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Case Report

Prostatic stromal sarcoma: A case report and literature review ☆

Vu-Thi Phuong, MD[a], Bui-Thi My Hanh, MD[b], Nguyen-Thi Hang, MD[a], Tran Duy Thanh, MD[c], Dau Quang Lieu, MD[d], Nguyen Minh Duc, MD[e,]*

[a] *Department of Laboratory, Pathology Division, Vinmec Times City International Hospital, Hanoi, Vietnam*
[b] *Department of Pathology, Hanoi Medical University, Hanoi, Vietnam*
[c] *Department of Anatomic Pathology- Cytology and Forensic Medicine, Vietnam- Germany Hospital, Hanoi, Vietnam*
[d] *Department of Internal Medicine, Hanoi Medical University Hospital, Hanoi, Vietnam*
[e] *Department of Radiology, Pham Ngoc Thach University of Medicine, 2 Duong Quang Trung Ward 12 District 10, Ho Chi Minh City, 700000, Vietnam*

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ABSTRACT

Stromal sarcoma of the prostate is extremely rare. In this article, we report the case of a 43-year-old male admitted to the local hospital due to dysuria. Although the pathological findings from transurethral prostatic resection showed low-grade stromal sarcoma, the surgical specimen after radical prostatectomy revealed high-grade sarcoma with hypercellularity, marked atypical spindle cells, and high mitotic activity. This case study and literature analysis aim to emphasize its rarity and raise awareness about clinical and pathological diagnosis.

Introduction

Tumors of the specialized prostatic stroma are uncommon, accounting for less than 0.1% of primary prostate cancers in adults [1]. These hormone-dependent lesions originate from specialized mesenchymal cells and have specific histological features. While they should be considered neoplasms instead of atypical hyperplasia, some follow a benign clinical course [2]. Gaudin et al. first classified stromal proliferation of the prostate in 1984 into 2 subtypes: prostatic stromal tumors of uncertain malignant potential (STUMPs) and prostatic stromal sarcomas (PSSs) [3,4]. Despite the paucity of published cases, PSSs are also divided into low and high grades based on their morphological features. Here, we describe a case of high-grade PSS that was determined from the surgical specimen after radical prostatectomy.

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* Corresponding author.
E-mail address: bsnguyenminhduc@pnt.edu.vn (N.M. Duc).

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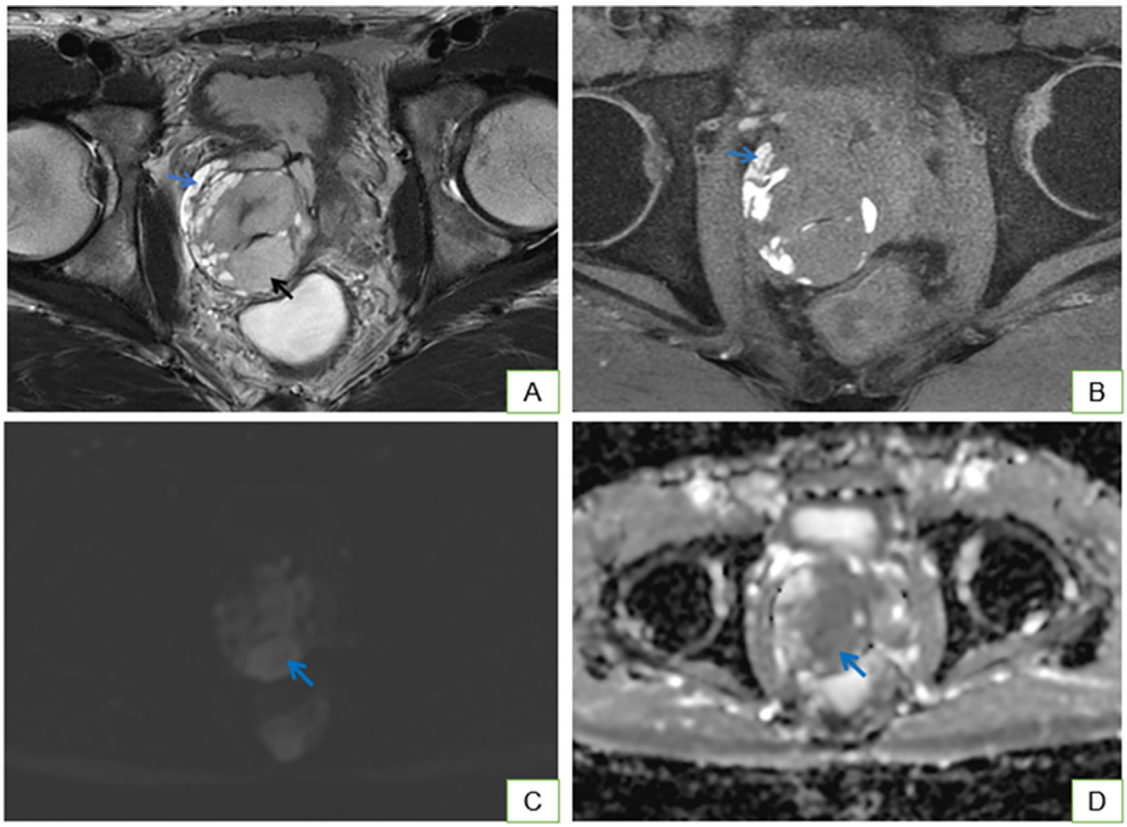

