Acta Dermatovenerol Croat 2010;18(4):271-275 CASE REPORT

Multiple Angiokeratomas of the Vulva: Case Report and Literature Review

Marija Buljan[1,3], Sanja Poduje[1], Mirna Šitum[1,3], Vedrana Bulat[1], Željana Bolanča[1], Davor Tomas[2,4]

[1]University Department of Dermatovenereology, [2]Ljudevit Jurak University Department of Pathology, Sestre milosrdnice University Hospital; [3]School of Dental Medicine, [4]School of Medicine, University of Zagreb, Zagreb, Croatia

Corresponding author:
Marija Buljan, MD
University Department of Dermatovenereology
Sestre milosrdnice University Hospital
Vinogradska cesta 29
HR-10000 Zagreb
Croatia
buljan.marija@gmail.com

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 naeviforme and angiokeratoma of Mibelli (angiokeratoma acroasphycticum digitorum) (2). Angiokeratomas of Fordyce are typically located on the scrotum, shaft of penis, labia majora, inner thigh, or lower abdomen (3). Angiokeratomas of the vulva are relatively rare finding and only few cases have been reported in the literature (4,5). Even though angiokeratomas are usually asymptomatic lesions

Acta Dermatovenerol Croat
2010;18(4):271-275

(6,7), 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 anesthesia, 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).

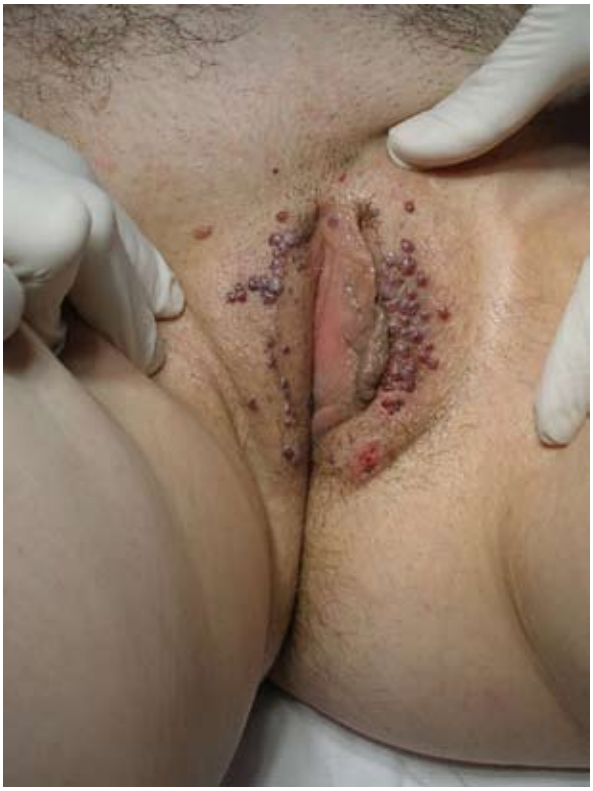

Figure 1. Patient with multiple angiokeratomas of the vulva before treatment.

