Case report:

Aggressive angiomyxoma of vulva in a pregnant female-a rare entity

Dyuti B[1], Ayan B[2], Pallab M[3]

Abstract:

Aggressive angiomyxoma (AA) is a rare slow growing vulvovaginal mesenchymal neoplasm with a marked tendency for local recurrence. We here present a twenty five year old thirty two weeks primigravida patient with a large pedunculated AA arising from left side of the vulval area. We delivered the baby by caesarean section and later completely excised the tumour successfully without any signs of recurrence even after two and half years follow up.

Keywords: Aggressive angiomyxoma (AA); neoplasm; recurrence.

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Introduction:

Aggressive angiomyxoma (AA) is a slow growingvulvovaginal mesenchymal neoplasm with a markedtendency for local recurrence, but with a low tendency to metastasize. AA was first described by Steeper and Rosai in 1983[1]. AA is also called deep angiomyxoma[2]. About one fourth of these tumours are pedunculated. Patients often complain of mass, dull aching pain and urinary symptoms such as dysuria, urinary retention. Angiomyxomas are of two types of which superficialangiomyxomas usually present in middleagedadults as a single nodule or a polypoidal lesion in the headand neck region that may be clinically confused with skin tag or neurofibroma. The stroma is made up ofmostly oedema with little myxoid material. On the other hand, AA occurs almost exclusively in the pelvic andperineal regions of women of reproductive age, but is occasionally reported in men (male to female ratio 1:6).[3]

Case Report:

A twenty five years old married primigravida woman came to OPD for antenatal check-up as she was carrying 32 weeks pregnancy. Excepting minor problems of pregnancy, she complained of about a gradually enlarging mass in her vulval area on left side involving labia minora and majora and extending upto groin (Figure 1& 2).

Fig 1- Patient's lesion before treatment/ Patients lesion at presentation.

Fig 2- Patient's lesion before treatment

1. Basu Dyuti, Calcutta Medical College, Kolkata
2. Basu Ayan, RMO cum Clinical Tutor, Department of Tropical Medicine, School of Tropical Medicine, Kolkata
3. Mistri Pallab, Associate Professor, Department of Gynaecology & Obstetrics, Calcutta Medical College, Kolkata

Correspondence to: Dr Dyuti Basu, Calcutta Medical College, Kolkata, Email : rilbong@yahoo.com

On detailed interrogation it was found that it was a small swelling since last 6-7 years before pregnancy. During pregnancy it had grown into very large size almost interfering her normal daily activities like walking and sitting. For the last one month she was experiencing dragging pain radiating to left groin. On examination a large (22 cm x16 cm x 9 cm) pedunculated mass with lobulated appearance almost obstructing vulval area and introitus was seen. The mass was nontender, soft &pigmented. All hernia sites were normal.

She was initially suspected to have a genital wart and referred to School of Tropical Medicine, Kolkata where she was undergone Human Papilloma Virus DNA testing and cervical pap smear test. Both were negative. HIV & HBsAg were nonreactive. Her Hb was 11.9 gm% and TLC-8900. Our patient was not subjected to detailed radiological investigation as its clinical appearance at presentation was warty superficial polyp of skin. However ultrasonography of whole abdomen and Trans Vaginal Sonography showed no intra-abdominal or pelvic extension.

She was admitted for biopsy from the mass in our department. Initial biopsy showed a lesion composed of spindle shaped cells and stellet cells lying in myxoid stroma containing delicate thin blood vessels. The stellate cells did not show any significant mitotic activity or nuclear anaplasia. The epidermis showed increase in melanocytes in basal cell layer. So there is a possibility of aggressive angiomyxoma. But they needed a larger biopsy specimen to comment. In the mean time she was near term and delivered by caesarean section as the mass could obstruct vaginal delivery. Two months later,when she attended for postpartum check-up, the whole lesion was excised and the specimenwas sent for biopsy again. Biopsy of multiple sections showed a tumour composed of widely scattered spindle to stellate shaped cells with ill-defined cytoplasm and thick and thin vascular channels lying in myxoid stroma rich in collagen fibres. Mitotic figures not seen. These features were compatible to aggressive angiomyxoma of vulva (Figure 3 & 4).