Fig. 1 – The lesion in the right part of the prostate on MRI. An axial T2-weighted image showed hyperintense cystic (blue arrow) and solid (black arrow) components (A). An axial T1 fat-saturated image indicated a hyperintense cystic component (blue arrow) suggestive of a late subacute phase hemorrhage (B). An axial (C) diffusion-weighted image (DWI) and (D) apparent diffusion coefficient (ADC) map show solid component restriction (blue arrow) with hyperintensity in the DWI and hypointensity in the ADC map.

Case report

Clinical data

A 43-year-old male presented to the nearby hospital because of dysuria. He has been having difficulty passing urine since a month ago. His past medical history was unremarkable. On physical examination, there were no abnormal findings. His blood and urine test results, including prostate-specific antigen (PSA), were in the normal range. Transurethral resection of the prostate (TURP) was performed in order to treat dysuria and collect samples for diagnosis when imaging revealed a suspicious prostatic tumor. The patient arrived at our hospital for consultation and treatment after receiving a pathological report indicating low-grade prostatic stromal sarcoma from the TURP specimen.

Imaging findings

A pelvic magnetic resonance imaging (MRI) scan showed a 5.2 × 4.9 × 5.6 cm well-demarcated, heterogeneous lesion in the right part of the prostate with cystic and solid components without any extension through the prostate capsule or bladder. Two seminal vesicles were dilated but not invaded by the prostatic lesion (Fig. 1).

Surgical pathological findings

The patient's treatment plan was discussed carefully by the hospital's multidisciplinary tumor board. With no surgical contraindications, he was advised to undergo a radical prostatectomy.

On gross examination, the prostate was 6.0 × 5.5 ×4.5 cm with 2 seminal vesicles measuring 3.5 × 1.5 × 1.0 cm and 2.0 × 1.5 × 1.0 cm. The surface was inked with different colors (right by black; left by blue; anterior stripe by yellow; posterior stripe by orange) (Fig. 2). After sectioning the surgical margins and seminal vesicles, the remaining specimen was cut serially into 0.3 cm-thick sections from the apical side to the basal side. The tumor was 4.0 × 3.5 × 2.5 cm in size, located mainly in the right and partly in the left lobe with an irregular border, tan-yellow cut surface without necrosis or invasion into the urethra or seminal vesicles.