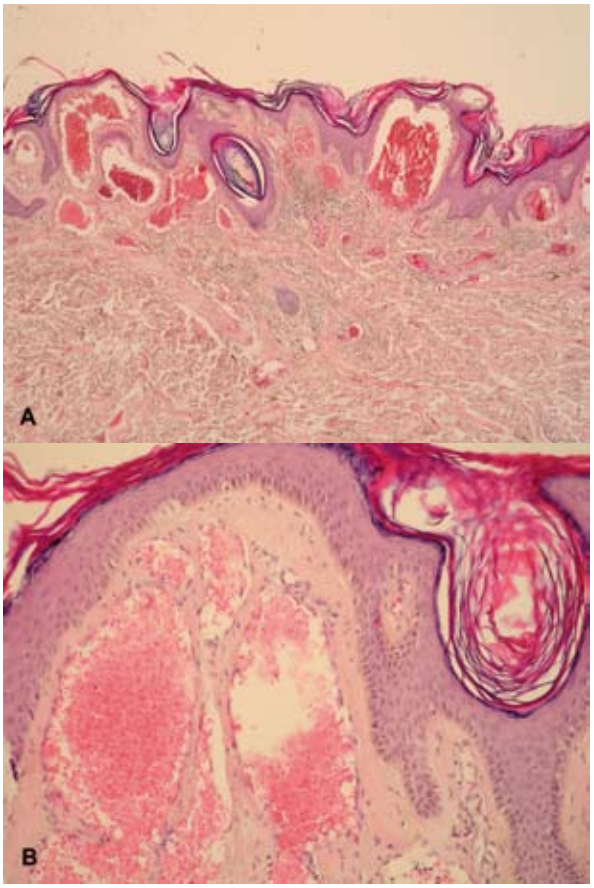

Figure 2. (A) Histopathology showing dilated blood-filled vessels in the papillary dermis (H&E, X40); (B) dilated blood vessels with overlying hyperplastic and hyperkeratotic epidermis (H&E, X200).

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

Acta Dermatovenerol Croat
2010;18(4):271-275

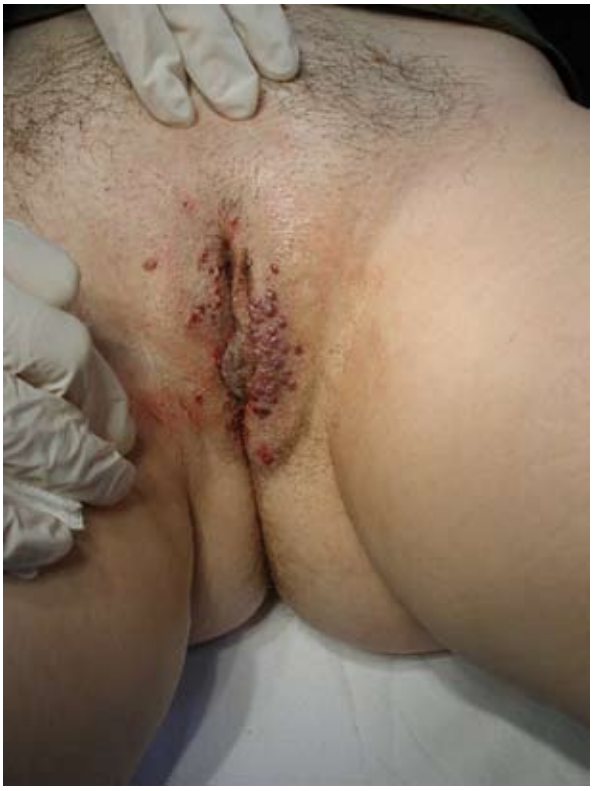

Figure 3. Clinical finding after the first treatment with electrocauterization.

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

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

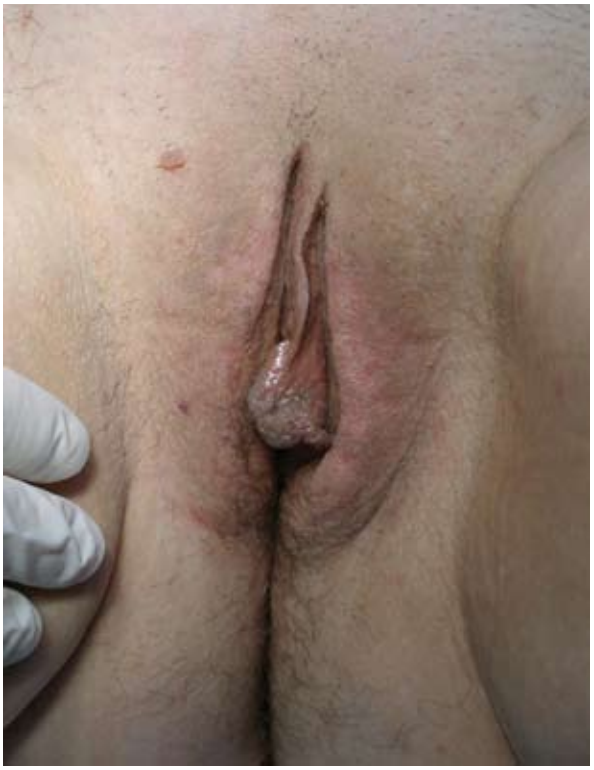

Figure 5. During one-year follow-up, there were no relapses and the patient remained asymptomatic.

