Case Report

Prostatic aggressive angiomyxoma (AAM) with initial presentation of urinary obstruction: a rare case report and literature review

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Background: Aggressive angiomyxoma (AAM) is a rare benign mesenchymal tumor known for its aggressive behavior and high recurrence rates, with male cases of AAM being less frequently reported. This study presents a rare case of primary prostatic AAM characterized by a prostatic urethral mass obstructing the bladder outlet, resulting in acute renal dysfunction.

Case Description: The 51-year-old male patient presented with lumbar pain, nausea, frequent urination, urgency, and incomplete urination. Laboratory test indicated increased levels of serum creatinine and potassium. Emergency bedside ultrasound found giant bilateral hydronephrosis. The patient received emergent potassium-lowering therapy and bilateral nephrostomy. Computed tomography (CT) revealed a 3.7 cm × 2.3 cm low-density soft tissue mass in prostate. Transurethral resection of the prostatic mass was performed. Pathological examination identified the presence of deep-seated AAM. For fear of recurrence, he underwent radical prostatectomy. Postoperative histopathology showed no sign of residual tumor. He recovered uneventfully and is now under regular follow-up.

Conclusions: AAM is a rarely reported locally aggressive mesenchymal neoplasm characterized by high recurrence, which was even rarer in prostate. As we know, this is the first case of prostatic AAM treated by both transurethral resection and further radical resection. In this case, the tumor was completely excised surgically, but long-term follow-up is required to monitor for recurrence.

Keywords: Aggressive angiomyxoma (AAM); prostate; hydronephrosis; renal dysfunction; case report

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Introduction

Aggressive angiomyxoma (AAM) is a rare mesenchymal tumor primarily found in the female pelvic cavity and perineum, characterized by mucinous and vascular components and exhibiting a high recurrence rate (1). Although rarely reported, there have been several documented cases of AAM originating from the male prostate (2-5), characterized by invasive growth and potential infiltration of surrounding tissues, often with similar symptoms of benign prostatic hyperplasia (BPH). As a result, AAM originating from the prostate is frequently misdiagnosed as BPH. In spite of this, standard treatment for this type of cancer has not been established, due to its atypical presentation and variable clinical course. This article reports a case of prostatic AAM with initial manifestation of bilateral hydronephrosis and severe renal dysfunction. Relevant literature is also briefly reviewed. We

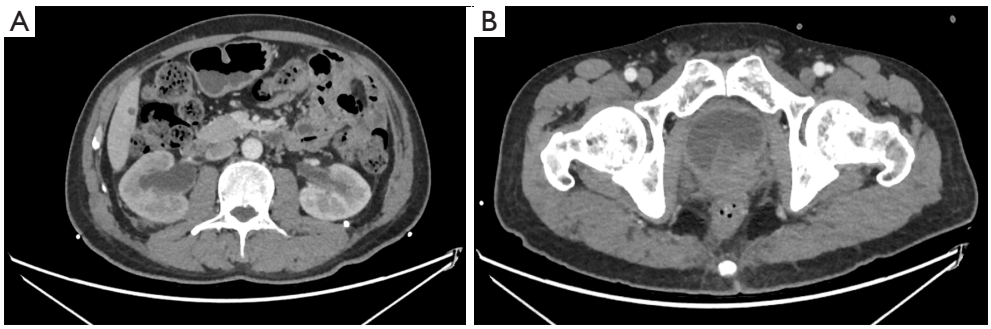

Figure 1 Preoperative enhanced CT scan of the urinary system. (A) Bilateral renal pelvic dilatation, bilateral ureters and bilateral hydronephrosis are observed. (B) Revealed the presence of prostatic hyperplasia, with a 3.7 cm × 2.3 cm low-density soft tissue mass protruding to the posterior wall of the bladder. CT, computed tomography.

present this article in accordance with the CARE reporting checklist (available at https://tau.amegroups.com/article/view/10.21037/tau-24-425/rc).

Case presentation

In September 2023, a male patient aged 51 years sought medical attention at Peking University People's Hospital presenting with a one-month history of left lumbar pain, along with nausea, frequent urination, urgency, and incomplete urination. Laboratory tests indicated elevated levels of serum creatinine at 661 μmol/L and potassium at 5.87 mmol/L, while other blood parameters and prostate-specific antigen were within normal limits. Considering the patient's acute upper urinary tract obstruction with severe renal dysfunction and hyperkalemia, which posed a life-threatening situation, emergent bilateral nephrostomy and potassium lowering therapy were urgently performed to relieve acute urinary obstruction. Subsequent computed tomography (CT) of the urinary system revealed the presence of prostatic hyperplasia, with a low-density soft tissue mass (3.7 cm × 2.3 cm) protruding to the posterior wall of the bladder (*Figure 1*).

