Case Report

A case report on vulval angiokeratoma – rare entity at a rare site

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Angiokeratoma is a benign disease which belong to the group of vascular ectasia, which is the term given to dilated blood vessels. These benign tumors are seen as ectatic blood vessels in papillary dermis along with overlying hyperplasia. Here, we are reporting a case of 50 year old female who presented in dermatology OPD with a polypoidal swelling. Angiokeratoma of vulva is relatively rare finding and only limited cases are reported. The case is being reported due to its rarity.

KEY WORDS: Angiokeratoma, Vascular Ectasia, warts, condylomas

INTRODUCTION

Angiokeratoma is a benign vascular disease, characterized by superficial vascular ectasia and overlying epidermal hyperplasia. The prevalence of angiokeratoma is estimated to be 0.16% among general population.[1] It is comparatively uncommon for angiokeratroma of Fordyce to develop across the vulva. Clinically, it can be separated into variants depending on sites that are widespread and limited. Localized variants include solitary angiokeratoma, angiokeratoma of Fordyce, angiokeratoma circumscriptum naeviform, and angiokeratoma of mibelli. Widespread forms are typically linked to congenital errors of metabolism.

In the present article, we report a case of angiokeratoma of Fordyce over vulva.

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CASE REPORT

A 50-year-old otherwise asymptomatic female presented with a single, asymptomatic, soft, and greyish-purple papule of size 0.3 cm and was located over the labia majora. The patient complaint of intermittent pruritis and a sizable psychological strain. Other than that, the patient's family and personal medical history were normal. There was no history of surgery or injury over vulva. Palpation revealed non-tender and soft to firm papule. Both the colposcopic and per vaginal examinations of the vagina were normal. Perianal examination revealed no abnormalities. Clinically diagnosis of angiokeratoma was suspected and excisional biopsy under aseptic condition with local anesthesia was done. The excised papule was sent for histopathological examination. Histopathological investigation revealed dilated blood-filled veins in papillary dermis with overlying hyperplastic and hyperkeratotic epidermis thus a diagnosis of Angiokeratoma was given. There were no side effects or complications from the surgery. No relapses occurred during the 1-year follow-up period and the patient is still symptom-free.

DISCUSSION

John Addison Fordyce originally characterized angiokeratoma of Fordyce across the scrotal area in 1896.[2] Solitary angiokeratoma

(site-oral cavities and lower limbs), angiokeratoma of Fordyce (scrotal wall/vulva), mibelli angiokeratoma (on dorsal skin of fingers and interdigital area), and angiokeratoma corporis diffusum are the four different varieties of angiokeratoma (on lower abdomen, genitals, hips, and thighs). The most typical variety of angiokeratoma is Fordyce's angiokeratoma.[3] Angiokeratomas are mostly asymptomatic lesions that range in color from blue to red to purple and can be up to 1 cm in diameter. Males are more likely to develop angiokeratomas, which usually affect the scrotal wall, the shaft, and the glans of the penis.[4]

Differential diagnosis for angiokeratoma clinically includes hemangioma, common spider angioma, pyogenic granulomas, eruptive angiomas, hereditary hemorrhagic telengiectasia, molluscum contagiosum, warts, and condylomas is among the differential diagnoses for angiokeratoma.[5] Dark colored angiokeratoma may mimic malignant melanoma. Therefore, it is always advised to do a biopsy and histopathologic examination to confirm the diagnosis.

Histopathological analysis is characterized by hyperkeratosis, papilomatosis, acanthosis, and dilated vasculature in papillary dermis. In perivascular elastic tissue, degenerative alterations are seen and may play a role in the etiology of vulvar angiokeratoma. The primary causative factor in the etiology of angiokeratoma is capillary ectasia in papillary dermis. Friction is the primary cause of epidermal alterations in all types of angiokeratoma.

Angiokeratomas are typically asymptomatic lesions that progress and generally do not need to be treated. There have been reports of sporadic bleeding, pruritus, soreness, a burning feeling, and dyspareunia. However, therapy is necessary for women who exhibit symptoms. Depending on the severity of the lesion, treatment options may include surgical excision, electro cauterization, cryotherapy, or laser treatment (using an argon or carbon dioxide laser).

CONCLUSION

In our case, solitary vulvar angiokeratoma caused discomfort due to its location and serious concern to the patient; therefore, it was successfully treated by electrocautery. In asymptomatic patients, reassurance and follow-up should be sufficient.

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