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Case Study

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VULVAR ANGIOFIBROMA: CLINICAL AND HISTOLOGICAL ASPECTS OF THIS RARE AND STILL LITTLE-KNOWN ENTITY

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ABSTRACT

Cellular angiofibroma is a rare benign mesenchymal lesion with a predilection for the genital region, although rare cases have been reported in the pelvic and extrapelvic regions. It mostly affects women in their fifties. Histologically, Cellular angiofibroma is characterized by two principal components: bland spindle cells and abundant prominent small to medium-sized vessels. The characterization by immunohistochemistry is helpful in distinguishing cellular angiofibroma from other mesenchymal lesions. The treatment requires a simple local excision due to low recurrences of the local lesion and no chances of metastasizing. The current study concerns a 52-year-old patient who had two angiofibromas for one year, one on the mons pubis and the other on the right labia majora. The treatment consisted of a simple excision, and the follow-up was unremarkable.

KEYWORDS: Cellular angiofibroma - Vulva - Mesenchymal tumors – Histopathology – Immunohistochemistry.

INTRODUCTION

Cellular angiofibroma belongs to soft tissue tumors that mainly affects the distal genital tract of both genders: it affects the vulvo-vaginal region in women and the inguino-scrotal area in men[1,2], although extragenital localizations have also been described.[3–4]

Among the mesenchymal tumors involving the vulvovaginal region, we distinguish lesions that can occur at any site, and those more specific of this area.

The first group includes leiomyoma, lipoma and solitary fibrous tumor, while the second group includes cellular angiofibroma as well as aggressive angiomyxoma, angiomyofibroblastoma and fibroepithelial stromal polyp.[5,6]

CASE REPORT

We report the case of a 52years old woman, in perimenopause, presenting with two asymptomatic vulvar nodules for one year. The two lesions had progressively enlarged in size over the last few months. There was no history of pain or bleeding.

The patient had no particular medical history, no history of sexually transmitted diseases or genital warts, nor did she have a family history of similar lesions or gynecological cancer. She also did not report taking hormone therapy, smoking or drinking alcohol.

A physical examination revealed two similar masses **(Figure1)**, one on the mons pubis (4 cm) **(Figure 2)**, and the other on the right labia majora (3 cm) **(Figure 3)**,. The two masses are well circumscribed, solid, mobile and painless. The rest of the gynecological examination was normal. No palpable inguinal lymph nodes were determined.

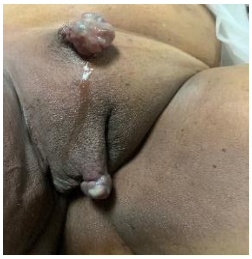

Figure 1: Vulvar lesions

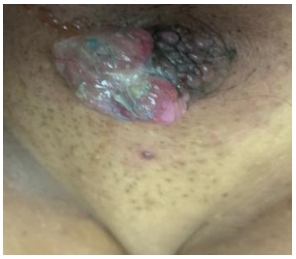

Figure 2: Lesion on the mons pubis

Figure 3: Lesion on the right labia

An ultrasound showed two superficial, solid, well-demarcated soft tissue tumors.

The hematological and biochemical parameters were normal. The cervicovaginal smear was also normal. A biopsy of the lesion located at the mons pubis was performed, and the results of the histological examination were consistent with the diagnosis of cellular angiofibroma. The immunolabeling objectified positive anti CD 31 and CD 34. The anti HHV8 and anti-cytokeratin were negative.

We proceeded to the simple excision **(Figure 4)** of the two vulvar masses, without intraoperative or postoperative complications.

