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Case Report

Magnetic resonance imaging presentation of prostatic stromal sarcoma ☆,☆☆

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ABSTRACT

Prostatic stromal sarcoma (PSS) is a rare malignant tumor that often occurs in young people. Despite the fact that their clinical pathological characteristics are well known, our understanding of the imaging characteristics still seems to be limited. In our search of the literature, PSS articles are mainly present as case reports. In this case series of PSS, we aimed to thoroughly describe the magnetic resonance imaging (MRI) features, histopathological findings, and distinguishing hallmarks from prostate cancer (PCA).

Introduction

Prostatic stromal sarcoma (PSS) is a rare malignant tumor that accounts for 0.1%–0.2% of primary prostate malignancy [1,2]. In 1998, Gaudin et al. first classified stromal proliferation into 2 groups: stromal tumors of uncertain malignancy potential (STUMP) and PSS [4,5]. Most PSS is well-known to originate from the mesenchymal tissue but there is a minority of them that can arise from the stromal components of the prostate gland. Despite a small number of published cases, PSS is also divided into low and high grades based on their morphological features. Furthermore, this neoplasm can be subdivided into several subtypes like leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, and unclassified sarcomas. Rhabdomyosarcoma is the most common form, accounting

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for 42% of PSS, and occurs mainly in children and adolescents [3,4]. Leiomyosarcoma accounts for 25% of PSS and occurs primarily in adults (40-78 years) [3]. Other subtypes are less likely to be mentioned.

Clinical symptoms of PSS are nonspecific and depend on the degree of invasiveness and mass-effect of lesions, including dysuria, urinary retention, hematuria, and abdominal pain. The imaging characteristics of these tumors are rarely noted in the literature and mainly appear in case reports. However, imaging features have an essential role in disease management and treatment. In this article, we report 2 cases of PSS examined by magnetic resonance imaging (MRI) to characterize features that may help distinguish them from prostate cancer (PCA).

Case presentation

Case 1

A 15-year-old male patient came to the hospital because of dysuria, hematuria, urinary retention, and urinating twice a night, which gradually increased over 2 months. He denied any history of trauma or other medical problems. On physical examination, the patient had mild abdominal distention, bilateral inguinal lymph nodes, and the digital rectal figured out a hard mass in the prostate area. Prostate-specific antigen (PSA) value was unremarkable (free PSA: 0.063 ng/mL, total PSA 0.344 ng/mL); Blood analysis showed normal white blood

