Cellular angiofibroma of the vulva: Report of a case

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Summary

A case of vulvar cellular angiofibroma in a 50-year-old woman was immunohistochemically examined. She presented with a right labial mass which was noticed four years before. Surgical excision of the mass was performed. Histopathological examination revealed typical characteristics of cellular angiofibroma. Immunohistochemically, the lesion was CD34 positive but non-reactive for desmin, smooth muscle actin and S-100 protein. In this article, we present a case of vulvar cellular angiofibroma, a lesion that should be considered in the differential diagnosis of vulvar soft tissue tumors.

Key words: Vulva; Cellular angiofibroma; Immunohistochemical staining; Aggressive angiomyxoma.

Introduction

Cellular angiofibroma of the vulva is a recently described benign mesenchymal tumor [1]. It occurs mostly in the vulvar region of middle-aged women; however a case of scrotal cellular angiofibroma has been reported [2]. The differential diagnosis is of specific importance as it could easily be confused with more aggressive lesions such as aggressive angiomyxoma. Patients with cellular angiofibroma are mostly middle-aged premenauposal woman. Lane *et al.* [3] described a 77-year-old woman with vulvar cellular angiofibroma.

We present a case of vulvar cellular angiofibroma that occurred in a 50-year-old woman who had had tamoxifen therapy for breast cancer for two years.

Case

A 50-year-old woman presented to our outpatient clinic with the complaint of a right labial mass. The mass was first noticed four years before and had not significantly changed in size. There was no pain but just a little feeling of itching. She was hospitalised with the initial diagnosis of a "right labial mass".

The patient was gravida 2, para 2, and having irregular menses for the previous two years. She had been an active smoker for 25 years. On her medical history, she had left mastectomy and lymph node dissection diagnosed as infiltrative ductal carcinoma in 1993. She had radiotherapy after the surgical intervention and she was on tamoxifen therapy for two years, between 1993 and 1995.

Her gynecological examination revealed a right sized labial mass which was mobile, rubbery and approximately 7 x 6 x 5 cm in size. Her inner and outer genitalia were otherwise normal. Her hematocrit level was 32.2% and erythrocyte sedimentation rate was 36 mm/hour. The mass was removed by local surgical excision. The mass extended deep under the right labia major up to the level of the right pubic arc.

On macroscopic examination the mass was solid and well-circumscribed by a thin capsule. The capsule was well-vascularised and yellowish-gray in color. It did not show any sign of infiltration to the surrounding tissues. The cut surface of the mass was yellowish-tan colored and had a lipoid appearance. There was no focus of necrosis or hemorhage.

Microscopically, the specimen was well-circumscribed and did not include the surrounding tissues. The tumor was incapsulated 1 mm in thickness in some areas. The tumor was highly cellular with thick-walled hyalinized vessels, loose myxoid tissue and in some areas a minor component of adipose tissue. The majority of the cells were spindle shaped, pale eosinophilic cytoplasmic cells with fusiform nuclei. No tumor necrosis or significant pleomorphism was seen. Immunohistochemically the tumor cells were reactive for CD34 but did not demonstrate immunoreactivity for smooth muscle actin, desmin or S-100 protein.

Discussion

A spectrum of vulvovaginal soft tissue tumors ranging from fibroepithelial stromal polyps to aggressive angiomyxoma has been described [4]. The accurate clinical diagnosis is of specific importance as these tumors differ in biological behavior but share the same phenotype. The vulvovaginal mesenchyme is their common origin. Accurate diagnosis for the proper treatment is necessary and can sometimes be made by routine hematoxylin and eosin slides as these lesions may exhibit similar immunohistochemical and ultrastructral properties.

Cellular angiofibroma is a clinically benign neoplasm. As its name implies it is cellular and well vascularised. It occurs as a solid, discrete mass mostly seen in premenopousal women. There may be some mitotic figures but the stromal cells are uniform and lack nuclear atypia. The vascular walls are thick and sometimes hyalinized. The stromal cells of cellular angiofibroma do not express markers for neural or muscle differantiation.

