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CASE REPORT

An Unusual Case of Primary Prostatic Extragastrointestinal Stromal Tumor

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Extragastrointestinal stromal tumors (EGISTs) originate from several anatomic sites, except the digestive system. We describe an early prostatic EGIST case with lower urinary tract symptoms and a palpable lesion on digital examination. The patient received transrectal ultrasonography-guided biopsy of the prostate, and the pathological finding demonstrated positive staining for DOG-1 and CD117. Treatment included only imatinib, which shrank the tumor and led to complete remission. Because of enlarged prostate volume with tumor progression, the initial common manifestations are lower urinary tract symptoms. Surgical resection is the recommended management, particularly in bulky lesions. However, imatinib still plays an important role in long-term therapy. We also review the associated reported cases of prostatic EGISTs.

Key words: Extragastrointestinal stromal tumor, prostate, imatinib

INTRODUCTION

Gastrointestinal stromal tumor (GIST) is a type of soft tissue tumor occurring primarily in the digestive system. GISTs arising from the outside of the alimentary tract are grouped into extragastrointestinal stromal tumors (EGISTs), and prostate is a rare presentation site.[1] The reported EGISTs are diagnosed by immunohistochemical staining, including CD34, CD117, and DOG-1. In particular, CD117 is strongly marked with the interstitial cells of Cajal, and GIST is believed to be transformed from the interstitial cells of Cajal. These cells stimulate smooth muscle contraction in the alimentary tract. It has been hypothesized that interstitial cells of Cajal could be found in the prostate and lead to EGISTs.

CASE REPORT

A 60-year-old male presented with symptoms of hesitancy, weak urine stream, and nocturia for 1 year. Digital examination showed a distinctive 2-cm hard mass fixed in the prostate. The prostate was within Grade I–II enlargement and had a firm consistency with the prostate-specific antigen level was measured to be 1.84 ng/mL. Meanwhile, colonoscopy revealed a small 0.2 cm × 0.2 cm polyp that was 20 cm away from the anal verge. Magnetic resonance imaging (MRI) of the abdomen revealed a nodular lesion measuring approximately 1.8 cm between the rectum and prostate [Figure 1]. He underwent transrectal ultrasonography of the prostate, and a hypoechoic nodule measuring 1.8 cm × 1.1 cm in size was identified within the peripheral zone in the left lobe of the prostate [Figure 2]. A biopsy was performed, which confirmed the pathological diagnosis of GIST based on the prostatic tissue biopsy strips. The immunohistochemical findings were positive for c-kit and DOG-1 [Figure 3], with a low Ki-67 proliferative index (5%). A whole-body bone scan showed no evidence of bony metastasis. He refused to undergo radical prostatectomy and agreed to take imatinib (400 mg/day) after consultation with the oncologist. During follow-up, an MRI of the pelvis showed remarkably complete remission 1 year later, and there was no tumor progression or metastasis. We rechecked the findings using digital examination, which

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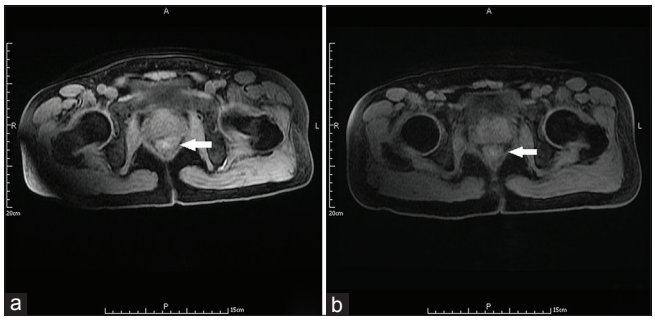

Figure 1: An magnetic resonance imaging of the abdomen showed a nodular lesion about 1.8 cm in size between the rectum and prostate with persistent enhancement during the dynamic study (a); compared with the magnetic resonance imaging taken 1-year earlier, a nodular lesion within the prostate could not be identified at follow-up (b)

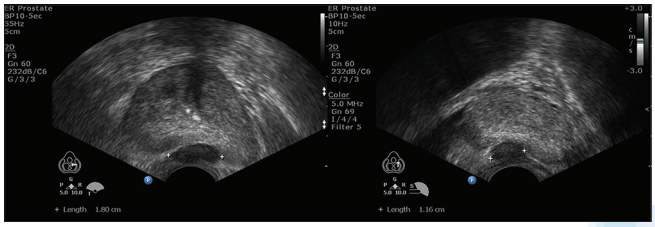

Figure 2: A hypoechoic nodule 1.8 cm × 1.1 cm in size was fixed within the left lobe peripheral zone of the prostate in the findings of transrectalultrasonography

