



Angiomyofibroblastoma of the Vulva: A Case Report

Sultanoglu B* and Demirbakan K

Department of General Surgery, Dr. Ersin Arslan Training and Research Hospital, Turkey

Abstract

Angiomyofibroblastoma (AMFB) is benign mesenchymal tumor and the incidence of AMFB is very rare. AMFB is usually occurs in the vulvo-vaginal area. It is frequently seen in middle age women. It is often mistaken for other benign disease such as Gartner's duct cyst, epidermal inclusion cyst, leiomyoma, inguinal hernia and fibroepithelial polyps. It is most important to distinguish AMFB from Aggressive Angiomyxoma (AA). AA has strong tendency to recur. On microscopic examination, AMFBs generally show higher cellularity, more numerous blood vessels and more frequent plump or short spindle-shaped cells whereas AAs have sparsely and diffusely distributed cells without the characteristics of altering density and aggregation around small blood vessels and also AAs show more distinctive myxoid degeneration than AMFB. AMFBs main characteristics are expression of vimentin, desmin and CD34.

Introduction

Angiomyofibroblastoma (AMFB) is benign mesenchymal tumor and the incidence of AMFB is very rare. AMFB is usually occurs in the vulvo-vaginal area. It is frequently seen in middle age women. [1]. AMFB usually measures less than 5 cm, but in literature review it is reported up to 23 cm in size [2]. AMFB can affect fallopian tube, ischiorectal fossa, cervix and bladder it is often misdiagnosed as Bartholin cyst in women [3]. We report a case of Angiomyofibroblastoma of vulva in 35 years old woman.

Case Presentation

A 35 years old woman was admitted to our hospital with complaint of swelling in the vulvar region for 4 months that made her feel uncomfortable. On physical examination, a soft, mobile and painful mass, measuring approximately 7 cm × 4 cm × 4 cm, was seen in the right labia majora. Preoperative routine blood tests were normal. Abdomen ultrasound (US) was performed and showed a complex mass and for further imaging tests computed abdomen tomography, was recommended. Computed Tomography (CT) was performed for further examination (Figure 1). Surgical excision was performed for patient under spinal anesthesia. Peroperative findings of the tumor were in the subcutaneous tissue measured 7.5 cm × 4 cm × 3 cm, non-capsulated, lobulated surface and non-vascular. The tumor was brownish, in the cut section was gray-whitish and medium firm (Figure 2 and 3). Histologically, the tumor was notable for hypocellular edematous myxoid stroma tissue alternating with walled small to medium sized vessels, consistent with angiomyofibroblastoma. Tumor cells were round to spindle shaped with eosinophilic cytoplasm. Mitosis were rare and had immunoreactivity for desmin. Estrogen and progesterone receptors were positive and CD34 and protein S100 were negative.

OPEN ACCESS

*Correspondence:

Sultanoglu B, Department of General Surgery, Dr. Ersin Arslan Training and Research Hospital, Gaziantep, Turkey, E-mail: drbulentsultan@hotmail.com

Received Date: 27 Nov 2018

Accepted Date: 10 Jan 2019

Published Date: 15 Jan 2019

Citation:

Sultanoglu B, Demirbakan K.

Angiomyofibroblastoma of the Vulva: A Case Report. *World J Surg Surgical Res.* 2019; 2: 1091.

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Figure 1: Computed Tomography of abdomen.



Figure 2: Removal of subcutaneous tissue.



Figure 3: Non-capsulated subcutaneous tissue.

Discussion

AMFB was first described by Fletcher et al. [4]. Clinically, these tumors can be misdiagnosed as other benign cases. Therefore Bartholin's gland cyst, inguinal hernia, leiomyoma, lipoma or liposarcoma must be kept on mind in differential diagnosis [5]. Angiomyofibroblastoma has characteristic histomorphologic features and shows myofibroblastic differentiation. AMFBs are generally well demarcated by a thin fibrous pseudocapsule and have alternating hypo- and hypercellular areas with abundant thin-walled blood vessels which are surrounded by stromal cells within an oedematous to collagenous matrix [6]. Differential diagnosis for AMFB includes Gartner's duct cyst, epidermal inclusion cyst, leiomyoma, inguinal hernia and fibroepithelial polyps among the more common etiologies. It is important to distinguish AMFB from Aggressive Angiomyxoma (AA). AMFB and AA can commonly be misdiagnosed because of their anatomic and pathologic similarities. AA is known to recur in 33% to 72% of cases and is locally invasive, often entrapping nerves and mucosal glands. In literature review, it is reported that AA has no distant metastasis [7]. Surgical approach to AA requires wide local excision due to the infiltrative nature of the lesion while simple excision

is enough for AMFB. AA is easily confused with AMFB. Because age, presentation, location and clinical manifestations are similar. But AA is a malignant and locally infiltrative, non-metastasizing stromal neoplasm. AA often recurs. On microscopic examination, AMFBs generally show higher cellularity, more numerous blood vessels and more frequent plump or short spindle-shaped cells whereas AAs have sparsely and diffusely distributed cells without the characteristics of alternating density and aggregation around small blood vessels and also AAs show more distinctive myxoid degeneration than AMFB [4,8]. Expression of vimentin, desmin and CD34 is seen in AMFBs. Most of AMFBs have positive progesterone and estrogen receptors. In our case, desmin expression, estrogen and progesterone receptors were positive, but CD 34 was negative. In the literature review, AMFBs have been found to relapse within 2 years [4].

Conclusion

The patients with vulvar or inguinal mass must be evaluated carefully and it is important to consider AA among differential diagnosis, particularly, because, while the treatment of AMFB requires simple excision, AA requires wide local excision. Histological examination must be performed in postoperative period for definitive diagnosis. We aim to announce AMFB as a rare tumor and to consider AMFB and AA among differential diagnosis.

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