



Case Report

## Aggressive Angiomyxoma - An Uncommon Cause of Vulval Mass: A Case Report

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### ABSTRACT

Aggressive angiomyxoma is a slow growing mesenchymal tumor. It is locally aggressive with a low rate of metastasis. Aggressive angiomyxoma arising from vulvovaginal region is a rare. As it is infiltrative in nature and should be differentiated from other locally occurring mesenchymal tumor like lipoma.

**Keywords:** aggressive angiomyxoma, mesenchymal tumors, vulva

### INTRODUCTION

Aggressive angiomyxoma was first described by Steeper and Rosai in 1983. [1] Aggressive angiomyxomas are slow growing mesenchymal tumors with locally aggressive tendency and a low rate of metastasis. It has a higher predisposition to women though it is rarely seen in men also. In the female aggressive angiomyxoma arises commonly from the vulvovaginal region. [2] It mostly presents as a vulvovaginal polyp but can also occur in perineum and urinary bladder in women of the reproductive age group. Many modalities of management have shown beneficial effects. Long term follow up is necessary to prevent recurrence. Literature search has shown approximately 150 cases till date.

### CASE REPORT

A 40 year old lady presented with complaints of slow growing mass on the left

side of her external genitalia since one month. The swelling gradually increased in size and was painless. It was not associated with discharge, discoloration, pruritus. She did not have any constitutional symptoms. Her general and systemic examination did not reveal any abnormality. Local examination showed a pedunculated, polypoidal mass of 6\*4cms arising from the left lower 3<sup>rd</sup> of labia majora. The mass was non-tender, mobile, soft and doughy in consistency (Fig 1). The inguinal nodes were not enlarged. Provisional diagnosis of degenerating left labial lipoma was made. Her blood investigations and ultrasound abdomen revealed no abnormality. Ultrasound of the pedunculated mass showed a cyst with multiple internal septations. Cyst wall thickness was around 3mm. Under regional anaesthesia, the pedunculated labial mass of 6\*4 cms was excised in toto and sent for histopathological

examination. Postoperative period was uneventful. Histopathological examination revealed that the tumor was composed of spindle and stellate shaped cells in myxoid matrix with variable sized thin walled capillaries and thick walled vascular channels suggestive of aggressive angiomyxoma (Fig 2).

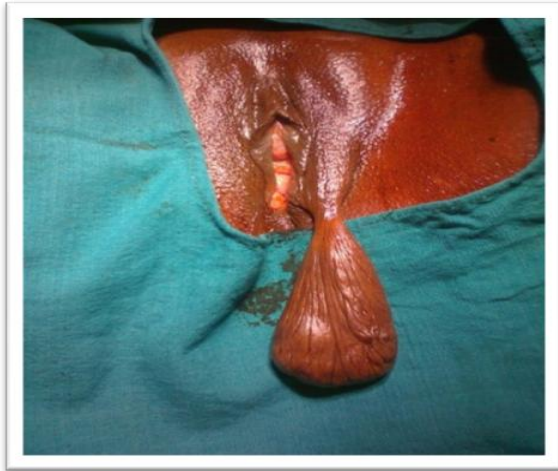


Fig 1. A well defined polypoidal pedunculated mass of 6\*4cms arising from the left lower 1/3<sup>rd</sup> of the labia majora.

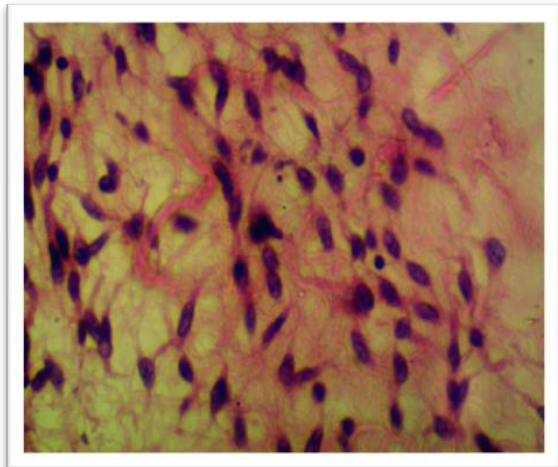


Fig 2. Tumor composed of spindle and stellate shaped cells in a myxoid matrix (H and E 100 X)

## DISCUSSION

Aggressive angiomyxoma is a soft tissue neoplasm which is a rare mesenchymal tumor. It is usually benign but has infiltrative potential into skeletal muscle and fat. It most commonly arises from

vulvovaginal region, perineum, and pelvis of the women. Mostly it is a slow growing, asymptomatic tumor. Sometimes it presents with regional pain, feeling of local pressure or dyspareunia. Clinically, they may be misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, levator hernia or sarcoma. Fibro-epithelial stromal polyp, superficial angiomyxoma, angiofibroma, angiofibroblastoma, cellular angiofibroma and smooth muscle tumors also need to be considered in the differential diagnoses of a polypoidal mass in the perineum.<sup>[2]</sup>

Aggressive angiomyxoma, despite the name, is not really aggressive, with only a 30% chance of recurrence. Recurrences may occur from months to several years after excision (2 months to 15 years).<sup>[3]</sup> Study has been done in 150 patients, of which only two showed metastatic changes.

It usually grows up to 10cms in diameter. Macroscopically it is lobulated and may adhere to soft tissue surrounding it. MRI will show contrast enhancement reflecting their inherent vasculature, and tend to displace and grow around structures rather than infiltrating them.

Microscopically cells are spindle or stellate shaped, embedded in loose matrix consisting of wavy collagen and edema. Tumor cells are characteristically positive for estrogen and progesterone receptor, suggesting a hormonal role in the development of tumor.<sup>[3]</sup> Immunohistochemically these tumors are positive for desmin, smooth muscle actin, muscle specific actin, vimentin, estrogen and progesterone receptors. Some tumors are positive for CD34, whereas S100 is invariably negative. Genetic studies have shown chromosomal translocation of 12q13-15 band involving the HMGA2 gene has been described.<sup>[4]</sup>

Recurrences may occur from months to several years after excision (2 months to 15 years). Recurrence is surgically treatable

by excision with a 1 cm margin. Most of the patients have only one recurrence. In view of late recurrence patients need to be informed of the need for long term follow up. Radiation therapy and chemotherapy are considered less-suitable options due to its low mitotic activity. 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. [5] Angiographic embolization may also help in subsequent resection by shrinking the tumor as well as making it easier to identify it from surrounding normal tissues. [6] The most common treatment of recurrence will be surgical excision.

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