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Angiokeratoma of the Clitoris: Case Report.

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Abstract

Angiokeratoma of the clitoris is an extremely rare lesion of the vulva caused by the dilatation of the blood vessels of the papillary dermis. We present a case of a 78-year-old patient with an angiokeratoma of the clitoris, which to our knowledge and reviewed of the literature represent the fist case described in America. It is of great importance to know this lesion to avoid misdiagnosis and to determine the real incidence of this pathology and help clinicians understand the disease and avoid biopsies and unnecessary management.

Keywords: Angiokeratoma, vascular lesion, vulvar lesions, clitoris lesions

1. Introduction

Angiokeratomas are uncommon benign vascular lesions. It is more frequently diagnosed in males than females, the actual incidence of vulvar angiokeratomas is unknown, with less than 50 cases reported by 2002. Angiokeratoma of the clitoris is extremely rare, only 2 cases have been reported in adults and 1 in a 14-year-old girl. Angiokeratomas were first described as localized lesions in the scrotum in 1896 by American dermatologist John Fordyce. These tumors are due to the dilation of the blood vessels of the papillary dermis, accompanied by a hyperkeratotic epidermis, angiokeratomas are usually asymptomatic although in some cases they can cause intermittent bleeding, pruritus, and pain. Angiokeratomas are characterized histologically by dilated ectatic, blood-filled vascular spaces in the papillary dermis associated with acanthosis, hyperkeratosis, and papillomatosis. Incidence of these lesions in the vulva is extremely rare and is easily mistaken for other benign and malignant conditions such as condyloma, melanoma, and pyogenic granuloma. This article reports a case of a 78-year-old patient with angiokeratoma of the clitoris. To our knowledge, this is the first case of angiokeratoma of the clitoris in Latin America.

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Case Description

A 78-year-old multipara woman, with history of hysterectomy and medical record of Diabetes Mellitus type 2 and Hepatopathy Child A, was admitted to our service referring stress urinary incontinence without accompanying symptoms. On physical examination of the vulva an 8 mm painless, black and soft tumor was observed, involving all the clitoris and part of the right labia minora (Fig 1), cystocele grade I and rectocele grade II were also observed with no evidence of urinary incontinence when performing a Valsalva maneuver. A biopsy of the lesion was taken and sent for histopathological study. The anatomopathological study reported two biopsies, the largest of 0.3 cm, with irregular edges and shapes, dark brown color, soft consistency, reporting angiokeratoma of the clitoris (Figure 2).

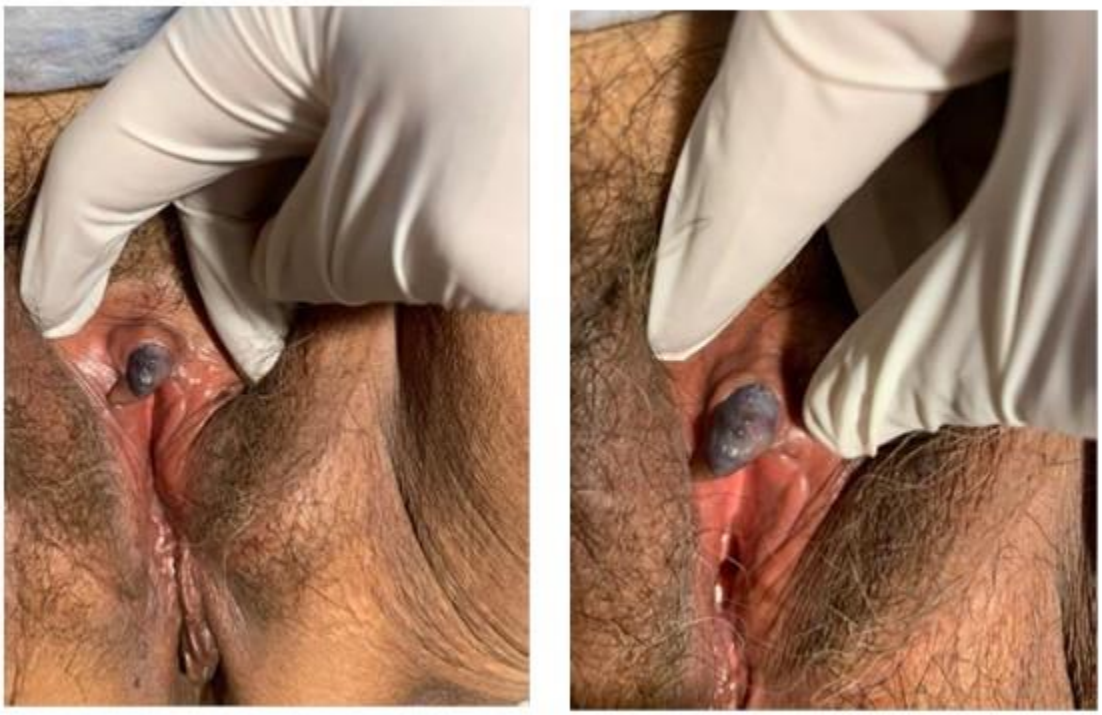

Fig. 1 An 8mm soft tumor located in the clitoris.

