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Case Report

Aggressive (Deep) Angiomyxoma of Vagina: A Rare Case Report

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Abstract

Aggressive angiomyxoma (AAM) is a rare, locally aggressive mesenchymal tumor that has a high propensity for local recurrence (exceeding 35%). It is extremely rare to metastasize and involves mainly pelvis, perineum, vulva, vagina and urinary bladder in women of reproductive age group with a peak incidence in 3rd and 4th decades. However, a few cases of its occurrence outside the pelvis have also been reported. AAM was identified as a distinct clinicopathologic entity in 1983 and since then fewer than 250 cases of this rare tumor have been reported in the world of literature. Angiomyxoma is called as aggressive due to neoplastic nature of blood vessels, locally infiltrative behavior and high risk for local recurrence; however, metastasis is extremely rare. Diagnosis is mainly made on histopathology after surgical resection and because of its rarity, it is often initially misdiagnosed. Surgical resection is the main treatment modality of angiomyxoma. We report a case of AAM in 45-year-old pre-menopausal female presented with a vaginal mass on anterior vaginal wall, in which diagnosis was only made after histological examination.

Key words: Aggressive angiomyxoma, Pelvic, Rare to metastasize, Reproductive age, Vulvo-vaginal neoplasm

INTRODUCTION

Case of aggressive angiomyxoma (AAM) was first described as a distinct clinic pathologic entity by Steeper and Rosai in 1983.[1] AAM is a rare tumor of mesenchymal origin. About 90% of the patients are women in reproductive age group with a peak incidence between 3rd and 4th decades of life.[1]

AAM usually arises in deep soft tissue of vulvo-vaginal region, perineum, pelvis of young adult females and analogs sites including scrotum and the inguinal area in males.[2] It is a slow growing tumor but problematic due to frequent local recurrence (30-72%).[3] Female to male ratio is 6.6:1.[4]

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However, metastasis is very uncommon.[4] Misdiagnosis is a very frequent problem.[3] Diagnosis is mostly made on histopathology following surgical resection.[3]

We report a case of AAM in pre-menopausal 45 years female presenting with vulvo-vaginal mass on anterior vaginal wall.

CASE REPORT

A 45-year-old premenopausal female patient, para$_2$ live$_2$ visited to gynecology outpatient department with complaints of a painless mass in the vagina gradually increasing in size over the last 8 years, now protruding out of the vagina and dyspareunia since the last 1 year. The patient had no relief of the symptoms on seeking medical advice from the local practitioner. No bladder or bowel complaints.

Menstrual History

Past menstrual cycles were regular. Last menstrual period was 7 ss before.

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Obstetrical History

Para$_2$ live$_2$, all full term normal delivery, last child birth 14 years before.

General and Systemic Examination

The patient was averagely built, afebrile, pulse was regular, mild pallor, and no icterus. Cardiovascular and respiratory system revealed no abnormality, per abdomen soft with no guarding and rigidity. No systemic signs of thyroid disorder found on examination.

Local clinical examination revealed a well-defined firm irregular mass of 7 cm × 6 cm × 5 cm, which was non tender, arising from anterior vaginal wall in the suburethral region, 4 cm below the urethral meatus with the wide base as shown in Figure 1. Posterior vaginal wall and cervix were healthy as seen in Figure 2. Uterus was anteverted and normal size, fornices free and non tender, per rectal examination revealed no mucosal involvement.

Investogram

Her investigations revealed hemoglobin: 11.8 g/dl, total white blood cell count (total leukocyte count): 12,680/cumm, platelet: 4.2 lakhs/cumm, serum bilirubin: 0.6 mg/dl, blood sugar random: 104 mg/dl, serum glutamic-oxaloacetic transaminase: 26 IU/L, serum glutamate pyruvate transaminase: 25 IU/L, blood urea: 29 mg/dl, and serum creatinine: 1 mg/dl. HIV and hepatitis B surface antigen were nonreactive. X-ray chest was within normal. Histopathology report of the pre-operative biopsy (done twice) showed inflammatory changes with no tissue diagnosis. Contrast-enhanced computed tomography (CECT) was performed for better characterization and the extension of tumor. It revealed an ill-defined heterogeneously enhancing soft tissue mass lesion of size 10.5 cm × 5.3 cm × 7.8 cm, arising from anterior vaginal wall, extending inferiorly in subcutaneous plane in the vulval region. Superiorly extending up to anterior lip of cervix. Fat planes between this mass and bladder were well maintained. Hence, surgery was planned after evaluation for possibility bladder involvement by the surgeons.

