

# Vulvar Lymphangioma Circumscriptum Secondary to Crohn Disease



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A 56-year-old woman presented to the dermatologist for worsening vulvar papules and edema for 3 years. She had Crohn disease in remission status post colectomy with ileostomy and vaginal fistula repair. Prior nondiagnostic biopsies performed by the gynecology service showed squamous hyperplasia, lichenoid features, acute and chronic inflammation, and dermal fibrosis. The labia were markedly edematous, with clear to hemorrhagic papules and vesicles coalescing to a papillomatous plaque (Figure 1). These findings were suggestive of acquired lymphangioma circumscriptum (LC) secondary to Crohn disease. This diagnosis was confirmed by a skin biopsy, which showed cystically dilated vascular lymphatic spaces in the dermis (Figure 2). Clobetasol 0.05% cream daily, initiated by the gynecologist, resulted in significant improvement during several weeks (Supplemental Figure, available online at <http://www.mayoclinicproceedings.org>).

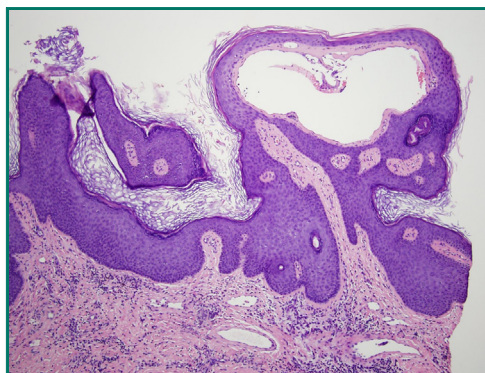
Lymphangioma circumscriptum, a benign lymphatic malformation, is most commonly seen in children on the trunk and extremities.<sup>1,2</sup> It is less commonly acquired in Crohn disease or from radiation therapy for pelvic malignant disease.<sup>3</sup> Acquired LC typically arises on the vulva as pink papillomatous or vesicular papules and plaques in the fifth decade.<sup>3,4</sup> Pruritus, oozing, and lymphedema are not uncommon.<sup>4</sup> Histopathologic evaluation, demonstrating dilated lymphatic channels in the papillary dermis with overlying epidermal hyperplasia, confirms the diagnosis.<sup>1,2</sup> Treatment options include laser therapy, cryotherapy, sclerotherapy, surgical excision, and labiectomy. The lesions tend to recur. Rarely, lymphangiosarcoma arising in long-standing LC has been described.<sup>4,5</sup> This case is unique because treatment with a superpotent topical steroid led to clinical improvement, although a multimodal treatment approach will likely be required.



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**FIGURE 1.** Grouped pink and gray verrucous vesicles and papules on an erythematous base involving the bilateral vulva with marked edema.



**FIGURE 2.** A vulvar biopsy specimen shows cystically dilated lymphatic spaces in the dermis with coagulated lymph and scattered red blood cells (hematoxylin-eosin stain, magnification  $\times 100$ ).

Recognizing vulvar LC early is important to establish the correct diagnosis and management.

#### SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at <http://www.mayoclinicproceedings.org>. Supplemental material attached to journal articles has not been edited, and the authors take responsibility for the accuracy of all data.

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