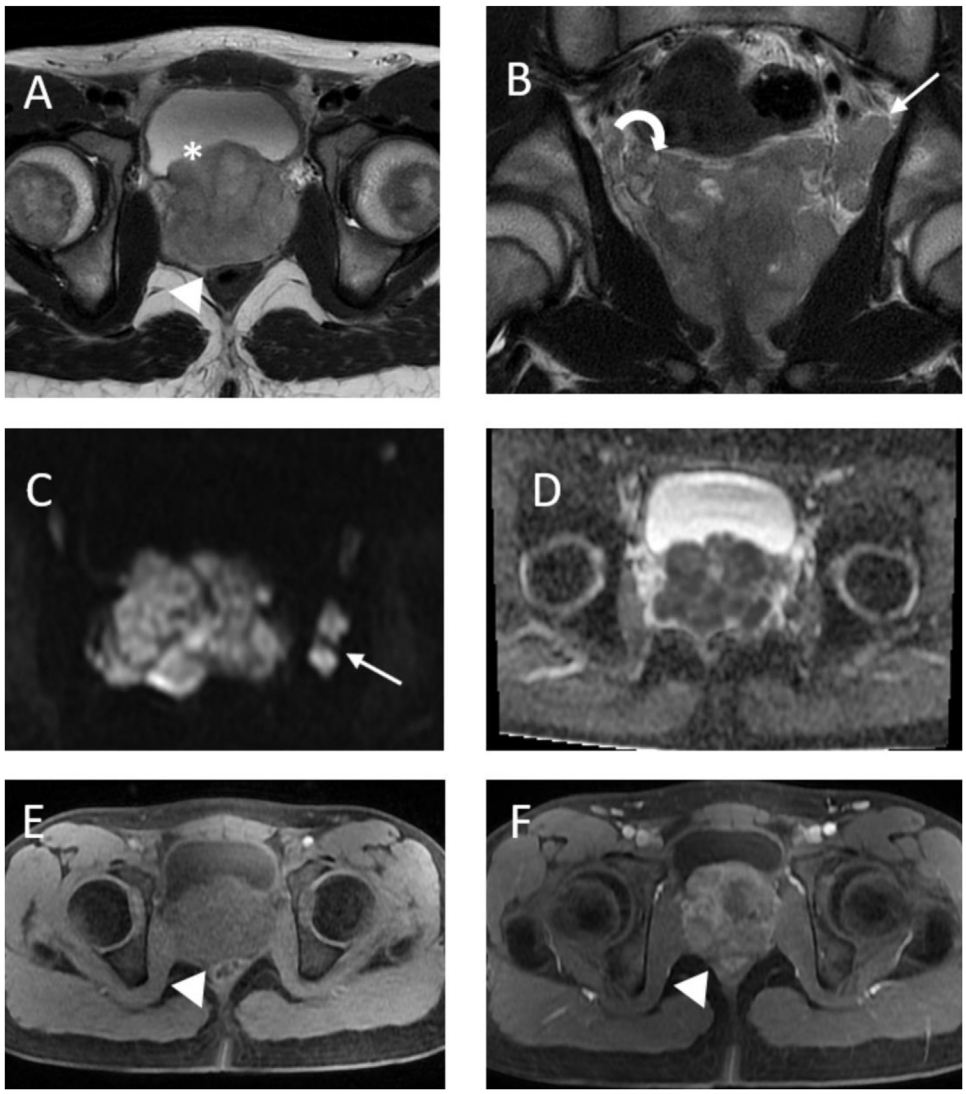

Fig. 1 – Contrast-enhanced pelvis MRI: (A) axial T2WI, (B) coronal T2WI, (C and D) DWI/ADC, (E and F) T1WI pre- and postcontrast. Enlarged prostate with whole parenchyma is heterogeneous hyper-signal on T2WI (A, B), hypo-signal on T1WI (E), restricted diffusion on DWI/ADC (C and D), postcontrast sequence (F), irregular rough surface, bilateral seminal vesicles invasion (curved arrow) and bladder neck (asterisk) compression and the rectum is pushed backward (triangle—A, E, F). Right external pelvic, left internal pelvic and left inguinal lymph nodes (straight arrow—B, C).

