Clinical Case Report

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Aggressive angiomyxoma of the prostate
A case report

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Abstract

Rationale Aggressive angiomyxoma (AAM) of the prostate should be considered as a differential diagnosis for prostatic tumor presenting with classical symptoms of benign prostatic hypertrophy.

Patient concerns A 55-year-old man experienced persisting symptoms of prostatic enlargement associated with urinary frequency and urgency and nocturia. Computed tomography images showed low density in the enlarged prostate.

Diagnoses The diagnosis of AAM of the prostate was confirmed based on histopathological findings.

Interventions The patient underwent transurethral resection of the prostate.

Outcomes The patient was enrolled into a watchful waiting protocol. His condition was fine without signs of recurrence on magnetic resonance imaging at the 8-month follow-up.

Lessons AAM of the prostate should be considered a possible cause of urinary difficulty, including retention, although this may be extremely rare. A reliable diagnosis and complete tumor removal enabled optimal treatment and prevention of tumor recurrence.

Abbreviations: AAM = aggressive angiomyxoma, BPH = benign prostatic hypertrophy, CT = computed tomography, IHC = immunohistochemistry, MRI = magnetic resonance imaging.

Keywords: aggressive angiomyxoma, misdiagnose, prostate

1. Introduction

Aggressive angiomyxoma (AAM) is a soft tissue tumor that affects the pelvis and perineum of women of reproductive age.[1] Of the limited cases detected among men, only a few AMM cases have indicated a prostatic origin. Due to the low incidence of AAM of the prostate, precise and clear clinical information on the condition is still not available, and AAM is often misdiagnosed as benign prostatic hypertrophy (BPH) because of the classic symptoms of prostatic enlargement.[2] Surgical excision is an effective treatment for AAM despite the high risk of recurrence. However, systematic standardized treatment was not demonstrated in previous study.

This study aimed to analyze the symptoms and mechanism underlying AAM of the prostate and to focus on the surgical treatment strategy of this rare disease for the first time.

2. Case report

Ethical approval was obtained from the Ethics Committee of the Shaoxing People's Hospital (Shaoxing Hospital, Zhejiang University School of Medicine). A 55-year-old man suffered from ongoing symptoms of prostatic enlargement with urinary frequency, urgency, and nocturia. Abdominal ultrasonography revealed a mildly enlarged prostate (46 mL) with residual urine of about 168 mL (Fig. 1). Computed tomography (CT) showed a soft tissue mass in the prostate with slight separations but did not show any tumor in the pelvis or perineum (Fig. 2). The prostate-specific antigen (PSA) was 2.225ng/mL, and preoperative flow studies showed a maximum flow rate of 6.8 mL/sec.

BPH treatment did not alleviate the symptoms, and the patient was treated by transurethral resection of the prostate under a clinical diagnosis of BPH. A 2-cm polypoid tumor protruding into the cavity was located in the top wall of the bladder neck. The inner part of the tumor was jellylike, fairly transparent, elastic, and soft. The boundary between the tumor and prostatic tissue was indistinct. Pathological examination of the AAM specimen revealed that the tumor consisted of scattered spindle cells and myxoid stroma that stained positively with alcian blue, without a definite boundary discriminating the surrounding prostatic glands. Immunohistochemistry (IHC) staining was positive for vimentin and CD34, but negative for desmin, myoglobin, and S-100 protein. The Ki-67 reaction demonstrated a very low proliferative index of 1% (Fig. 3).

After the operation, urinary symptoms greatly improved, and the maximum flow rate of urine was 13.4 mL/sec. The patient was then arranged into a watchful waiting protocol. Eight months

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Informed consent was obtained from the patient for publication of this case report.

The authors have no conflicts of interest to disclose.

