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CASE REPORT

# A Vulvar Angiomyofibroblastoma in A Child: The Youngest Case And Literature Review

## Kız Çocuğunda Vulvar Anjiomyofibroblastoma: En Genç Olgu ve Literatür Tarama

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## *ABSTRACT*

***Introduction:*** *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 vulva 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. The treatment of this rare tumor is generally a simple surgical excision.*

***Case-Discussion:*** *In the following case, an AMFB that was seen in childhood for the first time according to the literature with it's clinical and histopathological characteristics, was explained.*

***Keywords:*** *angiomyofibroblastoma; child; vulvar*

## *ÖZET*

***Giriş:*** *Anjiyomyofiblastomlar (AMFB), genital stromal tümörler içerisinde yer alan, nadir izlenen benign mezenkimal tümörlerdir. Çoğunlukla orta yaşlı kadınlarda izlenmekle birlikte, özellikle vulvayı nadiren de vajinayı tutabilmektedir. AMFB benzeri tümörler adı altında, nadiren erkekler de görülebilmektedir. Klinik olarak yerleşim yeri ve klinik prezentasyonu ile bartolin kistleri, alt genital tract lipomları veya nadiren de agresif anjiyomiksomalar ile karışabilmektedir. Bu nadir tümörlerin tedavisi genellikle basit cerrahi eksizyondur.*

***Olgu-Tartışma:*** *Bu olguda, literatür bilgimize göre çocukluk çağında olan 11 yaşında ki kız çocuğunda ilk kez izlenen AMFB, klinik ve histopatolojik karakteristiği ile bahsedildi.*

***Anahtar Kelimeler:*** *anjiyomiyofibroblastoma; çocuk; vulvar*

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## INTRODUCTION

Angiomyofibroblastoma (AMFB) which was defined by Fletcher et al in 1992, is a rarely seen mesenchymal tumor and it is categorized in genital stromal tumors [1]. It was observed that its main area of effect is vulva especially the labia majora; but it is rarely in the vagina [2]. AMFB-like tumors are also rarely seen in male patients affecting the scrotum, perineum or spermatic cord [1, 3].

AMFB is common in women predominantly in their premenopausal period within the age interval of 40-50 [2]. However, AMFB observed in a 17- year- old teenager patient was placed in the literature as a youngest case by Qublan et al. [4].

The simple surgical excision is the main treatment of AMFB, and no recurrence is observed in the clear margin of the lesion [5].

In the following case, AMFB case observed in childhood for the first time according to the literature was explained.

## CASE REPORT

11- year old child, was consulted to our clinics from pediatric services, because of a painless vulvar mass that was felt by hand. It was determined in the anamnesis of the patient's parents that she had applied to a pediatrician five months ago because of the same complaint, not been treated due to the pre- diagnosis of vulvar lipoma and applied to the Gulhane Military Medical Academy, Pediatric Department regarding the growth in the mass.

The patient's gynecological examination showed a solid, mobile mass with 2.5x1 centimeters in diameter, at the level of left labia majora. In lower abdomen ultrasonographic examination (General Electic Logiq S6® , 1.5-4.5 MHz probe, Waukesha, WI USA) the uterus 4x6x3 centimeters in diameter; the uterine myometrium was homogeneous, no focal lesion was seen within it; the endometrial three

layer pattern measured at 3 millimeters and the bilateral adnexal areas were assessed as normal with ovarian follicular activity. An operation was planned to the patient because of the growth in the vulvar mass.

An operation was performed for the patient with the pre- diagnosis of vulvar lipoma. A vertical incision was performed to the skin tissue over the mass. After dissection of subcutaneous vulvar tissue, the mass with the color of white- pink was excised easily with the clear margin of the lesion and then the operation was ended.

After a histopathologic observation of the mass, it was reported that the mass was conformed with AMFB morphological observations. In the examination of the case, vimentin, destin, S-100 protein, SMA (smooth muscle actin), estrogen and progesteron receptor antibodies were used for immunohistochemical approach, while alcian blue dye was used for histochemical examination (Fig.1). There was no recurrence in the 6- month follow up.

