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Aggressive Angiomyxoma Involving Penis and Urethra – A Case Report

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ABSTRACT

Aggressive angiomyxoma is a rare benign mesenchymal stromal tumour, characterized by locally infiltrative nature and a tendency for recurrence. Only a few cases of penile involvement have been reported in the literature so far. We report a case of aggressive penile angiomyxoma in a sixty-two-year-old obese, diabetic male patient. He presented with obstructive lower urinary tract symptoms (LUTS) and diffuse enlargement of the penis and scrotum. He was managed with excision, reduction scrotoplasty, internal urethrotomy, followed by Leuprolide therapy for prevention of recurrence. He is on follow up for 20 months without recurrence and obstructive symptoms.

Introduction

Aggressive angiomyxoma is a rare mesenchymal stromal tumor first described by Steeper and Rosai in 1983, as a benign soft tissue tumor occurring in female pelvis and perineum. This tumor had a prominent angiomatous component, characterized by large thick walled vessels without arborization. Later reports attribute this tumor to adult females exclusively, occurring in pelvis, perineum, vulva and buttock.[1] Aggressive angiomyxoma occurs rarely in men and only a very few cases involving the penis and urethra have been reported in the literature so far. We report a case of a 62-year-old obese and diabetic male, who presented with diffuse enlargement of the penis and scrotum and obstructive LUTS.

Case history

A 62-year-old obese man (92 kg – BMI 33) presented with an enlarging penile and scrotal mass and severe obstructive lower urinary tract symptoms (IPSS score 31) to our OPD. He was diabetic and had multiple comorbidities including coronary artery disease, hypertension, hyperlipidemia and a diabetic foot ulcer. On examination, his penis was grossly enlarged and deformed. The prepuce was part of the diffuse swelling and was adherent to the glans

Figure 1. Preoperative picture of aggressive AAM involving the penis and scrotum. The meatus was completely occluded and patient had severe obstructive LUTS.

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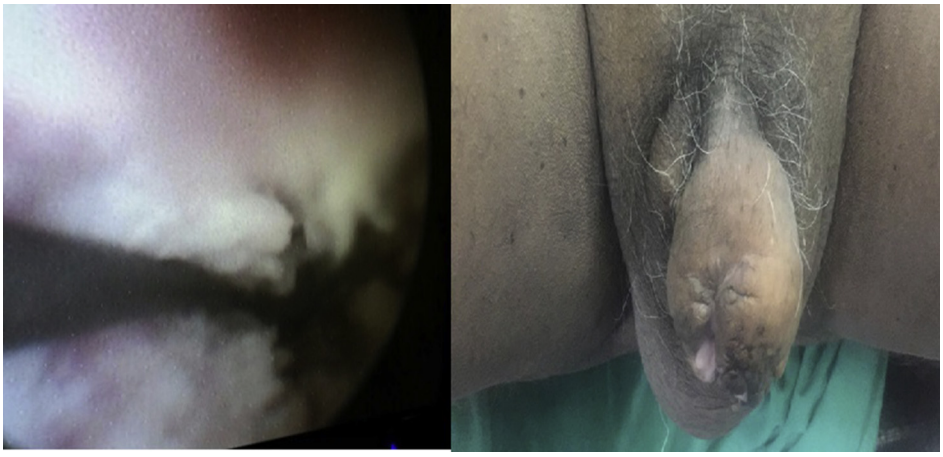

Figure 2. Postoperative picture after a limited resection, ventral meatotomy and scrotoplasty. Urethroscopy showing AAM tissue inside penile urethra.

(Fig. 1). Since the meatus was inaccessible for cannulation, a retrograde urethrogram could not be done. The scrotum was also moderately enlarged and had a similar consistency as that of penis. The testes were barely palpable. Lower limb examination was done to rule out a concurrent filarial lymphedema, as the penile mass was suspicious for a filarial ram horn penis and since the patient came from an area endemic for filariasis. However, a filarial work up was negative.

In view of the poor performance status of the patient and multiple comorbidities including a poorly controlled diabetes, it was decided to do excise the involved tissues for biopsy and do a primary closure. During surgery, resection of the prepuce was technically demanding, as the angiomyxoma had infiltrated the glanular tissues. Hence, a limited resection of preputial tissue was done and a ventral meatotomy was done. This was followed by a reduction scrotoplasty and primary closure (Fig. 2).