Two weeks later, the patient's general condition improved considerably. A diagnostic cystoscopy under spinal anesthesia was performed which unveiled obstruction of the prostatic urethra by a 4 cm mass protruding into the bladder. Transurethral resection of the prostatic mass was carried out. Pathological examination revealed spindle-shaped and oval tumor cells in the submucosa and most areas, extensive edema and mucinous degeneration in the stroma, abundant interstitial blood vessels, and irregularly dilated vessels in some areas. The tumor exhibited unclear boundaries with surrounding tissues. Immunohistochemistry (IHC) demonstrated positive staining for Vimentin and CD34 (vascular+), smooth muscle actin (SMA), Desmin, p53, and Ki-67 (5%+), suggesting the diagnosis of deep-seated AAM (*Figure 2*).

Given the uncertainty of R0 resection of transurethral resection, the high local recurrence rate of AAM,

Highlight box

Key findings

- Prostatic aggressive angiomyxoma (AAM) needs to be distinguished from benign prostatic hyperplasia (BPH). The primary objective in managing AAM patients is complete tumor resection and long-term disease control to prevent recurrence.

What is known and what is new?

- AAM is a rare, locally invasive tumor, with male AAM being even less common. Currently, there are no clear diagnostic and treatment standards for AAM, making the management of patients with genitourinary AAM a significant challenge for doctors.
- We report the first case of prostatic AAM treated with both transurethral resection and radical resection, emphasizing the need for long-term follow-up due to the tumor's high recurrence potential.

What is the implication, and what should change now?

- Prostatic AAM should be considered in male patients presenting with painless nodules and BPH symptoms, particularly when imaging reveals progressive enhancement and a whirlpool-like appearance. These features may indicate invasive AAM causing urinary obstruction.
- Local surgical resection and extensive radical resection remain the primary treatments for prostatic AAM. Hormone therapy is a promising treatment option with the potential to control the disease and complement surgery, but further clinical evidence is needed to support its efficacy.

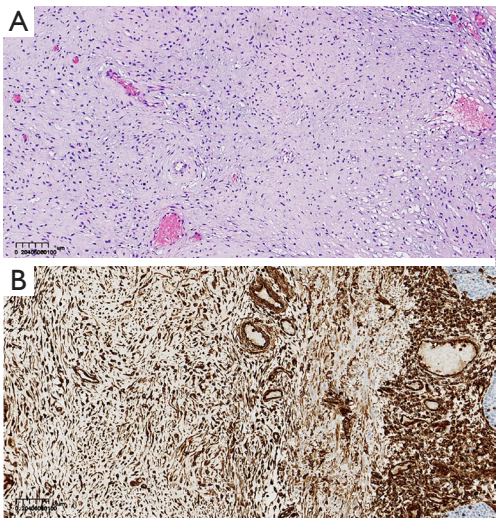

Figure 2 Pathological findings of tumor. (A) H&E staining (×100), spindle-shaped and oval tumor cells in the submucosa and most areas, extensive edema and mucinous degeneration in the stroma, abundant interstitial blood vessels, and irregularly dilated vessels in some areas. The tumor exhibited unclear boundaries with surrounding tissues. (B) IHC (×100) showing the expression of Vimentin (+++). H&E, hematoxylin and eosin; IHC, immunohistochemistry.

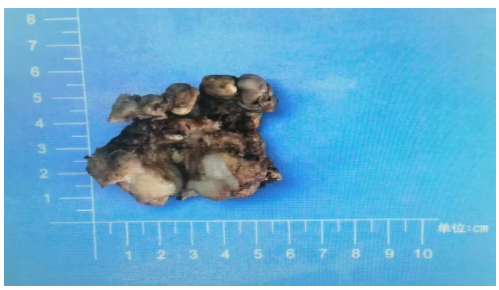

Figure 3 Gross image of the mass.

particularly within the initial 3 years after surgery (6), and the presence of adverse pathological characteristics (p53+, Ki67 5%+), the likelihood of postoperative recurrence is heightened. Following comprehensive deliberation with the patient concerning treatment alternatives and their respective risks, the patient opted for definitive surgery. Accordingly, laparoscopic radical prostatectomy was performed one month later. Examination of the postoperative specimen indicated the presence of normal prostate tissue devoid of any residual tumor (*Figure 3*). The patient experienced alleviation of voiding difficulties and renal failure, with no evidence of recurrence until now.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

AAM is a rare mesenchymal tumor that primarily affects the female pelvic and perineal regions, with even fewer reported cases in males [male-to-female incidence ratio approximately 1:6.6 (6)]. In males, AAM typically involves the perineum, male genital organs, and deep soft tissues in the inguinal region (7,8). Despite being classified as a benign tumor by the World Health Organization (WHO), AAM is distinguished by its rich vascular content, invasiveness, and tendency for recurrence. Local recurrence rates can be as high as 47%, with 85% of patients experiencing recurrence within the initial 5 years following surgery (6). Nevertheless, distant metastasis is rare, and to date, no cases of metastatic AAM in males have been reported. Therefore, it is classified as a locally invasive benign tumor.