Microscopically, the spindle cell tumor developing from the mesenchymal component was diverse in appearance, with both low-grade and high-grade regions. The surgical slides showed high-grade features, that were absent from

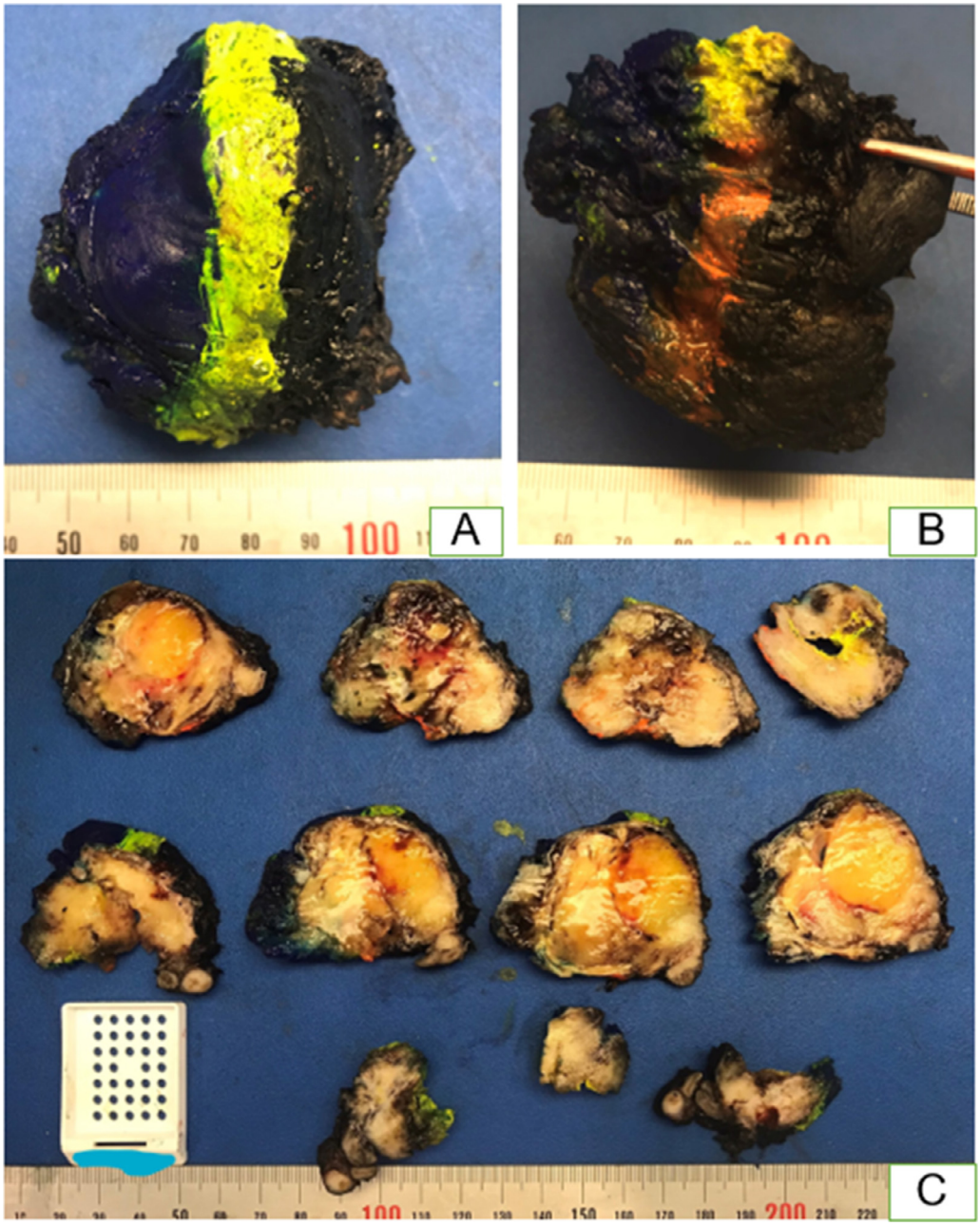

Fig. 2 – Macroscopic features from the prostatectomy specimen. Inked anterior prostate (A). Inked posterior prostate (B). The cut surface of the tumor was tan-yellow, with an irregular border, measuring 4 cm in the largest diameter (C)

