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Aggressive Angiomyxoma – A Rare Tumor Often Misdiagnosed

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Abstract
Aggressive Angiomyxoma is a rare mesenchymal tumour which is locally invasive most commonly occurring in women of reproductive age group with high chance of recurrence. This paper reports a case of 46 year old female with right vulvar swelling and 35 year old female with mass descending per vaginum both of these patients underwent surgical excision and histopathology reported as aggressive angiomyxoma. In view of high chance for recurrence patient is being followed up every 6 months.
Keywords: *aggressive angiomyxoma, vulval swelling, mesenchymal tumour.*

Introduction

Aggressive Angiomyxoma is a slow growing mesenchymal neoplasm found commonly in the vulval, perineal and pelvic region with a high propensity in women of reproductive age group.[1] It is locally aggressive with a 30% chance of recurrence even after several years of excision of the tumor.[2] Rarely distant metastasis of this tumor has been reported.[3] Complete resection with tumor free margin is the accepted mode of treatment but incomplete resection is very common because of its infiltrating nature and absence of a definite capsule.[4] Interventions like hormonal manipulations, radiotherapy, angiographic embolization, etc have been tried as adjunctive therapy to prevent and treat recurrence of this tumor.

Case Report

We report two cases of Aggressive Angiomyxoma which were misdiagnosed clinically as lipoma and vaginal wall cyst respectively and was later confirmed as Aggressive Angiomyxoma by histopathological diagnosis

Patient 1 was a 46 yrs old female presenting with a pedunculated swelling from the left labia majora which had progressively increased in size for the past 5yrs. On examination a well-circumscribed pedunculated swelling of size 9x6 cm arising from labia majora loosely hanging with a pedicle which was non-tender and soft in consistency. A clinical diagnosis of lipoma was made and excision was done. Gross examination showed a grey-white to brown soft tissue mass measuring 9.5x8.3.5cm which was grey-white on the cut section (Fig. 1). HPE revealed thick-walled vessels with few inflammatory cells and collagen. The Tumor was composed of myxoid areas, stellate cells and myoid bundles, thick-walled blood vessels and, collagen suggestive of Aggressive Angiomyxoma.

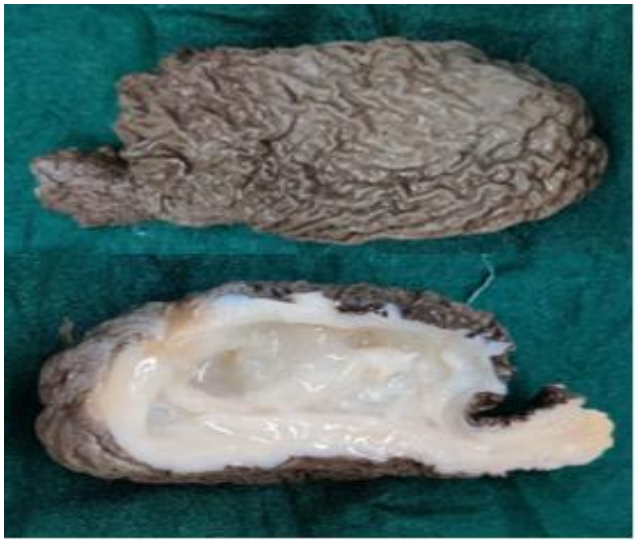

Fig.1: Well circumscribed pedunculated skin covered mass (above)
Cut section of the specimen showed glistening, gelatinous surface (below)

Patient 2 was a 35year old female presenting with mass descending per vaginum. On clinical examination, a soft tissue mass of size 9x6cm arising from the posterior vaginal wall and protruding through the introitus was found (Fig.2). A diagnosis of posterior vaginal wall cyst was made and excision was planned. USG imaging of this mass showed exophytic heterogeneous cystic lesion extending from introitus. On HPE after excision, multiple sections showed a well-circumscribed tumor composed of spindle cells with round to oval nuclei with few showing cytoplasmic processes. Stroma showed myxoid material with focal collagen fibers, congested and dilated thick-walled blood vessels of varying caliber suggestive of Aggressive Angiomyxoma (Fig.3 & Fig.4)
Both patients are under regular follow-up once in 6months to detect recurrence.

