CASE REPORT

An aggressive angiomyxoma of vulva - A rare entity - A case report

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ABSTRACT

Aggressive angiomyxoma is a rare, locally invasive mesenchymal tumor predominantly presenting in women of reproductive age and also having a moderate-to-high risk for local relapse. Hence, it needs to be differentiated from other mesenchymal tumors occurring in this region. We present here a case of a 40-year-old female presenting with a large, fleshy, pedunculated mass on the right labia majora.

Key Words: Aggressive angiomyxoma, labia majora, mesenchymal tumor

INTRODUCTION

Aggressive angiomyxoma (AA) was first described by Steeper and Rosai.[1] The tumor was named aggressive due to its characteristically slow and insidious growth as well as carrying a moderate-to-high risk of local relapse. Usually, it presents as a vulval polyp clinically and is diagnosed only on histopathology. It is a rare local mesenchymal tumor of unknown etiology usually affecting vulva, perineal region, buttocks, or pelvis of women in reproductive age.[2-6] Less than 250 cases have been reported till 2010.[7] Estrogen and progesterone receptors are commonly found in AA.[2] Thus, it is likely to grow during pregnancy and respond to hormonal manipulation. Considering its nature of aggression and chance of local relapse, appropriate management and long-term follow-up are necessary to diagnose early recurrence. No single modality of treatment of recurrence has been found to be of proven benefit till now. However, complete surgical excision – when possible – should be sought. Radiotherapy and chemotherapy have been used as adjunctive therapies but are unlikely to be useful as it has few mitotic activity.[8-12] Despite the availability of many options of treatment, recurrence of AA is reported to be as high as 72%.[9]

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CASE REPORT

A 40-year-old female para 2 presented with a swelling on the right labia majora with a duration of 3 years growing slowly throughout and increased in size for 6 months [Figure 1].

There was no history of any vulval discharge, bleeding, sexual difficulty, or pain, except a sensation of weight hanging while standing. Menstrual cycles were regular with a normal flow. Local examination revealed a well-circumscribed pedunculated fleshy polypoidal mass measuring 18 cm × 10 cm. It was soft, spongy in consistency, and nontender. The overlying skin had patches of pigmentation probably carrying evidence of healed ulceration. The inguinal lymph nodes were not enlarged bilaterally. Her blood reports and ultrasound of abdomen

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showed no abnormality. Ultrasonography (USG) of the swelling revealed heterogeneous hyperechoic areas with peripheral vascularity, thick echoes, and nonvascular central areas. Computed tomography (CT) scan of pelvis revealed no pelvic disease inside and had similar findings of the mass like USG. With a clinical diagnosis of a vulvar fibroepithelial polyp or lipofibroma, she underwent local excision of the tumor with ligation of the stalk [Figure 2]. There was moderate bleeding during the procedure. The cut surface revealed a glistening, gelatinous, and soft homogeneous appearance [Figure 3]. On histopathology, the tumor was composed of spindle- and stellate-shaped cells scattered in a myxoid background [Figures 4 and 5]. These cells had eosinophilic cytoplasm and lacked significant nuclear atypia or mitosis. In lipofibroma, there would be spindle cells in fatty background with minimum of vessels unlike AA. There were many variable-sized thin-walled and thick-walled vascular channels in the histopathological specimen, which were diagnostic of AA. A 6 monthly follow-up has been done for more than 2 years now without any specific therapy for the prevention of recurrence, yet showing no sign of any relapse so far.

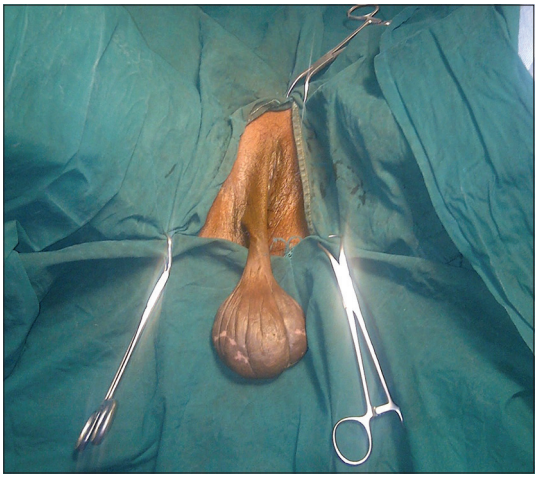

Figure 1: Pedunculated large, partly encapsulated growth from vulva

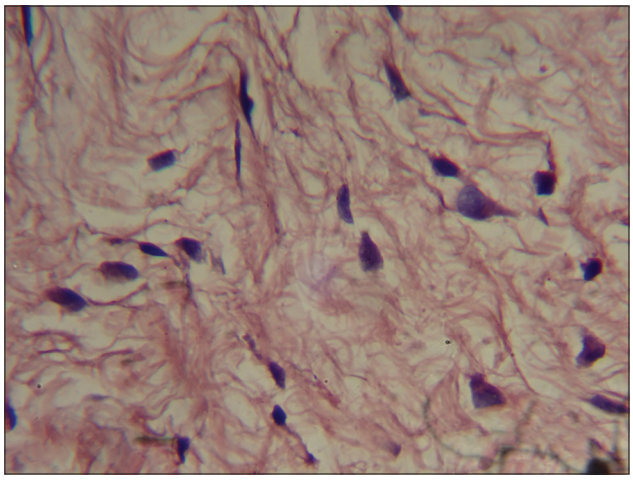

Figure 3: High power picture of angiomyxoma, (H and E, ×40)