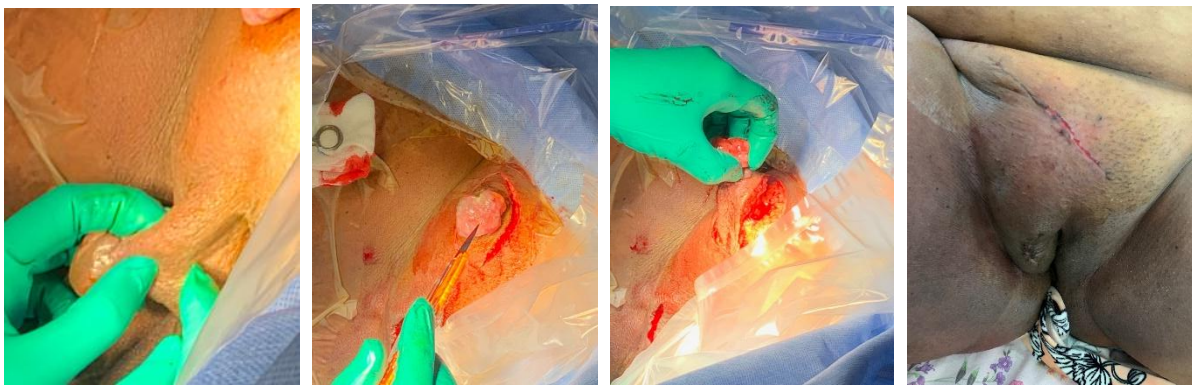

Figure 4: Excision chirurgicale des 2 tumeurs.

Macroscopic examination showed well circumscribed, solid, whitish and shiny tumors, measuring 4 and 3 cm in long axis. Under the microscope, the tumors were composed of uniform, short, spindle-shaped cells, proliferating in an edematous to fibrous stroma and numerous small to medium-sized thick-walled vessels, thus confirming the diagnosis of cellular angiofibromas.

At six months after surgery, the patient is doing well with no sign of recurrence.

Informed consent was obtained prior to reporting the case

DISCUSSION

Cellular angiofibroma is an uncommon benign mesenchymal lesion, first described by Nucci et al. in 1997 in a series of 6 cases that occurred almost exclusively in the vulva of middle-aged women.[1] In 1998, Laskin et al. described 11 cases of a histologically similar lesion which affects the adult men in the inguino-scrotal area.[2]

The World Health Organization classification has grouped this type of lesion in both females and males, under the term “cellular angiofibroma”, since there are no reproducible morphological differences between the two sexes.[7]

Women are affected most often in the fifth decade, earlier than men, mainly affected in the seventh decade[3], although a case of vulvar angiofibroma has been described in a 20-year-old patient.[8]

Etiopathologically, cellular angiofibroma pathogenesis is still unclear. The fact that cellular angiofibroma is mainly present in the menopausal and postmenopausal periods suggests that pathogenic development may be hormonally influenced. some authors suggested that these lesions are stem cell–derived, with a capacity for adipose and myofibroblastic differentiation in accordance with the influence of hormones, microenvironments, cytokines and growth factors.[9]

Cellular angiofibroma most often occurs in the vulvovaginal region, although other sites, pelvic and extra-pelvic have been reported: pelvis, perineum, left hip, knee, armpit, breast, hypochondriumlocation of the paravesical region.[3,4,5,10,.11,12]

Clinically, vulvar angiofibroma usually occurs as a small (usually less than 4 cm) with well-circumscribed margins AND slowly growing lesion. IT tends to gradually increase in size after about 1-2 asymptomatic years, inducing women to seek medical attention long after the tumor onset. the largest cellular angiofibroma described in the literature measures 20 cm.[13]

Cellular angiofibroma is often confused with a Bartholin's gland, vulvar cyst, leiomyoma, lipoma.[5]

Histologically, cellular angiofibroma is usually well circumscribed, consisting of two main components: bland spindle cells and relatively abundant small to medium-sized vessels with mural hyalinization.[3] however, wide morphological variations with atypia, foci of lipogenic differentiation and sarcomatous transformation have been described but these variations did not seem to predispose to a malignant mode.[3,14,15]

There are many mesenchymal tumors that enter into the differential diagnosis with cellular angiofibroma, such as spindle cell lipoma, solitary fibrous tumor, breast-like

myofibroblastoma, angiomyfibroblastoma, aggressive angiomyxoma, and smooth muscle tumor. These tumors may have overlapping morphological, immunohistochemical, and cytogenetic features, and differential diagnosis is therefore mandatory.[16,17]

Cellular angiofibroma appears to behave in a benign manner, based on the limited clinical follow-up available.[3,14,18,19] No metastasis has been reported in the literature and recurrence has only been reported in one case.[11]

The treatment of choice is therefore surgical and consists of a simple local excision with negative margins.[11,14]

CONCLUSION

To date, cellular angiofibroma still remains a poorly known lesion that needs further investigations to closely define its clinical and pathological features.

In women, it represents a distinct benign tumor with a wide anatomical distribution, with a predominance in the vulvovaginal region.

Although it is a benign tumor and shows different phenotypic characteristics, simple local excision is adequate, because it demonstrates no recurrence or metastasis.

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