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Dr. Pinki
2nd Year Resident Doctor,
Department of Obstetrics and
Gynaecology, JLN Medical College,
Ajmer, Rajasthan, India

Dr. Suchitra Narayan
Associate professor, Department of
Obstetrics and Gynaecology, JLN
Medical College, Ajmer, Rajasthan,
India

Dr. Purnima Panchor
Senior Professor, Department of
Obstetrics and Gynaecology, JLN
Medical College, Ajmer, Rajasthan,
India

Dr. Dharmendra Fathehpuria
Associate professor, Department of
Obstetrics and Gynaecology, JLN
Medical College, Ajmer, Rajasthan,
India

Corresponding Author:
Dr. Suchitra Narayan
Associate professor, Department of
Obstetrics and Gynaecology, JLN
Medical College, Ajmer, Rajasthan,
India

Angiomyofibroblastoma (AMFB) of the vagina: A case report

**Dr. Pinki, Dr. Suchitra Narayan, Dr Purnima Panchori and
Dr. Dharmendra Fathehpuria**

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Abstract

Angiomyofibroblastoma (AMFB) is a rare histopathologic finding of the female lower genital tract. It is a rare benign soft tissue mesenchymal subcutaneous vulvovaginal tumour. The vaginal location of this tumour is very rare. This tumor belongs to the group of mesenchymal tumors.

We reporting AMFB arising from the vagina to draw the attention of pathologists the fact that this rare neoplasm can occur outside the vulva, even in this study, a case of vaginal cellular angiofibroma reported in a 51-year-old female presenting with mass coming per vagina. The case was diagnosed based on morphology and immunohistochemistry and was treated surgically.

Keywords: Angiomyofibroblastoma, mesenchymal tumors, vagina

Introduction

Angiomyofibroblastoma is a rare, recently described, benign well circumscribed soft tissue tumour that occurs mainly, but not exclusively, in the vulval region of premenopausal women [1]. While majority of the tumors are encountered in the vulva, 10% to 15% of them occur in the vagina, occasionally in the perineum and the inguinal region while rare reports of fallopian tube lesions are also found.

These tumors are usually seen premenopausal or peri menopausal women [1]. Differential diagnosis of mesenchymal neoplasms in the vulvovagina includes an array of lesions that are bland and overlapping in both their morphology and immunohistochemistry. In addition to their rarity, this creates a significant dilemma in making the correct diagnosis. Among these, aggressive angiofibroma and cellular angiofibroma are the important mimickers of AMF, Bartholin gland cyst.

Case Report

A 51-year-old woman, para 3 (vaginal deliveries), presented at the gynecology outpatient ward at the R.M.C Hospital with a presumed diagnosis of genital prolapse. She complained of dyspareunia, vaginal bulge for three years, which had progressively grown until it was exposed in the vaginal introitus.

On gross Examination tumor was well circumscribed of 12 × 15 cm in size soft to rubbery consistency & bulging pink with lobulated surface arising from distal postero-lateral vaginal wall (Figure 1&2). There was no prolapse. The tumorous mass was fibroelastic and painless.

On bimanual examination – Cervix mobil downward uterus RVRFBULKY B/L Fx free.

Rectal examination did not show abnormalities or compression due to the mass, surgically removed after biopsy confirmation. Histologically, alternating hypercellular and hypocellular edematous areas were seen, and the spindle plump oval stroma of fibro-connective tissue with oedematous stroma and no mitotic activity, abundant blood vessels of various wall thickness.

Discussion

AMF is a rare, benign, soft-tissue tumor that shows myofibroblastic differentiation and represents neoplastic proliferation of stromal cells [1]. AMF was first described by Fletcher *et al.* in 1992. AMF is composed of myofibroblastic cells and prominent thin-walled blood vessels within a fibromyxoid stroma [2]. Following that, several reports had been published where

Majority was vulvo-vaginal origin [3, 4]. Its cause and pathogenesis are unknown at present. Since AMF is rare, no statistical data are available about its incidence in the general population. Most cases of AMF are benign, and only one case with sarcomatous transformation has ever been reported. A rare lipomatous variant of AMF with local invasion has also been reported.

In most reports, AMF has occurred in women aged between 20 and 50 years, i.e., during the reproductive years. In our case, the patient is aged 41 years old.

There are very rare reports of AMF occurring in male patients. In most patients, AMF presents as a painless neoplasm located in the superficial regions of the lower female genital tract, such as the vulva and vagina, and are therefore easily detected. Tumors arising in the cervix, uteri, or urethral region sometimes present with obstructive symptoms. Tumors in the pelvis, iliac fossa or peritoneal cavity are extremely rare, and usually grow insidiously and reach a massive size before they are detected. Their greatest dimensions have ranged from 3.8 cm to 25 cm. To our knowledge, only four cases of pelvic AMF have been reported at least so far [5].

Several reports have analyzed the imaging features of AMFs. On perineal ultrasonography, AMFs appear as a soft-tissue mass with inhomogeneous mixed echogenicity, which corresponds to the cellular inhomogeneity found on histopathological examination.

Therefore, the ultrasonographic characteristics of AMFs may help to differentiate them from other mesenchymal neoplasms. On CT imaging, AMFs most likely show moderate-to-strong enhancement, which may reflect the prominent vascularity of these tumors.

Immunohistochemistry showed that the tumor expressed estrogen and progesterone receptors, which suggests that it might have originated as a neoplastic proliferation of hormonally responsive mesenchymal cells. In our case, we report a case of vaginal AMF. It was firm, pearly white in color. Characteristic histological features of angiomyofibroblastoma were appreciated. Immunohistochemical staining showed strong positivity of the stromal cells for desmin and vimentin. There was no staining of stromal cells for actin and S 100 protein. The blood vessels were well-highlighted by staining with SMA. Diffuse strong positivity of stromal cells for ER and PR receptors was noted.

