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Case Report

A case report of aggressive angiomyxoma of vulva: first appearance deceives many

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ABSTRACT

Aggressive angiomyxoma (AA) of the vulva is a rare, benign tumour of mesenchymal origin. It has marked tendency for local recurrence but rarely metastasize. We report a case of 35-years old, nulliparous women presented with bilateral, large vulval masses. Local examination revealed approximately 20×20 cm mass arising from right labia majora, nodular, firm, non-tender with overlying skin intact. Left vulval mass was approximately 15×10 cm arising from labia minora, nodular, firm, non-tender. Inguinal lymph nodes were not palpable. After thorough evaluation, she underwent excision of bilateral vulval masses with reconstruction. Histopathology revealed tumour composed of ovoid to spindle-shaped cells in a fibrous and myxomatous stroma with loosely interspersed collagen fibres suggestive of aggressive angiomyxoma. On immunohistochemistry, the tumour cells were focally positive for smooth muscle actin and desmin. Stains for acid-fast bacilli and fungus did not reveal any organisms. Her postoperative period was uneventful with good recovery. She is on follow up and no recurrence has been seen so far in follow-up period of 15 months. AA should be kept in mind as one of the differential diagnoses in evaluation of large vulval masses. Keeping this rare entity in mind and relevant investigations may help in reducing the misdiagnosis and avoiding any mutilating surgery. Multidisciplinary approach can be crucial in management of such cases.

Keywords: Aggressive, Angiomyxoma, Vulval masses, Local recurrence

INTRODUCTION

Aggressive angiomyxoma (AA) is a rare, benign tumour of mesenchymal origin and was first described by Steeper and Rosai in 19831. It is a slow-growing tumour with marked tendency for local recurrence but rarely metastasize. It mostly affects women during the reproductive years of life and originate from soft tissues of the pelvis and perineum. The term "aggressive" denotes its propensity to infiltrate the local tissue at the site of origin. Though the different treatment options have been reported in literature, surgical excision with negative margins and close follow-up remains the standard treatment approach. AA of the vulva may mimic the malignancy and can lead to misdiagnosed by the clinician. Knowledge about this entity can help the clinician to make appropriate diagnosis and avoid operative morbidity due to delay in diagnosis. We discuss a case of young female presenting with AA of vulva successfully managed with surgical excision only with a brief literature review.

CASE REPORT

A 35-years old, nulliparous lady presented to our outpatient department with painless, bilateral masses of the vulva for last 5 years. These masses were initially of pea-nut size and gradually increased over time to reach present size of approximately 20 centimeters. She complained of coital difficulty because of these masses

but these were not associated with any menstrual complaints, itching, discharge per vaginum or other systemic symptoms of weight loss and anorexia. General and systemic examination were unremarkable. Local examination revealed bilateral, large and fleshy vulval masses. Right vulval mass was approximately 20×20 cm arising from labia majora, nodular, firm, non-tender with overlying skin intact. Left vulval mass was approximately 15×10 cm arising from labia minora, nodular, firm, non-tender. Inguinal lymph nodes were not palpable. Labia minora and urethral opening were not involved (Figure 1a, 1b). Her blood investigations were normal. Husband evaluation was also done for any sexually transmitted disease and did not reveal any significant findings.

On evaluation, biopsy from the mass were suggestive of benign lesion. Magnetic resonance imaging (MRI) pelvis was suggestive of large heterogenous hyperintense lesion arising from both labia. Bilateral enlarged inguinal lymph nodes with largest lymph node of 1.3 cm. After evaluation, she underwent excision of bilateral vulval mass with vulval reconstruction (Figure 2a, 2b). Postoperative period was uneventful.

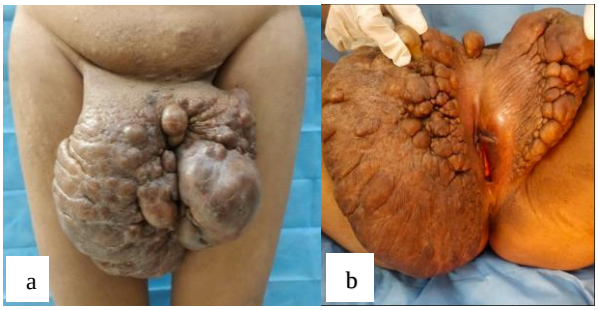

Figure 1: (a) Standing view (b) lying down view of right and left vulval masses.

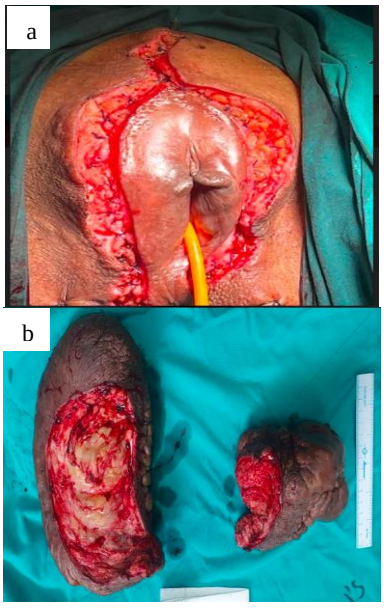

Figure 2: (a) Operative images (b) excised bilateral vulval masses vulval masses.

