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REVIEW ARTICLE: AGGRESSIVE ANGIOMYXOMA OF VAGINA

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ABSTRACT

Aggressive angiomyxoma is a rare, slow-growing soft tissue tumor that usually arises in the pelvis and perineal regions of women in reproductive age. It has marked tendency for recurrence. Surgical resection is the main treatment modality of aggressive angiomyxoma. We describe a case of a vaginal aggressive angiomyxoma in a 40-year-old woman in which the diagnosis was only made after histological examination. Pre-operative diagnosis is difficult due to rarity and absence of diagnostic features, but it should be considered in every mass in genital, perianal and pelvic region in a woman in the reproductive age.

KEYWORDS: Aggressive Angiomyxoma, Vagina.

INTRODUCTION

Aggressive angiomyxoma (AA) is a rare tumor of mesenchymal origin first described in 1983 by Steeper and Rosai.[1] They reported nine cases in young female patients. Since then, less than 150 cases have been reported in the literature[2]

AA is a rare locally aggressive myxoid mesenchymal tumor, usually arising in the deep soft tissues of the vulvovaginal region, perineum and pelvis of young adult females of reproductive age group, especially between third and fourth decade of life. It is a slow growing tumor and frequent local recurrences (30 - 72%) are noted.[1,6,7] However, the recurrence rate in patients with narrow surgical margins is not higher than the one in patients with wide surgical margins.[8] However, metastases are very uncommon[3] and misdiagnosis is a very frequent problem.[4] Diagnosis, more often than not, is made at histological examination following surgical resection.[5]

We describe a case of vaginal mass in a 40-year-old woman in which the diagnosis of AA was made after histological examination, and discuss management options and subsequent follow-up.

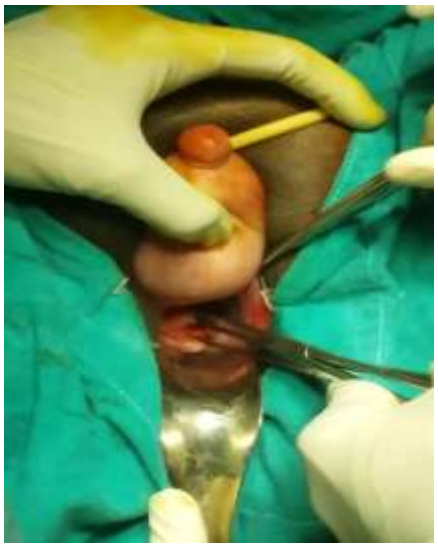
CASE REPORT

A 40-year-old female patient visited the gynecological clinic in December 2017 with chief complaint of mass coming out of vagina since 6 months. She had history of previous two caesarean sections. She underwent Bilateral Tubal ligation 15 yrs back. She had menopause since June 2017.

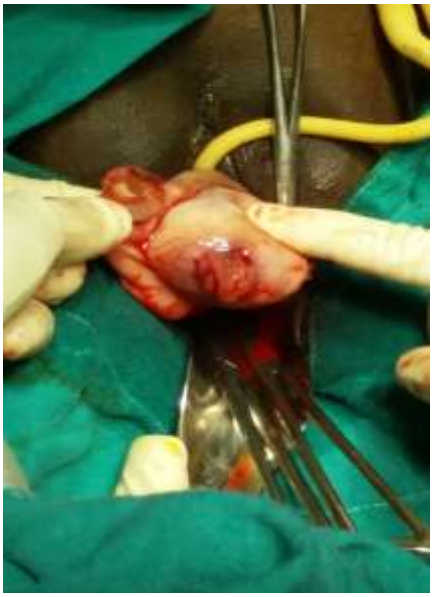
On Local examination, there was a nontender cystic mass in the upper vaginal wall. Per speculum examination was done. Findings were 5cm x 5cm mass seen protruding out of vagina with 1cm x 1cm area of erosion over it and no vaginal rugosities. White discharge was present. Sample taken for Routine Microscopy examination PAP smear was taken. Per vaginal examination Cervix was patulous, high up in position and was present behind the mass as shown in Picture 1. Uterus was normal sized and Retroverted. Complementary investigations were performed, such as Pap test, blood count, pelvis ultrasonography, which did not reveal any pathologic findings. Routine microscopy of white vaginal discharge revealed presence of pus cells. Patient was given vaginal pessary and asked for follow up. Provisional diagnosis of Gartner cyst was made. Decision of excision of cyst was taken. Pre aneasthetic Evaluation was done and patient was posted for surgery.

Under spinal anaesthesia, surgical excision of cyst was done Intraoperatively, 8cm x 6cm x 6cm cystic mass was seen arising from vaginal mucosa just inferior to bladder neck from 10'o clock to 2'o clock position, Pictures 2,3,4. It was sent for Histopathological examination. Report was suggestive of Aggressive Vaginal Angiomyxoma.

The patient was advised for follow up in case of recurrence. There was no local recurrence.



Picture-1.



Picture 2:

Picture-3.

Picture-4.