Since the first reported case of cellular angiofibroma in 1997, not many cases have been published [1]. There is no information about the long-term follow-up of these cases so the effectiveness of the only treatment choice –

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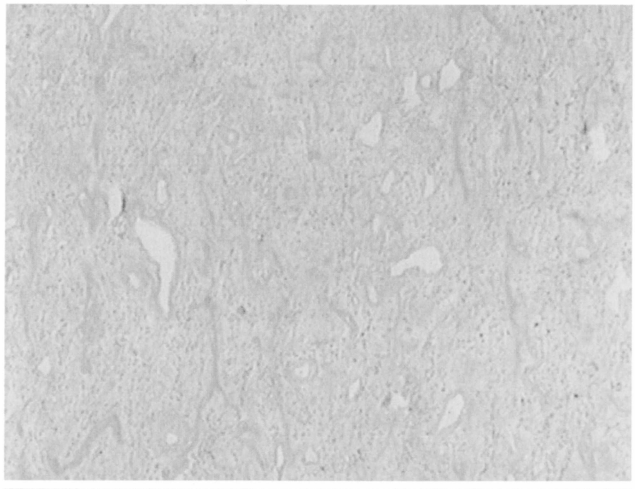

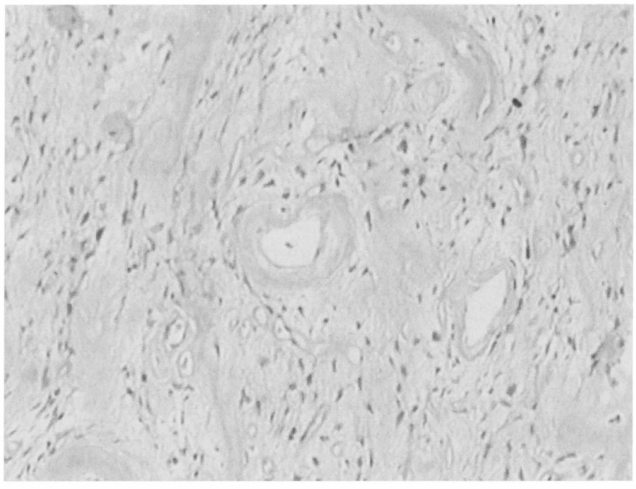

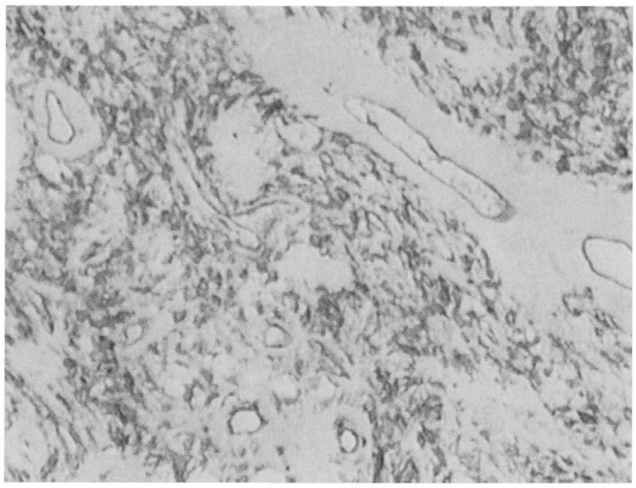

Figure 1. — Low-power view of cellular angiofibroma. Thick-walled hyalinized vessels are surrounded by loose myxoid tissue (Hematoxylin and eosin, x 25).
Figure 2. — Hyalinized vessels and spindle-shaped tumor cells with pale eosinophilic cytoplasm (Hematoxylin and eosin, x 100).
Figure 3. — CD34 positivity of the tumor cells (anti-CD34, x 100).

local excision – has not been confirmed. These tumors are known as non-recurrent or metastasizing.

The differantial diagnosis of cellular angiofibroma includes aggressive angiomyxoma, angiomyofibroblastoma, smooth muscle tumors and spindle cell lipoma [4].