TURP slides, including hypercellularity, atypical nuclei, and 14 mitoses per 10 high-power fields. However, there was no evidence of necrosis. The tumor was confined to the prostate with no extension into other surrounding organs. Immunohistochemical staining showed focal or weak positivity for cluster of differentiation 34 (CD34), progesterone receptor (PR), and desmin (DES). These histopathological and immunohistochemical findings are consistent with a high-grade PSS (Figs 3 and 4).

After radical prostatectomy, the patient made a good recovery, and the subsequent follow-up revealed no signs of recurrence.

Discussion

Prostate sarcomas account for less than 0.1% of all malignant prostate tumors [3]. They are believed to arise from the mesoderm in the reproductive tract [1,4]. In the past, these tumors have been referred to using several names, including atypical stromal hyperplasia, phyllode type of atypical stromal hyperplasia, and cystic epithelial-stromal tumors [5].

To our knowledge, the first PSS case report was reported in 1998, and only around 30 cases have subsequently been published in English [1,4]. The majority of patients were diagnosed

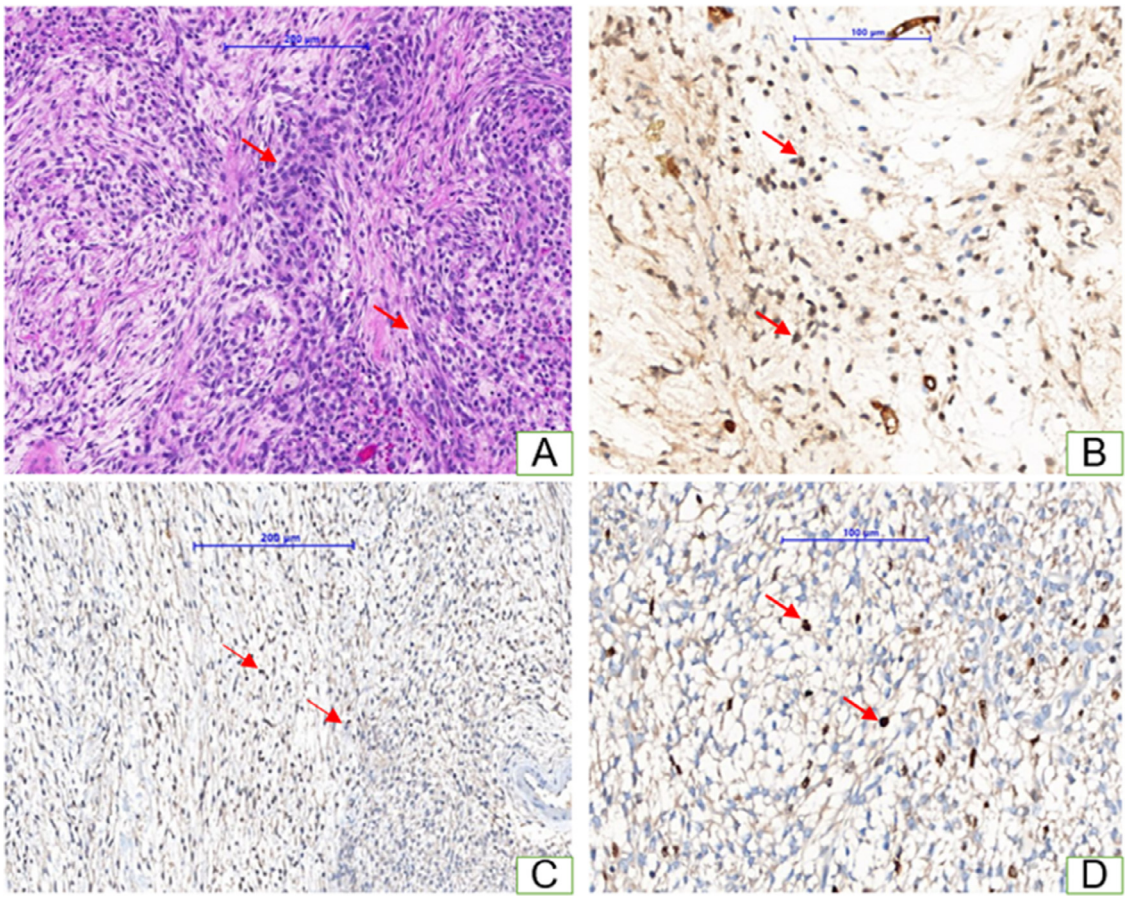

Fig. 3 – Microscopic features from the transurethral resection (TURP) specimens. The medium magnification showed the proliferation of spindle cells with mild atypia (red arrows) and rare mitotic activity (A. HE x 100). The tumor cells were focally positive for CD34 (B. CD34x200, red arrows), PR (C. PR x 100, red arrows), and revealed the low index proliferation (D. Ki67 x 200, red arrows).

at a mean age of 58 years, ranging from 27 to 83 years [6]. The most common presenting symptom is urinary tract obstruction, followed by hematuria and dysuria. There is no predilection for any prostatic zone. Also, the plasma PSA level is usually normal. Because there are no specific symptoms and only a limited number of indicators, the definitive PSS diagnosis is frequently reached after a TURP or open prostatectomy [3,7–9]. No standard treatment protocols have been released owing to the rarity of this tumor. Metastasis to the lung and bone is often observed in the aggressive course. The prognosis is generally poor, with only 10% of patients living longer than 3 years [3].