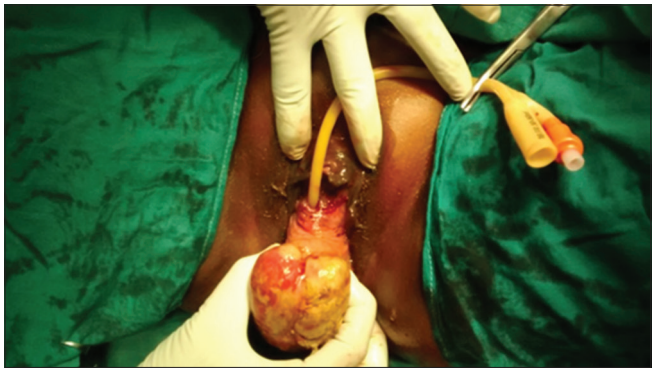

Figure 1: Anterior vaginal mass 7 cm × 6 cm × 5 cm size

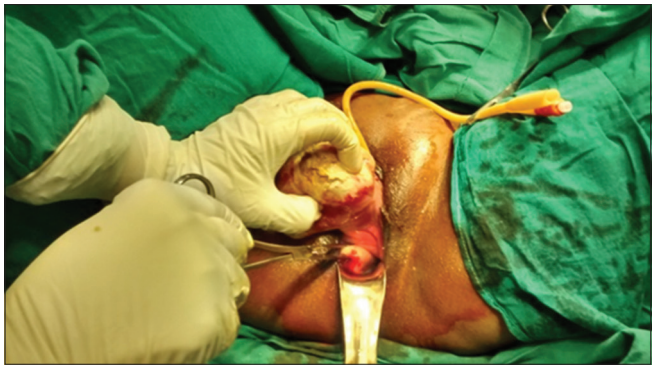

Figure 2: Cervix and its relation to mass 4 cm away from urethra

Procedure Performed

Removal of the vaginal mass was done in the lithotomy position. Under spinal anesthesia per speculum and per vaginal examination were done to confirm pre-operative findings. The patient was catheterized to facilitate the surgery and to prevent trauma to urethra. Mass was identified 4 cm below from urethral meatus. To develop the tissue planes normal saline was infiltrated around the mass, and the circumferential incision was taken at the base of mass (bladder approximately 2.5 cm away from the mass). The base of the pedicle was separated with sharp dissection, and the base of mass was dissected from the vaginal wall in the region of the pedicle. The dissection was easy. The base of the pedicle was reached posterior to symphysis pubis. The base of the pedicle was clamped as close to the hind surface of the symphysis pubis and the polyp growth was removed. At the end of the procedure, there was no visible or residual palpable mass. Hemostasis was achieved and the incision was closed with 2-0 vicryl. Vagina was packed and the pack was removed after 24 h catheter was removed after 7 days.

On gross examination, the mass was well circumscribed measuring 8 cm × 8 cm × 7 cm in size (Figures 3 and 4) and weighing 65 g.

On cut section, it appeared gelatinous, glistening, and bluish-gray.

On histopathology, hematoxylin and eosin stain revealed tumor tissue composed of small and large vessels lined by endothelium surrounded by smooth muscle coat with myxomatous stroma. Tumor covered with thin layer of epidermis with fairly uniform, moderate cellularity containing small, stellate-shaped and spindled cells, in myxomatous stroma suggesting angiomyxoma (Figrues 5).

