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## Case Report

# Aggressive angiomyxoma of the vulva: an uncommon entity

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### ABSTRACT

AA is a rare locally invasive mesenchymal tumor predominantly presenting in women of reproductive age group, having a moderate to high risk for local relapse. Hence, it needs to be differentiated from another mesenchymal tumor occurring in this region. We present a case of a 50 year old female with a large, polypoidal, spongy mass on the right labia majora.

**Keywords:** Aggressive angiomyxoma, Labia majora, Mesenchymal tumor

### INTRODUCTION

Aggressive angiomyxoma is a rare tumor with higher risk of infiltration and recurrence. The AA was first described by Steeper and Rosai in 1983.<sup>1</sup> Tumor is aggressive due to its characteristically slow and insidious growth as well as carrying a moderate to high risk of local relapse. This tumor pathogenesis is unclear.<sup>2</sup> It occur predominantly in vulvo-vaginal, perineal and groin region. Diagnosis is made by histology examination.<sup>3-6</sup>

Most of the cases reported have been in women at reproductive age and those in forth decade of life. The AA can affect the women and men in proportion of 6:1. Estrogen and progesterone receptor are commonly found in AA thus it is likely to grow during pregnancy and respond to hormonal manipulators. Considering its nature of aggression and chances of local relapse, appropriate management and long term follow up are necessary to diagnose early recurrence.

No single modality of treatment has been found to be proven benefit till now. However complete surgical excision- when possible should be sought. Where fertility is to be preserved or surgery is likely to be extensive and

mutilating, incomplete resection is acceptable as local recurrences can be treated with further resection.<sup>7</sup> Radiotherapy and chemotherapy have been used as adjunctive therapies but are unlikely to be useful as it has few mitotic activity.<sup>8-12</sup> Despite of availability of many options of treatment, recurrence of AA is reported to be as high as 72%.

### CASE REPORT

A 50 Year old perimenopausal female (P2L2) presented with a slow growing mass on right labia majora since 6 month. She had history of similar right labial swelling 12 years back for which she underwent excisional surgery. Details are not available. There was no history of any vulval discharge, bleeding, sexual difficulty, or pain, except a sensation of weight hanging while standing. Local examination showed a well circumscribed polypoidal mass measuring 12\*6 cm. The mass was nontender, soft and spongy in consistency (Figure 1).

She is diabetic and on oral hypoglycemic drug. Blood investigations revealed uncontrolled sugar level. Ultrasonography of local part showed a large heteroechoic lesion measuring 52\*70\*98 mm in right labia majora

with internal vascularity. Few prominent ovoid lymph nodes with preserved fatty hilum seen in right inguinal region.



**Figure 1: A well-defined polypoidal 12\*6cm right labia majora swelling.**

CT suggested multiloculated polypoidal complex cystic lesion in right labial region. With a clinical diagnosis of a vulvar fibroepithelial polyp or lipofibroma, she underwent for local excisional surgery (Figure 2).



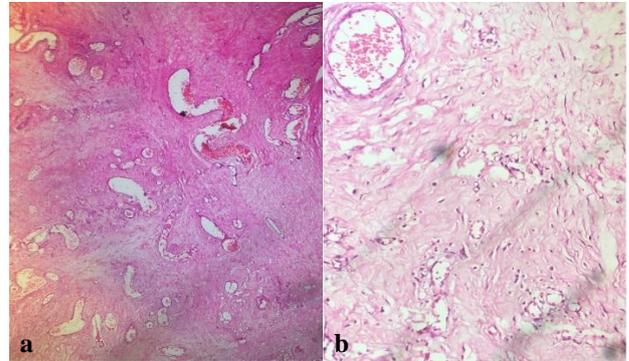
**Figure 2: Local excisional surgery.**

There was moderate bleeding during the procedure. The cut surface revealed a glistening, gelatinous, and soft homogeneous appearance (Figure 3).



**Figure 3: Specimen of angiomyxoma showing cut open homogenous white area.**

On histopathology, Tumor composed of spindle fibroblast in hypocellular myxoid stroma. Many prominent dilated thick-walled blood vessels are seen. Admixed component of mature adipose is also noted. No nuclear atypia, hemorrhage or necrosis seen (Figure 4 a and b).