Gaudin et al. [3] divided prostatic stromal tumors into 2 subtypes (STUMPs and PSSs) based on the extent of cellularity, mitotic figure, necrosis, and stromal overgrowth. STUMPs mostly do not show metastatic behavior, and prostatectomy with clear margins is curative while PSSs may need adjuvant therapy in cases of high-risk features [7]. However, STUMPs occasionally recur after resection, possibly related to PSS at the time of diagnosis or progression to PSS [6].

In terms of radiology, there has only been limited published data regarding computed tomography (CT) and MRI features of PSS. Most cases described PSS as a solitary, hypervascular, heterogeneous pelvic mass. PSS may show a slight enhancement on CT and a slightly hypointense mass on the axial, unenhanced, T1-weighted MRI. Hypodense or cystic areas on radiological images usually correspond with necrosis or degeneration on microscopic examination [10].

With regard to pathology, PSS may measure from 2 to 18 cm on gross examination and they are not related to any clinical behaviors [7]. Microscopically, PSSs exhibit the proliferation of spindle or oval stromal cells with a variety of growth patterns, including storiform, fascicle, rhabdoid, and phyllode-like [11]. PSSs are subdivided into low-grade and high-grade based on the degree of atypia, mitotic activity, and necrosis. High-grade PSSs reveal the characteristics of significant atypia, increased cellularity, frequent mitoses, and necrosis [12]. Nevertheless, no existing criterion is known to have a distinct cutoff separating low-grade and high-grade PSSs. In our case, the TURP specimens displayed low-grade features, with tumoral cells exhibiting minor atypia, a very low mitotic index, and no signs of necrosis. The prostatectomy specimen did not have any evidence of necrosis, but other histological findings, such as substantial atypia and mitosis, were consistent with a high-grade sarcoma.

