

# Angiomyofibroblastoma of Vulva: Case Report with Histomorphological Differential Diagnosis and Review of Literature

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## Article Info

doi: [10.30699/jogcr.8.2.181](https://doi.org/10.30699/jogcr.8.2.181)

Received: 2022/10/01;

Accepted: 2022/12/22;

Published Online: 22 Feb 2023;

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## ABSTRACT

Angiomyofibroblastoma (AMF) is a benign mesenchymal neoplasm. They are typically well-circumscribed lesions, more commonly occurring in the vulvo-vaginal region of reproductive-age females. It is curable by complete excision. They do not have a characteristic clinical and radiological feature. It is an exclusive histopathological diagnosis. However, many mesenchymal neoplasms are predominantly perineal in location, with similar clinical and morphological features and may be confused with angiomyofibroblastoma. We present a 40-year-old married female with a 5.5 × 3.5 × 2 cm left vulval mass, clinically considered to be a Bartholin's cyst. Magnetic resonance imaging (MRI) was suggestive of infected cystic lesion. Histologically, it was an AMF. Histomorphological features are compared with other similar entities occurring in the vulva.

**Keywords:** Angiomyofibroblastoma, Aggressive angiomyxoma, Mesenchymal tumor, Vulva



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## Introduction

Angiomyofibroblastoma (AMF) is a rare, benign, non-recurrent mesenchymal neoplasm occurring mainly in the pelvi-perineal region of reproductive age females (1). They are composed of two major components, namely prominent blood vessels and stromal cells (2). In the literature, there is one case of AMF recurrence (3), and one case with sarcomatous transformation (4). Preoperative diagnosis of AMF and distinction from other soft tissue tumors is often difficult, as there are no characteristic imaging findings. It is often mistaken for a Bartholin's cyst, causing delays in diagnosis and treatment (1, 5). AMF needs to be differentiated from other mesenchymal tumors of the vulva (2, 6); more importantly from aggressive angiomyxoma (AAM) because of differences in their biological behavior (2). Local excision of the tumor mass is an adequate treatment for AMF. This report presents a case of AMF of the labia in a 40-year-old female. We have laid special emphasis on the differential diagnosis and review of literature.

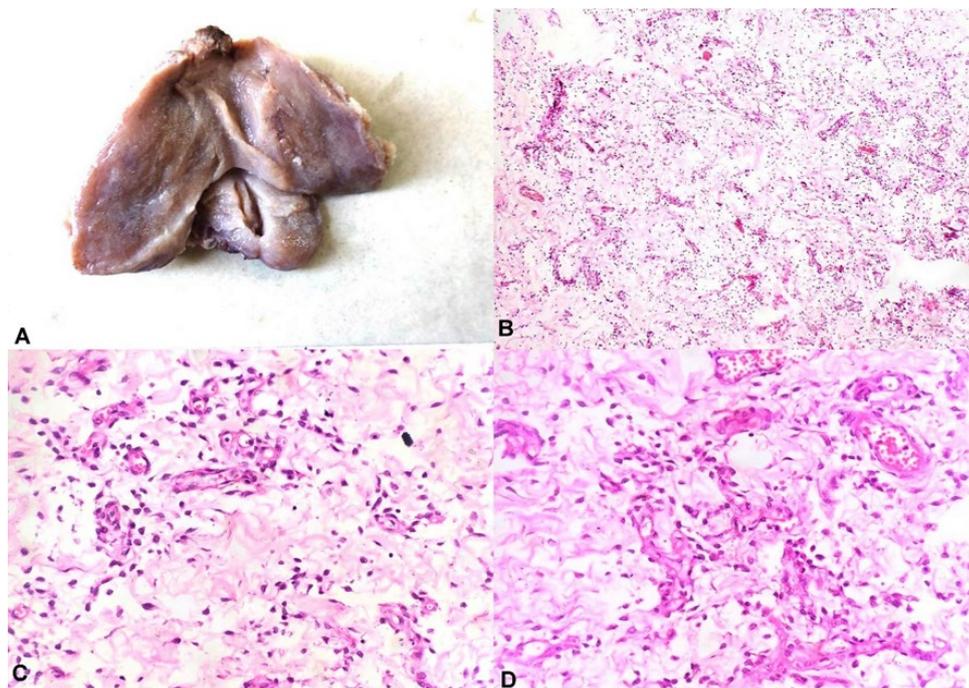
## Case Report

A 40-year-old married female presented in the obstetrics and gynecology clinic with menorrhagia and a slow-growing, painless mass in her left vulva, which gradually increased in size over a period of four months. She complained of dyspareunia. No other significant past, family, obstetric or gynecologic history was identified. She denied history of sexually transmitted disease, changes in appetite, bowel movements, dysuria, hematuria, fevers, chills, night sweats or weight loss. Laboratory investigations were within normal limits. On local examination, a 6×2 cm painless, cystic mass was identified in the left labia majora and was clinically diagnosed as a Bartholin's cyst. Ultrasonography (local part) showed a 6.5×2.4 cm well-defined heterogeneously iso-hypoechoic lesion with multiple dense echoes. Magnetic resonance imaging was subsequently performed for further characterization which revealed a 5.9×3×1.7 cm well-defined, heterogeneous altered

signal intensity lesion involving the left labia majora; hypointense on T1W and hyperintense on T2W and STIR images. Possible radiological differentials included an infected hydrocele of the canal of Nuck, Bartholin's cyst and epidermoid vulval cyst. Complete excision of the cystic swelling was performed with clear margins.