Though the tumour was known for high recurrence but our patient did not show any signs of recurrence even after two and half years follow up (Figure 5).

Ethical Approval: This case report was published after getting approval from the local Ethics Committee.

Discussion:

AA presents as a vulval polyp clinically and is diagnosed on histology. Oestrogen and progesterone

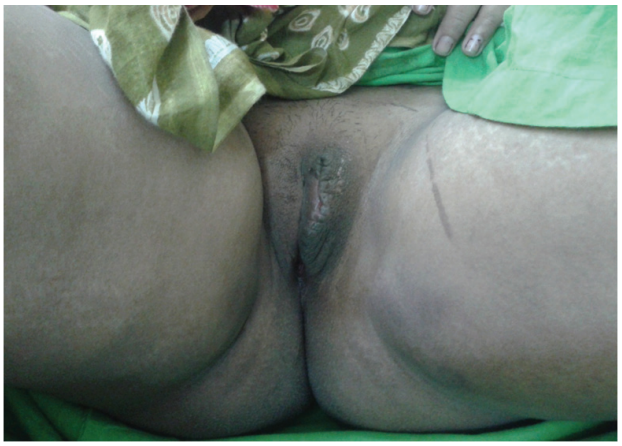

Fig 3- Patient after treatment/ Figure demonstrate affected area after treatment.

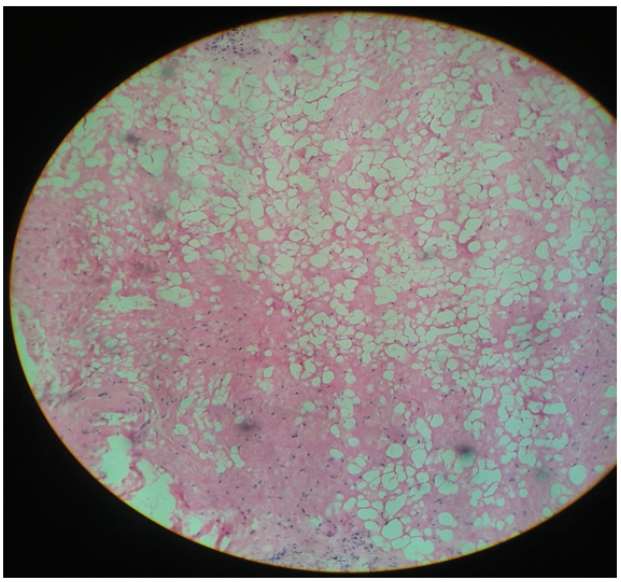

Fig 4- Aggressive angiomyxoma under high power microscope

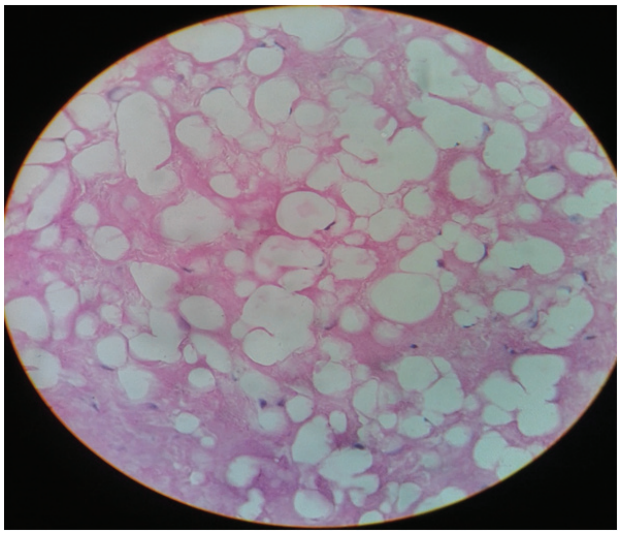

Fig 5- Patient after treatment/ Figure demonstrate affected are after treatment.

