Dermatology Practical & Conceptual

Lichen Sclerosus or Vulvar Angiokeratoma: A Diagnosis That is not always so easy

Vincenzo De Giorgi[1,2], Gabriella Perillo[1], Pietro Antonini[3], Giovanni Cecchi[1], Biancamaria Zuccaro[1], Elisabetta Magnaterra[1], Eleonora Castellacci[4], Daniela Massi[5]

1 Section of Dermatology, Department of Health Sciences, University of Florence, Florence, Italy
2 Cancer Research "Attilia Pofferi" Foundation, Pistoia, Italy
3 Section of Pathology, Department of Diagnostic and Public Health, University of Verona, Verona, Italy
4 Section of Gynecology, Villa Donatello Hospital, Florence, Italy
5 Section of Pathology, Department of Health Sciences, University of Florence, Florence, Italy

Key words: Mucosal Lesion, Vascular Lesion, Dermoscopy, Ecchymotic Macules

Citation: De Giorgi V, Perillo G, Antonini P, et al. Lichen sclerosus or vulvar angiokeratoma: a diagnosis that is not always so easy. *Dermatol Pract Concept*. 2025;15(1):4691. DOI: https://doi.org/10.5826/dpc.1501a4691

Accepted: September 2, 2024; **Published:** January 2025

Funding: None.

Competing Interests: None.

Authorship: All authors have contributed significantly to this publication.

Corresponding Author: Vincenzo De Giorgi, MD, Department of Dermatology, University of Florence, Via Michelanielo 41, 50124 Firenze, Italy. Tel/Fax: 0039-055-6939632. E-mail: vincenzo.degiorgi@unifi.it

Introduction

Vulvar lichen sclerosus, a chronic inflammatory condition, can mimic angiokeratoma both clinically and dermoscopically, presenting diagnostic challenges. Initial misdiagnosis of vulvar lichen sclerosus as angiokeratoma can lead to inappropriate management and delayed treatment. This case highlights the importance of considering a broad differential diagnosis when evaluating vulvar lesions, particularly those with vascular characteristics.

Angiokeratoma is a benign cutaneous lesion derived from the dilatation of blood vessels in the papillary dermis, associated with hyperkeratosis and acanthosis of the epidermis. Clinically, it appears as a papule or plaque that is soft upon palpation and blue or violet in color, with a hyperkeratotic surface. Histologically, it shows vascular ectasia of the papillary dermis which may appear to extend into the epidermis, with overlying epidermal hyperplasia characterized by acanthosis, elongation of the rete ridges and hyperkeratosis, with the epidermis encircling the dilated vascular spaces.

Angiokeratoma could also be associated with other diseases, creating difficulties in differential diagnosis or in distinguishing it from an underlying disease.

Case Presentation

A 74-year-old woman was referred for evaluation of a vulvar blue plaque. The lesion had been present for 4 months and initially appeared as a small papule. Over time, it extended along the right entire labia minora. The patient reported the onset of symptoms such as itching and inconstant pain in the genital region. Examination showed a soft red-blue plaque

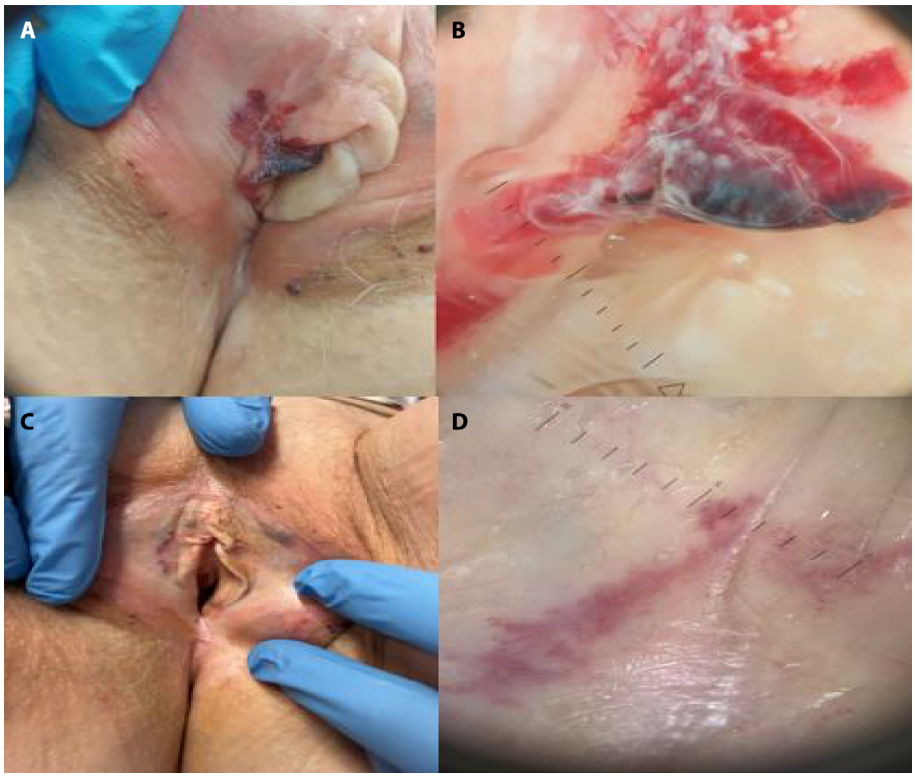

Figure 1. 74-year-old woman diagnosed with vulvar hemorrhagic lichen sclerosus. (A) Clinical examination showed a soft, red-blue plaque with irregular edges and clear demarcation from the surrounding normal skin. (B) Dermoscopy revealed blue-black lacunae with homogeneous red peripheral areas. Pearly white regions, skin atrophy with shiny white streaks and dots were observed on the left labia minora. (C) Significant clinical improvement was observed after one month of treatment with clobetasol propionate. (D) Post-treatment dermoscopy revealed only a residual reddish area with distinct boundaries.

with irregular edges and clear delimitation from the normal adjacent skin. In addition, there were also some other isolated papules measuring 2 mm, similar to the main lesion, over the vulva and labia majora (Figure 1A).

