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## Case Report

# A rare case of aggressive angiomyxoma of vulva

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### ABSTRACT

Aggressive angiomyxoma is a rare, slow-growing soft tissue tumor developing from myxoid cells that usually arises in the pelvis and perineal regions of women in reproductive age, with a marked tendency to local recurrence but a low tendency to metastasize. It is often initially misdiagnosed because of its rarity. It has a marked tendency for local recurrence. Surgical resection is the treatment modality of choice for aggressive angiomyxoma. We describe a case of an aggressive angiomyxoma of vulva in a 75 year old postmenopausal woman with an asymptomatic mass on the genitalia since 3 years. Excision of the mass was done under local anaesthesia and the diagnosis was only made after histological examination. . The patient has been kept under follow-up. The etiology, presentation, diagnosis and management of this rare tumor are outlined. Angiomyxoma of vulva and vagina refers to a rare disease. Pre-operative diagnosis is difficult due to rarity and absence of diagnostic features, but it should be considered in every mass in genital, perianal and pelvic region in a woman in the reproductive age.

**Keywords:** Aggressive angiomyxoma, Mesenchymal tumor, Vulval tumor

### INTRODUCTION

Angiomyxoma is a rare tumor that develops from myxoid cells, which are a type of cells found in the connective tissue of the body. There are two types of angiomyxomas -superficial, which grows near the surface, and aggressive -which involves the deeper structures. AA was first described by Steeper and Rosai in 1983 for morphologically distinctive, slowly growing locally infiltrative tumor.<sup>1</sup> It mainly involves the pelvis, often located in the vagina, vulva, perineum or buttocks.<sup>2</sup> Female to male ratio is 6.6/1, usually of reproductive age, with a peak incidence between the third and fourth decades of life.<sup>3-6</sup> It's uniqueness is in being benign structurally although it is locally malignant behaviourally. Estrogen and progesterone receptors are commonly found in AA. It is thus likely to grow during pregnancy and respond to hormonal manipulation.

Clinically it presents as a vulval polyp and it is diagnosed on histology. Sizes range from a few centimetres to more than 20 cm. In view of its locally aggressive nature, appropriate management and long-term follow-up should be considered.

### CASE REPORT

A 75 year old woman post-menopausal since 20 years referred to our tertiary care hospital i/v/o? Ca vulva. She had a mass in left labia since 3 years, initially small and slowly increased in size to attain the current size of 10 x 3 cms. There was no similar illness in the past, no fever or genital ulcer disease, discharge or ulceration of the growth. The patient was a known case of hypertension with moderate-severe Aortic stenosis on T Amlodipine 5 mg BD. Her general and systemic examination was within normal limits. On examination, there was a single

pedunculated mass of size 10 cm × 3 cm arising from the left labium majus (Figure 1). It was lobulated in appearance with variable consistency (soft to firm). There were no visible pulsations or cough impulses. Skin over the swelling was not pinchable. It was a non-reducible and non-compressible swelling. Cervix and vagina were healthy on per speculum examination. Rest of the external genitalia was normal. The diagnosis of vulval neoplasia was made and differential diagnosis of neurofibroma, fibrolipoma, filariasis, and lymphogranuloma venereum were considered. Blood, urine analysis, X-ray chest, ECG, USG (A+P), were normal. PAP-Menopausal smear (negative for intraepithelial malignancy) lipid profile, Thyroid profiles were normal. 2D ECHO was suggestive of severe concentric left ventricular hypertrophy. Sclerodegenerative Aortic Valve disease, with severe AS with good LV function. Pt was given fitness for surgery with due cardiac risk and advised to avoid hypotension and tachycardia in intra-operative and post-operative period.



**Figure 1: Pre-operative presentation of the vulval angiomyxoma at left labia.**



**Figure 2: Microscopically, it is composed of numerous thin-walled blood vessels and myofibroblasts in a collagenous matrix. It may have alternating zones of hypo and hypercellularity.**

Local USG was suggestive of heterogeneously hyperechoic lobulated mass lesion seen arising from vulval region. It showed internal vascularity on color Doppler. Findings were suggestive of neoplastic etiology

FNAC Aspirate revealed only hemorrhagic and few squamous epithelial cells. No representative cellularity was seen. The mass was totally excised and was sent for histopathological examination (Figure 2). The section revealed a skin covered tumour. The epidermis showed acanthosis. Tumor was comprised of fibromyxoid tissue of scattered stellate and spindle shaped cells. Various blood vessels of varying sizes were also noted. There was no mitotic activity. Features were suggestive of aggressive angiomyxoma.



**Figure 3: The postoperative follow-up was uneventful. No signs of recurrence after surgery.**

Post-operative course was uneventful (Figure 3). The patient has been coming for follow up in opd. There have been no signs of recurrence

## DISCUSSION

Although the tumour is benign and does not invade the neighbouring tissue, it has a tendency to recur after surgical excision; hence it is termed as aggressive. The recurrence can be as close as six months from initial resection. It is polypoid and is mostly associated with Carney's complex which is a triad of spotty pigmentation, cardiac myxomas, and endocrinal over activity.<sup>7</sup> As observed in light microscope, the lesion has lobular or multi-nodular appearance at low magnification. It is poorly circumscribed with extension into subcutaneous tissue. A sparse proliferation of spindle and stellate-shaped cells is seen in extensive myxoid stroma. There is often a prominent vasculature that is focally arborizing. A mixed cellular infiltrate particularly of neutrophils is a feature.<sup>8</sup> Clinically it should be differentiated from cyst, skin tag, and neurofibromatosis. Complete surgical removal is the treatment of choice, but it has a recurrence rate of 30-40%. Grossly, they are soft and partly circumscribed. On cross section they have gelatinous appearance. Microscopically the tumor is composed of widely scattered stellate and spindle-shaped cells with variable-sized thick and thin-walled vascular channels in myxoid stroma. Clinically it needs to be differentiated from Bartholin's cyst, periurethral cyst, and

hernia. It needs complete surgical removal but has a local recurrence rate of 30-40. It is more of a pathological diagnosis than a clinical one.

## CONCLUSION

This is one such an example of non-venereal disease occurring on genitalia causing apprehension to the patient. In view of its locally aggressive nature, appropriate management and long term follow-up should be considered.

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