receptors (ER & PR) are commonly found in AA. It is thus likely to grow during pregnancy and respond to hormonal manipulation. Hormone dependency has been suggested based on a single case report of a lesion that grew rapidly during pregnancy[4].In addition, other reports have demonstrated ER and/or PR positivity within aggressive angiomyxoma. It is of interest that positivity with androgen receptors has been demonstrated in aggressive angiomyxoma in a male patient, suggesting that in men the neoplasm might also be hormone dependent. It involves mainly the pelvis, vulva, perineum, vagina and urinary bladder in adult women in the reproductive age [5]

The term "aggressive" denotes its propensity for local aggression and recurrences after excision. Usually, this tumour is nonmetastasizing, but there are reports of multiple metastases in women treated initially by excision and who later succumbed to metastatic disease [6,7]. About one fourth of these tumours are pedunculated .Clinical differential diagnosis is Angiomyo fibroblastoma, Bartholin gland cyst, Sarcoma botryoides**,** Superficial angiomyxoma**,** Vulvar hypertrophy lymphedema**.** Histologic differential diagnosis is Myxoma, Myxoid liposarcoma, Myxofibrosarcoma, Nerve sheath myxoma, Sarcoma botryoides, Vulvar hypertrophy with lymphedema, other soft tissue tumours with secondary myxoid change.

On computed tomography (CT) scan, these tumours have a well-definedmargin with attenuation less than thatof the muscle. On MRI, these tumours show high signal intensity on T2weightedimages. The attenuation onCT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high watercontent of angiomyxoma that of a benign polyp[8].

Wide surgical excision is the traditional treatment ofchoice. However, organs, such as the rectum and bladder to which the tumour may be attached, are spared asthe morbidity of extensive surgery may not be justified due to its high recurrence rate even after completeresection. Where fertility is to be preserved or surgery is likely to be extensive and mutilating, incompleteresection is acceptable as local recurrences can be treated with further resection.The tumour tends to grow around the structures of pelvic floor without penetrating musculature of vagina or rectum [9]Recurrences may occurfrom months to several years after excision (2 months to 15 years).

Recurrences generally occur in the first 5 years after primary surgery, and about 70% occur in the first 3 years, but late recurrences up to 14 years have been reported[10]. Five cases of AA of the vulva have been reported in the Korean literature; four cases were local recurrence of AA[11-13] and one case first occurred in the adolescent period. The case was followed up for one year without recurrence[14].

AA, despite the name, is not thataggressive, with only a 30% chance of recurrence, which is eminently treatable by excision with a 1 cmmargin. Most of the patients have only one recurrence. Radiation therapy and chemotherapy are considered lesssuitableoptions due to low mitotic activity. Hormonal manipulation with tamoxifen, raloxifene and gonadotropin releasing hormone analogues has been shown to reduce the tumour size and may help to makecomplete excision feasible in large tumours and in the treatment of recurrence. Angiographic embolizationmay also help in subsequent resection by shrinking the tumour as well as making it easier to identify it fromsurrounding normal tissues. As late recurrences are known, all patients need to be counselled about theneed for longtermfollowup.Magnetic resonance imaging is the preferred method for detecting recurrences. Wide surgical resection as an en-bloc specimen is usually accomplished by a combined transperineal and abdominopelvic approach, with preservation of surrounding structures.Preoperative vascular embolization may decrease the tumour size.Lifetime follow up to monitor for recurrence is mandatory.Chemotherapy and radiotherapy are ineffective due to the low proliferative potential of the lesional cells, but high dose radiotherapy has been used successfully. Hormonal therapy with gonadotrophin-releasing hormone (GnRH) inhibitors such as leuprolide is useful for recurrent tumours that are positive for oestrogen and progesterone receptors.Non-steroidal selective oestrogen receptor modulators (SERM) such as raloxifene have also been used.But our case was treated successfully by simple resection and even after two and half years follow up no signs of recurrence.

Conflict of interest: None declared

Acknowledgement: Department of Gynaecology & Obstetrics, Calcutta Medical College, Kolkata

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