Because of their location, AMFBs could be mistaken for Bartholin cysts or lower genital tract lipomas, causing delays in diagnosis and treatment [6].

Differential diagnosis of the vulvar mass includes other benign tumors such as cellular angiofibroma, fibroepithelial stromal polyp, or superficial angiomyxoma. These benign mesenchymal tumors are mostly found at the vulvovaginal area and organize a distinctive disease entity [7].

Aggressive angiomyxoma (AAM) is most likely to be confused with AMF because it shares many features with AMF including age at presentation, location, clinical manifestations, and pathological entities. However, AAM presents as a malignant, locally infiltrative, non-metastasizing stromal neoplasm with a strong tendency to recur. On microscopic examination, AMFs generally show much higher cellularity, more numerous blood vessels and more frequent plump or short spindle-shaped cells (Figure 3) in contrast, AAM cells are sparsely and diffusely distributed, without the characteristics of alternating density and

aggregation around small blood vessels [4]. AAMs show more distinctive myxoid degeneration than AMFs. AMFs are characterized by the expression of vimentin, desmin and CD34, suggesting an undifferentiated mesenchymal tumor with preferential myofibroblastic differentiation. Desmin expression was previously thought to be specific for AMFs, but positive expression of desmin has been found in some cases of AAMs. Most AAMs are also positive for estrogen and progesterone receptors [8]. Hence, desmin, estrogen receptor and progesterone receptor are no longer considered reliable markers for distinguishing AAMs from AMFs. After surgical treatment, 30% of AAMs have been found to relapse within 2 years [9, 10].

Since the morphology and IHC markers of AMF are similar to those of AAM, differentiation between these two tumors is largely based on the appearance of the tumor margins (AMFs AMFB's treatment is always simple excision. It is curable with surgical simple excision and no recurrence in the follow-up [11], which confirms the benign nature of AMF. AMF shows no propensity for infiltrative growth [12].



Fig 1: On gross Examination tumor was well circumscribed of 12 × 15 cm in size soft to rubbery consistency & bulging pink with lobulated surface arising from distal postero-lateral vaginal wall



Fig 2: On gross Examination tumor was well circumscribed of 12 × 15 cm in size soft to rubbery consistency & bulging pink with lobulated surface arising from distal postero-lateral vaginal wall

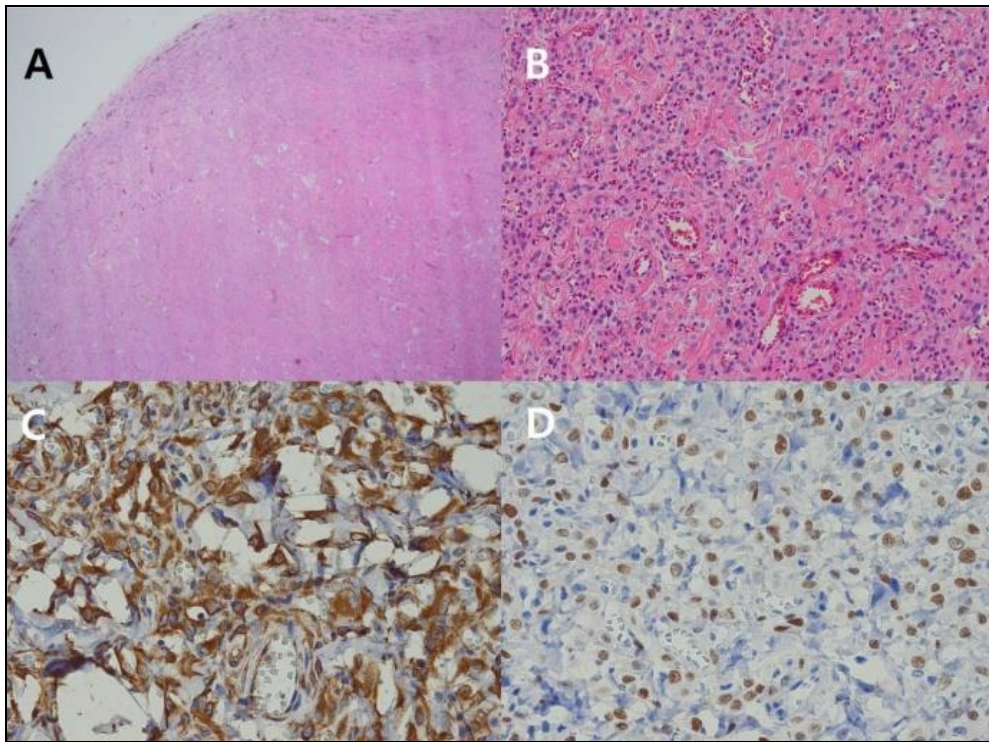


Fig 3: (A) The tumor mass showing well circumscribed border (hematoxylin-eosin, $\times 12.5$). (B) High cellularity of plump bland-looking stromal cells and abundance of blood vessels (hematoxylin-eosin, $\times 200$). (C) Diffuse strong positive finding of stromal cells in vimentin immunohistochemical stain ($\times 400$). (D) Strong nuclear positive finding of stromal cells in estrogen receptor immunohistochemical stain ($\times 400$).

Conclusion

Although the exact nosologic position of AMF is still surrounded by some controversy and requires further elucidation, we conclude that our case represents the eighth report of AMF of the vagina, based on its conventional histopathologic and immunophenotypic features. Simple excision appears sufficient for the surgical management of AMF, whereas wide excision after histological examination is needed for the management of the related AAM, which is associated with a high risk of local recurrence and infiltration.

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