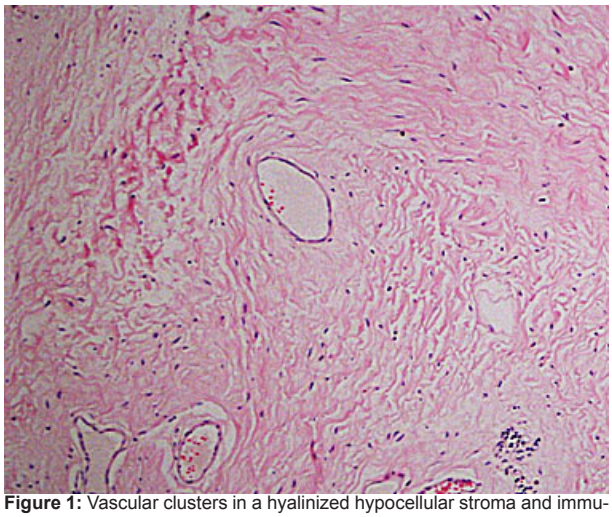

**Figure 1:** Vascular clusters in a hyalinized hypocellular stroma and immunoreactivity for vimentin were seen.

## DISCUSSION

Genital stromal tumors are primarily observed in vulvar region and they are seldom seen [1]. AMFBs could be mistaken with bartholin cysts or lower genital tract lipomas due to their locations, which then causes delays in diagnosis and treatment [5].

AMFB is generally seen women in premenopausal period within the age interval of 40-50 [2] and rarely seen early in life [4]. The 17-year-old female patient was the youngest patient having AMFB and 86-year-old patient was the oldest one that were evaluated by Sims et al. in a review containing 71 cases [6]. With our case, the patient's age range from 11 to 86 years.

In English medical research literature, there are approximately 65 cases about AMFB that had been published between 1997 and 2014. According to literature, malign transformation was only observed in 1 case among 65 cases [7].

In the macroscopic examinations of AMFB cases, it is observed that they are solid lesions, being well-demarcated, pink- colored and with the diameters ranging between 0.5-12 centimeters [8]. Vessel clusters can be seen in the hipocellular stroma in microscopic observation. It is considered that CD34 positive stem cells normally place around vessel clusters [9]. Nuclear atypia as well as an increase in mitosis are not commonly seen [8]. In the immunohistochemical analysis, most of the tumor cells that are tested, is strongly positive for desmin and vimentin. Beside this, the tumor cells reveal differentiated expressions of muscle actin and are often positive for estrogen and progesterone receptors [8].

Simple excision is considered as the main approach for AMFB treatment. AMFB is curable with simple surgical excision and no recurrence is observed in the follow-up [10, 11]. AMFB cases, as mentioned above, can be misdiagnosed as bartholin cysts, lower genital tract lipomas or aggressive angiomyxomas. Thus, unnecessary medical treatments are mistakenly given to the patients who have been pre- diagnosed as bartholin cyst or have outpatient care by the pre-diagnosed with lower genital tract lipoma. That is why, it is essential to mention about some strategies that are important for avoiding misdiagnosis of AMFB cases with bartholin cyst and lipomas. Basically, bartholin cyst is 1 to 4 centimeter in size, fluid-filled swelling that do not cause any problems. However, bartholin cysts being more than 4cm in size, have potential to cause pain or discomfort especially during walking, sitting or sexual intercourse [12].

When bartholin cysts are infected, patients usually complain about severe pain and pus-filled swelling, and among these, they maybe have fever and skin tissue hyperemi over the abscess formation [12].

Vulvar lipomas generally present as a single, slowly growing, nearly always benign, painless and mobile soft tissue swelling with a characteristic feeling as dough [13]. However, AMFB is solid, well-demarcated lesion and larger AMFB with the diameters of 12 cm may be painless [8]. In conclusion, AMFB is one of the circumscribed solid lesions of the external genitalia.

Since aggressive angiomyxoma, lower genital tract lipoma and bartholin cyst are mainly located in the vulva, they are generally misdiagnosed with AMFB.

That is why, it is crucial that gynecologists and pediatricians should know the symptoms of AMFB and be aware of the differential diagnosis of the three entitles mentioned before.

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### *Disclosure of Interests*

The authors declare that they have no competing interests.

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