ORIGINAL RESEARCH PAPER

Gynaecology

ANGIOMYXOMA OF THE VULVA

KEY WORDS:

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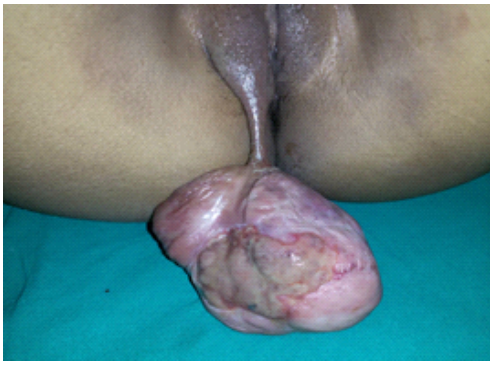
BACKGROUND

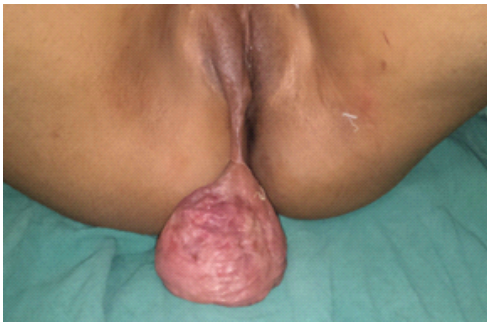
Angiomyxoma is a myxoid tumor involving the blood vessels. Aggressive angiomyxoma (AA) is an unusual mesenchymal tumor. AA occurs most commonly in women of reproductive age.This is a distinct soft tissue tumor that has a prominent myxoid matrix and numerous thin-walled blood vessels and may have an aggressive local recurrence. The tumors have the characteristics of large size (usually greater than 10 cm) and slow growth, and are not painful. It can affect the vulva , perineum and other parts of the pelvis. It is a benign tumor and does invade adjoining tissues. The recurrence can be as close as six months from initial resection There is no proven medical therapy. Wide surgical excision is the traditional treatment of choice.Here we present a case of aggressive angiomyxoma of the vulva in 15 year old girl.

CASE REPORT

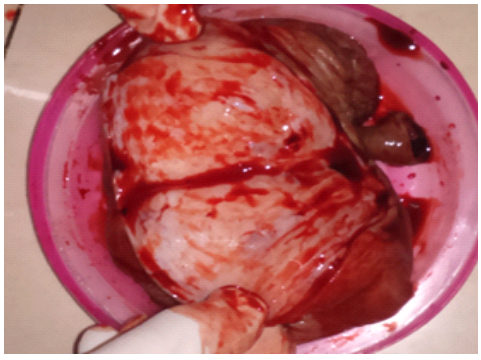
15 yr,. unmarried girl presented to the OPD as a case of swelling arising from the vulva. History dates back to 3yrs when she saw a small swelling about the size of a peanut on the right labia. It gradually increased over 3 yrs . Now there Is an abrupt increase in the size in last one week . Associated with pain and discharge from the swelling. Her menstrual cycle are regular. LMP 7-4-15. H/O dysmennorhoea. Family history , past history not significant. On examination she is average built . On local examination there is a large soft boggy mass of the right vulva seen. An ulcer is seen on the swelling.It was pedunclated mass measuring 7x6cm .on GPE pallour mild , no lympadoema , no thyroid enlargement, chest and cvs normal. Hb 9gms,BT-1'50", CT-5'60".RFTS and LFTS normal. PT 15SEC PTI- 90.5%. USG shows uterus A/V normal size ,ET normal, B/L ovaries normal.



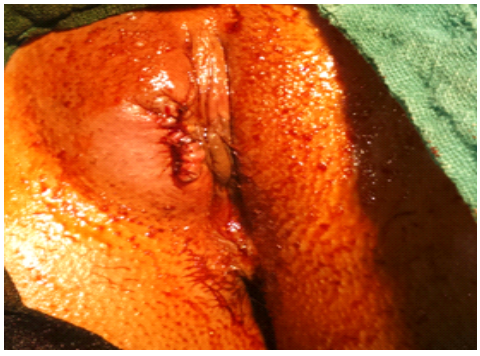
ULCER OVER THE VULVAL MASS(right labia)



A PEDUNCALATED VULVAL MASS



CUT SECTION – HOMOGENOUS MASS



POST EXICSION

Patient got admitted for exicsion of the vulval mass.It was 7x6 cm in size soft without any solid areas.The vulval mass was held by two clamps at its peduncle and excised.Hemostatic sutures were applied with vicryl.Cut section of the mass showed homogenous texture .The secimen was sent for histopathology. HPE reports showed histological features of ANGIOMYXOMA.

DISCUSSION

Angiomyxomas are classified either as superficial (also called as cutaneous myxoma) or AA. Superficial angiomyxomas usually present in middle-aged adults as a single nodule or a polypoidal lesion in the head and neck region that may be clinically confused with skin tag or neurofibroma. On the other hand, AA occurs almost exclusively in the pelvic and perineal regions of women of reproductive age, but is occasionally reported in men (male-to-female ratio 1:6) (1) . AA was first described by Steeper and Rosai in 1983.(2) It presents as a vulval polyp clinically and is diagnosed on histology. Estrogen and progesterone receptors are commonly found in AA.(3) It is thus likely to grow during pregnancy and respond to hormonal manipulation. Less than 250 cases of aggressive angiomyxoma have been reported till 2010.[4]

The term "aggressive" denotes its propensity for local aggression and recurrences after excision.Clinically, AA may be misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, levator hernia , superficial angiomyxoma, angiomyofibroblastoma and smooth muscle tumors as a differential diagnoses of a polypoidal mass in the perineum.(5) On CT scan, these tumors have a well-

defined margin with attenuation less than that of the muscle. On MRI, these tumors show high signal intensity on T2-weighted images. These features are likely to be related to the loose myxoid matrix and high water content of angiomyxoma (6). No single modality of treatment of recurrence has been found to be of proven benefit till now. AA, despite the name, only a 30% chance of recurrence, which is eminently treatable by excision with a 1 cm margin. Wide surgical excision is the traditional treatment of choice. Hormonal manipulation with tamoxifen, raloxifene and gonadotropin-releasing hormone analogues has been shown to reduce the tumor size and may help to make complete excision feasible in large tumors and in the treatment of recurrence(7). Angiographic embolization may help in subsequent resection. by shrinking the tumor as well as making it easier to identify it from surrounding normal tissues.(8). Radiotherapy and chemotherapy have been used as adjunctive therapies but are unlikely to be useful as it has few mitotic activity (9,10). As late recurrences are known patients need to be counselled about the long-term follow-up. MRI is the preferred method for detecting recurrences.

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. Vulval AA—can be optimally treated by surgical excision only. 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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