Vulvar Angiomyofibroblastoma: A Case Report

Vulvar Anjiyomiyofibroblastom: Bir Olgu Sunumu

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ABSTRACT

Angiomyofibroblastoma (AMFB) is a rare seen mesenchymal tumor that is categorized as a genital stromal tumor. It is commonly seen in the middle-aged women usually affecting the labia majora and rarely the vagina. A variant called AMFB-like tumors are also rarely seen in male patients. AMFB with its clinical presentation and location can be wrongly diagnosed as an aggressive angiomyxomas, bartholin cyst or lower genital tract lipomas. In the patients who are applying because of the vulvar mass, AMFB should not be forgotten for the pre-diagnosis. The treatment is generally simple surgical excision. In this case, AMFB was reported with a wrongly pre-diagnosis as a bartholin cyst to which a medical treatment had been given.

Key words: Angiomyofibroblastoma; vulvar; bartholin cyst.

INTRODUCTION

Angiomyofibroblastoma (AMFB) is a rare seen mesenchymal tumor that is categorized as a genital stromal tumor and was first defined by Fletcher et al. in 1992¹. It is seen commonly in females between 40-50 ages in their premenopausal stage, usually affecting the labia majora but rarely the vagina². AMFB-like tumors are also rarely seen in male patients affecting the scrotum, perineum or spermatic cord¹³.

AMFB which is clinically asymptomatic, can be observed as a lesion that is painless and grows slowly commonly in the vulvar region²⁴. Aggressive angiomyxoma, bartholin cyst or lower genital tract lipomas are predominantly vulvoperineal in location so may be confused with AMFB⁵. That is why, patients who are applying because of a vulvar mass, AMFBs that are one of the benign genital stromal tumors, should be evaluated for pre-diagnosis¹⁵. The treatment of AMFB is simple excision, and it is cured by this
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procedure and no recurrence with the clear margin of the lesion⁵.

In this case, AMFB was reported with a wrongly pre-diagnosis as a bartholin cyst to which a medical treatment had been given in another medical center.

CASE REPORT

A female multipar patient aged 50, applied to our clinics because a vulvar mass could be felt by hand. It was determined in the anamnesis of the patient that she had applied to a gynecologist six months ago because of the same complaint, been treated with antibiotherapy due to the pre-diagnosis of bartholin cyst and applied to the Gulhane Military Medical Academy, Obstetrics and Gynecology Department regarding the growth in the mass.

The patient's gynecological examination showed a semi-solid, mobile mass with 5x3 centimeters in diameter, at the level of left labia majora. Bimanual examination was observed normal. In transvaginal ultrasonographic examination (General Electric Logiq S6®, 1.5-4.5 MHz probe, Waukesha, WI U.S.A.) the uterus 6x9x4 centimeters in diameter; the uterine myometrium was homogeneous, no focal lesion was seen within it; the endometrial three layer pattern measured at 4 millimeters and the bilateral adnexal areas were assessed as normal.

An operation was performed on the patient with the pre-diagnosis of vulvar lipoma. A vertical incision was performed to the skin tissue over the mass (figure 1A). After dissection, the mass with the color of white-grey was excised easily with the clear margin of the lesion (figure 1B) and the operation was ended.

After a histopathologic observation of the mass, it was reported that the mass was benign stromal tumor, and conformed with AMFB morphological observations (figure 1C). The histopathologic observation was compatible with immunoreactivity for vimentin and vascular clusters in a hyalinized hypocellular stroma were seen.

There was no recurrence in the 6-month follow up.

Figure1 A: Vertical incision, 1B: Well-demarcated, solid mass dissection, 1C: Vascular clusters in a hyalinized hypocellular stroma were seen and immunoreactivity for vimentin (HE x100).
DISCUSSION

Although genital stromal tumors are rarely seen, they are especially observed in vulvar region[1]. It is commonly seen in females between 40-50 ages in their premenopausal stage, and it usually involves the labia majora and rarely involves the vagina2. AMFB-like tumors are also rarely seen in male patients affecting the scrotum, perineum or spermatic cord1,3.

Because of their location, AMFBs could be mistaken for bartholin cysts or lower genital tract lipomas, causing delays in diagnosis and treatment5. As seen in our case, misdiagnosis of AMFBs resulted in unnecessary antibiotherapy that was given to the patient after having been wrongly pre-diagnosed with bartholin cyst.

In English medical research literature, there are nearly 65 cases about AMFB published between 1997 and 2014. According to literature, malign transformation was seen in only 1 out of 65 cases6.

In the macroscopic examinations of AMFB cases, it is seen that they are solid lesions that are generally well-demarcated, in the color of pink and with the diameters of 0.5-12 centimeters7. In microscopic observation, vessel clusters can be observed within the hypocellular stroma. It is usually believed that CD34 positive stem cells normally reside around vessel clusters5. Nuclear atypia and a rise in mitosis are rarely seen7. In the immunohistochemical process, most tumor cells tested strongly positive for desmin and vimentin. The tumor cells also show variable expression for muscle actin and are often positive for estrogen and progesterone receptors7.

AMFB's treatment is always simple excision. It is curable with surgical simple excision and no recurrence in the follow-up9,10.

Cases of AMFB, as mentioned above, can be wrongly diagnosed as bartholin cysts, lower genital tract lipomas or aggressive angiomyxomas. That is why, as in our case, unnecessary medical treatments are mistakenly given to the patients who have been pre-diagnosed with bartholin cyst. We would like to mention about some strategies for avoiding misdiagnosis with bartholin cyst. Bartholin cyst is usually 1 to 4 centimeter in size fluid-filled swelling, which may cause no problems. However, a larger bartholin cyst may cause some pain or discomfort when walking, sitting or sexual intercourse11. When bartholin cysts become infected, patients usually present with severe pain and pus-filled swelling, maybe have fever and skin tissue hyperemi over the abscess formation11. But AMFB is solid, well-demarcated lesion and larger AMFB with the diameters of 12 cm may be painless7.

In conclusion, AMFB is one of the circumscribed solid lesions of the external genitalia. Aggressive angiomyxoma, lower genital tract lipoma and bartholin cyst are predominantly located in the vulva and may be confused with AMFB. So, gynecologists would like to know AMFBs' symptoms and differential diagnosis with the three entities mentioned before.

REFERENCES


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