Multiple Angiokeratomas of the Vulva: Case Report and Literature Review

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Received: July 7, 2010
Accepted: October 28, 2010

SUMMARY Angiokeratomas of the vulva are relatively rare finding and a limited number of cases have been reported in the literature. Clinically, angiokeratomas of the vulva are benign vascular lesions usually occurring in middle-aged or older women. Microscopically these lesions are characterized by epidermal hyperkeratosis, papillomatosis, acanthosis, and marked dilatation of the papillary dermal vessels. In most patients, genital lesions are asymptomatic; however, bleeding, dyspareunia and other symptoms have been described. We report a case of a 45-year-old woman with numerous blue-to-red, scaly papules that spread over the entire area of both labia majora. The patient reported occasional pruritus and burning sensations, discomfort during the intercourse, and significant psychological burden. Histopathologic analysis of the lesion confirmed the diagnosis of angiokeratoma, and all lesions were electrocauterized under local anesthesia. The results of the treatment were very satisfactory, with no side effects or complications. During one-year follow-up, no relapses were noted and the patient remained asymptomatic. Therefore, dermatovenereologists should be aware of angiokeratomas and respective therapeutic options when examining a patient with pruritic, painful, or bleeding lesions in the genital region.

KEY WORDS: angiokeratoma, multiple angiokeratomas, vulva

INTRODUCTION

Angiokeratomas are benign vascular lesions presenting as blue-to-red papules with a scaly surface (1). Angiokeratomas are classified into widespread forms (angiokeratoma corporis diffusum), which are usually associated with a congenital error of metabolism, and localized forms including solitary angiokeratoma, angiokeratoma of Fordyce, angiokeratoma circumscriptum nae-
therapy is sometimes required, especially if they bleed and cause anxiety (3). We report a case of a 45-year-old woman with numerous angiokeratomas spread over the entire area of the labia majora.

**CASE REPORT**

A healthy 45-year-old Caucasian woman presented with numerous grayish-purple macular and papular lesions, ranging in size from 0.1 to 0.4 cm, spread over the entire area of both labia majora (Fig. 1). Several lesions were centrally eroded. The patient complained of occasional pruritus and burning sensations, but discomfort and pain during the intercourse as well as significant psychological burden were her major concerns. The patient's family and personal medical history was otherwise unremarkable. Clinically, the diagnosis of angiokeratomas was suspected and excisional biopsy of one papule was performed. Histopathologic analysis showed dilated blood-filled vessels in the papillary dermis with overlying hyperplastic and hyperkeratotic epidermis, and the diagnosis of angiokeratoma was confirmed (Fig. 2A, B). All lesions were electrocauterized under local anesthetic, on a couple of sessions (Figs. 3 and 4). Besides the initial moderate pain during the application of local anesthetic, there were no side effects or complications. The patient was extremely satisfied with the outcome. During one-year follow-up, there were no relapses and the patient remained asymptomatic (Fig. 5).

**DISCUSSION**

In 1896, a localized form of angiokeratomas in the scrotal area was first described by John Addison Fordyce (1). Angiokeratomas are typically asymptomatic, blue-to-red papular lesions with slightly keratotic surface, measuring less than 1 cm in diameter (8). Angiokeratoma is more common in males, in whom it most commonly occurs on the scrotal wall (8), with occasional affection of the shaft and glans of penis, and rarely leg, especially lower leg area and bulbar conjunctiva (1). An equivalent form can occur in females, generally on the vulva, while the clitoris is an extremely rare location (8,10). Angiokeratomas of the vulva

![Figure 1. Patient with multiple angiokeratomas of the vulva before treatment.](image)
are uncommon benign vascular lesions (7,8), and some authors suggest that they usually occur in older women (11), while other report that they occur before the age of 50 years (6,12).

Histopathologic analysis of these lesions is characterized by epidermal hyperkeratosis, papillomatosis, acanthosis, and dilated vasculature in the papillary dermis (6,13). Degenerative changes in the perivascular elastic tissue are observed and may contribute to the pathogenesis of vulvar angiokeratomas (6). In all forms of angiokeratoma, epidermal changes are secondary to friction (1). Capillary ectasia in the papillary dermis is the central part in the pathogenesis of angiokeratomas (2). Different entities causing vessel ectasia lead to the many clinical variants of angiokeratoma (2). Hence, the vessel wall weakness, either for acquired or congenital reasons, can cause the formation of these lesions (8). In addition, a case of angiokeratoma of the vulva, on both labia majora, following chronic HPV-6 viral infection in the vulva has been reported in the literature (14).

Differential diagnosis of angiokeratomas includes hemangiomas, papular spider angiomas, pyogenic granulomas, eruptive angiomas, hereditary hemorrhagic telangiectasia, molluscum

Figure 3. Clinical finding after the first treatment with electrocauterization.

Figure 4. Clinical finding after the second treatment with electrocauterization.

Figure 5. During one-year follow-up, there were no relapses and the patient remained asymptomatic.
Angiokeratomas of the vulva

In particular, dark-colored, solitary angiokeratoma may resemble malignant melanoma (12). Therefore, biopsy and histologic analysis is recommended to confirm the diagnosis. Clinically, infections, inflammatory lesions, vascular conditions, and epithelial tumors must be differentiated (6).

Likewise, in cases of diffuse distribution of hundreds of tiny angiokeratomas, one should always investigate to exclude Anderson-Fabry’s disease (16,17) and other metabolic causes of angiokeratomas, such as fucosidosis (18). Fabry’s disease is a rare, X-chromosome-linked lysosomal storage disease caused by a deficient alpha-galactosidase, a lysosomal enzyme leading to the accumulation of glycolipids in the cells of different tissues (16,19). Clinical manifestations start at early age and include angiokeratoma, acroparesthesia, hypohidrosis, heat/exercise intolerance, gastrointestinal pain, diarrhea, and fever. The main complications of Fabry’s disease are more prominent after the age of 30, when the kidney, heart and/or cerebrovascular disorders appear (16). As mentioned above, our patient’s medical history was unremarkable, with no cases of cardiac or cerebrovascular disorders in the family; therefore, no additional examinations were conducted. However, cases of patients with Fabry’s disease without typical lesions of angiokeratomas have been described (19).

In general, angiokeratomas are asymptomatic lesions and do not require treatment, which is the case with genital lesions, too. However, intermittent bleeding, pruritus, pain, burning sensations and dyspareunia have been described (20). In our case, genital lesions caused significant psychological burden and affected sexual activity of the patient. Therefore, in asymptomatic patients, management should only include reassurance and follow-up observation (6). However, in women who have symptoms therapy is indicated, and treatment modalities include surgical excision, electrocauterization, cryotherapy or laser treatment (argon or carbon dioxide laser), the choice of which would largely depend on the size of the lesion (1,6,7,15). Recently, a series of successfully treated multiple angiokeratoma patients with pulsed dye laser have been reported, and the treatment was performed without the need of local anesthesia (7).

CONCLUSION

In the present case, multiple vulvar angiokeratomas caused discomfort and concern to the patient; therefore, they were successfully treated with electrocauterization. In asymptomatic patients, reassurance and follow-up should be sufficient. However, angiokeratomas may occur as a cutaneous marker of Fabry’s disease or other metabolic disorders. Therefore, in case of diffuse distribution of numerous tiny angiokeratomas, patients should be extensively analyzed to exclude the possible underlying systemic disorder.

References


Nivea cream – for winter sports; year 1935. (From the collection of Mr. Zlatko Puntijar)