Case Report

AGGRESSIVE ANGIOMYXOMA OF SCROTUM PRESENTING AS AN INGUINAL HERNIA

Safia Rehman\(^1\), Rooh-ul-Muqim\(^2\), Tamjeed Gul\(^3\), Mohammad Aziz Wazir\(^4\), Muhammad Zarin\(^5\)

ABSTRACT

Aggressive Angiomyxoma (AAM) is a rare mesenchymal tumor with a locally aggressive course. The most common location is in the pelviperineal region and occurs mostly in females of childbearing age. In males, it is extremely rare and thus is apt to be mistakenly diagnosed as an inguinal hernia or hydrocele. We report a case of 45 year old man who had right sided scrotal swelling for about four years. It was diagnosed as an inguinal hernia clinically. On further investigations, it was found to be a soft tissue tumor of the scrotum. The patient underwent scrotal exploration. At surgery, a huge lobulated gelatinous mass surrounding the right testicle was found. The mass was excised including the testicle and sent for histopathological analysis. Histologic examination of the excised mass revealed a nonencapsulated lesion composed of sparse spindle-shaped cells and a myxoid stroma with a prominent vascular component. The cells showed focal and weak positivity for desmin. The findings were thus consistent with aggressive angiomyxoma of the scrotum. The margins of the specimen were clear. The patient we report has so far done well with no evidence of recurrence.

KEY WORDS: Aggressive angiomyxoma, Myxoma, Myxoid tumours, Soft-tissue tumor, Scrotal tumor, Inguinal hernia.

INTRODUCTION

Mesenchymal neoplasms of the modified genital skin and mucosa are uncommon. This entity encompasses benign as well as malignant lesions and neoplasms with locally aggressive nature. Leiomyosarcoma and aggressive angiomyxoma are the most frequent histologic types. They are mostly seen in the pelvi-perineal region of the females.\(^1\) Very uncommonly they may present in male external genitalia.

Steeper and Rosai first described aggressive angiomyxoma (AAM) in 1983.\(^2\) Around 150 cases have been described in literature since then.\(^3\) Due to its locally infiltrative course and frequent recurrences it has been termed aggressive. It is called angiomyxoma because of its myxoid nature and prominent vascularity.
It is a mesenchymal tumor found mainly in the pelvis and perineum. Aggressive angiomyxoma is a non-metastasizing slow growing and locally infiltrative soft tissue tumor with a high rate of local recurrence. Clear surgical margins are thus mandatory and there must be a long term follow up. Because of its rarity, it is often initially misdiagnosed. In females, it is mistaken to be gynecological malignancy. While in males, it is confused with either a hernia or hydrocele when it presents in the scrotal region. Detailed radiological work-up may be helpful in detection, but histology is the gold standard for establishing the diagnosis.

**CASE REPORT**

A 45 year old man presented to us with a large right sided scrotal swelling for about four years which was diagnosed to be inguinal hernia previously. Now the swelling was noticed by the patient to have increased in size. There was also complaint of pain in the swelling for the past ten days. Past medical and surgical histories were unremarkable. The patient had five children and was leading a healthy marital relationship.

On examination, there was large right scrotal swelling about 15.0 × 8.0 cm. It was elastic, tender and did not trans-illuminate. The temperature of the swelling was normal. There was no cough impulse and it seemed irreducible. We were unable to palpate the right testicle separately from the swelling. Our first impression was that of inguinal hernia as the patient had been clinically diagnosed previously. Due to concerns about the exact nature of the swelling, we sent the patient for ultrasonography. It showed the swelling to be a homogeneous, hypo-echoic lesion with no calcification merging with scrotum and surrounding the right testicle. Pelvic computed tomography (CT) showed heterogeneous mass of right scrotum consistent with a soft tissue tumor. The patient was subjected to routine laboratory and radiological workup which were all normal. Chest X-ray, liver and renal function tests also revealed no abnormality. Alpha fetoprotein, β hCG and LDH were also found to be normal. A diagnosis of right para-testicular soft tissue tumor was thus being made and patient was counseled properly. A well-informed written consent was taken for wide excision including orchidectomy if need arose.