Based on the findings of the literature review, it is evident that AAM tends to exhibit slow growth within the deep pelvic and perineal regions. Patients commonly manifest with asymptomatic perineal or pelvic masses, while some may encounter challenges in urination and persistent pain as a result of the mass exerting pressure on neighboring organs such as the bladder, rectum, ureter, and uterus. Consequently, the potential for misdiagnosis in clinical settings is a significant concern. The clinical presentation in this case is consistent with existed literature (2,4,5,9), with symptoms of urinary frequency, urgency, and incomplete voiding likely due to compression of the prostatic urethra and bladder by the enlarged tumor. Prolonged urinary obstruction may progress to acute upper urinary tract obstruction, leading to significant renal impairment and hyperkalemia, thereby creating a potentially life-threatening scenario.

The challenging nature of diagnosing AAM in males is compounded by the unclear pathogenesis, lack of distinctive clinical presentations, and rarity of the condition. Early detection of AAM is complicated by its deep-seated nature, necessitating the use of preoperative imaging to

assess the extent of the tumor. While current imaging techniques may not definitively distinguish AAM from other conditions, there are identifiable features that can aid in diagnosis. In ultrasound examination, AAM may exhibit a combination of echogenicity patterns, layered structure, and alternating hypoechoic and hyperechoic tissue layers (10). Magnetic resonance imaging (MRI) is considered a more effective diagnostic tool for AAM, as it typically shows low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Contrast-enhanced scans may reveal a distinct "whirlpool sign" within the lesion, which holds diagnostic significance. Early enhancement is generally mild, with delayed persistent enhancement observed (11). On CT scans, AAM frequently presents as a homogeneous mass with distinct borders and decreased density relative to muscle tissue. Contrast-enhanced scans commonly reveal delayed enhancement within the lesion, characterized by whirlpool or layered alterations. In cases of suspected AAM, additional enhanced MRI evaluation is advised (11). In this case, a preoperative urinary system CT scan was exclusively performed, without the inclusion of a contrast-enhanced MRI. This choice was informed by the identification of AAM following cystoscopy and transurethral resection of the prostate. The ultimate diagnosis still relies on surgical exploration and postoperative pathology, primarily derived from morphological assessments and IHC. The tumor displays a notable mucinous background and elongated fibrous-like cells with limited cytoplasm and an absence of malignant cells. Tumor cells are interspersed with normal connective tissue in the mucoid stroma. IHC results reveal strong positivity for Vimentin and CD34 (most significant), moderate positivity for SMA, Desmin, estrogen receptor (ER), and progesterone receptor (PR), and negativity for S-100, CD68, and p53, with Ki-67 (<1%), indicating a mesenchymal origin of the tumor (12). The presence of ER and PR positivity suggests that the patient's tumor is hormone-dependent (12). Although ER and PR testing were not conducted in this case, other pathological and immunohistochemical markers align closely with existing literature, despite the presence of unfavorable pathological characteristics (p53 positivity, Ki67 expression exceeding 1%), potentially indicating an increased risk of postoperative recurrence. Therefore, vigilant monitoring and follow-up are warranted.

Currently, there is no standardized international protocol for treating AAM. Treatment strategies primarily include local excision and extensive tumor resection, with the latter being preferred but subject to debate. The choice of surgical approach is influenced by factors such as the extent of the remaining tumor, the anatomical features of adjacent organs, and patient fertility requirements. In cases of larger lesions extending along the genitourinary or rectal tract, complete resection may lead to severe complications, necessitating partial resection (4). Conversely, previous research indicates no significant difference in recurrence rates between patients with negative and positive margins, casting doubt on the efficacy of wide complete tumor resection in reducing recurrence rates (6). In this case, the patient underwent cystoscopy and transurethral resection of the prostate, confirming the diagnosis of AAM. The patient then opted for radical surgery, and postoperative pathological findings showed no residual tumor, suggesting that transurethral resection may also achieve complete resection of prostatic AAM.