Acta Dermatovenerol Croat
2010;18(4):271-275

contagiosum, warts, and condylomas (15). 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

1. Pande SY, Kharkar VD, Mahajan S. Unilateral angiokeratoma of Fordyce. Indian J Dermatol Venereol Leprol 2004;70:377-9.
2. Schiller PI, Itin PH. Angiokeratomas: an update. Dermatology 1996;193:275-82.
3. Ozdemir M, Baysal I, Engin B, Ozdemir S. Treatment of angiokeratoma of Fordyce with long-pulse neodymium-doped yttrium aluminium garnet laser. Dermatol Surg 2009;35:92-7.
4. Clark JR, Wheelock JB. Angiokeratoma of the vulva. A case report. J Reprod Med 1988;33:473-4.
5. Liapis A, Bakalianou K, Salakos N, Iavazzo C, Dalainas I, Kondi-Pafiti A. Vascular tumors: rare neoplasms of the female genital tract. Eur J Gynaecol Oncol 2009;30:220-2.
6. Cohen PR, Young AW Jr, Tovell HM. Angiokeratoma of the vulva: diagnosis and review of the literature. Obstet Gynecol Surv 1989;44:339-46.
7. Oni G, Mahaffey P. Treatment of angiokeratoma of the vulva with pulsed dye laser therapy. J Cosmet Laser Ther 2010;12:51-2.
8. Yigiter M, Arda IS, Tosun E, Celik M, Hicsonmez A. Angiokeratoma of clitoris: a rare lesion in an adolescent girl. Urology 2008;71:604-6.
9. Yamazaki M, Hiruma M, Irie H, Ishibashi A. Angiokeratoma of the clitoris: a subtype of angiokeratoma vulvae. J Dermatol 1992;19:553-5.
10. McNeely TB. Angiokeratoma of the clitoris. Arch Pathol Lab Med 1992;116:880-1.
11. Namazi MR, Maghsoodi M. Association of angiokeratoma of the vulva with angioma serpiginosum. J Drugs Dermatol 2008;7:882-3.

12. Kontogianni-Katsaros K, Kairi-Vassilatoy E, Grapsa D, Papadias K, Hasiakos D, Kondi-Pafitis A. Angiokeratoma of the vulva: a rare benign vascular tumor mimicking malignancy - case reports. Eur J Gynaecol Oncol 2006;27:632-3.
13. Kwon NH, Kim SY, Kim GM. A case of post-injury angiokeratoma circumscriptum of the chest. Ann Dermatol 2010;22:66-8.
14. Baruah J, Roy KK, Rahman SM, Kumar S, Pushparaj M, Mirdha AR. Angiokeratoma of vulva with coexisting human papilloma virus infection: a case report. Arch Gynecol Obstet 2008;278:165-7.15.
15. Braun-Falco O. Dermatology. 3rd completely rev. ed. Berlin; New York: Springer; 2009.
16. Rozenfeld PA. Fabry disease: treatment and diagnosis. IUBMB Life. 2009;61:1043-50.
17. Albano LM, Rivitti C, Bertola DR, Honjo RS, Kelmann SV, Giugliani R, *et al*. Angiokeratoma: a cutaneous marker of Fabry's disease. Clin Exp Dermatol 2009 Oct 19.
18. Venencie PY, Pauwels C, Malherbe V, Perie G, Landrieu P. [Angiokeratoma and fucosidosis. Immunohistochemical and ultrastructural study]. Ann Dermatol Venereol 1995;122:432-5.
19. Mirceva V, Hein R, Ring J, Mohrenschlager M. A case of multiple angiomas without any angiokeratomas in a female heterozygote with Fabry disease. Australas J Dermatol 2010;51:36-8.
20. Novick NL. Angiokeratoma vulvae. J Am Acad Dermatol 1985;12:561-3.

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