Dermoscopy revealed blue-black lacunae with homogeneous red peripheral areas (Figure 1B). At first sight, this lesion seemed to be a vulvar angiokeratoma, but on the left labia minora, there were pearly white areas, skin atrophy with shining white streaks, and dots upon dermoscopic examination (Figure 1B). Despite the main lesion, these elements suggested a clinical suspicion of lichen sclerosus. The plaque was biopsied, and the microscopic findings were consistent with the diagnosis of lichen sclerosus with concomitant haemorrhage (Figure 2). Thus, the patient was treated topically with 0.05% clobetasol propionate. After 1 month of treatment with clobetasol propionate she reported the disappearance of symptoms such as genital pain and itching. The examination revealed that the ecchymotic macules, vascular lesions, and the erosion on the vaginal vestibule had almost disappeared, both clinically and dermoscopically (Figure 1, C and D).

Conclusions

The initial diagnostic suspicion of angiokeratoma was disproven by anamnestic, clinical and dermoscopic evidence. The final diagnosis was hemorrhagic lichen sclerosus, which can sometimes mimic angiokeratoma, both clinically and dermoscopically, creating difficulties in differential diagnosis.

Khatib and colleagues reported a case of extragenital hemorrhagic bullous lichen sclerosus [1]. The mechanism underlying the onset of hemorrhagic bullae could be explained by the binding of IgG autoantibodies with basal membrane proteins such as extracellular matrix protein 1 (ECM1), which is fundamental for the interaction of collagen and the elastic fibres of the dermis at the basal membrane [2]. An other reason for the formation of hemorrhagic bullae is the oedema of the papillary dermis secondary to the inflammation and lymphocytic infiltrate, which interferes with maintenance of the structures of dermal collagen fibers [3].

This case highlights the critical importance of differentiating between vulvar lichen sclerosus and angiokeratoma, as accurate diagnosis is essential for effective treatment and management.

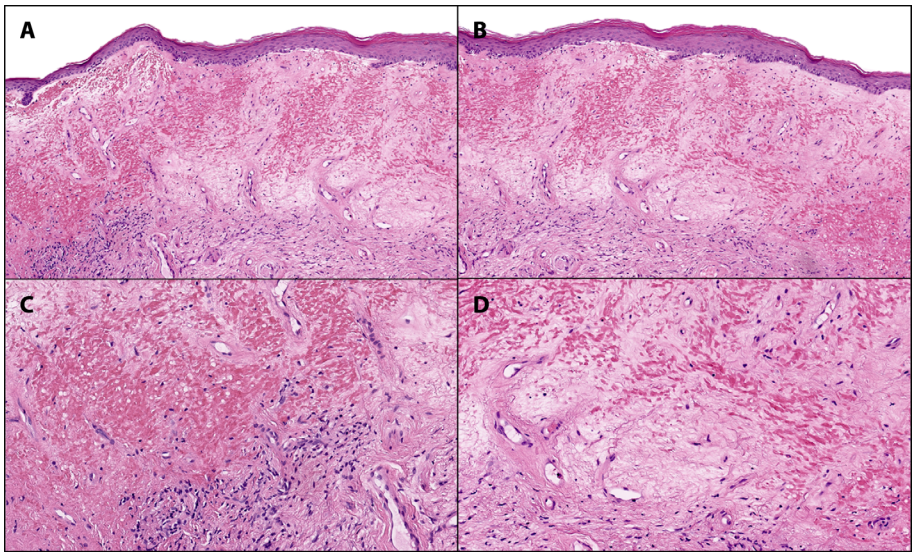

Figure 2. (A-D) Histopathological examination shows epidermal thinning with loss of rete ridges, hyperkeratosis and dermal sclerosis. Notable is the presence of superficial dermal hemorrhage with extravasated red blood cells. (A) H&E, original magnification X10. (B) H&E original magnification X10. (C) H&E original magnification X20. (D) H&E original magnification X20.

References

1. Khatib J, Wargo JJ, Krishnamurthy S, Travers JB. Hemorrhagic Bullous Lichen Sclerosus: A Case Report. *Am J Case Rep.* 2020;21:e919353. DOI: 10.12659/AJCR.919353. PMID: 33024070. PMCID: PMC7552941.
2. Chan I, Oyama N, Neill SM, Wojnarowska F, Black MM, McGrath JA. Characterization of IgG autoantibodies to extracellular matrix protein 1 in lichen sclerosus. *Clin Exp Dermatol.* 2004;29(5):499-504. DOI: 10.1111/j.1365-2230.2004.01573.x. PMID: 15347336.
3. Sauder MB, Linzon-Smith J, Beecker J. Extragenital bullous lichen sclerosus. *J Am Acad Dermatol.* 2014;71(5):981-4. DOI: 10.1016/j.jaad.2014.06.037. PMID: 25088813.