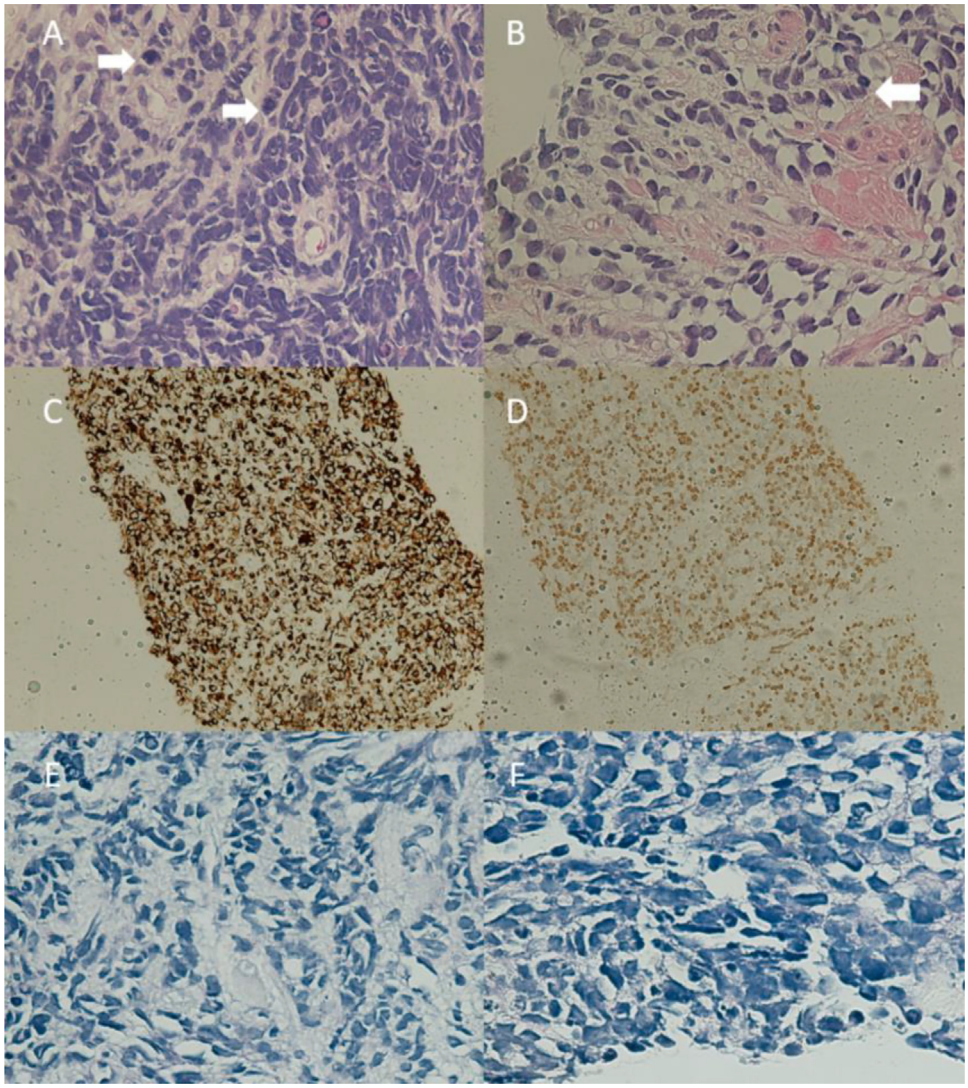

Fig. 2 – Histopathological images and immune-histochemical examination result. (A)The hyper-cellular tumor tissues, not observed normal prostate tissue (H&E, 5X). (B) At high magnification (40X) the tumor cells show a rhombic or oval nucleus with a high proportion of divided nuclei (arrows- A, B) and mucous fibrous stroma. Tumor cells are arranged into rafts, bands, or clusters that invade the stromal tissue. Tumor cells react positively with markers Desmin (C) and Myogenin (D), negative with markers CKAE1/AE3 (E) and TLE1 (F).

cell (WBC): 6.9 g/L (Neutrophilia 50.5%), normal Hemoglobin: 155 g/L, all other blood routine indexes were within the normal limits.

Pelvic transabdominal ultrasound revealed the enlarging prostate with an irregular rough surface, asymmetry, and loss of boundaries between regions. The whole parenchyma was heterogeneous, hypoechoic, and hypervascularity on Doppler mode. Notably, the mass demonstrated several signs of unclear border with surrounding tissues, bilateral seminal vesicles, and bladder neck wall, which is consistent with prostate cancer.

On pelvic MRI, the prostate gland weighs about 107 grams, nondistinctive between the peripheral zone and transitional zone. The lesion is hyper-signal on T2-weighted images (WI) (Figs. 1A and B), restricted diffusion on diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) (Figs. 1C and D) (suggest high cellularity) and heterogeneous enhancement on postcontrast sequence (Fig. 1F). The mass invades bilateral seminal vesicles and the bladder neck, pushing the rectum backward. MRI also detected several lymph nodes in the bilateral pelvis and left inguinal region (the largest 15 × 20 mm) with an irregular rough surface and heterogeneous enhancement on the postcontrast sequence. The aforementioned features strongly raise concerns about PSS invading surrounding structures and metastasis to the bilateral pelvis and left inguinal lymph nodes.

Transrectal ultrasound-guided 12-core prostate biopsy results are rhabdomyosarcoma prostate (Fig. 2). The patient is subsequently managed by chemotherapy.

