

# Angiomyofibroblastoma of the Vulva

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## Abstract

Angiomyofibroblastoma (AMF) is a rare benign soft-tissue tumor that most frequently affects the lower genital tract of young to middle-aged women. It mainly consists of two components: stromal cells and prominent vasculature. Clinically, it is usually asymptomatic and resembles Bartholin's cyst. Although it is a benign tumor, cases with recurrence and sarcomatous transformation have been reported. Due to the overlapping of histopathological picture, diagnostic perplexity often arises between AMF and aggressive angiofibroma (AAM). AMF being benign in nature is treated by local excision, whereas AAM is a more infiltrative lesion that has a higher tendency for local recurrence.

**Keywords:** Angiomyofibroblastoma, pedunculated mass, vulva

## INTRODUCTION

Angiomyofibroblastoma (AMF) is a rare benign mesenchymal neoplasm that most frequently affects the lower genital tract of young to middle-aged women.<sup>[1]</sup> It commonly arises in the vulva but can also occur in perineum, vagina, retroperitoneum and inguinoscrotal region, scrotum, and occasionally spermatic cord in men.<sup>[2]</sup> Clinically, it is usually asymptomatic and resembles Bartholin's cyst or labial cyst. AMF is a well-circumscribed, nonencapsulated soft tumor, and microscopically, it consists of two components: stromal cells and numerous thin-walled blood vessels.<sup>[3]</sup> We report herein a case of AMF of vulva that presented as a large pedunculated mass.

## CASE REPORT

A 42-year-old Asian female patient presented with asymptomatic pedunculated mass in the vulva, which she noticed a year back and was gradually increasing in size since then. On examination, the mass was measuring 6.5 cm × 6 cm × 4.5 cm, soft in consistency, and having a stalk arising from labia majora

[Figure 1]. No inguinal lymph nodes were palpable. No other family or medical history was present. Simple excision was performed from the site.

## Gross

The surgically resected specimen was a well-circumscribed and nonencapsulated pedunculated mass measuring 6.5 cm × 6 cm × 4.5 cm. Cut surface showed homogenous gray–white areas. No areas of hemorrhage, necrosis, and cystic changes were noted.

## Microscopy

The excised tissue showed a tumor underlying stratified squamous epithelium with focal ulceration. The tumor was well circumscribed, nonencapsulated with hypercellular and hypocellular areas set in a collagenous stroma. Tumor showed numerous thin-walled capillaries surrounded by spindle-shaped cells and myxoid changes in the stroma. No nuclear atypia, mitosis, or necrosis was noted in the tumor. A diagnosis of AMF was thus rendered [Figure 2].

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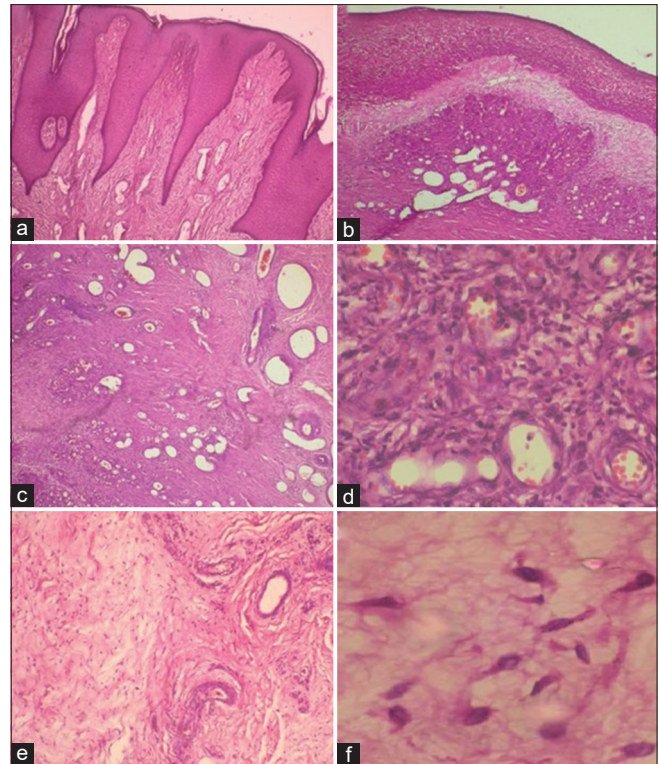


