Case Report

An aggressive angiomyxoma of vulva, an uncommon entity: a case report

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Abstract

Introduction: Aggressive angiomyxoma is a rare, locally invasive mesenchymal tumour predominantly presenting in women of reproductive age and also having a moderate to high risk for local relapse. So it needs to be differentiated from other mesenchymal tumours 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.
Keywords: Aggressive angiomyxoma, mesenchymal tumour, labia majora.

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INTRODUCTION

Aggressive Angiomyxoma (AA) was first described by Steeper and Rosai[1]. The tumour 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 tumour 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]. Few cases were reported in adult male and children (8-13years) also[8,9]. 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 is necessary to diagnose early recurrence. No single modality of treatment of recurrence have been found to be of proven benefit till now. However complete surgical excision – when possible- should be sought. 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]. Adjunct hormonal treatment with tamoxifen, raloxifene and GnRH analogues , has been described with varying degrees of success ranging from noresponse to complete remission of primary and recurrent AA[8,9,11]. Radiotherapy and chemotherapy have been used as adjunctive therapies but are unlikely to be useful as it has few mitotic activity[8,12]. Due to its rarity, the role of sentinel lymph node biopsy and lymphadenectomy in AA is still debatable. Despite availability of many options of treatment, recurrence of AA is reported to be as high as 72%[9].

CASE REPORT

A 40 year old female para 2 presented with a swelling on the right labia majora with duration of 3 years and growing slowly through out and increase in size since 6months[fig 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 normal flow. Local examination revealed a well- circumscribed pedunculated fleshy polypoidal mass measuring 18cmx10cm. It was soft,

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spongy in consistency and non-tender. The overlying skin had patches of pigmentation probably carrying evidence of healed ulceration. The inguinal lymphnodes were not enlarged bilaterally. Her blood reports and ultrasound of abdomen showed no abnormality. USG of the swelling revealed heterogenous hyperechoic areas with peripheral vascularity, thick echoes and non-vascular central areas. CT scan of pelvis revealed no disease inside and had similar findings of the mass like USG. With a clinical diagnosis of a vulvar fibroepithelial polyp or lipofibroma, she underwent a local excision of the tumour with ligation of the stalk. [fig2]. There was moderate bleeding during the procedure. The cut surface revealed a glistening, gelatinous and soft homogeneous appearance. [fig3]. On histopathology, the tumour was composed of spindle and stellate-shaped cells scattered in a myxoid background. [fig 4]. These cells had eosinophilic cytoplasm and lacked significant nuclear atypia or mitosis. There were also many variable-sized thin walled and thick walled vascular channels. This was diagnostic of AA. A six-monthly follow-up has been done for more than 2 years now and without any specific therapy for prevention of recurrence showing no sign of any relapse so far.

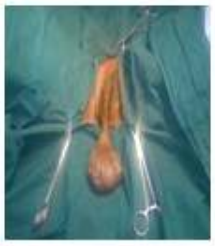

Figure 1: Pre-Operative picture of Vulval aggressive angiomyxoma (right vulva)

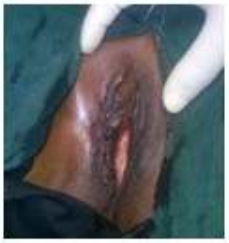

Figure 2: Picture of Vulva after surgical excision of the tumour mass

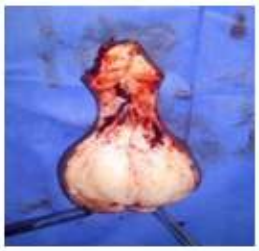

Figure 3: Cut open section of the tumour mass (Vulval tumour): well circumscribed and well capsulated fleshy gelatinous mass

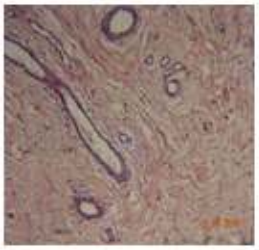

Figure 4: Low Power (10X) H and E slide showing widely scattered spindle cells within a myxoid background with accompanying vascular structures

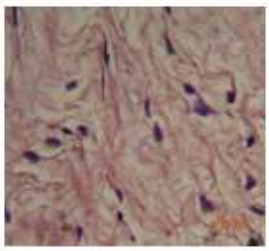

Figure 5: High Power (40X) HandE slide showing widely scattered spindle cells within a myxoid background with accompanying vascular structures

Figure 6: Low Power Immuno Histochemistry showing CD 34+ cells

Figure 7: Low Power Immuno Histochemistry showing Desmin +

DISCUSSION

The tumour AA 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 tumour is not always aggressive and recurrence rate is about 30percent.Clinically AA may misdiagnose as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst etc. Superficial angiomyxoma, angiomyofibroblastoma, cellular angiofibroma and smooth muscle tumours also need to be considered in the differential diagnosis of a polypoidal mass in the perineum. AA is an infiltrative tumour whereas angiomyofbroblastoma is well-circumscribed. Also, AA has thick-walled vessels, which are less numerous than thin-walled vessels in angiomyofibroblatoma. On computed tomography (CT) scan, these tumours have a well-defined margin with attenuation less than that of them muscle. The attenuation on CT and high signal intensity on magnetic resonance imaging (MRI) are likely to be due to presence of loose myxoid matrix and high water content of AA[13]. Usually, this tumour does not metastatize, but there are reports of multiple metastases in women treated initially by excision and ultimately succumbing to it.[3,4]. There is no consensus regarding the pathogenesis of AA. This hormonally responsive tumour is believed to arise from specialised 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, CEA 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 (HMGIC), which encodes protens involved in the transcriptional regulation, appears to have a role in the pathogenesis of this tumour. 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 AA, our case was completely encapsulated without any breach in continuity and without any projections into the neighbouring tissues. The pathologist reported negative margins on excised mass. This may be the reason why there is no recurrence in the last 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 CD 34, 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 post-operatively 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 has been reported 20 years after surgery as well[4]. Han-Guerts *et al*[2] propose 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 tumour, 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 periodic evaluations with physical examination and MR imaging 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 but should always be considered especially when it in an insidious painless fashion, particularly in premenopausal women in their 3rd-4th decades of life. High level of suspicion needed to make a clinical diagnosis. All relevant haematological and radiological studies including MR imaging or CT scan should help in reducing the number of misdiagnosed cases of AA preoperatively. Once its anatomical location and extension if any, is defined, any vulvar tumour—particularly AA- can be optimally treated by surgical excision only, while avoiding any mutilating surgery. If a 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 post surgery, it is evident that AA requires close and long-term follow-up.

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