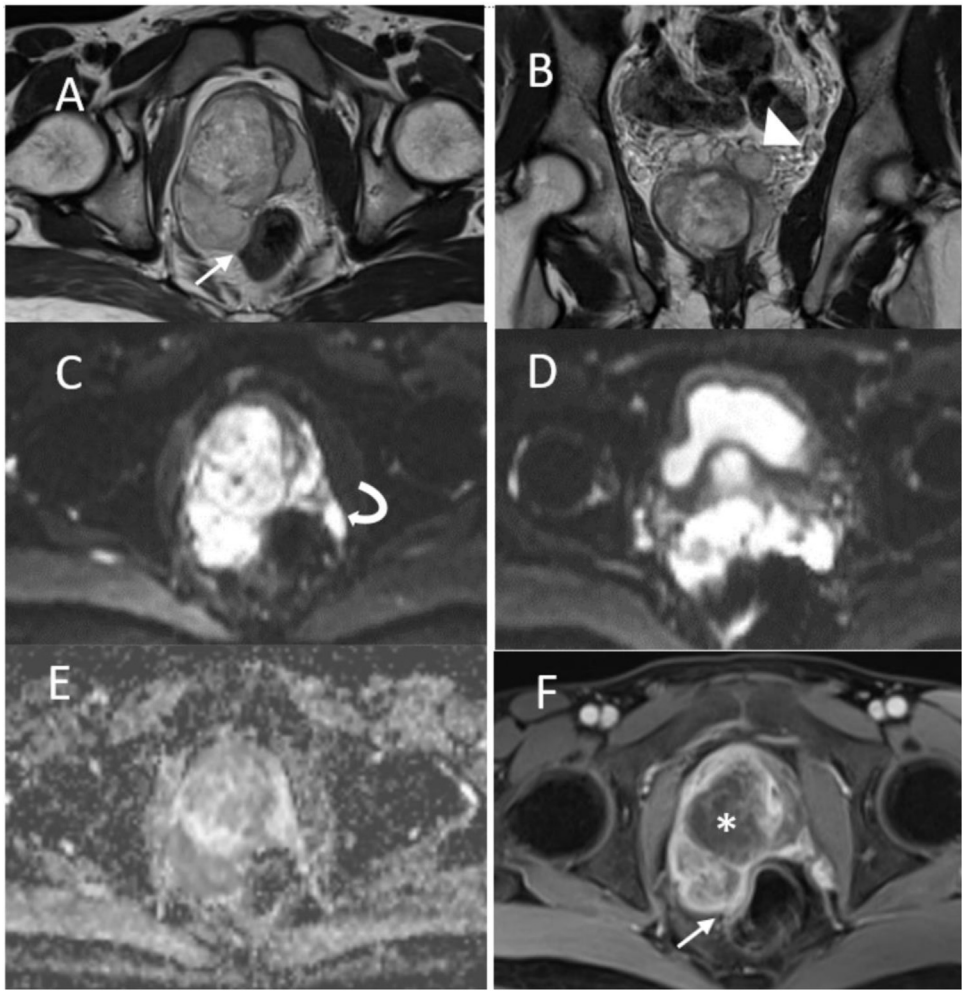

Fig. 3 – Contrast-enhanced pelvis MRI: (A) Axial T2WI, (B) Coronal T2WI, (C and D) DWI, (E) ADC, (F) T1WI postcontrast. The mass is on the right lobe of the prostate, hyper-signal on T2WI (A, B), loss of boundaries between the peripheral zone and transitional zone, uncontinuous capsule of the prostate gland, invading and pushing the rectum to the left (A, F - straight arrow). The neoplasm is hyper-signal intensity on DWI (C, D), hypo-signal on ADC (E), peripheral enhancement postcontrast sequence(F), with a nonenhanced center (asterisk). The mass invades the seminal vesicle (B- triangle) and several pelvic lymph nodes (C - curved arrow).

Case 2

A 31-year-old male came to the hospital because of dysuria, which gradually increased over 3 months. He denied any history of trauma or other medical problems. The patient had no abdominal pain or lymph nodes in the inguinal region on the physical examination. The digital rectal examination figured out a hard mass on the right of the prostate area. PSA value was unremarkable (free PSA: 0.09 ng/mL, total PSA 0.45 ng/mL); Blood analysis was within normal limit, WBC: 8.2 g/L (Neutrophilia 60%).

A transrectal ultrasound revealed that the prostate gland weighs about 100 grams with an irregular rough surface, heterogenous echogenicity, indistinct boundaries between regions, and hypervascularity on Doppler mode.

The prostate on pelvic MRI is enlarged with a mass on the right lobe, which is hyper-signal on T2WI, restricted diffusion on DWI/ADC, and peripheral enhancement on the postcontrast sequence. The boundaries between the peripheral and transitional zones, the prostate capsule, and the rectum walls are all blurred (Fig. 3). This neoplasm also invades bilateral seminal vesicles with several pelvic lymph nodes.

