A rare benign genitourinary tumor in a Japanese male: urinary retention owing to aggressive angiomyxoma of the prostate

Yugo Sawada,[1] Fumio Ito,[1] Hayakazu Nakazawa,[1] Nobuhiko Tsushima,[1] Hikaru Tomoe,[1] Motohiko Aiba[2]

[1]Department of Urology and [2]Department of Surgical Pathology, Tokyo Women's Medical University Medical Center East, Tokyo, Japan

Abstract

Close examination of a 67-year-old Japanese man, who complained of persistent nocturia, revealed that a semitransparent polypoid tumor had developed from the bladder neck to the prostatic urethra obstructing the internal urethral meatus, which resulted in excessive urinary retention and post-renal dysfunction. The tumor was resected by a transurethral procedure and a pathological examination of specimens revealed aggressive angiomyxoma (AAM) of the prostate. AAM usually develops in the intrapelvic and perineal organs of females. So far as we know, this is the second case of primary prostatic AAM reported in the English literature, and is the first case where the patient encountered urethral obstruction.

Introduction

We experienced a case where urinary retention was caused by a tumor of the prostate of unusual origin. This neoplasm appeared translucent and characteristically gelatinous on sectioning. Pathological examination identified the neoplasm as an aggressive angiomyxoma (AAM), which is a benign non-epithelial tumor with a high incidence in the pelvis and perineum of premenopausal females.[1] Of the limited cases found in males, only one AAM case has indicated a prostatic origin to date.[2] Although AAM does not generally cause urethral, rectal, vaginal, or vascular obstruction,[3] we encountered a case with urinary retention owing to AAM. In this report, we pursued the clinical course of a patient with this extremely rare disease and briefly reviewed the relevant literature.

Case Report

In March 2007, a 67-year old Japanese man without previous physical or surgical history visited our hospital after suffering from intractable nocturia for six months. Although digital rectal examination showed a mildly enlarging prostate, transrectal or transabdominal ultrasonic examination revealed excessive urinary retention with subsequent bilateral hydronephrosis where a polypoid mass (size: 2 cm) protruding into the cavity was located in the bladder neck. According to routine blood tests, the serum creatinine level had increased to 1.71 mg/mL and prostatic specific antigen registered 0.364 ng/mL. The results of remaining blood tests were within the normal range and urine cytology showed no atypical cells. Flexible cystoscopy demonstrated that a marshmallow-like translucent protuberance developed from the left prostatic urethra and protruded into the bladder cavity (Figure 1A and B).

In April 2007 we performed transurethral resection under a clinical diagnosis of benign prostatic hypertrophy (BPH) without conducting other diagnoses, although its external appearance was quite contrary to the BPH image. The inner part of the tumor was jelly-like, fairly transparent, elastic, and soft. Some fine arteries from the prostate were seen penetrating the tumor body in straight lines (Figure 1C). Since the boundary between the tumor and prostatic tissue was indistinct and arteries could have coursed from the prostate tissue directly to the tumor through the distinct border, the tumor was most likely to have developed from deep within the prostatic tissue. Therefore, the tumor and surrounding normal tissue were resected as extensively as possible in our surgical intervention.

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 (Figure 2). Additionally, immunohistochemical staining demonstrated that the spindle cells reacted positively for vimentin, androgen receptor, and progesterone receptor, while was weakly positive for MIB-1 and negative for desmin, the estrogen receptor, CD34, and S-100 protein.

After surgical intervention, the patient was relieved from urinary difficulty and renal failure, and follow-up examinations (up to the time when the present manuscript was under preparation; >15 months) indicated no signs of recurrence.

Correspondence: Fumio Ito, Department of Urology, Tokyo Women's Medical University Medical Center East, 2-1-10 Nishiogu, Arakawa-ku, Tokyo 116-8567, Japan.
E-mail: fitour@dnh.twmu.ac.jp

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Discussion

AAM is a benign non-epithelial tumor, which was originally reported by Steeper and Rosai in 1983.[1] The adjective "aggressive" added to the pathological name in spite of its benign nature is because of its high risk of local recurrence after resection.[1] Except in a few cases,[4] the prognosis of AAM is generally favorable owing to a lack of distant metastases. AAM, characterized by components including myxoid stroma and scattered spindle-shaped cells, is believed to be of fibroblast or myofibroblast origin by ultrastructural analysis. According to a recent immunohistochemical profile, the response of relevant tumor cells is positive for vimentin; variable for desmin,[5] muscle-specific actin,[1,5,6] and CD34;[5] and negative for S-100 protein.[5]

The following myxoid soft tissue tumors need to be distinguished from AAM: organ-nonspecific stromal tumors including intramuscular myxoma, neurofibroma, neurothekeoma, angiomyofibroblastoma, superficial angiomyxoma, myxoid liposarcoma, myxoid malignant fibrous histiocytoma, and embryonal rhabdomyosarcoma;[7] and prostate-specific stromal tumors such as pure specialized stromal tumors of the prostate unassociated with sarcomas,[8] and prostatic stromal hyperplasia with atypia.[9] However, the clinicopathological features of our case were conclusive evidence to diagnose and discriminate AAM from other previously mentioned myxoid tumors; as follows: a gross appearance that resembled jelly; low cellularity; lack of cytological atypia; lack of aggressive behavior suggesting malignancy, such as necrosis and mitosis; an ill-defined border; lack of immunoreactivity for desmin,[9] S-100, and CD34;[10] and characteristic features of other stromal components, such as numerous size-

