Aggressive angiomyxoma presenting as urinary retention in a male: a case report and literature review

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We present a case of pelvic aggressive angiomyxoma presenting as urinary retention in a male. A 46-year-old male presented with urinary retention and was found on computed tomography (CT) scan of the pelvis to have a large pelvic tumor. A transrectal ultrasound guided needle biopsy of the tumor and prostate revealed a myxoid tumor; low volume, low grade prostate cancer was also detected. The patient underwent radical prostatectomy and excision of the pelvic tumor which was diagnosed as aggressive angiomyxoma (AAM). The patient was free of recurrence after 1 year of follow up.

AAM is a benign myxoid tumor seen very rarely in males. Treatment consists of surgical excision with negative margins. Tumors variably express estrogen and progesterone receptors. Immunohistochemistry should be used to exclude other benign and malignant tumors. Patients should be followed with axial imaging as recurrence is common.

Key Words: aggressive angiomyxoma, male, myxoid tumor, mesenchymal tumor

Introduction

Aggressive angiomyxoma (AAM) is a rare, benign, locally infiltrative myxoid tumor most commonly arising in the pelvic and genital soft tissues of adult females.[1] It has a 6-fold higher incidence in females, and has a high rate of recurrence, even after complete extirpation.[2] There have been fewer than 50 cases of AAM reported in males, usually presenting as scrotal masses or mimicking groin hernias. This tumor rarely metastasizes and often expresses hormone receptors.[3]

Case report

We report the case of an otherwise healthy 46-year-old Caucasian male who initially presented to an area emergency department in acute urinary retention. A computed tomography (CT) scan of the abdomen and pelvis without contrast revealed a distended urinary bladder and an 11 cm well demarcated soft tissue structure posterior to, and anteriorly displacing the urinary bladder, Figure 1. A Foley catheter was placed, and a urologic referral was made.

The patient reported sporadic, brief episodes of slowing of stream over the past year. Digital rectal examination (DRE) of the left lobe of the prostate was partially obscured by a soft structure. Cystoscopy was unremarkable, and a transrectal ultrasound (TRUS) revealed what appeared to be a cystic structure arising from the left seminal vesicle. This structure was evaluated and biopsied using TRUS; a sextant biopsy of the prostate was also performed secondary to abnormal DRE.

The biopsy of the pelvic mass was interpreted as fibromuscular soft tissue with myxoid change. The patient was also found to have a small volume Gleason 6 (3 + 3) adenocarcinoma of the prostate on the right. The patient underwent excision of the pelvic mass and radical prostatectomy. The lesion was firm in nature and appeared to arise deep within the pelvis,

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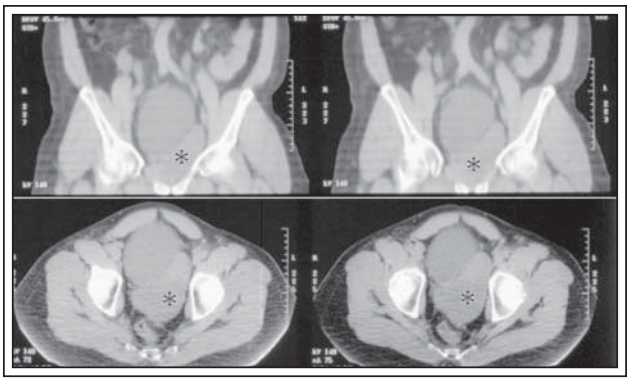

Figure 1. CT scan showing mass posterior and lateral to urinary bladder (asterisks).

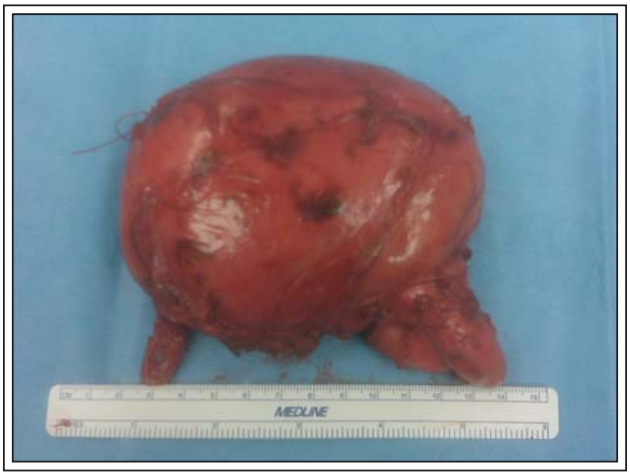

Figure 2. Gross specimen measuring 11 cm in greatest dimension.