After completion of pre-operative workup, the patient was explored through inguinal incision. A large, lobulated, yellowish mass was found that was surrounding the right testicle. The tumor had soft and myxoid appearance on cut section and was extending from subcutaneous tissue under the skin. Since the testicle was fully surrounded and not separable, so orchidectomy along with wide excision was done in order to be sure of complete excision. The wound was not closed primarily and later on scrotoplasty was done for the patient. The specimen was sent for histopathological analysis. On receipt of the report, it was found to be aggressive angiomyxoma of 13.0 × 7.0 × 6.0 cm size with no capsule. There was no evidence of cytologic atypia, mitotic activity or necrosis. Desmin immunostain showed focal and weak positivity in spindle cells. It was hypocellular and composed of spindled to stellate cells with abundant collagen, loose to myxoid stroma with numerous associated capillaries. There were
also thin walled dilated arterioles with mild peri-vascular chronic inflammation. The excised testicle had seminiferous tubules with germ cells but decreased spermatogenesis. It was surrounded by 3.0 × 2.0 × 1.5 cm of the mass. The margins of the excised specimen were clear. The patient was discharged home after full recovery and is well on follow up for the past two months.

**DISCUSSION**

Aggressive angiomyxoma (AAM) is a rare vulvovaginal mesenchymal neoplasm with a marked tendency for local recurrence, but it usually does not metastasize. It is reported in 3rd to 5th decades of life with above 95% preponderance in females. A few cases have been described in males, usually in the scrotum. It arises from the soft tissues of the pelvic region, perineum, vulva and buttock in females. In a study reported from Pakistan, it was seen to comprise 10% of the benign tumors of the vulva. In men, AAM is usually derived from the mesenchymal tissue of scrotum, spermatic cord, perineal and gluteal regions and intrapelvic organs including bladder. AAM in the scrotal region may present as a scrotal mass, wrongly diagnosed as hernia or hydrocele as has been previously reported in few cases. In both males and females, the gross and microscopic appearances and clinical course are similar. Grossly it is usually large, poorly circumscribed gelatinous mass. It clinically simulates a Bartolin gland cyst or an inguinal hernia. It is usually painless, slow growing and locally infiltrating and has a high risk of local recurrence after excision. These recurrences may present even decades after the excision. This tumor usually does not metastasize but few cases have been reported in literature where metastasis have occurred.

On microscopic appearance, the tumor is infiltrative and composed of fibromyxoid stroma with sparse bland-looking spindled and stellate cells with delicate cytoplasmic processes.

Scrotal MRI is the best imaging modality. However, most cases have been radiologically diagnosed by computed tomography. In this regard, US-guided needle biopsy is not of much help. The differential diagnosis includes a series of benign and malignant soft tissue tumors so histo-pathology after radical excision is the gold standard for definitive diagnosis.

On Immunohistochemical studies, estrogen and progesterone receptor protein may be positive in females. While in males, androgen as well as progesterone receptor protein have been shown to be positive in a number of cases. This may serve as basis for anti-receptor therapy in future and needs further research. The neoplastic cells may show positivity for vimentin, muscle specific actin, desmin and CD34. There may be chromosomal translocation leading to rearrangement of the HMGIC gene.

Wide local excision with histologically clear margins is the recommended treatment and prognosis of such tumors is good. Long-term follow-up by pelvic MRI is necessary in all cases. Gonadotropin-releasing hormone agonist treatment has also been shown to be effective in reducing the size. The overall prognosis is quite good. In many cases with recurrence, further resection can be carried out. Reports about metastasis needs further research as there is very little data available in this regard.

**CONCLUSION**

Aggressive angiomyxoma (AAM) being a rare disease entity, is apt to be misdiagnosed. This is especially true of the aggressive angiomyxoma of the scrotum because of lack of awareness. So it must be considered in the differential diagnosis of scrotal swellings and careful preoperative workup should be carried out in suspicious cases.

Because of its rarity, there is no defined clinical protocol. Resection with clear margins if possible and radiographic follow-up is currently recommended.

**REFERENCES**