**Figure 4: a-Angiomyxoma, b-High power picture of angiomyxoma (H and E).**



**Figure 5: Post-operative picture.**

## DISCUSSION

The AA tumor commonly presents as an asymptomatic mass in the genital area of women in their reproductive life. The term aggressive denotes its propensity for local aggression and recurrence after excision. Clinically, AA may misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, etc. Superficial angiomyxoma, angiomyofibroblastoma, cellular angiofibroma, and smooth muscle tumors also need to be considered in the differential diagnosis of a polypoidal mass in the perineum. AA is an infiltrative tumor whereas angiomyofibroblastoma is well circumscribed. In addition, AA has thick-walled vessels, which are less numerous than thin-walled vessels in angiomyofibroblastoma.

On CT scan, these tumors have a well-defined margin with attenuation less than that of muscle. The attenuation on CT and high signal intensity on magnetic resonance imaging (MRI) are likely to be due to the presence of loose myxoid matrix and high water content of AA.<sup>7</sup>

Usually, this tumor does not metastasize, but there are reports of multiple metastases in women treated initially by excision and ultimately succumbing to it.<sup>3,4</sup>

Partial excision may have to be done in view of high operative morbidity.<sup>9</sup> Unfortunately, recurrence may still occur with negative margins.<sup>10</sup> This may necessitate multimodal therapies using surgical and medical means to treat recurrent AA.<sup>11</sup> There is no consensus regarding the pathogenesis of AA. This hormonally responsive tumor is believed to arise from specialized mesenchymal cells of the pelvic-perineal region or from the multipotent perivascular progenitor cells, which often display variable myofibroblastic and fibroblastic features.<sup>5</sup> Immunohistochemically, most AA express different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth muscle actin CD34, and CD44, but all are invariably negative for S-100, carcinoembryonic antigen, and keratin.<sup>9,11,12</sup>

Recent cytogenetic and molecular studies have revealed a variety of genetic alterations, involving the chromosome 12, in the region 12q13-15. A gene in this region, called high-mobility group protein isoform I-C (HMGI-C), which encodes proteins involved in the transcriptional regulation, appears to have a role in the pathogenesis of this tumor. Detection of inappropriate HMGI-C expression using the immunoperoxidase technique with anti-HMGI-C antibody may potentially be a useful marker for microscopic residual disease.<sup>6</sup>

Our patient had previous history of similar swelling 12 years before exact data not available but considering it as a relapse of AA we decided to start preventive therapy, i.e., GnRH agonist, etc.

Han-Geurts et al proposed the following guideline for treating AA:

- complete excision of the lesion when possible, avoiding mutilating surgery,
- adjunct therapy using arterial embolization and/or hormonal treatment needed in case of partial resection of the tumor, and
- radiotherapy is reserved for cases that are resistant to embolization and/or hormonal therapy and still symptomatic.<sup>2</sup>

There are no specific guidelines for postoperative management of vulvar AA; however, due to high recurrence rate and potential morbidity associated with undiagnosed recurrences, several authors recommend a periodic evaluation with physical examination and MRI up to 15 years after excision.<sup>2,8</sup>

CT and MR imaging accurately reveal extension of these tumors across the pelvic diaphragm and are thus valuable in determining the surgical approach.<sup>13</sup>

## CONCLUSION

This case report illustrates the challenges that a physician might face when dealing with a vulvar mass which may be an AA. Though it is a rare entity, it should always be considered, especially when it is an insidious painless lesion. High level of suspicion is needed to make a clinical diagnosis. All relevant hematological and radiological studies including MRI or CT scan should help in reducing the number of misdiagnosed cases of AA preoperatively. Once its anatomical location and extension, if any, are defined, any vulvar tumor particularly AA-can be optimally treated by surgical excision only, while avoiding any mutilating surgery. If complete resection is possible under the circumstances, one should expect lowest recurrence rate. AA is rarely life-threatening, and therefore one can afford to have a partial resection when high operative morbidity is anticipated. Irrespective of treatment modalities instituted postsurgery, it is evident that AA requires close and long-term follow-up.

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