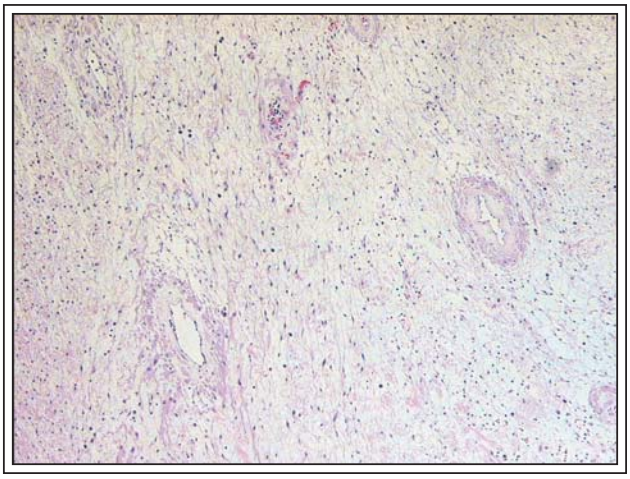

Figure 3. Photomicrograph of aggressive angiomyxoma showing myxoid stroma and numerous thick-walled vessels. (200x, H&E)

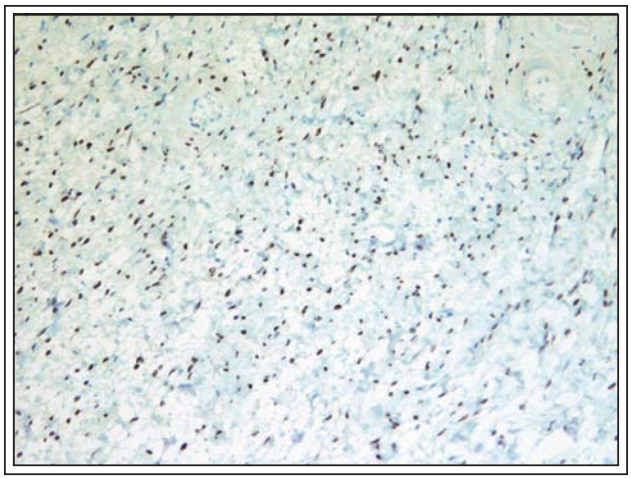

Figure 4. Photomicrograph of aggressive angiomyxoma with immunohistochemical stain showing estrogen receptor positivity (blue). (400x)

lateral to the prostate and the dorsal venous complex. The specimen measured 11 cm x 9.5 cm x 3.6 cm and weighed 300 grams, Figure 2.

The pelvic mass, diagnosed as aggressive angiomyxoma, was described by the reviewing pathologist as a low grade mesynchymal tumor with numerous mast cells, a myxoid stroma and abundant blood vessels, Figure 3. On immunohistochemical analysis, the tissue was found to stain positive for smooth muscle actin (SMA), estrogen receptors (ER), and progesterone receptors (PR), Figures 4 and 5. It stained negative for desmin and S-100 protein. The final prostate specimen revealed T2cNxMx Gleason 6 (3 + 3) adenocarcinoma.