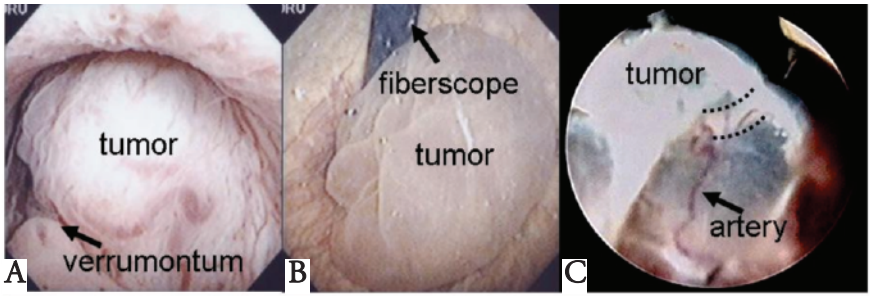

Figure 1. Macroscopic aspects of the tumor. (A) The view from the distal to the proximal urethra showed that the tumor developed from the left side of the prostate and had almost occluded the prostatic urethra. (B) The turn-around view with flexible cystoscopy depicted a translucent polyp overspreading the bladder neck. (C) The view taken in the course of a transurethral resection showed that the tumor body was fairly transparent so that the resectoscope loop (dotted curves) and arteries coursing vertically inside the tumor (arrow) could be seen through the tumor.

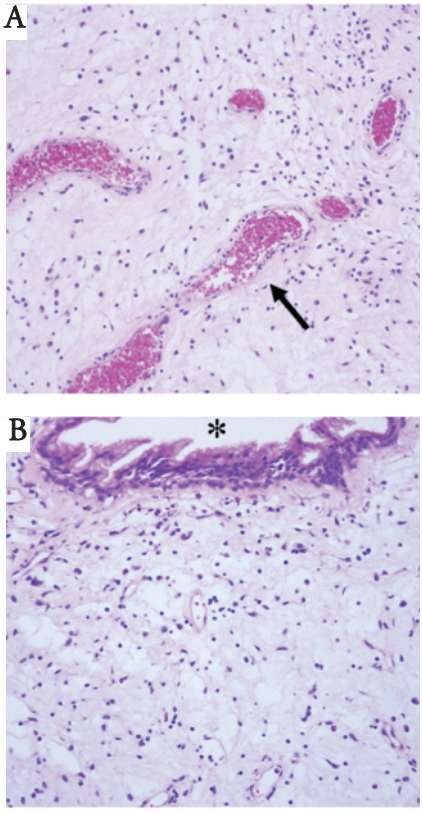

Figure 2. Light microscopic aspects of the tumor. (A) Spindle-shaped tumor cells and vessels of various sizes were scattered randomly in the myxoid stroma. Especially large vessels were accompanied with hyalinization around them (arrow). (B) Although hyperplasic prostatic glands (*) were seen in the periphery of the figure, there was no apparent boundary between the myxoid lesion and proper prostatic tissue. (Hematoxylin and eosin stain.)

variable vessels with thin to thick walls and hyalinization of larger vessels.

Although AAM often develops in the female pelvis or perineum,[1,4] twenty-four male cases have been reported cumulatively in the English literature until 2006.[2,3,6,7,11,12] The affected common sites in these patients include the scrotum, spermatic cord, inguinal region, and pelvis.[13] AAM of prostatic origin is rare, and there have been no reports except a Bastian's case.[2] The AAM sites are limited to the reproductive areas, and the predominance of AAM in premenopausal females suggests that certain hormonal factors may participate in the development or proliferation of AAM. In fact, estrogen and progesterone receptors have been identified and confirmed in more than 90% of female tissues.[4] Additionally, Htwe *et al*. have reported a case where AAM is aggravated by pregnancy.[11] However, AAM expresses androgen instead of estrogen receptors in some male cases[12] and in our case. Interestingly, the age range of both the Bastian's case and our patient overlapped that most relevant to BPH. Additionally, coexistent BPH tissue in our case also exhibited androgen receptors. Therefore, a certain androgen-dependent mechanism might have promoted proliferation and growth of AAM as well as BPH via the androgen receptor.

According to these findings on the hormonal environment, conservative treatment using gonadotrophin-releasing hormone (Gn-RH) agonists has been attempted against recurrent tumors with encouraging results.[13] Although such favorable findings have been established, the current therapy prefers a local extensive resection per se.[6] The approach with such an extensive surgical intervention, however, remains controversial. Since AAM generally grows slowly as a benign neoplasm and it is difficult to excise extensively around the pelvis-perineal region without inflicting injury on the adjacent organs, urgent and complete resection is not always established. Furthermore, postoperative options in follow-up treatment are controversial. According to Behrankawala and Thomas,[14] long-term follow-up by careful clinical examination is necessary, although regular imaging is not a prerequisite.

In our present case study, any symptoms or signs of recurrence were screened carefully and verified accordingly, using periodical surveys that included transrectal ultrasonography, because it might be difficult to discriminate symptoms and/or signs of AAM from those of BPH in the event that our patient complained of any such AAM symptoms.

In conclusion, we need to take AAM of the prostate into consideration as a possible cause of urinary difficulty including retention, although this may be extremely rare. A reliable diagnosis of AAM of the prostate may lead to an optimal treatment and prevention of its recurrence.

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