Thoracic and abdominopelvic computed tomography (CT) scans highly recommended liver metastasis (Fig. 4A), bone metastasis (Fig. 4B), multiple lung metastases (Fig. 4C).

The patient was scheduled for surgery to remove the tumor. Macroscopically, the large mass has extended through the prostatic capsule, invading the bladder neck, and rectum. The patient underwent radical prostatectomy and cystectomy with cutaneous ureterostomy. Histopathological result: Rhabdomyosarcoma prostate (Fig. 5).

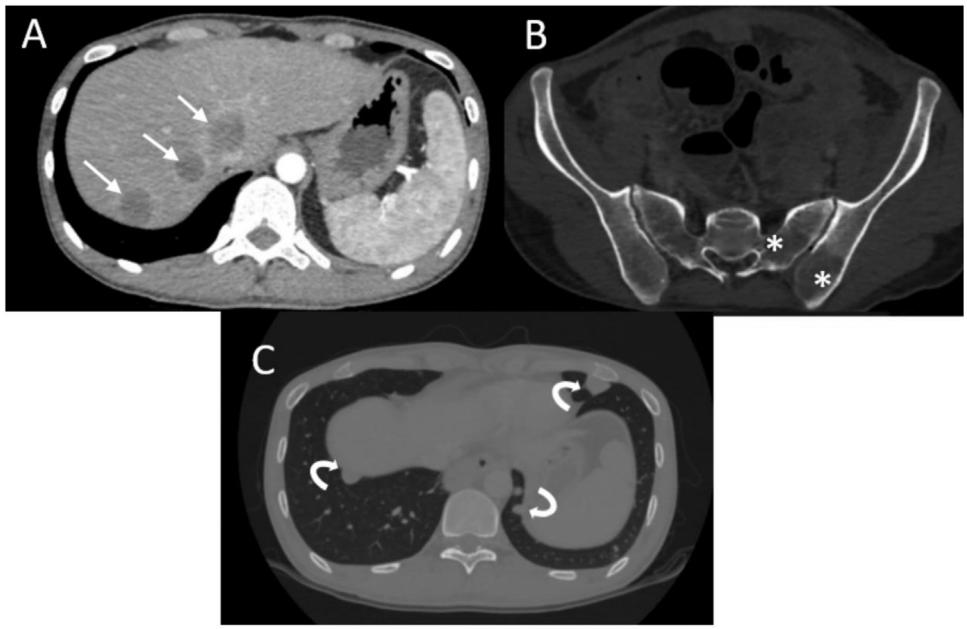

Fig. 4 – Abdominopelvic CT scan, axial abdominal window (A), axial bone window (B) and pulmonary window (C), several liver metastases (A- straight arrow), bone metastasis (B – asterisk), pulmonary metastasis (C- curved arrow).