DISCUSSION

Aggressive angiomyxoma of the genital region is a rare condition that has been reported to exhibit a peak incidence in young women in the third and fourth decades of life.[3]

Patients usually present with complaints of a noticeable mass and rarely have pain.[4] The size of AAs can fluctuate widely, but most of them are >10 cm.[5,13] The largest AA was described in 1998 by Chen et al.,[12] who published a case report concerning a 38-year-old woman with a mass of 57x47x23 cm that had developed slowly over about 8 years, but particularly during pregnancy. There is no correlation between the disease and the ethnic origins, but the majority of patients are white.[4] The tumor preferentially involves the pelvis, often located in the vagina, vulva, perineum or buttocks.[13] The tumor characteristically grows slowly and insidiously. It usually takes 2 months to 17 years for the patients to go to the hospital with a painless mass after the initial discovery. It may grow and occupy the whole pelvic region and invade paravaginal and pararectal spaces displacing pelvic structures.[5,15]

Genetics

Chromosomal translocations involving 12q13-15 have been reported in a variety of mesenchymal neoplasms, including lipomas, liposarcomas, leiomyomas, and pulmonary hamartomas. These translocations involve the high mobility group A (*HMGA2*) gene. *HMGA2* belongs to a family of transcription factors that function during embryogenesis and are not usually detected in adult tissues. Cytogenetic analysis and fluorescent in situ hybridization have confirmed the presence

of *HMGA2* gene rearrangement in AAM and have shown that AAMs, or at least a subset of them, have the same molecular genetic background as other common mesenchymal tumors.

Pathological Features

On gross examination, these tumors are characteristically soft, bulky masses. The external surface is smooth and usually appears not to be encapsulated. Some have finger-like projections that extend into neighboring tissues. A bluish grey tumor of homogeneous, gelatinous consistency with focal areas of congestion and hemorrhage is seen on cut section. AAs generally displace rather than invade adjacent viscera, and they are rarely destructive. Although invasions are rare, AAs can invade the bladder, bowel and pelvic bone.[3,9,10,15,18,19,20]

Histologically, the tumor consists of a rather hypocellular population of small, somewhat stellate and spindle-shaped neoplastic cells with thin cytoplasmic processes.[3,5,15,19] The cells are scattered in a loose myxoid matrix composed of delicate wavy collagen fibrils and hyaluronic acid, which gives the tumor a pale-pink color by eosin staining.[15,18,19] There is also an accompanying prominent vascular component, ranging from tiny capillary-like vessels to larger and thick-walled vessels with a distinct smooth muscle cell but with no evidence of anastomosis or arborization.[3,10,15] The histogenesis of aggressive angiomyxoma has been controversial. It is likely that aggressive angiomyxoma arises from specialized stromal cells of the lower genital tract. Immunohistochemically, the tumor cells are positive for vimentin and weakly positive for desmin but not for myosin. These findings suggest a fibroblastic origin and differentiation of this tumor.[15,21] AA usually expresses estrogen (ER) and progesterone receptors (PR). However, dermal fibroblasts in normal vulvar skin and stromal cells in a variety of vulvar lesions can also be positive.

Differential Diagnosis

The differential diagnosis with other soft tissue tumors of the female genital tract is not always clear. Among the most common are: cyst of the bartholin gland, leiomyomas, vulvar lipomas, angiomyofibroblastoma, malignant fibrohistiocytoma, myxolipoma, myxoid neurofibroma, myxoid leiomyoma, myxofibrosarcoma, leiomyosarcoma, lymfangioma, neurofibroma malignant mesenchymoma, sclerosing hemangioma and botryoid rhabdomyosarcoma, among others. The distinctive striking vascular component in AA helps in ruling out most of above-mentioned neoplasms.[15,21]

It is a slow growing tumor, but local recurrences are frequent (30-72% of the cases), sometimes even years later. The term "aggressive" was chosen to emphasize the neoplastic nature of the blood vessels, its locally infiltrative nature and the high risk of local recurrence, not to indicate its malignant nature.[1,15,21] However, there are two reports of metastasized disease in the literature. Siassi et al.[22] described a case of a 63-year-old woman with pulmonary and mediastinal metastatic disease. Blandamura et al.[23] described the other case in a young woman with multiple local recurrences and metastases in the lungs.

Owing to its similar clinical presentation to common lesions such as bartholin duct cyst, lipoma, vulvar mass or vulvar abscess, edema caused by chronic venous stasis in the pelvis due to compression, Gartner duct cyst, vaginal cyst, vaginal prolapse or levator hernia, it is often misdiagnosed.[3] So, most cases are diagnosed on histology after primary surgical excision[18] just like in our case. It has been reported that the misdiagnosis rate of AA varies from 70 to 100%.[14] Even after sonography, computed tomography (CT) and MRI preoperatively, the diagnosis of AA can be missed.[24]

Radiological Features

MRI, which has a "swirled" pattern visible in the angiomyxoma, is more specific than CT and is thus the imaging study of choice for these lesions. On T1, the tumor shows isosignal compared to the muscles, similar to presentation on CT, and moderate contrast enhancement on a gadolinium scan. On T2-weighted MRI, the tumor has high signal intensity. The attenuation on CT and high signal intensity on MRI are likely related to the loose myxoid matrix and high water content of angiomyxoma. Both CT and MRI show the translevator and the transdiaphragmatic extent of these tumors.[3,9,10,12,13,19,25,26]

In the present case, preoperative MRI of the patient was not done, as the clinical appearance was suggestive of a vaginal cyst.

Treatment

One should aim for complete resection, but incomplete or partial resection is acceptable, especially when high operative morbidity due to extensive surgery is anticipated and preservation of fertility is an issue. Long-term follow-up and careful monitoring with imaging techniques are essential for timely identification of recurrence.[3,5,8,9,10,28,29]

A newer approach to the treatment of AAM involves hormonal therapy. Given that the tumor occurs primarily in premenopausal women and is positive for estrogen receptor and progesterone receptor, estrogen and progesterone may play a role in its development. In one patient, a vulvar AAM was discovered during the first trimester of pregnancy and progressively increased in size throughout the pregnancy. Several case reports using a gonadotropin-releasing hormone agonist as medical management for AAM showed complete radiographic resolution of the tumor. Although surgery remains the standard of care, the medical treatment of AAM with a gonadotropin-releasing hormone agonist in the primary or adjuvant setting may offer an alternative to radical

surgery.[11] Long-term follow-up by careful clinical examination is necessary for detection of recurrence.

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