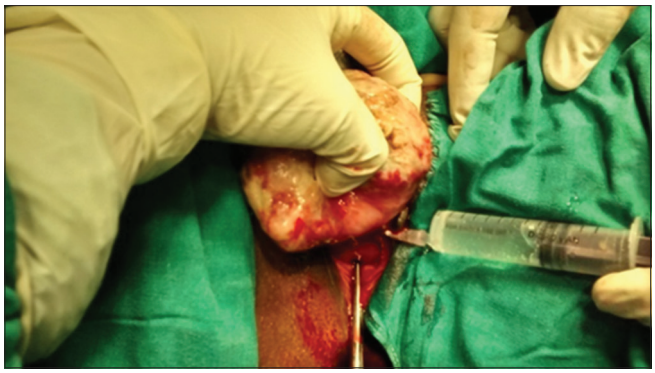

Figure 3: Normal saline infiltration

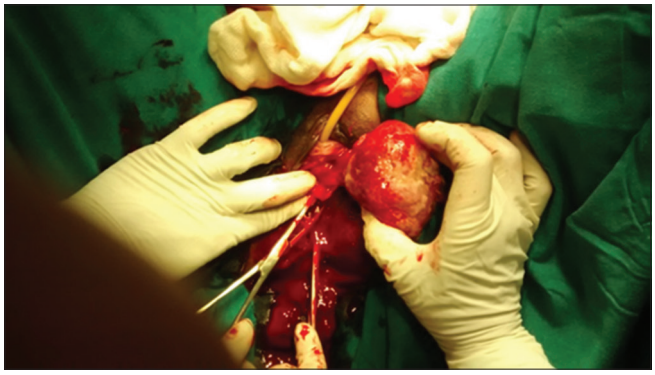

Figure 4: Mass enucleation

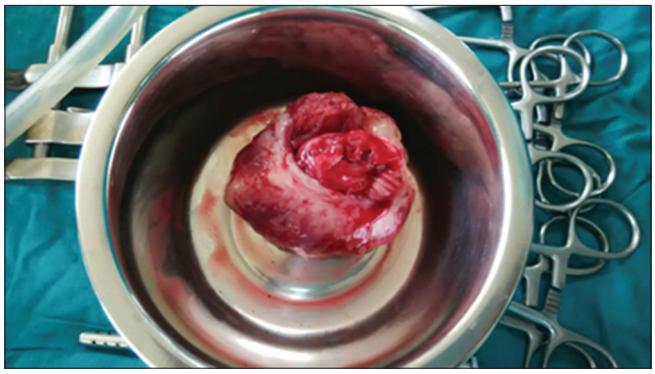

Figure 5: Excised angiomyxoma

Immunohistochemistroy was not done in this case as the patient was poor and at Government Medical College and Hospital, Aurangabad, this test is not available.

Post-operative patient was observed in the hospital for bleeding and urinary leaks for 7 days and was treated with prophylactic antibiotics. The follow-up was uneventful with no signs of recurrence (Figure 6).

Histopathology

Histopathology revealed thin uniform epidermis, small and large vessels lined by endothelium surrounded by smooth muscle coat. Stroma is myxomatous spindle and stellate cells seen (Figures 7 and 8).