DISCUSSION

The AA tumor commonly presents as an asymptomatic mass in the genital area of women in their reproductive life, but is occasionally reported in men (male-to-female ration being 1:6).[2] The term "aggressive" denotes its propensity for local aggression and recurrence after excision. The tumor is not always aggressive, and recurrence rate is about 30%. 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 angiomyofbroblastoma is well circumscribed. In addition,

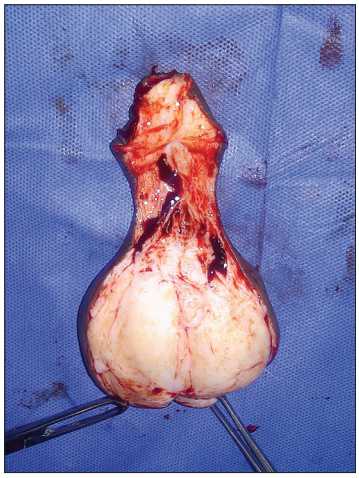

Figure 2: Cut open specimen of angiomyxoma showing homogeneous white area

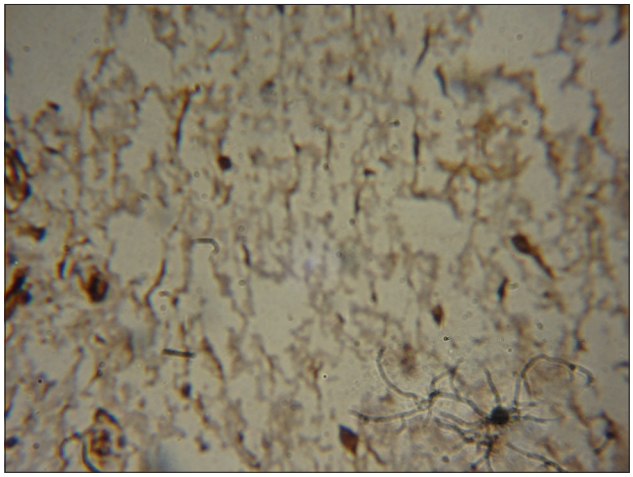

Figure 4: Immunohistochemistry picture of angiomyxoma, $CD34^{+}$ (low power)

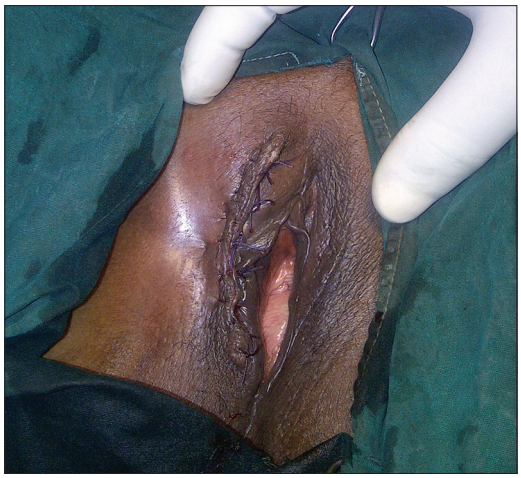

Figure 5: Postoperative picture of vulva after resection of angiomyxoma

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.[13] Usually, this tumor does not metastasize, but there are reports of multiple metastases in women treated initially by excision and ultimately succumbing to it.[3,4] Partial excision may have to be done in view of high operative morbidity.[9] Unfortunately, recurrence may still occur with negative margins.[10] This may necessitate multimodal therapies using surgical and medical means to treat recurrent AA.[11] 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.[5] 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.[9,11,12] 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 protens 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.[6]

Unlike most AAs, our case was completely encapsulated without any breach in continuity and without any projections into the neighboring tissues. The pathologist reported negative margins on excised mass. This may be the reason why there was no recurrence in the past 2 years or so. Considering her very poor economic status and the HPE report, we decided not to start any preventive therapy, i.e., GnRH agonist, etc., in her case. On H and E staining, the tissue resembled a typical AA, and immunohistochemically, it showed positive for CD34, desmin, and vimentin, hence proving beyond any doubt that it was a case of vulval AA.

Our patient required no additional treatment or any investigations postoperatively till date and has been asymptomatic and enjoying good health. Despite this fact, AA is notorious for local recurrence in approximately 70% of the cases after a period of 2 years postoperatively,[7] and it has been reported even 20 years after surgery as well.[4] Han-Geurts *et al*.[2] proposed the following guideline for treating AA: (1) Complete excision of the lesion when possible, avoiding mutilating surgery, (2) adjunct therapy using arterial embolization and/or hormonal treatment needed in case of partial resection of the tumor, and (3) radiotherapy is reserved for cases that are resistant to embolization and/or hormonal therapy and still symptomatic.

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.[2,8]

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, particularly in premenopausal women in their third to fourth decades of life. 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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Conflicts of interest

There are no conflicts of interest.

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