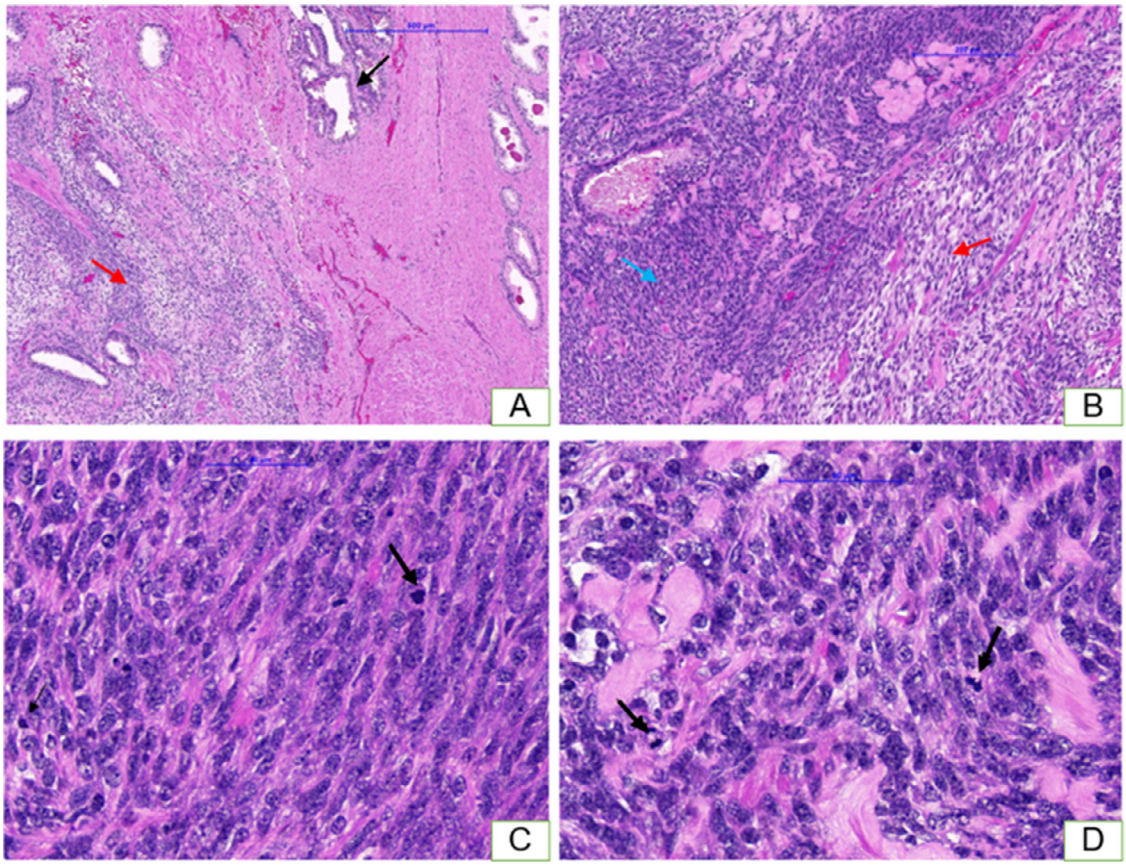

Fig. 4 – Microscopic features from the surgical (prostatectomy) specimen. The low-grade stromal tumor (the lower left, red arrow) and adjacent benign prostatic tissue (the higher right, black arrow) were observed at low magnification (A. HEx50). Both the high-grade area (the left side, blue arrow) and the low-grade area (the right side, red arrow) were seen in the same area (B. HEx100). The high magnification showed the features of a high-grade tumor with hypercellularity, markedly atypical spindle cells and mitotic activity (C and D. HE x40, black arrows).

