



Two Cases of Female Genital Angiomyofibroblastoma: Case Report and Review of the Literature

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Abstract

Angiomyofibroblastoma (AMFB) is a rare benign tumor of the female lower genital tract. It is a mesenchymal tumor which characterized by Spindle Cells. We report two cases of vaginal and vulvar angiomyofibroblastoma. The positive diagnosis was made on the basis of morphology and immunohistochemistry. Tumors were treated surgically. It is important to differentiate AMFB from aggressive Angiomyxoma because they have different treatment.

Keywords: Angiomyofibroblastoma; Mesenchymal tumors; Vagina

Introduction

Angiomyofibroblastoma is an uncommon benign mesenchymal tumor. It was described by Fletcher et al. [1]. Several locations have been reported in the literature. The vulvovaginal area is the most frequent location. The other locations reported are fallopian tube, broad ligament and less frequently in the male genital tract [2-4].

This tumor is seen especially in patients with genital activity and perimenopause [5]. The tumor is usually asymptomatic but patient can present pelvic discomfort, dyspareunia or vaginal bleeding. Physical examination usually finds a well-circumscribed elastic tumor typically sized <5 cm. Studies reported tumor size between 3 cm and 25 cm. Differential diagnosis includes both benign and malignant tumor. It is important to differentiate AMFB from Aggressive Angiomyxoma (AAM) because therapeutic care is different.

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Case Presentation

Case one reports a 41-year-old woman gravida 2, para 1. She consults for dyspareunia. The physical examination objectified a 6 cm tumor on the left posterior vaginal wall.

Pelvic and abdominal ultrasonography were normal. Perineal ultrasonography showed a 6 cm heterogeneous mass which prolapsed laterally in Douglas without extension to neighboring structures. The patient had a lymphectomy with simple suites.

Case two reports a 23-year-old woman gravida 2 para 1 presented with a painless vulvar tumor that appeared since two years. She underwent tonsillectomy 6 years ago and has no other medical or surgical history. Physical examination revealed an approximately 5 cm × 2 cm sized mass in the left labia majora without inflammatory signs (Figure 1). The patient underwent surgical resection of the mass. Hemostasis was difficult (Figure 2). The anatomo-pathological study concluded in an Angiomyofibroblastoma (Figure 3).

Discussion

AMFB is a rare pathology. Several reports analyzed the clinical, paraclinical and anatomopathological characteristics.

Clinical symptoms are non-specific.

Transvaginal Ultrasonography is the first imagery. It is an available and efficient exam. MRI is a sensitive examination but access is more difficult. It is recommended as a second line imaging. Computed Tomography (CT) is not recommended [6].

AMFBs appear hyperechoic on perineal ultrasound with irregular and small hypoechoic cystic areas interspersed in homogeneous echogenic stroma [7,8]. AAM appears as a hypoechoic mass with

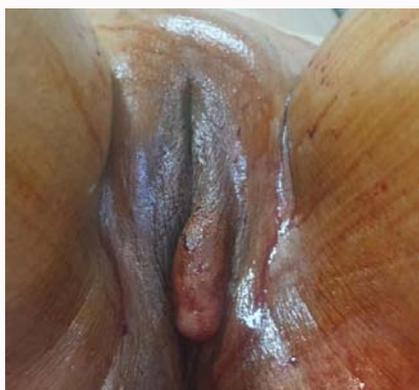


Figure 1: Vulvar tumor.



Figure 2: Intraoperative appearance of the tumor.

homogenous echogenicity and occasional echogenic septa correlating to fibrous bands [9,10]. The vascularization of the AMA is greater on the color Doppler than AMFB [11]. Most of the time MRI reveals a well circumscribed tumor hypointense on T1 weighted images and hyperintense on T2 weighted images.

Denomination AMFBs is based on the two components of this tumor: The blood vessels and stromal cells. Although most pathological characteristics of myofibroblast are lacking, the use of myofibroblastoma is justified because the tumor is desmin positive which is a reliable marker of myoid differentiation.

Histological features of AMFBs are spindle shape. They have a fascicular, wavy or palisade pattern, with areas of high and low cell density. They have also stromal proliferation of small vessels.

Immunohistochemically, AMFB cells are positive for vimentin, desmin estrogen receptor and progesterone receptor. But AMFB cells are negative for S-100 and α -Smooth Muscle Actin (α -SMA). Few cases of desmin-negative and α -SMA-positive have been reported [12-14].

In our study, we report a case of vaginal AMFB and a case of vulvar AMFB. Both showed characteristic histological features of Angiomyofibroblastoma and positivity of stromal cells for estrogen and progesterone receptors.

In the first case, it was a firm, pearly white mass. Stromal cells have strong positivity for desmin and vimentin. There were no

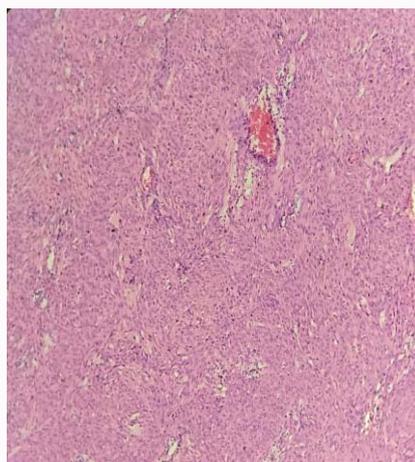


Figure 3: Spindle cells.

stromal cells for actin and S100 protein. The blood vessels were well highlighted by staining with SMA.

The second case showed a whitish lesion, fully circumscribed. Tumor cells were also positive for CD34 and negative for cytokeratin. Index of proliferation Ki 67 was 2%.

We report several differential diagnoses of AMFB such as cellular angiofibroma, fibroepithelial stromal polyp or superficial angiomyxoma. These tumors are found at vulvovaginal area and represent a disease entity [15].

AAM is the primary diagnosis to differentiate from AMFB. They have the same clinical presentation. Differentiation between these tumors is important since treatment and prognosis differ. While AMFB needs a complete excision without additional therapy, aggressive angiomyxoma needs a wide excision and may require hormonal treatment because of its risks of infiltration and recurrency. AAM is a non-metastasizing tumor.

AAM cells are sparsely and densely distributed, without the characteristics of alternating density and aggregation around small blood vessels. However, AMFBs have higher cellularity more numerous blood vessels and more frequent plump or short spindle-shaped cells (Figure 3) [4]. AAMs are characterized by myxoid degeneration.

AAMs can also have positive for estrogen and progesterone receptors and positive expression of desmin.

The expression of vimentin, desmin and CD34 in AMFB, suggest an undifferentiated mesenchymal tumor with preferential myofibroblastic differentiation.

Large AMFBs are characterized by well-developed blood vessels. The risk is an important intraoperative blood loss. To reduce blood loss preoperative embolization was reported.

Conclusion

Angiomyofibroblastoma is a rare tumor that occurs mainly, but not exclusively, in the vulvovaginal region. Clinical findings and radiology may guide the diagnosis. Confirmation is based on histopathologic exam and immunophenotypic features. Differential diagnosis includes an array of pathologies but it is mostly important to differentiate it from AAM.

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