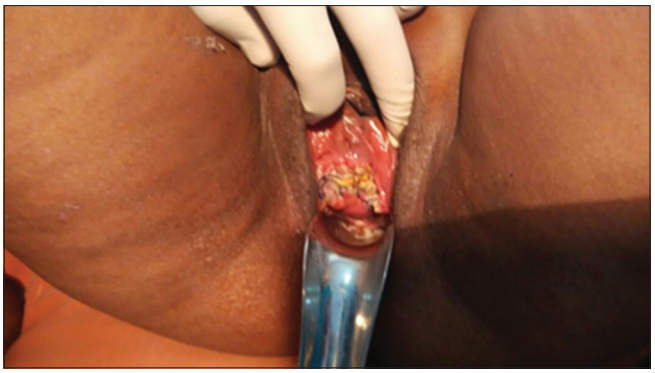

Figure 6: Wound after 7 days

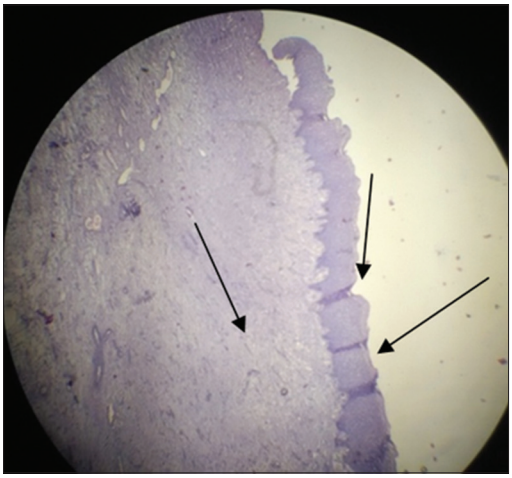

Figure 7: Slide showing epidermis with spindle cells In myxomatous stroma

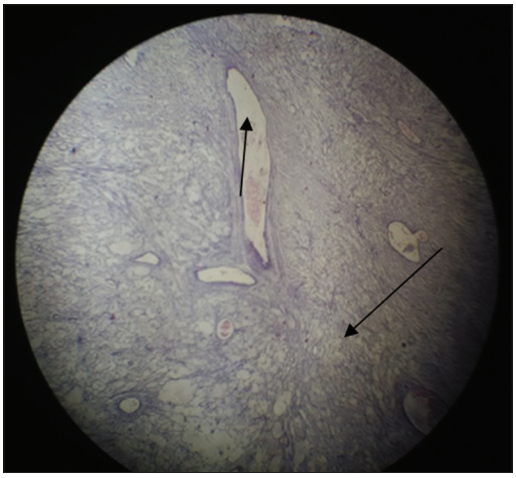

Figure 8: Slide showing prominant vascular componenet

DISCUSSION

The term AAM was coined by Steeper and Rosai in 1983 for a morphologically distinctive, slow growing, myxoid neoplasm that occurs mainly in genital, perineal, and pelvic

region of adult women.[1] It is of two types as follows:

1. Superficial which grows near surface[1]
2. Aggressive which grows and invades in deeper tissues and has tendency for recurrence,[1] 90% of patients are in reproductive age group.[2]

The patient usually presents with noticeable mass and rarely has pain.[3] The initial presentation varies from asymptomatic perineal or vulval nodule or polyp or perineal hernia to a pelvic mass diagnosed on imaging study.[3] In this case, premenopausal patient presented with painless mass increased in size over the last 8 years. Occasionally, tumor can be cystic and mistaken for bartholins, labial, or gartner duct cyst. The tumor characteristically grows slowly and insidiously. It usually takes 2 months to 17 years for patient to report to the hospital.[3]

Genetics

The pathogenesis of the AAM is poorly understood; however, genetic alterations along the 12 q chromosome region chromosomal abnormality involving chromosome 12q 13-15 have been implicated.[4] These translocations involve high mobility Group A protein (high-mobility group AT-hook 2 [HMGA2]).[5] HMGA2 belongs to a family of transcription factor that function during embryogenesis and usually not detected in adult tissues.[5] Cytogenetic analysis and fluorescent *in situ* hybridization have confirmed the presence of HMGA2 gene in the rearrangement in AAM.[5]

Immunohistochemically, most AAM express different combinations of estrogen and progesterone receptors and show immunopositivity for vimentin, desmin, smooth muscle actin, muscle specific actin, CD 34, and CD 44.[5,3] They showed strong immunoreactivity for actin but were negative for S-100 protein.[6]

Immunohistochemistry, in this case, was not done as the patient was poor and test is not available in Government Medical College, Aurangabad.

