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Large pedunculated angiomyofibroblastoma of the vulva with concomitant anemia: a case report and mini review of the literature

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Summary

Angiomyofibroblastoma is a rare myxoid tumor and is often seen in vulvar and vaginal area. Patients usually present with a mass on the vulva or labium majus at the premenopausal ages. The lesions are usually between 0.5 cm and ten cm. Lesions have quite cellularity with regular borders under the microscope. Lesions contain numerous veins and bulging stromal cells. Treatment of angiomyofibroblastoma is simple surgical resection. Angiomyofibroblastoma should be differentiated from the other vulvar neoplasms, which need radical surgical treatment, especially from the aggressive angiomyxoma. Hereby, the authors aimed to report a premenopausal women presented with vulvar mass and accompanying anemia. She was diagnosed with angiomyofibroblastoma.

Key words: Angiomyofibroblastoma; Large pedunculated; Vulva.

Introduction

Angiomyofibroblastoma is a rare tumor of the vulva and it was first described by Fletcher *et al.* in 1992. It is a benign mesenchymal tumor, that usually arises in the genital area, in particular in vulvar area of middle-aged premenopausal women [1]. It was initially conceived as a vulva specific neoplasm, but then was also shown to occur in the vagina, groin, rarely in the fallopian tube and rarely in the urethral area [8-10]. Angiomyofibroblastoma is rarely seen in men and occurs particularly in the scrotum, spermatic cord, and testicular tissue [11-14]. Its histopathology is not exactly clear, but the origin of the cells have been shown to be mesenchymal cells altered to the fibroblastic and myofibroblastic cells in the immunohistochemical analysis [15]. Benign solid tumors of the vulva are rarely seen. Hemangioma, lipoma, leiomyoma, myxoma, fibroma, and other pelvic masses showing myxoid changes should be considered in the differential diagnosis. Bartholin's cysts (seen frequently in the vulvar area), vaginal cysts, vulvar abscess, Gardner cyst, and Nuck duct cysts should also be kept in mind in the differential diagnosis. It is particularly important to distinguish aggressive angiomyxoma from angiomyofibroblastoma, because of its aggressive and invasive pattern. Angiomyofibroblastoma differs from aggressive angiomyxoma by regular borders, high cellularity, containing plenty of blood vessels, minimal stromal mucin, and rarely erythrocyte extravasation. Angiomyofibroblastomas are treated with local excisions. On the other hand, aggressive angiomyxomas tend to relapse and require deeper resections because of their infiltrative characteristic [16].

Case Report

A 44-year-old female patient was admitted to the present clinic because of a palpable vulvar mass, which do not produce any symptoms and existed for the last two years. The mass had grown rapidly in the recent months. Physical examination revealed a mobile, soft, right labium majus originated and sessile mass with regular borders. The mass was ten cm in diameter, and had a five-cm stalk. In addition, the patient had a type 2 female genital circumcision history. There were some lesions, which might possible due to friction, observed on the mass (Figure 1). Ultrasonography showed a well-circumscribed mass with internal vascular areas and heterogenous echogenicity. The mass was totally excised under general anesthesia. The cut surface of the mass was grey in color and homogenous (Figure 2). Vulvar anatomy became normal after surgical excision (Figure 3). Pathological examination of the lesion revealed a lesion containing thin-walled dilated blood vessels in a variable cellular connective tissue. Oval bulging stromal cells with eosinophilic cytoplasm were detected around the vessels of the stroma. Meanwhile, stuck mucosal glands or nerve fibers were not observed. There were also no mitosis and no multinucleated giant cells (Figure 4). As a result, she as diagnosed with angiomyofibroblastoma. She had also anemia. Laboratory tests revealed 8.3 mg/dl of hemoglobin level with low MCV level. There were hypochromic microcytic erythrocytes without any blasts on the peripheral smear.

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Figure 1. — Preoperative appearence of vulvar mass.

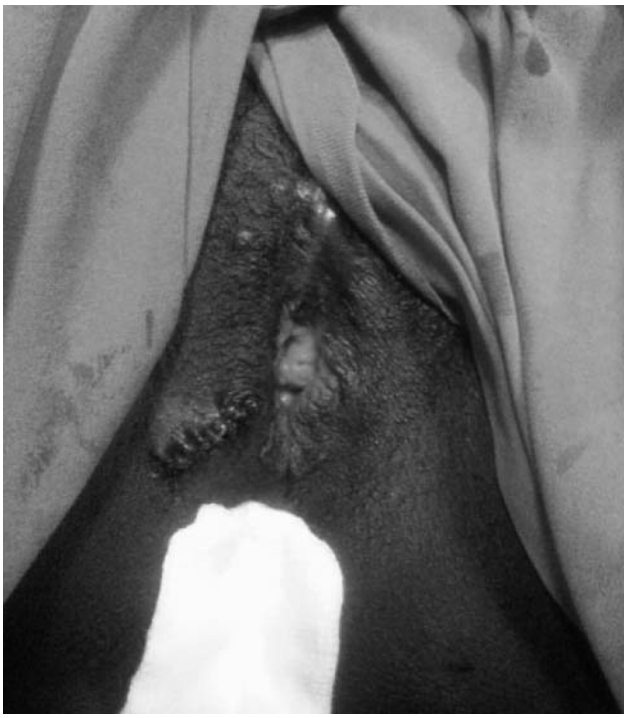

Figure 2. — Postoperative macroscopic appearence of mass.