The specimen was a 5.5×3.5×2 cm single, nodular, pseudo-encapsulated mass with a pale-pink, soft, solid, homogenous and fleshy cut surface (Fig 1 a). There was no evidence of cystic areas, necrosis or hemorrhage. Microscopic examination revealed a tumor with alternating hypercellular and hypocellular

areas, comprising spindle cells with bland, hyperchromatic nuclei, indistinct nucleoli and eosinophilic cytoplasm that appeared to be clustered around numerous capillary-sized thin-walled blood vessels. The stroma was loose, edematous and collagenous (Fig. 1 b-d) and showed the presence of scattered chronic inflammatory infiltrate. Occasional intralesional adipose tissue was seen. However, there was no evidence of mitosis, necrosis, perivascular hyalinization, atypia or malignancy in the sections studied and a diagnosis of AMF was given. No evidence of recurrence was seen in the three months since excision.



**Figure 1.** a) Gross photograph of single, nodular, pseudo-encapsulated mass with soft, solid, homogenous, fleshy, pale-pink cut surface; b) Photomicrograph showing alternating hypocellular and hypercellular areas (Hematoxylin and Eosin stain, 40×); c, d) Photomicrograph showing spindle cells with bland, hyperchromatic nuclei, indistinct nucleoli and eosinophilic cytoplasm that appeared to be clustered around numerous capillary-sized, thin-walled blood vessels against a loose edematous and collagenous stroma. (Hematoxylin and Eosin stain, 100×).

## Discussion

AMF is a rare, benign, non-recurring mesenchymal tumor composed of myofibroblasts and blood vessels. It was first recognized by Fletcher *et al.*, in the early 1990s (5). It occurs almost exclusively in the subcutaneous tissue of the vulvo-vaginal region of women but can also occur occasionally in the ischio-rectal fossa, cervix, and bladder, as well as similar tumors have been reported in the spermatic cord, scrotum, and perineum of men (7). It is thought to be derived from the subepithelial mesenchyme of the lower genital tract (1).

It presents as a well-circumscribed, 2 to 8 cm, painless mass. The cut surface is usually soft, tan-white and solid. Histologically, it is a well-demarcated neoplasm composed of an admixture of numerous delicate, thin-walled, capillary-sized vessels and plump

round to spindle-shaped stromal cells. The spindle-shaped cells are typically clustered around the prominent vasculature. The stromal cells have nuclei with fine chromatin and inconspicuous nucleoli and moderate amounts of eosinophilic cytoplasm; set within a variably edematous to collagenous matrix with alternating zones of cellularity. Occasionally, intralesional adipocytes may be present and lack mitoses. The stromal cells are typically desmin positive and show variable positivity for actin (1, 2, 6).

Though the principal differential diagnostic consideration is AAM, other common vulval mesenchymal lesions include superficial angiofibroma, cellular angiofibroma, fibroepithelial stromal polyp, mammary-type myofibroblastoma,

superficial (cervicovaginal/vulvovaginal) myofibroblastoma (SCVMF) and glomus tumor (2, 6).

AAM is most likely to be confused with AMF because they share many features including age at presentation, location, clinical manifestations and pathological features, but AAM has infiltrative margins, pauci-cellularity, large and thick-walled blood vessels and stromal mucin. They are potentially destructive lesions with a local recurrence rate of 30-50% (2, 6). On the other hand, superficial angiomyxoma has a more lobulated and distinct margin. Histologically it can be differentiated from AMF due to the hypocellularity and presence of delicate, elongated and thin-walled blood vessels (6). Cellular angiofibromas most commonly occur in the vulva. It also occurs at reproductive age and is a well-circumscribed subcutaneous mass like AMF. However, unlike AMF it is more uniformly cellular and shows presence of large thick-walled vessels and perivascular hyalinization (2, 6). Fibroepithelial stromal polyp is typically an exophytic polypoid lesion with variable cellularity and shows the presence of large, thick-walled blood vessels within the central core (2). Mammary-type myofibroblastoma occurs typically after 40-years-of-age. Though it is well-circumscribed, it shows variable cellularity and is composed of bland spindle cells with short, stubby nuclei admixed with collagen bundles and adipocytes. It lacks the prominent vascular component of AMF (2, 6). SCVMF is more superficially located than AMF and composed of moderately cellular spindle, stellate to epithelioid cells with scant cytoplasm, arranged in a reticular pattern. They have inconspicuous vascularity (6). The

perivascular clustering of cells in AMF may suggest a glomus tumor. However, glomus tumor has a uniform cellularity and is composed of sheets of round-shaped cells with indistinct borders and rounded, sharply punched-out nucleus and eosinophilic cytoplasm (2).

AMF has no risk of recurrence or metastasis, therefore; simple total surgical excision of the lesion is the treatment of choice.

## Conclusion

In conclusion, AMF is a distinctive neoplasm of reproductive age females. A wide variety of lesions can present with similar clinicoradiological features. Recognition of this entity is important to avoid misdiagnosis and confusion with other vulval mesenchymal neoplasms.

## Acknowledgments

None.

## Conflict of Interest

The authors declare that they have no conflict of interest.

## Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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### How to Cite This Article:

Shetty, D., Khurana, R., Bhalekar, S. Angiomyofibroblastoma of Vulva: Case Report with Histomorphological Differential Diagnosis and Review of Literature. *J Obstet Gynecol Cancer Res*. 2023; 8(2):181-3.

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