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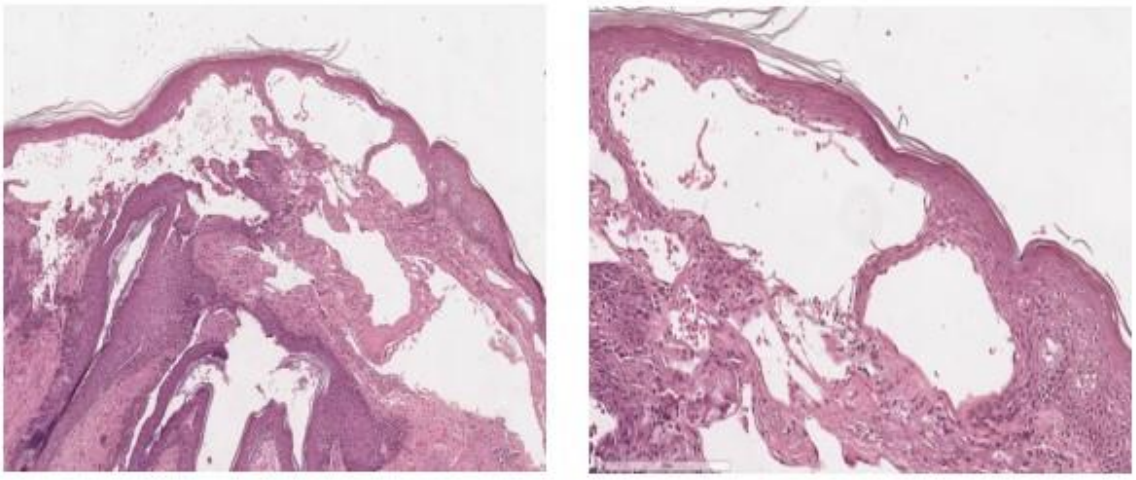

Fig. 2 Dilatation and ectasia of superficial dermal blood vessels partially surrounded by elongated rete ridges.

Discussion

Angiokeratoma of the vulva is an uncommon and rare benign vascular lesion and is characterized by ectasia of blood vessels in the papillary dermis associated with acanthosis and hyperkeratosis.

Vulvar lesions are commonly associated with hemorrhoids, varicose veins, patients with history of hysterectomy and salpingectomy, oral contraceptive use, malignant ovarian tumors and pregnancy due the proliferation of capillaries attributed to an increase in venous pressure and elevated serum levels of progesterone. In our case, the patient presented some of these risk factors such as multiparity with eleven pregnancies, history of hysterectomy and a liver disease.

Its more frequently presented in 20- to 40-year-old women during the reproductive age, our 78 years-old patient is the first case reported in this age group; a physical examination reveals a unilateral painless red to bluish papule and is usually asymptomatic although intermittent bleeding, pain and pruritus may occur.

Due the wide range of similar pathological features, the initial diagnosis is often inaccurate. Inguinal and pyogenic granuloma, condylomas, melanoma and fungal infection are among the differential diagnosis.

There are approximately 50 cases of vulvar angiokeratoma reported in the literature and only 3 clitoral angiokeratomas have been described: two cases in Japan and one in Turkey. We are presenting the first case of angiokeratoma of the clitoris in America.

Treatment usually consists of expectant following for asymptomatic patients. Surgical excision, cryotherapy or electrodesiccation should be performed in cases where bleeding or pain are present. In all cases, biopsy of the lesions should be performed to rule out malignancy and avoid

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misdiagnosis. In our case, two incisional biopsies were performed due to a suspected diagnosis of melanoma.

It is of great importance to know this lesion to avoid misdiagnosis and to determine the real incidence of this pathology and help clinicians understand the disease and avoid biopsies and unnecessary management.

Conflict Of Interest

The authors declare no conflict of interest.

Consent

Written informed consent was obtained from the patient for the publication of this case report.

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