CASE REPORT

Angiokeratoma of the vulva

Angioceratoma da vulva

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Abstract: Angiokeratomas are benign tumors characterized by ectasia of blood vessels in the papillary dermis associated with acanthosis and hyperkeratosis of the epidermis. Dermatological examination of angiokeratomas of Fordyce is characterized by papular keratotic lesions of erythematous-violet color. They are more common in the scrotum, and vulvar involvement is rarely reported. Histopathology is particularly important to distinguish them from other benign and malignant tumors. The article reports the case of a middle-aged black woman with a history of chronic constipation, varicose veins of the lower limbs and cesarean section performed 20 years ago who had had multiple vulvar angiokeratomas for three months.
Keywords: Blood vessels; Hemangioma; Skin neoplasms

Resumo: Os angioceratomas são tumores benignos caracterizados por ectasia dos vasos sanguíneos da derme papilar associada à acantose e hiperceratose da epiderme. Ao exame dermatológico, os angioceratomas de Fordyce caracterizam-se por lesões papulo-ceratóticas de coloração eritemato-violácea. São mais comuns no escroto, sendo raramente relatada a localização vulvar. A histopatologia é particularmente importante para diferenciá-los de outros tumores benignos e malignos. O artigo relata o caso de uma mulher negra de meia idade com história de obstipação intestinal, varizes de membros inferiores e cesárea há 20 anos que apresentava múltiplos angioceratomas vulvares há três meses.
Palavras-chave: Hemangioma; Neoplasias cutâneas; Vasos sanguíneos

INTRODUCTION

Angiokeratomas are benign tumors characterized by numerous dilated vessels in the superficial dermis with epidermal hyperplasia and hyperkeratosis. They are classified as [1] disseminated - angiokeratoma corporis diffusum (Fabry disease) - and localized, which includes angiokeratoma of the scrotum, penis and vulva (angiokeratoma of Fordyce), circumscribed angiokeratoma and angiokeratoma of Mibelli. Angiokeratomas [2,3,4] of the scrotum are common and their incidence increases with age. Incidence in the vulva is rare; at this site the lesion is easily mistaken for other benign and malignant conditions such as condyloma, verruca vulgaris, melanoma, and pyogenic granuloma. [3,5,6] This article reports the case of a middle-aged black woman with multiple vulvar angiokeratomas, history of previous gynecological surgeries and varicose veins of the lower limbs.

CASE REPORT

Black woman, 46 years old, had been complaining of papular vulvar lesions occasionally and slightly pruritic, which bled upon friction with clothing, for three months.

She complained of chronic constipation and sporadic abdominal colic pain. She denied having hemorrhoidal varices. She reported history of three pregnancies, the last 20 years ago, with two births; the first, by vaginal delivery and the second, by cesarean section, and an abortion. She also referred conization of the cervix by lesion associated with HPV five years ago.

Clinical examination revealed papular angiomatous lesions in the labia majora and labia minora, some with hyperkeratosis on the surface (Figure 1). She was not overweight, but microvarices were seen in her lower limbs. The largest vulvar

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lesion, on the left, measured 0.4 cm in diameter and was removed by shaving. The smallest lesions were treated with trichloroacetic acid 70%, with partial involution. After six months, there were no signs of recurrence of the excised lesion; the others had reduced in size and there was no more bleeding.

Anatomopathological examination of the lesion revealed mild hyperkeratosis and epidermal hyperplasia, involving highly dilated blood vessels and, sometimes, with organized thrombus in the lumen (Figure 2).

DISCUSSION

Clinically, angiokeratomas of the vulva present as keratotic papules of slow evolution whose color varies between red, reddish-blue and brown. They measure less than 10 mm, usually between 2 to 5 mm. The lesions are often unilateral, multiple, and asymptomatic, although pruritus, pain, burning and bleeding are reported. [3,5-7] The majority of patients affected are between 20 and 40 years of age. [3,5] Clinical findings in this patient are in agreement with what has been described, but her lesions were recent.