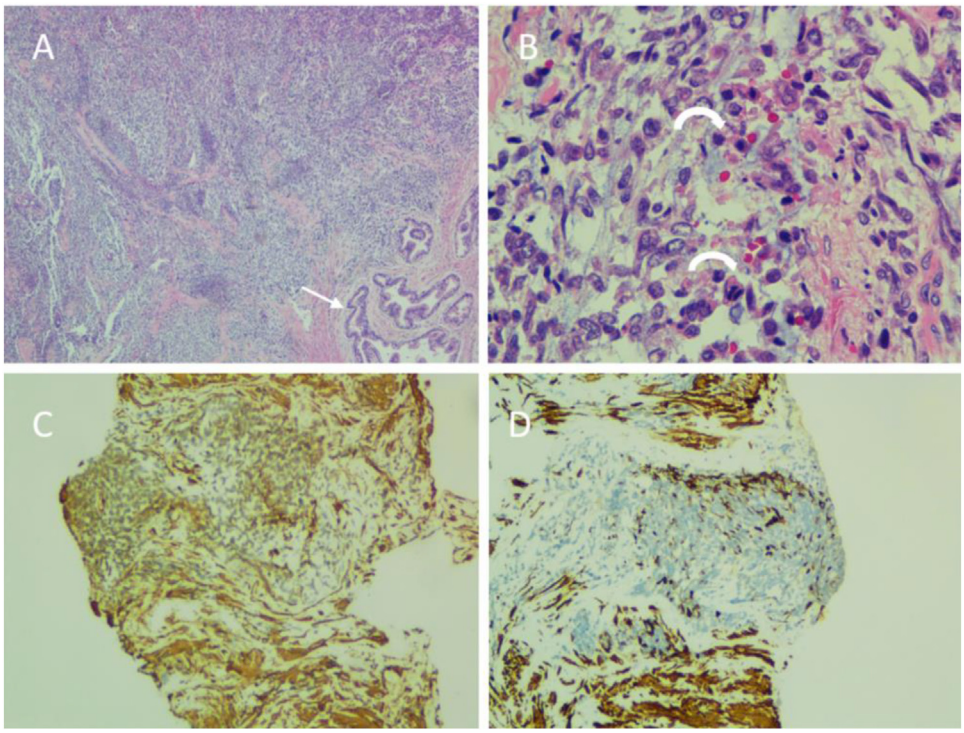

Fig. 5 – Histopathological images and immune-histochemical examination result. (A)The hyper-cellular tumor tissues, take note that the lower right corner (straight arrow) is residual normal prostate tissue (H&E, 5X). (B) At high magnification (40X) the tumor cells show a rhombic or oval nucleus with a high proportion of divided nuclei (curved arrows) and mucous fibrous stroma. Tumor cells are shown to express smooth muscle actin (SMA) (C) and Desmin marker (D).

Discussion

PSS is an atypical neoplasm encompassing 2 common subtypes: rhabdomyosarcoma (42%) and leiomyosarcoma (25%). While PCA is commonly present in older adults, PSS tends to onset in young people. Leiomyosarcoma occurs primarily in adults between 40 and 78 years of age, while rhabdomyosarcoma is more likely to occur in children and adolescents.

Symptomatic PSS is often large in size at the time of diagnosis. In a study of 25 patients, the average tumor size was 9.5 cm [6]. Due to its exceeding shape, radiologically distinguishing between PSS and bladder tumors can be difficult. Our 2 prostate tumors weighed about 100 and 107 grams. Moreover, PSS usually rapidly grows and invades adjacent structures. Therefore, local symptoms such as urination disorders are the most common symptom, which accounts for 72% [6]. Both of our patients came in with dysuria and frequent urination. The second patient also had hematuria due to the tumor invading the bladder neck. However, none of the aforementioned findings are specific to PSS [3]. Serum PSA values are usually normal, which is unsurprising as this entity has a nonepithelial origin [3]. Hence, this feature is valuable in distinguishing PSS from PCA. Both of our patients had total and free PSA within normal limits, which helped to navigate our preoperative diagnosis toward PSS.

Due to closer access, transrectal prostate ultrasound is more advantageous in high-resolution images than abdominal ultrasound. PSS lesions on ultrasound often have great size, abnormal structure, irregular margins, hypoplasia, extracapsular component, and hypervascular on Doppler mode. Nearby metastatic nodes can also be observed.

Tumor shapes may vary from rounded, lobular, or irregular and indistinct lesions. Both cases in this report present heterogeneous masses with irregular margins, occupying the vast majority of the prostate gland and losing their normal anatomical structure (as seen on MRI). The extension beyond the cortex of the tumor was also seen in both cases of our report but can be found in only 35% of PCA [1]. Despite its locally invasive features, PSS can also exert significant mass effects on nearby structures such as the bladder and rectum. PCA, on the other hand, purely develops invading behavior.