**Figure 1:** Gross appearance: Well-circumscribed pedunculated mass measuring 6.5 cm × 6 cm × 4.5 cm and on cut section homogenous gray–white areas noted

## DISCUSSION

The most commonly encountered tumors in the vulva are hemangioma, lipoma, fibroma, and leiomyoma.<sup>[4]</sup> AMF is a rare tumor of the vulva that was first described by Fletcher *et al.* in 1992. In 2015, Wolf *et al.* described 125 cases of female AMF in their review of literature, and most of the cases were either vulvar or vaginal in origin.<sup>[5]</sup> Since 2015, there have been additional fourteen cases of AMF reported in the English literature. Out of these eight cases were located in the vulva, rest in the broad ligament, cervix, inguinal hernia, glans penis, spermatic cord, and foot each.

AMF is a slow-growing benign tumor that mainly arises in the vulva. The size of AMF usually varies from 0.5 to 12 cm.<sup>[4]</sup> Only two cases of pedunculated AMF have been reported in the English literature to date.<sup>[3]</sup> It presents as a well-circumscribed, nonencapsulated mass in the subcutaneous tissue of the vulva. Microscopically, the tumor is characterized by myxoid stroma, numerous blood vessels, and plump stromal cells.<sup>[6]</sup> Other vulvar masses such as cellular angiofibroma, fibroepithelial stromal polyp, and superficial, aggressive angiofibroma (AAM) may resemble clinically and microscopically with AMF.<sup>[4]</sup> Its circumscribed border, alternating hypocellular and hypercellular areas, and thick-walled blood vessels help to distinguish it from AAM.<sup>[7]</sup> AMF has low tendency for local recurrence, whereas AAM has high chance of local recurrence when incomplete excision is done. However, Weiss *et al.* reported few cases of AMF with recurrence where the tumor was not completely excised or showed sarcomatous differentiation.<sup>[8]</sup> Immunohistochemically, the cells of AMF express immunoreactivity for vimentin, desmin, ER/PR, and occasionally for CD34, whereas AAM is much more frequently immunoreactive for CD34 and actin and negative for desmin.



**Figure 2:** Microscopy: (a and b) Tumor lined by stratified squamous epithelium with focal ulceration (H and E, ×40). (c and d) Numerous thin-walled capillaries surrounded by spindle-shaped cells (H and E, ×100 and ×400, respectively). (e and f) Myxoid stroma and spindle-shaped cells (H and E, ×100 and ×400, respectively)

The cell origin of AMF remains unclear. Some studies consider AMF to be from myofibroblastic origin, where strong immunoreactivity for vimentin and variable expression for desmin and alpha-smooth muscle actin support this hypothesis.<sup>[9]</sup> It has also been proposed that AMF might originate from perivascular stem cells with a capacity for myofibroblastic and fatty differentiation which is supported by capillary-like pattern demonstrated by stromal cells and obvious CD34 positivity around the vessels.<sup>[10]</sup> A recent study has demonstrated strong CD34 immunoreactivity in many other tumors including spindle cell lipoma, solitary fibrous tumor, inflammatory myofibroblastic tumor, dermatofibrosarcoma protuberans, and gastrointestinal stromal tumor. Hence, stem cell origin of AMF may not be supported by CD34 positivity alone. Since this tumor shows positivity for ER/PR, it provides the possibility of role of hormonal manipulation in its management.<sup>[11]</sup>

## CONCLUSION

AMF is the mesenchymal benign tumor of vulvar region rarely presenting as a large pedunculated mass. Complete surgical excision is the mainstay of the management of this tumor. AMF has to be differentiated from AAM, as the latter is a locally invasive tumor having infiltrative margins and requires wide excision.

### Ethical statement

The ethical approval of this study is exempted by the Institutional Ethics Committee of MVJ Medical College and Research Hospital.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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