

Aggressive angiomyxoma of the vulva- An Enigma in Clinical Practice

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Abstract

A 25 years old nulliparous regularly menstruating woman presented in ShSMC Hospital with the complaints of swelling in the right side of vulva and Dysparunia for 4 months. The swelling was initially small and painless but later on it became painful. The swelling recurred after surgical treatment. Examination revealed a swelling in the right Labium majus which was about 4x3 cm, bulging into right side of vagina. It was diagnosed as a case of recurrent Bartholin cyst but during surgical procedure it was apparent that the swelling was not Bartholin cyst but some other fleshy structure. The removed mass was 7x4 cm. Histopathology revealed benign lesion containing hypocellular structure with large blood vessels and no malignancy. It was diagnosed as Aggressive angiomyxoma. Aggressive Angiomyxoma is a very rare mesenchymal tumor, upto now only 200 cases have been reported in the world. It is a slow growing benign tumor which is typically located in the pelvis and perineum. It often presents with asymptomatic perineal or vulval mass and may be confused with Bartholin's cyst, lipoma or hernia. Local recurrence is upto 70%; metastasis is very rare. Treatment is surgical resection. Histology reveals a mass of mixed mesenchymal origin with low mitotic activity. It is a hypocellular and highly vascular tumor with myxoid stroma.

Introduction:

Aggressive angiomyxoma is distinctive soft tissue tumour of pelvis & perineum. It was 1st described by Steeper & Rosae having a distinctive histological characteristics with tendency to local infiltrate & recurrence in 1983¹. It is to diagnose this condition because the tumour is locally infiltrative & requires wide excision & follow up.²

The following is a case report of a woman who presented in our hospital with a swelling in the labia majora. Due to rarity of this condition and diagnostic dilemma it presents, the case has been reported

Case report:

A 25 years old regularly menstruating nulliparous lady hailing from Mirpur, presented in our admission unit of gynaecological department with the complaints of swelling on right side of vulva and dyspareunia for 4 months. Initially the swelling was small &

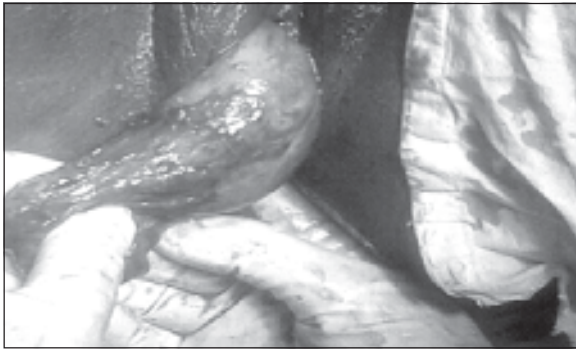
symptomless, then it gradually increased in size and became painful. She gave no history of fever or per vaginal foul smelling discharge. There was no per urethral discharge of her husband. She was admitted with this complaint on 14th Feb 2013; treated surgically and discharged. After 1 month swelling recurred & she was readmitted on 30th March 2013. On inspection of vulva, there was a swelling in posterior part of right labium majus, overlying skin was normal. There was no discharging sinus and no impulse on coughing. On palpation the swelling was 4x3 cm in diameter, bulging into the right side of vagina, tensely cystic, mildly tender, smooth surfaced. Fluctuation test negative. Speculum examination revealed cervix apparently healthy, no discharge present. On bimanual examination uterus was normal sized, fornices free. It was diagnosed as a case of Recurrent Bartholin Cyst. After some investigations local excision was planned. A vertical incision was given over the swelling but it

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was apparent that fleshy tissue was found underneath skin. The fleshy mass was gradually dissected from underlying tissue taking healthy tissue in margin. Dissection continued until upper limit of fleshy mass was reached. The mass was about 7x4 cm and was removed completely keeping healthy tissue in base. The patient was discharged on the 8th day. Patient is on regular follow up