A diagnostic urethroscopy showed diffuse narrowing in the penile urethra, with segmental stricture in the mid penile urethra (Fig. 2). The narrowed portion was lined by white, vascular tissue which had a gelatin like consistency. We performed an internal urethrotomy, in view of his comorbidities. Post procedure, a 16 French silastic catheter was introduced into the bladder and was retained for 2 weeks. Histopathological examination of the penile and scrotum specimens confirmed the tumor to be an aggressive angiomyxoma (Fig. 3).

Due to the high likelihood of recurrence of aggressive angiomyxoma, the patient was started on LHRH agonist therapy (Leuprolide 3.75 mg IM once a month). The tumor had been shown to express androgen receptors and found to be susceptible to hormone therapy from previous studies.[2] The patient has opted out of a definitive surgical resection due to his comorbidities and is on regular follow up for the past 20 months (monthly physical and office based urethroscopy once every 3 months).

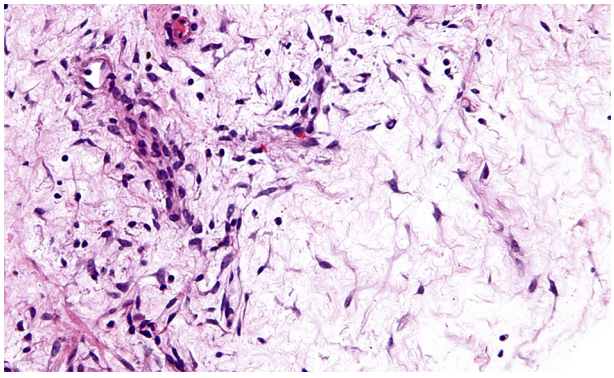

Figure 3. Histopathology showing characteristic thick walled blood vessels and dense, relatively acellular stroma.

Discussion

Aggressive angiomyxoma is a rare benign myxomatous tumor of males. Previous reports of AAM in males, attribute scrotum (38%), spermatic cord (33%), perineal region (13%), and intrapelvic organs such as the bladder (8%) to be the most commonly involved.[3] Involvement of penis is very rare. To the best of our knowledge only two cases of penoscrotal involvement by AAM have been reported in the literature so far. Rocco et al reported a case of massive recurring angiomyxoma of the scrotum obscuring the penis, in an obese man, which was managed by scrotal resection, bilateral orchidopexy and transposition of the penis. The patient soon developed a recurrence that involved the external meatus.[1] This was the only case reported so far with urethral involvement.

Xambre et al[4] reported on a case of aggressive angiomyxoma of the penoscrotum in a young male which was treated with wide local excision and plastic surgical reconstruction. Both the above mentioned reports highlight the infiltrative nature and tendency for local recurrence. Wang et al[5] have reported a case of superficial angiomyxoma of the penis, which unlike AAM had well defined borders and was easily resectable. In our case, the infiltrative lesion could only be partially resected form glanular tissues and a neomeatus was fashioned after the partial resection and a ventral meatotomy.

AAM is microscopically a stroma rich tumor, with low cellularity. Prominent blood vessels with hyaline thick walls and the anomalous muscular artery are the characteristic features. High proliferating cell nuclear antigen (PCNA) immunoexpression, together with the lack of expression of the p21 protein are associated with a high number of recurrences.[3] The tumor has been shown to express varying proportion of androgen, estrogen and progesterone receptors, making it amenable to hormonal manipulation.[2]

AAM should be distinguished from other benign tumors affecting the pelvis and the genital tract, such as intramuscular myxoma, myxoid neurofibroma, myxoid or spindle cell lipoma, superficial

angiomyxoma, angiomyofibroblastoma and angiomyolipoma; in the differential diagnosis one should also consider some malignant tumors with myxoid stroma, such as myxoid liposarcoma, myxoid malignant fibrous histiocytoma, and embryonal rhabdomyosarcoma.

Surgery is the mainstay of treatment for AAM. Incomplete resection can lead to recurrence. Recurrence rates can vary between 36 and 72 percent.[2] In our patient, multiple comorbidities precluded a complete excision and we had to resort to partial excision, scrotoplasty, neomeatus reconstruction internal urethrotomy, followed by hormonal therapy. Regular follow up is essential for AAM after surgery, in view of its locally recurrent nature and the optimal duration has to be determined from larger prospective studies.

Conflicts of interest

None.

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