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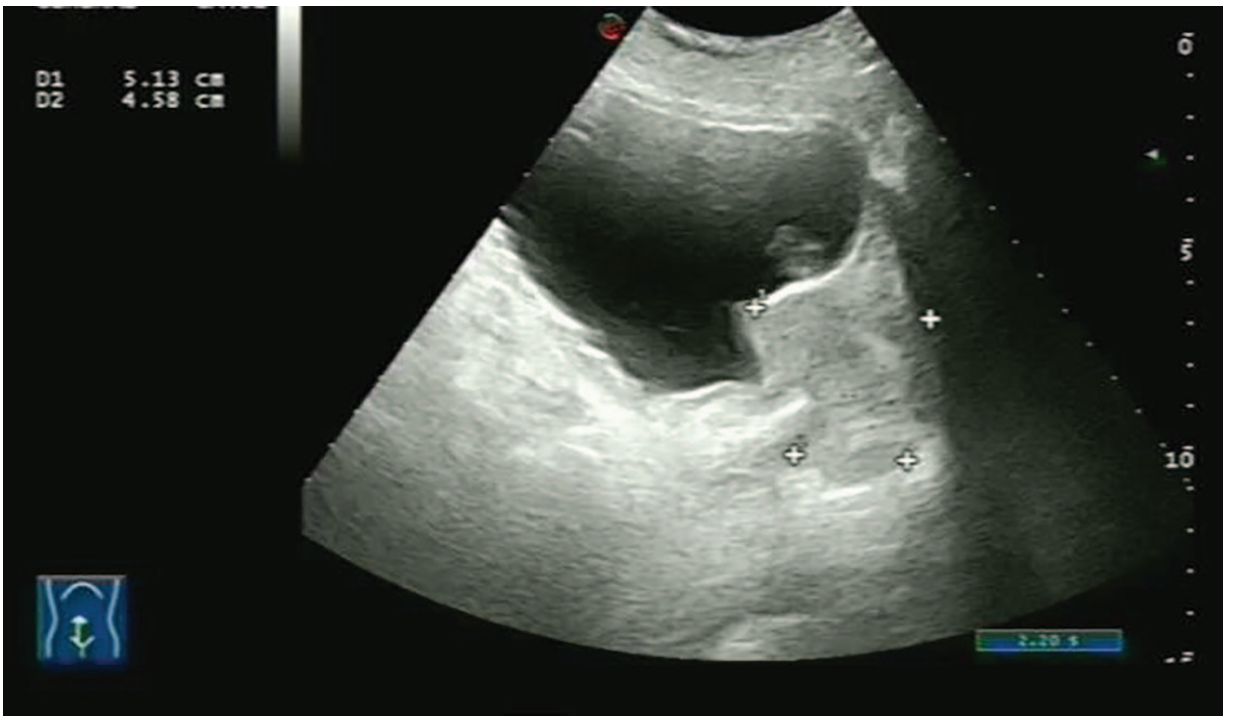

Figure 1. Abdominal ultrasonography image showing a mildly enlarged prostate (46 mL) with residual urine of about 168 mL.

postoperatively, the patient was fine without signs of recurrence, as determined by magnetic resonance imaging (MRI).

3. Discussion

AAM is a rare mesenchymal tumor that predominantly affects the perineum and pelvis of women, characterized by invasiveness and relapse.[3] It is considered an aggressive tumor since it is associated with a high risk of local recurrence after surgery. AAM involving the scrotum, spermatic cord, testis, and perineal region was reported in men,[4] but reports on prostate involvement are few as AAM is often misdiagnosed as BPH.

The etiology of AAM is still unknown. Cytogenetic studies show that 40% of mid-term cells in AAM patients have X chromosome loss (45, X, −X), whereas cytogenetic analysis of peripheral blood in AAM patients shows normal chromosome karyotype (46, XX). Therefore, the loss of chromosomal parts of the lesion is considered a reason for formation of new organisms. Recent studies reported that translocation of chromosomal segments between chromosomes 8 and 12, t (8; 12) (p12; q15), causes aberrant expression of the high mobility group box A2 gene in almost 50% of patients with AAM. AAM was reported to have the same cytogenetic background as other mesenchymal tumors, but the differences lie in the invasion and growth patterns, which may be associated with the activation of interleukin (IL)-5, PGF-R, and other genes.

Preoperative diagnosis is often challenging because of the rarity of these neoplasms, and the specific features associated with these