Gross Appearance

Tumor size ranges from 3 to 60 cm,[7] it can appear as polypoid, soft, bulky mass, or vaginal cyst.[7] External surface is smooth and usually neither encapsulated nor circumscribed.[8] It has gelatinous consistency with focal areas of congestion and hemorrhage on cut section.[5] It is usually homogeneous in consistency with no obvious nodularity.[7]

On gross examination, in this case, the mass was well circumscribed measuring 8 cm × 8 cm × 7 cm in size.

On cut section, it appeared gelatinous, glistening, and bluish-gray.

Microscopic Appearance

Spindle stellate cells separated with loose myxoid stroma focally rich in collagen fibrils, a prominent vascular component. Mitotic activity is extremely low.[9] In this case, histopathology reveal revealed tumor tissue composed of small and large vessels lined by endothelium surrounded by smooth muscle coat with myxomatous stroma. Tumor covered with thin layer of epidermis with fairly uniform, moderate cellularity containing small, stellate-shaped and spindled cells, in myxomatous stroma suggesting angiomyxoma.

Scanning

Several imaging modalities have been used in identifying and describing AAM.[4] Sonography usually reveals hypoechoic or cystic mass. CTss and magnetic resonance imaging (MRI) are useful in diagnosis and help in complete removal of tumor particularly arising in perineum, vulva, and bladder. AAM display unusual growth pattern with high signal density in T2 weighted MRI.[10] MRI scan shows a "swirled" pattern visible in the angiomyxoma and is more specific than CT scan, so imaging study of choice for these lesions.[3] In this case, CECT revealed an ill-defined heterogeneously enhancing soft tissue mass lesion of size 10.5 cm × 5.3 cm × 7.8 cm, arising from anterior vaginal wall, extending inferiorly in subcutaneous plane in the vulval region extending superiorly up to anterior lip of cervix. Fat planes between this mass and urinary bladder were well maintained.

Differential Diagnosis of AAM[3]

- Angiomyofibroblastoma
- Bartholin gland cyst
- Vaginal polyp
- Leiomyoma
- Leiomyosarcoma
- Lymphangioma
- Malignant fibrohistiocytoma
- Myxolipoma
- Myxoid leiomyoma
- Myxoid neurofibroma
- Slerosing hemagioma.

Treatment and Prognosis

The first line of therapy for AAM is surgery[5,7,10] although achieving negative resection margins are difficult because of the infiltrative nature of tumor and the absence of defined capsule.[5] Smaller and more superficial tumor of the vulva or vagina may be removed with the wide local excision, but larger, deep-seated tumors of the pelvis may require more extensive surgery with partial or complete resection of some pelvic organ.[5] The recurrence rate is very high (>35%). Most of the AAM tumor shows estrogen and progesterone receptors and is likely to be hormone

dependent.[11] Hormones can be given pre operatively to decrease tumor size. Pre-operative GnRH analogs has been used successfully in few instance in premenopausal women with AAM having positive estrogen and progesterone receptors.[2,11] Pre-operative use of GnRH analog decreases the tumor size and makes the complete removal of the tumor feasible.[2] Due to the low mitotic activity in these tumors radiotherapy and chemotherapy are of not much help.[3] Angiographic embolisation has been attempted to shrink the tumor.[2] Two cases of successful control of recurrent angiomyxoma with relatively high doses of external radiotherapy have also been reported.[2] Two cases of metastasis had been reported in literature as follows:

1. Pulmonary and mediastinal metastasis
2. Metastasis to the lung.

Recurrence is one of the unique characteristics of these otherwise nonmalignant tumors. The usual sites of recurrence reflect the site of the primary disease, i.e., perineum, pelvis. No definite relation between patient's age, size of tumor and rate of recurrence has been established so far.[2]

CONCLUSION

Many options for the treatment of recurrence such as repeat surgery radiotherapy and hormonal therapy have been tried with varying success, but no single modality is clearly beneficial over others. All patients need long-term follow-up, usually with the clinical examination and MRI to detect early recurrence. In our case, we resected the tumor with no visible or palpable residual tumor and there are no signs of recurrence after 3 months.

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