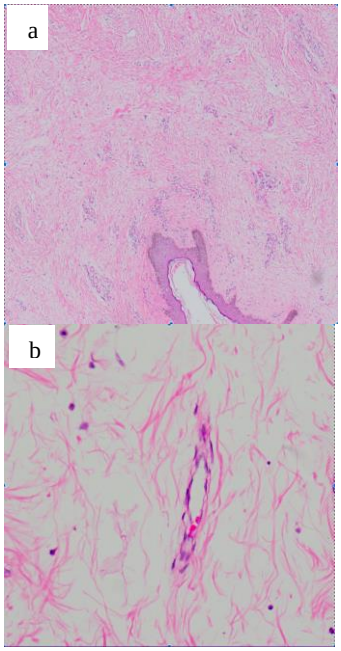

Figure 3 (a and b): Histopathological images of AA in our patient.

On histopathology report, sections from bilateral vulval masses showed tumour composed of ovoid to spindle-shaped cells in a fibrous and myxomatous stroma with loosely interspersed collagen fibres (Figure 3a, 3b). Blood vessels of varying caliber and thickness were noted, some of which show perivascular collagen and smooth muscle bundles. The tumour was infiltrating into the adjacent fibroadipose tissue. On immunohistochemistry, the tumour cells were focally positive for smooth muscle actin and desmin. Stains for acid-fast bacilli and for fungus did not reveal any organisms. It was reported as aggressive angiomyxoma.

DISCUSSION

Angiomyxoma are benign tumors of mesenchymal origin and are classified into superficial (cutaneous myxoma) and aggressive (deep) types. AA is predominantly seen in women of reproductive years, particularly in third to fifth decades of life.[2] Rarely, it can occur in postmenopausal females, children and males. In females, it mainly originates from vulva, pelvis and perineal region. In males, it particularly involves inguinal region and scrotum. Rare sites of involvement include lung, liver, larynx and orbits.[3]

Clinically, it presents as slow-growing swelling in vulva or perineal region. Patient may have non-specific symptoms like dull-aching pain, dyspareunia, difficulty in walking, dysuria etc. Though it is benign in nature, the term “aggressive” denotes its propensity to invade locally into the paravaginal and perirectal tissues. The exact pathogenesis of this tumour is yet to be clearly elucidated. However, it originates from specialized mesenchymal cells or multipotent perivascular progenitor

cells and expresses smooth muscle actin and desmin on immunohistochemistry.[4] HMGI-C gene (High mobility group protein isoform I-C) located on chromosome 12 has been found to play a role in occurrence of this tumour.[5] Differential diagnosis of AA include angiomyofibroblastoma, lipoma, Bartholin gland cyst, sarcoma botryoids, vulvar hypertrophy with lymphedema, sarcoma etc. Careful clinical examination, preoperative imaging and biopsy may help to make appropriate diagnosis. Imaging helps in knowing the extent of tumour to plan extent of surgical excision and follow up. Ultrasound can be done as primary imaging modality where it may appear as hypoechoic or cystic mass. However, MRI is investigation of choice for diagnosis and follow-up. It delinates the extent of tumour and its relation to surrounding structures better than CT scan. On MRI, these tumours have characteristic "swirled" linear low-intensity appearance on both T1 and T2-weighted images.[5]

Surgery remains the mainstay of treatment. Surgical resection aims to achieve wide local excision with negative margins. Sometimes, goal of achieving negative margins may not be fulfilled because of its locally infiltrative nature which may result in increased operative morbidity. In such cases, less radical surgery is advisable with close follow-up to pick recurrences. However, negative margins also do not ensure no recurrence. Chan YM et al, in his case series of 73 patients with AA has reported recurrence rate of 47% with no statistical difference between patients with positive and negative margins after surgical resection.[6] Other studies have also reported a varied recurrence rate ranging from 33-83% with most of them occurring during first 3 years of follow up.[7]

As the tumour is hormone-sensitive, use of GnRh agonists, raloxifene, tamoxifene etc as adjuvant therapy has been considered to shrink the tumour before surgery as well as to prevent recurrences after surgery8. However, these agents cannot be used as long-term therapy due to known side-effects of bone loss and menopausal symptoms. The role of radiotherapy and chemotherapy is limited in cases of AA due to its low-mitotic activity and need further exploration.

Arterial embolization has also been mentioned as a treatment option in literature but not often used because of multiple feeder vessels to the tumour. In a case report by Goyal et al, patient received 3 doses of monthly leuprolide injection after surgical resection to prevent recurrences. Our patient did not receive any adjuvant treatment after surgical excision and presently free of disease at 15 months of follow-up.[3] Hazzar et al, reported use of luteinizing hormone-releasing hormone (LHRH) agonist for treatment of recurrent AA in a premenopausal woman after surgical resection and successful reduction in size by its use.[9] However, effectiveness of this treatment option in postmenopausal women is not well elucidated. Fine et al reported a case of recurrent AA of the vulva which was treated solely by 3 months of GnRH agonist without needing any other medical therapy or surgery.[10] Gaurav A et al, performed laparoscopic bilateral oophorectomy as prophylactic measure for recurrence owing to its hormone-sensitive nature along with surgical resection of vulval AA3.

There is lack of recommendations regarding appropriate postoperative follow-up among patients with AA. Due to its high chances of recurrence, patients should be kept on long-term follow-up.

CONCLUSION

AA is a rare, benign but locally aggressive tumour. Clinical look of these large vulval masses may deceive the clinician. Keeping this rare entity in mind and relevant investigation may help in reducing the misdiagnosis and avoiding any mutilating surgery. AA can be optimally treated by surgical excision and require long-term follow -up.

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