Immunohistochemically, the spindle tumor cells usually express PR, CD34, and vimentin (VIM). The PR positivity appears to favor the tumor originating from hormonally responsive mesenchymal cells in the prostate. DES and alpha-smooth muscle actin (αSMA) may show variable reactivity. This tumor does not express S100 proteins, KIT proto-oncogene receptor tyrosine kinase (*KIT/CD117*), or the estrogen receptor (*ER*). Ki67 levels are often high in high-grade PSS but cannot distinguish STUMP from low-grade PSS [1,11,12]. Our immunohistochemical results showed positivity for PR, CD34, and DES with variable expressions, supporting the PSS diagnosis.

Due to the variations between treatment and prognosis, differentiating between PSS and other prostatic disorders plays an important role. The primary differential diagnosis for PSS is STUMP. On MRI, both tumors could have cystic and solid components, but necrosis, cystic change and hemorrhage are more likely to occur in PSS because of its rapid growth. Although our case revealed heterogeneous features, it is difficult to diagnose PSS based on MRI only because the nonsolid component was not predominant and the tumor did not invade the surrounding tissue. Using immunohistochemistry to distinguish PSS and STUMP is challenging because their expressions are similar. On H&E slides, STUMPs have few or no mitotic figures, minimal atypical nuclei, and no necrosis. Nevertheless, STUMP and low-grade PSS may be undistinguishable, especially on limited specimens. The most important indicator of malignancy on biopsy is the presence of a single atypical mitosis. Low-grade sarcoma is sometimes diagnosed by local infiltration or an atypical mitotic figure, regardless of its relatively benign histological appearance at low magnification [11]. The hypercellularity, nuclear pleomorphism and high mitotic activity in our case help rule out the possibility of STUMP and low-grade PSS.

There are also some other differential diagnoses. Adenocarcinoma, the most common malignant tumor of the prostate, also needs to be excluded. Adenocarcinoma usually arises in the peripheral zone of the prostate with increased PSA while stromal sarcoma may originate from any zone with normal PSA. This is because stromal sarcoma develops from nonepithelial cells, whereas epithelial cells produce PSA. On MRI, prostate adenocarcinomas are frequently multifocal as ill-defined regions with homogeneously low T1 and T2 signal intensities while sarcoma is characterized by a heterogeneous appearance. In contrast to most prostatic adenocarcinomas, which are organ-confined at the time of diagnosis,

prostatic sarcoma usually spreads beyond the prostate and invades surrounding organs [13]. Although the tumor in our case did not infiltrate outside the prostate, a large lesion occupying nearly all of the prostatic zone and the cystic change in the MRI did not favor adenocarcinoma. Histopathological findings help confirm the diagnosis since there is obviously a distinction between adenocarcinoma and sarcoma. In addition, benign prostatic hyperplasia and other mesenchymal prostatic tumors, such as solitary fibrous tumors, smooth muscle tumors, and gastrointestinal stromal tumors, were also ruled out in our instance based on the combination of both radiological, histopathological and immunohistochemical features.

Surgery is still the mainstay treatment modality with the aim of removing the entire tumor with a free margin. The role of adjuvant chemotherapy is currently debatable due to a paucity of data and scant clinical evidence. In high-risk sarcomas (large tumor size, high grade, R1 or R2 margin), adjuvant radiotherapy may be beneficial. Despite the high-grade of PSS, follow-up was used in our patient instead of adjuvant therapy because of other considerations such as being limited to prostate, having an R0 margin and the patient's expectations as well [14].

Conclusions

Stromal tumors are rare mesenchymal tumors of the prostate with nonspecific clinical symptoms for which resection is the primary treatment. It is divided into STUMPs and PSSs, which are difficult to distinguish in some circumstances. High-grade sarcomas show significant degrees of atypia, mitotic figures, and necrosis. PR, CD34, and VIM positivity may support the diagnosis.

Authors' contribution

Vu-Thi P, Bui-Thi MH, and Nguyen MD: Case file retrieval and case summary preparation. Vu-Thi P, Bui-Thi MH, and Nguyen MD: preparation of manuscript and editing. All authors read and approved the final manuscript.

Availability of data and materials

Data and materials used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2023.05.077.

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