While surgery remains the primary treatment for AAM, its recurrence rate can be as high as 50% (13). The primary objective in treating AAM patients is to manage local disease progression and prevent recurrence. At present, the management of AAM underscores a personalized, multidisciplinary strategy, predominantly incorporating surgical intervention in conjunction with adjuvant therapies, including hormone therapy, embolization, and radiotherapy. In instances where complete surgical resection is unattainable or in the presence of local recurrence, adjuvant therapy assumes a critical role. Preoperative embolization and radiotherapy may facilitate tumor size reduction, thereby enabling more extensive surgical excision. In cases of local recurrence, adjuvant radiotherapy may contribute to the eradication of residual tumor cells, thereby diminishing the likelihood of subsequent recurrence. Several case reports have indicated that radiotherapy can effectively achieve local tumor control and symptom relief for a duration of at least 3 years following treatment (14,15). Nonetheless, radiotherapy is not routinely employed as a standard treatment for AAM due to its severe side effects and the significant variability in individual patient responses, which continue to cast doubt on its overall efficacy. Radiotherapy may be more suitably applied in cases of recurrence that are unresponsive to hormone therapy.

In instances of disease advancement or recurrence, systemic hormone therapy has shown efficacy in controlling local disease. Given the prevalent expression of ER or PR in female AAM patients, systemic anti-estrogen therapy has emerged as the preferred initial treatment for the majority of recurrent or unresectable cases. The commonly used

drugs include gonadotropin-releasing hormone (GnRH) agonists (GnRHa), aromatase inhibitors (AIs), and estrogen and progestogen receptor blockers (13). However, male AAM patients are rare, and related treatment data are limited. Although ER, PR, and androgen receptor (AR) are also expressed in male AAM patients (16,17), the positive rates for ER and PR are relatively lower compared to female patients (16). Therefore, whether the currently used hormone therapy drugs can benefit these patients remains to be further evaluated.

A recent study documented a case involving a 59-year-old male presenting with perineal AAM, who tested positive for ER, PR, and AR. Based on his positive hormone receptor status, dual hormone blockade was recommended, but the patient opted out due to concerns regarding potential adverse effects.

Following a three-month course of letrozole therapy, the patient experienced a rapid progression of their condition, necessitating the cessation of treatment. Subsequent administration of bicalutamide, a singular AR blocker, yielded sustained disease stability without notable adverse effects as of the present time (17). This finding implies a potential heightened reliance of the tumor on the androgen pathway for its growth and dissemination.

Furthermore, previous research has demonstrated that adjuvant administration of GnRHa after local surgical excision or before recurrent surgery can effectively reduce the recurrence rate of AAM (12).

While hormone therapy presents potential for managing local advancement and averting recurrence in male AAM patients, additional investigation is warranted. This entails examining the selection of efficacious medications, the optimal duration of treatment, the potential benefits of combination therapy, the appropriateness of dual hormone blockade, and the protocols for perioperative adjuvant therapy.

Atypical prostate tumors like AAM often present with nonspecific symptoms and varying aggressiveness. Tumors like prostatic stromal sarcoma, leiomyosarcoma, and solitary fibrous tumor share a mesenchymal origin and recurrence potential but differ in aggressiveness and metastatic risk, requiring tailored management. Leiomyosarcoma, with its rapid growth and high metastatic risk, usually needs a combination of surgery, radiotherapy, and chemotherapy (18,19). As for solitary fibrous tumors typically grow slowly and are often treatable with local excision. Although radical excision is the main treatment, other options like radiotherapy, hormone therapy, or targeted therapy may be used depending on the tumor's pathology and receptor status (20).

Given the high risk of recurrence and potential for metastasis in these atypical tumors, long-term follow-up with regular MRI or CT scans is crucial. Creating standardized guidelines that consider histological subtypes, growth patterns, and individual risk factors would enhance management. Clinicians should be well-informed about these rare tumors and use a personalized, multidisciplinary approach to improve patient outcomes.

This study has limitations. Although all patients underwent transurethral resection and radical surgery, they did not undergo testing for ER, PR, and AR. Therefore, it remains unclear whether these patients are suitable for preoperative and postoperative adjuvant hormone therapy. More rigorous follow-up is necessary to prevent recurrence and progression.

Conclusions

In conclusion, male patients with painless nodules in the prostate or prostatic urethra, along with typical symptoms of BPH, should consider prostatic AAM as a potential cause of urinary obstruction and upper urinary tract obstruction. Imaging results indicating progressive enhancement and a layered whirlpool-like appearance should prompt further investigation for invasive AAM.

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Footnote

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tau.amegroups.com/article/view/10.21037/tau-24-425/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related

to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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