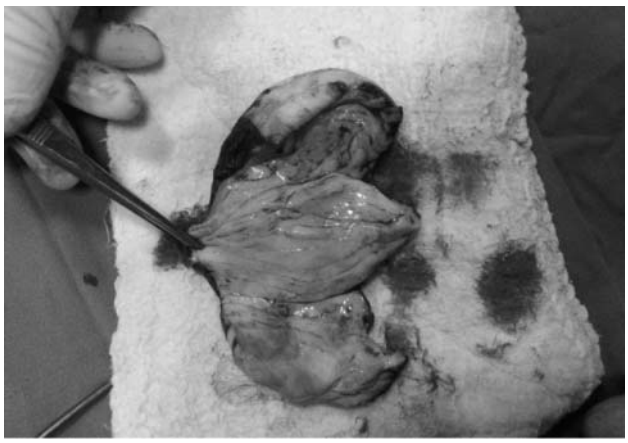

Figure 3. — Postoperative appearence in vulva.

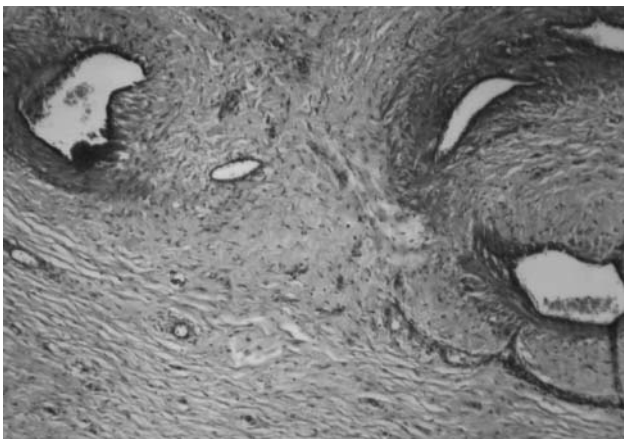

Figure 4. — Histopathological appearance (H&E x40).

Discussion

Benign solid tumors of the vulva are very rare. Other pelvic tumors like fibroma, lipoma, leiomyoma, hemangioma, and myxoma should be considered in the differential diagnosis. Fletcher *et al.* presented a ten case series of cellular variant angiomyxoma and named these cases as angiomyofibroblastoma [1]. Patients' ages ranged from 25 to 54 years (mean 36.3) [1]. The size of the vulvar masses in all patients ranged from 0.5 to 12 cm [1]. All had regular borders with myxoid homogeneous appearance under pathological examination [1]. Microscopic examinations revealed hypocellular hypercellular areas containing large number of thin-walled vessels without any interstitial hemorrhage [1]. All patients were treated with simple excision [1]. Many researchers have published studies with similar clinical and histological features of angiomyofibroblastoma after Fletcher *et al.* [2-7]. It is important to differentiate angiomyofibroblastoma from aggressive angiomyxoma because of different surgical approach and different prognosis. Angiomyofibroblastomas are generally smaller than five cm diameter, while aggressive angiomyxomas tend to be giant lesions. Angiomyofibroblastomas are lim-

ited in the superficial part of the vulva and have regular borders, but aggressive angiomyxomas invade into deeper tissues and therefore carry the risk of recurrence. Although mitosis are generally not seen in angiomyofibroblastoma, Takeshima *et al.* reported a mitotically active variant of angiomyofibroblastoma [3]. Nielsen *et al.* reported an angiomyofibroblastoma with sarcomatoid transformation and named that as angiomyofibrosarcoma [4].

Anemia is common in Sub-Saharan Tropical Africa. The major causes of anemia are iron deficiency anemia, vitamin deficiency anemia, sickle cell disease, and chronic disease anemia in this region. The authors do not think that anemia and angiomyofibroblastoma have a special relation and must be a coincidental situation.

In conclusion, angiomyofibroblastoma should be kept in mind in the differentiation of vulvar masses. Discrimination of angiomyofibroblastoma from aggressive angiomyxoma should be carefully done because of the need of aggressive surgical approach in the aggressive type.

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