CASE REPORT

Cellular Angiofibroma: Two Case Reports of a Rare Vulvar Tumor

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Introduction

Cellular Angiofibroma of the vulva is a rare benign mesenchymal tumor of middle-aged women, reported first in 1997 [1]. Although it has predilection for the vulvar region, few extravulvar cellular Angiofibromas have also been reported. [2, 3]. It has a limited potential for local recurrence and is usually treated with complete local excision. The differential diagnosis includes Aggressive Angiomyxoma, Spindle cell lipoma, Mammary-type Myofibroblastoma, Angiomyofibroblastoma, Fibrous tumors, Perineuroma, and Leiomyoma [1, 4–6].

We are reporting here two cases of cellular Angiofibroma, both arising from the labia major, treated by complete local excision and with no recurrence so far in the follow-up.

Case Report

Case No. 1

A 35-year-old female presented with a mass over the vulva for 3 years, which was progressively increasing in size for the past 1 year. It was painless. On examination, it was a globular pedunculated tumor of about 6 cm diameter arising from the right labia, and it was dark, smooth, soft, and compressible with no impulse on coughing, while ulcerated over the posterior surface. The rectal and vaginal examination was normal. The patient had no history of trauma, chronic infection, or insect bite. Her menstrual cycle was regular and she had 4 children, all delivered vaginally.

After routine investigations, the tumor was excised with the peduncle and sent for histopathology. The surgical specimen was a soft greyish-brown mass measuring $8 \times 6 \times 4$ cm and was flat, well delineated with a disorganized fascicular pattern on section. Slides were stained with hematoxylin-eosin. On microscopy, the lesion was well circumscribed, but not truly encapsulated. There was abundant small to moderate size blood vessel proliferation in the midst of the fibrous stroma having fusiform cells arranged in fascicles. There were areas of mitoses, myxomatous degeneration, and lymphocytic infiltration, but no area of necrosis or hemorrhage was seen. Immunohistochemistry was positive for vimentin and negative for actin.

The post-op period was uneventful and there was no recurrence in the follow-up till 32 months (Fig. 1).

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Fig. 1 Angiofibroma arising from the Right Labia major in Case 1

Case No. 2

A 57-year-old female, mother of six children, presented with a gradually increasing pedunculated mass of about 6 cm arising from left labia for the last 2 years. It was firm in consistency and was similarly ulcerated and excoriated over its posterior wall. The patient had no complaint except discomfort and apprehension. She had attained her menopause 10 years ago. Her physical examination and routine investigations were normal.

Local complete excision of the mass was done. The specimen was of a cricket ball size, dark brown in color, measuring 6 cm × 6 cm × 5 cm in all, and with a peduncle of about 2 cm. The surface was retractile on the section, and had an irregular fascicular pattern. Fixation and staining were done with hematoxylin-eosin. On microscopy, it was well circumscribed, and similarly composed of uniform fusiform spindle cells and numerous thick-walled hyalinized vessels along with mature adipocytes, haphazardly present throughout the fibrous stroma. Some mitotic activity was seen, but there was no cellular atypia or area of necrosis or hemorrhage. Immunohistochemistry was positive for vimentin.

There was no recurrence over 24 months of follow-up (Fig. 2).

Discussion

Cellular Angiofibroma is a benign mesenchymal lesion with a predilection for the vulva, presenting as a painless mass [1], although some reports of extravulvar cellular Angiofibroma have also been described. A very similar lesion has been reported in subcutaneous tissue of the chest in a 43-year-old lady. [2] Its variant, Giant-cell Angiofibroma, is a rare mesenchymal neoplasm which most commonly arises in the soft tissue near the orbit and in the

Fig. 2 Pedunculated Angiofibroma arising from the Left Labia major in Case 2

Fig. 3 Abundant small to moderate size blood vessel proliferation in the midst of the fibrous stroma (Case 1)

nasolacrimal-duct. A case of extraorbital Giant-cell Angiofibroma arising in the inguinal region has also been reported [3].

It is chiefly a tumor of middle-aged women [1], as in our two cases. Lane & Walker have reported a case in a postmenopausal 77-year-old woman who was on estrogen replacement therapy [5]. Laskin et al. [7] described 11 very similar cases in the male genital tract. Recently, Flucke et al. [6] reported 25 such cases of genital and extragenital cellular angiofibroma, out of which 17 were in females and the vulvar region was the most common site of origin (Fig. 3).

Histologically, cellular Angiofibroma has two principle components—fusiform cells and prominent blood vessels. Fusiform cells form small fascicles in the middle of collagen bundles, frequent blood vessels of small to medium caliber sometimes with a hyalinized wall, and a variable component of mature adipocytes. The tumor may present few mitotic activities and atypical cells in the stroma, but necrosis remains absent [4, 8], although few instances of sarcomatous transformation and nuclear pleomorphism have also been reported [6, 9, 10].

In the Immunohistochemical studies reported so far, the tumor has been characteristically and constantly found positive for vimentin, which suggests that the tumor cells are of fibroblastic origin, while negative for desmin and actin proteins [1, 4]. CD34 has been seen in 50 % of such cases [6].

These tumors are almost always positive with estrogen and progesterone receptors, suggesting they probably arise from the hormone receptor positive subepithelial mesenchymal layer within the lower female genital tract [5, 10].

Flucke et al. [6] tested the genetic relationship of these tumors by means of FISH analysis in 7 of their reported 25 cases, which exhibited monoallelic deletion of RB1 gene in all seven cases suggesting that cellular angiofibroma, mammary-type myofibroblastoma, and spindle cell lipoma form a spectrum of a single entity.

Cellular Angiofibroma should be differentiated from other aggressive soft tumors of the vulva with fibroblastic and myofibroblastic differentiation such as Aggressive Angiomyxoma and Angiomyofibroblastoma [4, 5]. Immunohistochemistry and FISH analysis could be used in such conditions.

Cellular Angiofibroma of the vulva appears benign since there are no reports of tumor metastasis, although the follow-up period in most of the studies is short [1, 7].

Also, local excision with a free margin is the appropriate treatment as so far, there are no reports of recurrence [1, 7].

Conflict of interest None.

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