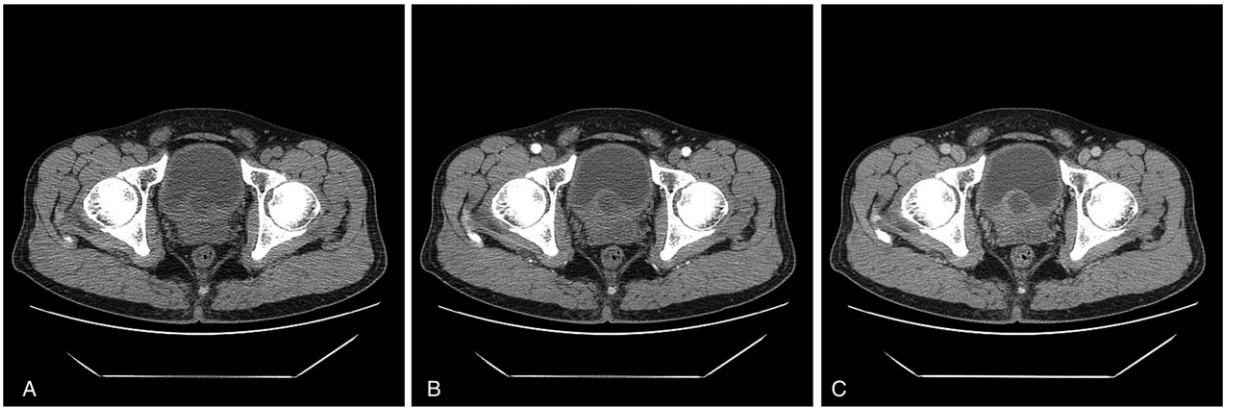

Figure 2. Conventional CT image showing a soft tissue mass in the prostate (A). Contrast-enhanced CT scan showing gradual enhancement of sections in the arterial phase (B) and venous phase (C). CT = computed tomography.

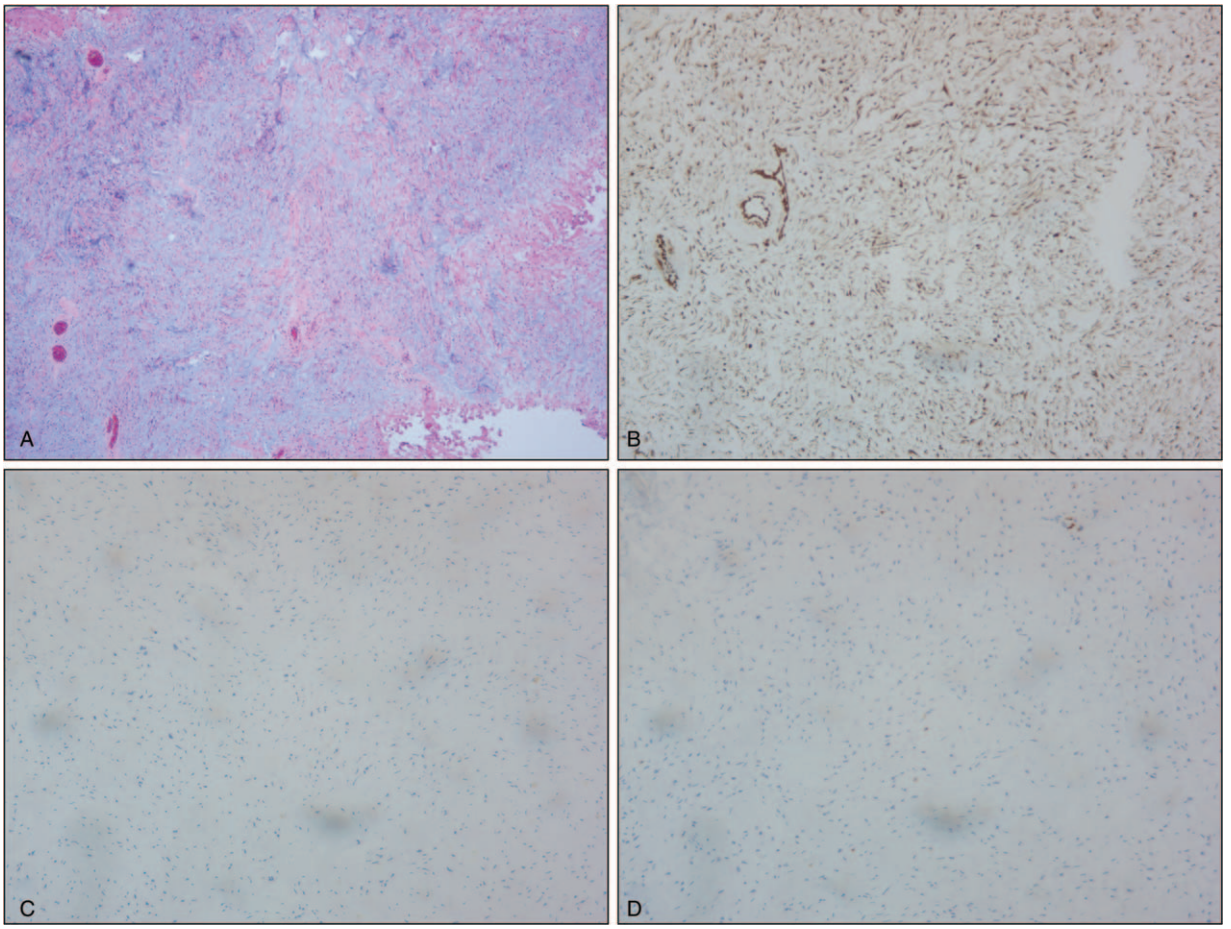

Figure 3. Pathological examination of the aggressive angiomyxoma specimen showing scattered spindle cells and myxoid stroma that stain positively with alcian blue (A); immunohistochemical examination showed that the tumor was positive for vimentin (hematoxylin and eosin staining, 100 ×) (B) and negative for S-100 (C). The Ki-67 reaction showed a very low proliferative index of 1% (D).