Histopathology Report

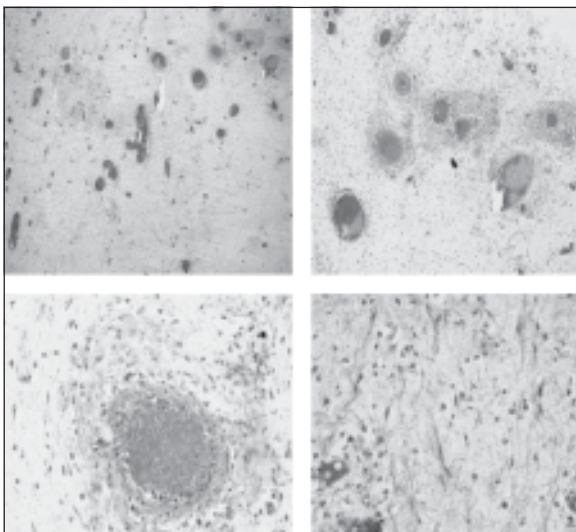
Gross examination



Specimen consists of a cyst like structure about 7.5 cm in maximum length. Cut surface is grey brown and solid.

Microscopic examination

Sections show a benign lesion containing hypocellular stroma with large sized blood vessels filled up with blood. No malignancy is seen.



Diagnosis: Aggressive angiomyxoma

Discussion

Aggressive angiomyxoma is a uncommon mesenchymal neoplasm occurring predominantly in pelvis & perineal region of adults. About 90% of patient are women, usually of reproductive age. ³Very rare, 1st described in 1983, ¹ Since then about 250 cases have been reported. ⁴

Genetic abnormality in chromosome 12 is found in some aggressive angiomyxoma underlying HMGA 2 gene. It presents as a poorly circumscribed gelatinous material mass & clinically simulates a Bartholin gland cyst or an inguinal hernia. On gross examination tissue are soft, rubbery with smooth external surface measuring about 3 & 60 cm in greatest dimension. ⁵

It can be mistaken both clinically & microscopically with other conditions such as myxoma, myxoid liposarcoma, myxoid variant of malignant fibrous tissue cytoma & other soft tissue tumour with secondary myxoid changes with rhabdomyosarcoma botrioides. Cut surface reveals a glistening soft homogenous appearance.

Recurrent tumours show more prominent area of haemorrhage & fibrosis.

Histopathologically, angiomyxoma is a mesenchymal tumour, composed of fibroblast within a strong myxoid background. Vascular proliferation is also prominent & virtually no mitoses are present ⁶. The vast majority of cases demonstrates positivity for desmin in the myxoid bundle & or stromal cells, while Actins & CD34 may be variably positive. ⁶ Oestrogen & progesterone receptor positivity suggests that aggressive angiomyxoma might be hormone dependent, as rapid growth has been observed during pregnancy. The tumour grows slowly & its benign nature is suggested by the histology & by the fact that it shows no tendency to metastasis. However it is locally aggressive & tends to recur (36-72%) after resection ⁷. Among the imaging techniques USG, CT scan & MRI have specific role to determine the extent of surgery. USG shows a mass that is hypoechoic or cystic. Angiography can detect a hypervascular mass. On CT Scan the tumour has a well defined margin & attenuation less than that of the muscle. On T₂ weighted MRI imaging, the tumour has high signal intensity ⁷.

Treatment is surgery in the form of wide local excision. Post operative angiographic embolization; postoperative external beam irradiation are useful to

decrease the chance of local recurrence⁸. Hormonal treatment with a Gonadotropin releasing hormone agonist can be applied for small primary lesion in addition to adjuvant therapy for residual tumour.

Conclusion:

Although a rare diagnosis, aggressive angiomyxoma can present with unusual features. Detailed radiological examination is helpful in suspecting the problem, but histology is gold standard for diagnosis. Wide local excision is curative and prognosis of such tumour is good.

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