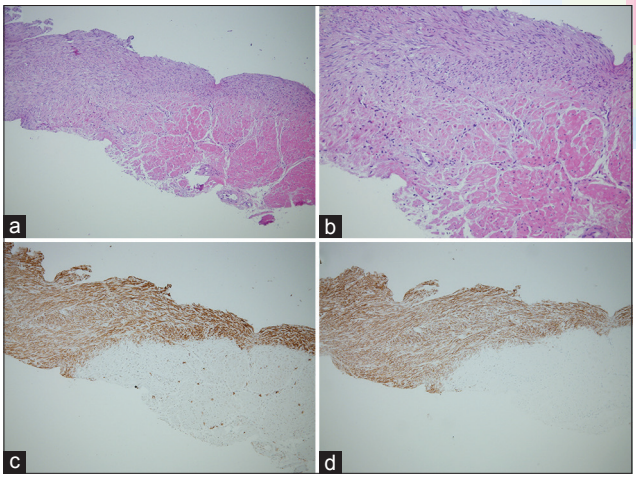

Figure 3: Tissue strips were composed of hypercellularity and bland-looking spindle tumor cell. The junction between tumour cell and prostatic stroma were distinctive in (a and b). Diffuse CD117 (c) and DOG-1 (d) staining on needle prostate biopsy of extragastrointestinal stromal tumor

showed that the palpable lesion diminished. The lower urinary tract symptoms were also well improved.

DISCUSSION

EGISTs are mesenchymal tumors that include smooth muscle tumors, fibromatosis, and neural tumors and commonly occur in the omentum, mesentery, and retroperitoneum; however, primary prostatic EGISTs are relatively rare.[2] A majority of the patients are diagnosed at a late stage when they exhibit excessive tumor growth and lower urinary tract symptoms. Transrectal ultrasound-guided prostate biopsy is the common diagnostic procedure initially, but the biopsy strips may occasionally get mixed with the rectal tissues when the needle passes through the rectal wall. Hence, colonoscopy is an essential procedure to exclude gastrointestinal lesions, and pathological findings can further clarify the diagnosis.

It is believed that GIST is transformed from the interstitial cell of Cajal, which is an important pacemaker cell that regulates smooth muscle contraction in the gastrointestinal tract. The interstitial cells of Cajal or related stem cell-like precursors are positive for CD34 or CD117 in the immunohistochemical staining. The diagnosed prostatic EGISTs show similar immunohistochemical features to those of GISTs; however, there is no strong evidence that all EGISTs correlate with the transformation of the interstitial cells of Cajal. Gevaert *et al*. showed that cells resembling the interstitial cells of Cajals are present in the subepithelial area, which is the area between the epithelial cells and smooth muscle bundles of the prostate stroma space. These interstitial cells are immunoreactive to CD34. In addition, positive immunohistochemical staining for c-kit (also known as CD117) compared to mast cells was found in the prostate stroma.[3] These findings might be correlated with the potential risks of prostatic EGISTs. CD117 is absolutely an important marker for diagnosis, and higher immunohistochemical labeling for CD117 indicates a greater therapeutic response in GIST cases.[4] In those cases with a weak or negative staining for CD117, DOG-1 is a reliable antibody that could be used in the diagnosis of GISTs and EGISTs. The Ki-67 proliferative index is a nonlocation-specific prognostic factor. The higher value of Ki-67 in the present case (cut-off point is 6%) indicates a poor prognosis, and it has more statistical significance than the original tumor site.[5]

The therapeutic strategy for prostatic EGISTs depends on individualized conditions and is applied based on the therapeutic management of GISTs. Imatinib, the target therapy for GISTs and a selective tyrosine kinase inhibitor, primarily acts on tyrosine kinase: Abl, c-kit, and platelet-derived growth factor receptor. Due to its specific mechanism, imatinib 400 mg/day is the major treatment for those cases with prostatic EGISTs, wherein improved outcomes have been reported.[6-8] However, during long-term imatinib use, tumor resistance is a concern and the therapeutic response in advanced stages is also controversial. Hence, surgery is recommended as the first-line of treatment because bulky lesions exhibit poor responses to targeted therapies and have a higher likelihood of secondary recurrence. Blanke *et al*. speculated that there

was no correlation between imatinib use duration and the possibility of secondary recurrence.[9]

Clinicians should be aware of prostatic EGISTs in clinical practice among differential diagnoses. Surgical resection is the major treatment in bulky lesions. While considering the surgical side effects and the patient's quality of life, administrating imatinib to patients with early prostatic EGISTs may be beneficial compared to immediate surgical intervention. The duration of imatinib use has no correlation with tumor recurrence but has a significant effect on the overall survival rate.

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Conflicts of interest

There are no conflicts of interest.

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