The etiopathogenesis is related to increased local venous pressure and consequent subepithelial vascular dilation. The labium majus is embryologically similar to the scrotum, since both derive from the labioscrotal folds. [3] Thus, the mechanisms attributed to the occurrence of angiokeratomas of the scrotum can be extrapolated to angiokeratomas of the vulva. The loss of support of the local vessels, secondary to congenital deficiency of elastic tissue, is one of the associated factors. Interestingly, racial or genetic predisposition is not described. The role of increased local venous pressure caused by venous malformations, varicocele or thrombosis, in addition to phlebectasia secondary to chronic inflammation, is also postulated . Dilation would therefore be induced by stasis, retrograde pressure or venous injury. [5,7] Although venous drainage of the scrotum and labia majora is different, both contain elastic tissue around the vessels, smooth muscle fibers and venous structures vulnerable to inflammation and obstruction, with consequent increase in local venous pressure. [5]

Therefore, excess weight, increased parity, hemorrhoids, pelvic inflammatory disease, prior hysterectomy, varicose veins, and vulvar varicosity are considered risk factors for the occurrence of angiokeratomas of the vulva. [3,5,7] The patient described in the present work showed as identifiable risk factors the presence of varicose veins of the lower limbs and constipation (even though she did not report hemorrhoids) as well as history of cesarean section and gynecological surgery (conization).

Dermoscopy can be a valuable tool for diagnosis, as documented by Zaballos et al., 2007, in a multicenter study in eight hospitals in five different countries. This study evaluated 32 patients with solitary angiokeratomas and the dermoscopic findings were compared with those obtained of other skin dermatoses. [8]

Biopsy of the lesion may be essential to confirm the diagnosis because it may clinically resemble melanoma, basal cell carcinoma, vulvar intraepithelial neoplasia (spinocellular carcinoma), other angiomas,

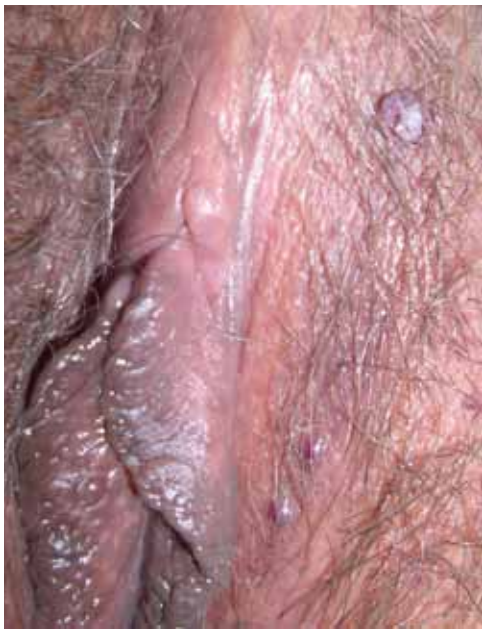

Figure 1: Fordyce angiokeratomas: papular-angiomatous lesions on the left labium majus. Note hyperkeratosis on the larger lesion

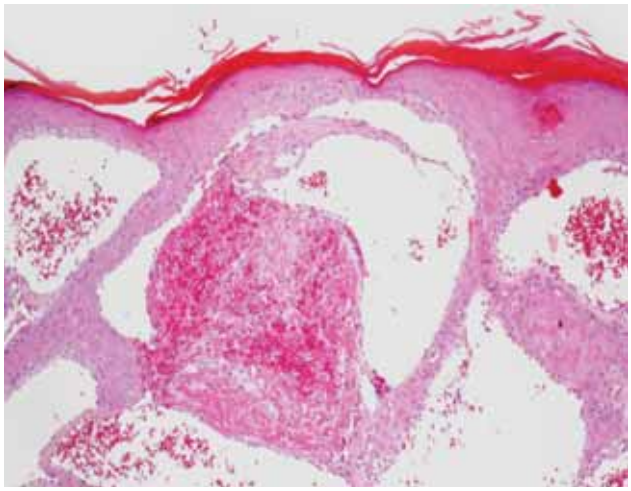

Figure 2: Mild hyperkeratosis and hyperplastic epidermis, involving severely dilated blood vessels, sometimes with thrombi in organization in the lumen

including pyogenic granuloma, lymphangioma, nevi, condyloma latum, condyloma acuminata, nodular prurigo and seborrheic keratosis. [5,6] As noted in this case, microscopic findings include hyperkeratosis, papillomatosis and dilation of the capillaries of the papillary dermis, without significant endothelial proliferation. Dermal inflammation is minimal. Occasionally, lymphangiectasia may occur. [1,2,5,7]

Treatment can be done through surgical excision or physical and chemical cauterization. It can also be more conservative, if the complaint is minimal, as in the case of this patient. [6]

In conclusion, although angiokeratomas are rarely reported in the literature, they can be mistaken for malignant tumors, especially melanoma, when located in the vulva, and be a cutaneous marker for increased local venous pressure, which may require investigation of pelvic diseases. ❑

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