MRI helps determine the location, local invasion, tumor characteristics, lymph node metastasis, and surgical planning. Prostate MRI protocol includes T1WI, T2WI, DWI, ADC maps, and dynamic contrast-enhanced (DCE) imaging to provide both imaging and functional information about the prostate [7]. Most PSS appear as homogeneous hypo-signals on T1WI and multiple heterogeneous hyper- or iso-signal regions on T2WI. This imaging feature on T2WI differs from PCA lesions, which are usually distinct hypo-signal nodules on T2WI. Our 2 cases showed diffuse heterogeneous signals. Necrosis and cystic changes in PSS are common due to their high malignancy and rapid growth rate [1,7]. On the MRI, the most common manifestations of PSS are a rim-like hypo-signal "capsule" on T2WI, which may be complete or incomplete. However, PSS does not contain fibrous capsules on histopathologic examination, so the term "pseudocapsule" may be more appropriate for the abrupt transition between tumor and background prostate tissue [1]. This PSS's imaging features have been previously described in the literature and viewed at the junction between this neoplasm and the pelvic fat layer on the upper and back of the mass [5]. The pseudocapsule results from the interference between the tumor that has spread beyond the prostate gland and the compressing soft tissues nearby. Despite its locally invasive nature, PSS often presses on nearby tissues or organs, as opposed to being directly invasive only in PCA [3]. The image of the 2 tumors above shows pressure on the rectum backward to the left and pushing the bladder forward (no fat layer in front of the bladder). PSS is rarely confused with PCA, which usually extends by infiltrating its adjacent tissues, so it is commonly ill-defined without a capsule on T2WI. However, leiomyosarcomas and rhabdomyosarcomas are more infiltrative than other types of PSS, perhaps with poorly defined tumor margins on MRI [8]. Contrast-enhanced MRI was performed on 2 patients in this report. In the second patient, the tumor was strongly enhanced on the periphery. This sign can be explained by histological findings, which show that the central component of the lesion consists of myxoid and hyalinated substances. Other studies show that heterogeneous contrast enhancement of PSS, reflecting the heterogeneous composition of the lesion, which may be solid or mixed, with areas of cystic and necrotic changes in the tumor, is consistent with the enhancement in the first patient. Therefore, the PI-RADs assessment may not be suitable for most cases of PSS [1].

The differential diagnosis of PSS includes PCA, benign prostatic hyperplasia (BPH), and prostate abscess, which share similar clinical symptoms but different imaging features [9]. Firstly, PCA often occurs in the periphery of the prostate, with low density, irregular enlargement, and strongly enhanced postcontrast. On MRI, PCA has a hypo-signal area on T2WI in the periphery (normal hyper-signal), while PSS is heterogeneously hyper-signal [7]. The central and transition regions of the BPH proliferate, flattening the thin periphery of the prostate and leaving clear boundaries between the regions. In addition, pseudocysts can be seen in the enlarged area. For prostate abscess, the fluid in the prostate abscess and the internal walls have decreased signal on T1WI, while the abscess wall has increased signal. The abscess has increased signal on T2WI, and the capsule and inner septum strongly enhanced on postcontrast. Clinical features should also be considered, PCA and BPH usually occur in older people with markedly increased PSA. In contrast, PSS occurs mainly in young people with normal PSA, and patients with prostate abscesses often have fever and chills.

The treatment of PSS includes surgery, radiotherapy and chemotherapy [9,10]. Surgery alone is not enough for long-term survival. Long-term survival, as well as freedom from local recurrence, is associated with clear surgical margins and the absence of metastases at diagnosis [11]. As soon as there is a recurrence, even if the risk is low, treatment should be aggressive [12]. 100% of cases with highly malignant tumors relapse and also have a high ability to metastasize [11]. Adjuvant radiotherapy may be beneficial for high-risk sarcomas (large tumor size, high malignancy) [3].

The main sites of distant metastases of PSS are the lungs, bones, and liver, and bone metastases are usually osteolytic [1,3]. In contrast, PCA metastases mainly occur in bone and are generally osteoblastic. Our second patient satisfied the above location, including pulmonary, hepatic, and osteochondral metastases. One series reported that 62% of patients had

distant metastases at diagnosis [8]. Five-year overall survival ranges from 11.3% to 38% [12]. The prognosis is generally poor, with only 10% of patients living longer than 3 years [4]. The median survival time in another study was about 23 months [6]. Rhabdomyosarcoma subtypes have a more invasive nature. The prognosis is not good and requires more aggressive chemotherapy [1]. Poor prognostic factors include age of >50, metastases, and lack of surgery [12].

Conclusion

PSS is a rare malignant tumor. Even though PSS's imaging characteristics often vary, they typically appear as large diffuse heterogeneous prostatic masses extending to the peripheral structures, heterogeneous hyperintensity on T2W images, restricted diffusion, peripheral and heterogeneous enhancement on MRI. In addition, a normal PSA index and young age patient are also characteristics that can help suggest prostate sarcoma rather than adenocarcinoma.

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.

Author contributions

Nguyen LH and Nguyen MD: Case file retrieval and case summary preparation. Nguyen LH and Nguyen MD: preparation of manuscript and editing. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Our institution does not require ethical approval for reporting individual cases or case series. Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Consent for publication

Not applicable.

Patient consent

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

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