Aggressive angiomyxoma is another type of vulvar soft tissue tumor. It is found in women of older age. It is a non-metastasing tumor that has potential for local recurrence. The lesion is well-circumscribed, deeply located, more infiltrative and less cellular. It has an abundant myxoid matrix. The cells are also spindle shaped and more unevenly distributed. The lesion has prominent capillary sized vessels. Immunohistochemically, it is desmin and sometimes actin positive. The lesion was cytogenetically studied and a translocation involving chromosome 12 was found. This translocation was found to be associated with HMGIC protein whose aberrant expression has been seen in many mesenchymal neoplasms including endometrial polyps, uterine leiomyomas and pulmonary hematomas.

Angiomyofibroblastoma is a tumor that almost exclusively occurs in the vulvovaginal region of women. It can also be seen in the scrotal region of men. As it is well-circumscribed, angiomyofibroblastoma can be easily confused with a Bartholin's gland cyst. It is usually less than 5 cm in size. Local excision is the treatment of choice and it is non-recurring. A sarcomatous transformation has also been reported. Histologically, the lesion has hyper- and hypocellular areas. It has an edematous collagenous matrix and the tumor cells have an epitheloid

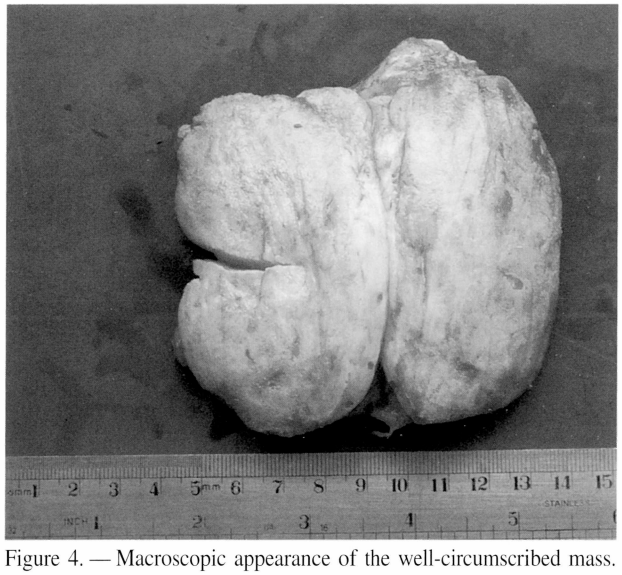

Figure 4. — Macroscopic appearance of the well-circumscribed mass.

appearance. Mitoses are not prominent as in cellular angiofibroma.

Spindle cell lipoma and cellular angiofibroma have similar histological features. In spindle cell lipoma there are floret-like multinucleated giant cells and lipoblasts. They are less circumscribed and non capsulated. The vessels in cellular angiofibroma are hyalinized and thick-walled.

Vulvar smooth muscle tumors must be distinguished from cellular angiofibroma. They occur most commonly in the fourth decades. The lesions are usually well circumscribed and have three histological patterns: spindled, epitheloid and myxohyaline. They have fewer mitoses than cellular angiofibroma and if they do, it may be a sign of malignancy. They are usually less than 50 mm in size.

Various immunohistochemical stainings have been performed to differentiate vulvar soft tissue tumors. Cellular angifibromas are vimentine positive and sometimes express reactivity for CD34. They are negative for desmin, actin, keratin, epithelial membrane antigen and S 100 protein. As the majority of the patients with cellular angiofibroma are premenopausal, a hormonal influence could be suggested.

References

[1] Nucci M.R., Granter S.R., Fletcher C.D.M.: "Cellular angiofibroma: a benign neoplasm distinct from angiomyofibroblastoma and spindle cell lipoma". *Am. J. Surg. Pathol.,* 1997, *21*, 636.

[2] Laskin W.B., Fetsch J.F., Mostofi F.K.: "Angiomyofibroblastoma-like tumor of the male genital tract. Analysis of 11 cases with comparison to female angiomyofibroblastoma and spindle cell lipoma". *Am. J. Surg. Pathol.,* 1998, *22*, 6.

[3] Lane J.E., Walker A.N., Mullis E.N., Etheridge J.G.: "Cellular angiofibroma of the vulva: Case report". *Gynecol. Oncol.,* 2001, *81,* 326.

[4] Nucci M.R., Fletcher C.D.M.: "Vulvovaginal soft tissue tumors: update and review". *Histopathology,* 2000, *36,* 97.

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