neoplasms were not detected by imaging studies. On CT, areas with AAM are hypodense relative to muscles with a layering or swirled pattern after contrast administration.[5] On MRI, owing to the high water content and loose matrix, areas with AAM are isointense relative to muscles in T1-weighted images, hyperintense in T2-weighted images, and enhanced markedly after the administration of gadolinium indicating hypervascularity.[6] CT and MRI aid in determining the location, scope, and blood supply of the tumor; under the CT and MRI guidance, complete tumor resection and decreased postoperative recurrence are possible.

Diagnosis of AAM is made based on a combination of histologic features, such as stellate and spindled cells with poorly demarcated cytoplasm and separated by loose myxoid stroma abundant in collagen fibrils.[7] AAM has a prominent vascular component with many thick-walled vessels. IHC features play a key role in the diagnosis of AMM. They can be positive for vimentin, CD34, and smooth muscle actin, but lack immunoreactivity for desmin and S-100 protein. Tumors also frequently exhibit positive IHC staining for estrogen and progesterone receptors and often respond to hormonal treatment.[8]

There is a high risk of local recurrence after AAM surgery at 36% to 72% and occurs in the first 3 years in more than 70% of cases.[9] This is due to the absence of a preoperative diagnosis and estimation of the actual extent of the tumor or to preserve the genitourinary function of patients. There are a few cases of metastasis in the lung, bone, pleura, and lymph nodes after AAM resection. Some studies found no significance for intraoperative lymphadenectomy. The traditional treatment for AAM is a wide surgical excision with clear margins. However, there is no obvious difference in recurrence rate in patients with negative margin of resection compared with those with positive margins. Incomplete resection is acceptable nowadays when radical resection is difficult or the patient demanded to preserve fertility, followed by long-term follow-up and careful monitoring for timely identification and prompt resection of relapse.[10] Considering the hormonal findings, conservative treatment using tamoxifen or gonadotropin-releasing hormone agonists has been considered to prevent the occurrence of recurrent tumors effectively.[11] In addition, angiographic embolization can be used to treat AAM with high risk of postoperative recurrence, special tumor location, or large volume of surgery, which helps reduce the tumor size and distinguish the scope of operation. Chemotherapy may not be an effective treatment, as patients with AAM demonstrate a low proliferative activity.[12] The effect of radiotherapy is still uncertain. A previous case report described that preoperative extracorporeal irradiation had been used to reduce recurrence. There are individual cases that AAM recurrence was controlled by high-dose external radiotherapy.[13]

Radiotherapy is currently used for patients who are unresponsive to hormone therapy and embolization.

Till date, there is no established treatment guideline because of the rarity of AAM involving the prostate. However, some therapeutic modalities can be used. Transurethral resection of the prostate is commonly used in cases of localized tumors. In case of larger and deeper masses with clear boundaries of normal tissues, radical prostatectomy should be performed as far as possible, and laparoscopic or robotic-assisted surgery may also performed. For extensive infiltration of pelvic organs, if complete resection is not feasible or may lead to severe surgical complications, embolization can be performed. However, when embolization could not be performed or had failed, hormone therapy may be attempted depending on the expression of estrogen and progesterone receptors in the tumor. If both treatment strategies fail and symptoms are not relieved, narrow-margin resection or radiotherapy can be used.

Long-term follow-up is required, because late recurrences have been reported, up to 14 years after treatment. There is no standardized follow-up regimen for AAM involving the prostate; history, physical examination, and imaging modality, usually MRI, are used.[7]

In summary, AAM should be considered as the differential diagnosis for prostatic tumor presenting with classical symptoms of BPH. This report described in detail the clinical characteristics of AAM involving the prostate to avoid misdiagnosis and discussed systematically radiotherapy, chemotherapy, and endocrine therapy for this rare disease. More importantly, this report discussed the surgical treatment strategy for AAM for the first time. A reliable diagnosis and complete tumor removal enable optimal treatment and prevention of tumor recurrence. All patients treated for AAM need close long-term follow-up, regardless of the primary surgical resection. Physicians should watch for symptoms of AAM of the prostate for early diagnosis and selection of appropriate therapeutic modalities.

Author contributions

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Writing – review & editing: Guiliang Tang.

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