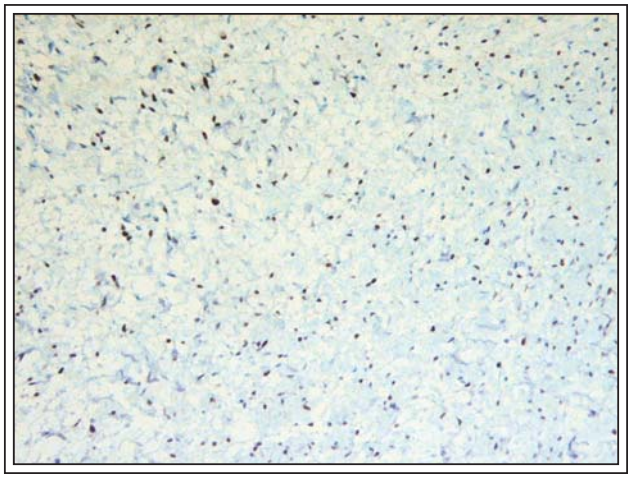

Figure 5. Photomicrograph of aggressive angiomyxoma with immunohistochemical stain showing progesterone receptor positivity (light blue). (400x)

Discussion

Since first being described in 1983 by Steeper and Rosai there have been approximately 250 published cases of AAM, the majority of patients being adult females.[4,5] Fewer than 50 cases have been reported in males, usually arising in the scrotum, pelvic wall, spermatic cord, perineal region, and intrapelvic organs.[1,2] The mean age of presentation in men is 46 years, with a range of 1 to 82 years.[6] CT scan shows a clearly delineated mass with moderate enhancement, and magnetic resonance imaging (MRI) reveals a characteristic swirling pattern in approximately 83% of cases.[7]

AAMs are typically large, most are between 2 cm and 17 cm in greatest dimension, however there have been reports of even larger lesions.[3] Gross inspection typically reveals a soft, smooth, gelatinous, gray-white tissue with occasional firm fibrous areas.[3] Microscopically, these tumors are bland with a hypocellular myxoid stroma.[3] Tumor cells are stellate or spindled in appearance with rare nuclear pleomorphism and mitotic figures.[3,8] Abundant blood vessels of varying size and type are frequently present and there is often a chronic inflammatory background within the stroma.[3]

Immunohistochemical (IHC) features play a key role in the diagnosis of AAM. Tumors usually stain positive for vimentin, variably for muscle specific actin and α-smooth muscle actin, and negative for desmin, S-100 protein, and CD34.[6] Greater than 90% of tumors in females are ER and PR postive. The rate of ER/PR positivity in men is unclear; in a series of four tumors, one stained positive for ER, and three for PR.[9] IHC evaluation should be used to distinguish AAM from other benign (including intramuscular myxoma, neurofibroma with myoid change, and angiomyofibromyxoma) and malignant (myxoid liposarcoma, myxoid malignant fibrohistiocytoma, and embryonal rhabdomyosarcoma) tumors.[6]

Local recurrence is common (36% to 72%), and occurs in the first 3 years in greater than 70% of cases.[6] It has been reported in a retrospective study of 106 cases that completeness of extirpation may not be significantly associated with recurrence rate in female patients, though wide complete excision with tumor free margins is recommended.[10] Metastasis is very rare with two reported cases in females, both culminating in patient death.[11]

Surgical excision has been the standard method of treatment, however there have been reports of use of radiotherapy, and angioembolization.[10] There is one reported case of complete resolution of an AAM recurrence using gonadotropin-releasing hormone agonist (GnRH) monotherapy in a female.[12] The role of long term hormonal therapy for treatment of AAM in males remains unclear.[6,12] There is no consensus regarding duration and nature of follow up, however most advocate the use of history, physical examination and an imaging modality, usually MRI.[5]

Conclusion

Aggressive angiomyxoma is a benign myxoid tumor seen infrequently in men. It usually arises in the pelvic and genital soft tissues and is treated by surgical excision. Local recurrence is common, and metastasis is rare. The role of GnRH treatment in men is unclear. There is no standard follow up regimen for this tumor in men, though it should include an axial imaging modality.

This case was reported secondary to the rarity of the tumor, the unique mode of presentation, and the presence of the coexisting condition of prostate cancer. Currently the patient is doing well, with no evidence of prostate cancer recurrence (serum PSA < 0.05 ng/mL). He is potent and voiding freely with improving post-prosatatectomy urinary incontinence. There is no clinical evidence of AAM recurrence. □

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