since been reported. The age of patients with PSS ranges between 30 and 40 years, which is younger than that of prostate epithelial tumours. There are no specific clinical symptoms of PSS although the majority of patients presents with dysuria, urinary retention and hematuria [6]. Their diagnosis is rarely suspected clinically because of their rarity and the nonspecific nature of clinical symptoms [7].

Due to this lack of typical clinical symptoms, the tumour is easily overlooked or misdiagnosed as benign prostatic hyperplasia (BPH), hence in many cases the prostate was significantly enlarged when the tumour was discovered [8].

It is essential to differentiate prostate sarcoma and prostate adenocarcinoma because of the differences in their treatment and prognoses. Patients with prostate sarcoma should be treated with chemotherapy, radiotherapy and surgery, whereas management of patients with prostate adenocarcinoma requires active surveillance, hormone therapy, chemotherapy, brachytherapy and surgery depending on the grade and stage of disease. The diagnosis should be made by biopsy and there is a rare small risk of seeding along the biopsy tract with tumour cells, however, this is not a concern when biopsying prostate adenocarcinoma. Once after the diagnosis of prostate sarcoma, staging should include CT imaging of the chest and liver to look for metastasis [9].

Essential criterial according to WHO 5th edition Male Genital Tract is Diffuse stromal growth with hypercellularity, cytological atypia, increased atypical mitosis and necrosis. Desirable criteria include IHC positive for CD34 and PR [10].

The most valuable investigations to diagnose PSS involves transrectal ultrasound (TRUS), CT scan and MRI. By TRUS, the prostate appears large with irregularity and accessible tool to get a biopsy from the prostate With Immunohistochemical studies, PSS is typically positive for vimentin, CD34, and progesterone receptors. However, in few cases CD34 are negative in the literature; therefore, although immunohistochemistry is crucial for diagnosing PSS, it cannot be the sole diagnostic criterion. Furthermore, a significant increase in the Ki-67 labelling index was reported in most cases [11].

Conclusion

PSS is a rare mesenchymal tumour of the prostate with nonspecific clinical symptoms and a highly aggressive tumour. Prostatic stromal sarcoma, may pose a diagnostic challenge, due to histological overlap between them and their rarity. High-grade sarcomas display significant degrees of atypia, mitotic figures, and necrosis. IHC markers with PR, CD34, and Vimentin positivity may support the diagnosis

Conflict of Interest: Nil

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