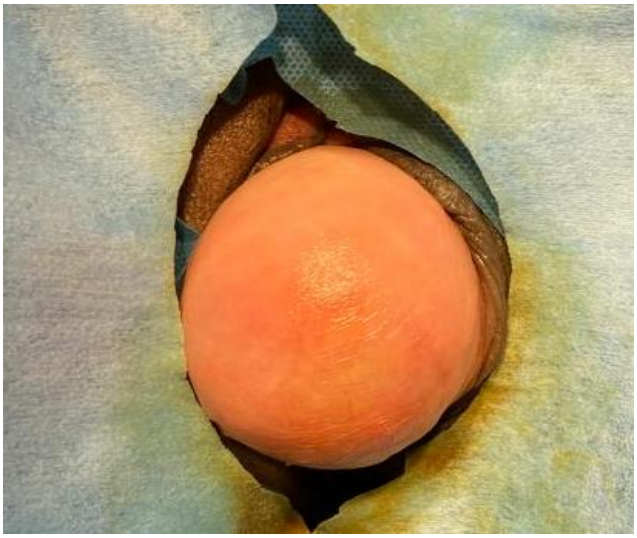

Fig.2: Soft tissue mass arising from the posterior vaginal wall

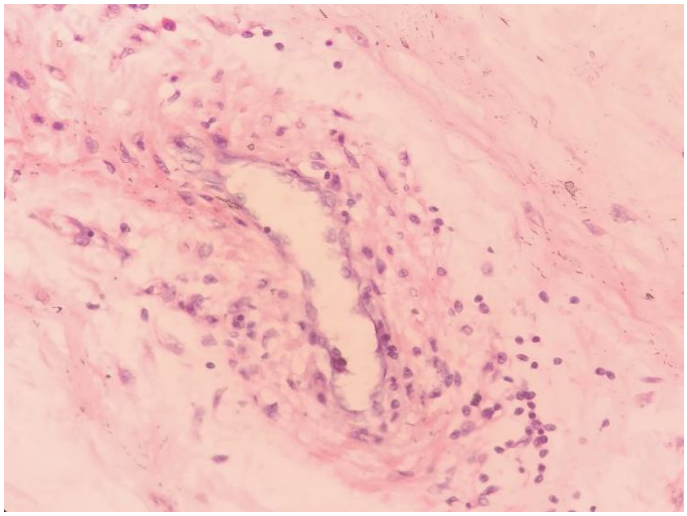

Fig.3: Thick-walled vessels with few inflammatory cells and collagen were seen

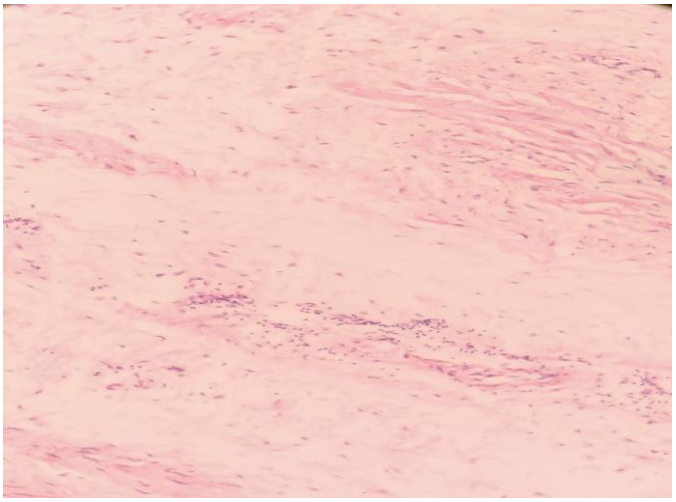

Fig.4: Tumor composed of myxoid area, stellate cells, myoid bundles, thick walled blood vessels and collagen

Discussion

Aggressive Angiomyxoma is a slow growing rare mesenchymal tumor with a locally aggressive nature with propensity for future recurrence.[5,6]

It is more common in women of reproductive age group with a female male ratio of 6:1.[7]

Translocation at the level of chromosome 12 where HMG A2 which is a high mobility group protein has been suggested by Nucei and Fletch as the pathogenesis of this tumor.[8,9] AA is derived from myofibroblast as a phenotypic variant of the basic fibroblast with a prominent vascular component.[10]

Microscopically the tumor is composed of widely scattered spindle to stellate shaped cells with ill defined cytoplasm and small round to oval hyperchromatic nuclei with small centrally located nucleoli embedded in a myxoid stroma which is rich in collagen. Another specific finding is the presence of vessels that range from thin walled capillaries to large vessels with secondary changes including perivascular hyalinization and medial hypertrophy.[11]

Immunohistochemistry shows strong positivity to desmin, vimentin, estrogen and progesterone receptor and negative for S-100 protein which helps in distinguishing this tumor from other mesodermal tumors.[12]

Preoperative imaging is useful to determine the true extent and size of the tumor. In ultrasound AA appears as a hypoechoic or cystic mass and is not specific for AA whereas CT and MRI shows specific features. In MRI AA shows a high intensity signal on T2 weighted images due to loose myxoid matrix and high water content of the tumor, the typical swirling pattern.[13] MRI also helps in detecting the invasion of the tumor above the pelvic diaphragm in order to plan appropriate surgery and also to diagnose recurrences.[14]

Clinical misdiagnosis of this tumor is very common. It is usually misdiagnosed as many benign condition of vulva and vagina like lipoma, bartholin's cyst, labial cyst, fibro epithelial stromal polyp and other smooth muscle tumors of the vulva.[15]

Surgical excision with a tumor free margin of 1cm to prevent recurrence is the mainstay of treatment of AA. But in spite of complete resection recurrences have been reported. Considering the possibility of distant metastasis reported in some studies complete resection should be performed.[16]

Multiple option have been tried as a adjunctive therapy to prevent and treat recurrences. Radiotherapy and chemotherapy have not been found to be effective due to the low mitotic activity of the tumor. As the tumor has estrogen and progesterone receptors hormonal manipulation like GnRH agonist, tamoxifen and raloxifene have been used as adjuvant with variable success.[17]

Angiographic embolization of the tumor has also been tried. These treatment modalities are also used in a neo adjuvant setting to shrink the tumor the mass prior to surgery with the aim of complete resection of huge masses.[18]

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