CASE REPORT

Vulvitis circumscripta plasmacellularis in pre-existing lichen sclerosus

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A B S T R A C T

Vulvitis circumscripta plasmacellularis (VCP) is a rare, benign vulvar disorder that presents with inflammatory patches and erosions. Patients with VCP usually experience vulvodynia, pruritus, or dyspareunia, which may cause severe discomfort. A 72-year-old woman presented with 10 years of vulvar pruritus and whitish patches on the labia minora and majora. A biopsy was performed on the labia minora, including the majora, the results of which revealed pronounced edema and homogenization of collagen in the upper dermis. Hyperkeratosis and follicular plugging in the epidermis and inflammatory infiltrate in the mid-
dermis were observed (Figure 2). Lichen sclerosus was diagnosed, and the patient was treated with a topical potent corticosteroid (clobetasol-17-propionate) for 2 weeks. Her condition improved, but her symptoms were aggravated after 5 months. She presented with severe pruritus and pain in the vulvar area. A physical examination revealed localized whitish to erythematous eroded patches on the labia minora (Figure 1B). A rebiopsy on the eroded patches of the labia minora was performed to rule out squamous cell carcinoma. The biopsy specimen showed epidermal thinning, absence of the stratum corneum, irregular acanthosis, and spongiosis in the epidermis. A dense dermal lichenoid infiltrate with many plasma cells and lymphocytes was observed (Figure 3). Results of an immunohistochemical analysis revealed that plasma cells were polyclonal for kappa and lambda light chains, and syphilis testing with venereal disease research laboratory was negative. Based on the results of the biopsy, VCP was diagnosed. After treatment with a systemic and topical corticosteroid, the patient’s condition temporarily improved; however, her symptoms recurred, and she was advised to apply imiquimod 5% cream two to three times a week. After 1 month, her condition slightly improved and she was lost to follow-up.

Discussion

VCP is a rare vulvar disorder, which causes vulvodynia, pruritus, and discomfort. Patients usually present with erythematous patches or plaques on the labia minora, majora, and other vulvar structures. The clinical appearance of VCP may be similar to extramammary Paget’s disease, squamous cell carcinoma, Bowen’s disease, pemphigus vulgaris, allergic contact dermatitis,
candidiasis, syphilis, sexual abuse, herpes simplex infection, erosive lichen planus, or fixed drug eruption.2,4 The etiology and pathogenesis of VCP remain unclear, but predisposing factors, such as infection, poor hygiene, heat, and constant friction have been suggested.5,6 In our patient’s case, due to the pruritic characteristic (which may trigger friction) of lichen sclerosus, the cause of VCP may have been lichen sclerosus.

Histological findings of VCP are as follows.2,7,8 The epidermis shows atrophy and often an absence of the horny and granular cell layers. It is composed of lozenge-shaped keratinocytes, and spongiosis in the epidermis is also present. The dermis shows a lichenoid infiltrate predominantly composed of plasma cells. In addition, blood vessels are dilated, and there may be extravasation of erythrocytes and deposits of hemosiderin. Reactive epithelial changes may be present, but true dysplasia is absent.

In our patient’s case, VCP was diagnosed in pre-existing lichen sclerosus. The concurrence of VCP with vulvar lichen sclerosus has been reported only twice, one of which involved a 60-year-old woman, who presented with a 1-year history of vulvodynia. Results of a biopsy confirmed the diagnosis of lichen sclerosus and VCP.2 The other case was a 60-year-old woman with a long-standing history of lichen sclerosus under local therapy; she developed soreness and a scaling erythematous lesion in the vaginal introitus. A biopsy confirmed the diagnosis of VCP.3

As a complication of long-standing lichen sclerosus, the risk of developing squamous cell carcinoma is in the range of 4–5%, and lichen sclerosus is also associated with genital verrucous carcinoma.9,10 Therefore, a biopsy should be performed when lichen sclerosus lasts for a long period and shows erythematous eroded

![Figure 1](A) Localized whitish patches on the labia minora and majora. (B) After 5 months, the patient’s symptoms were aggravated. She presented with localized whitish to erythematous eroded patches on the labia minora; a rebiopsy was performed on the eroded patches of labia minora (arrow).

![Figure 2](Biopsy specimen from the labia minora including the majora shows pronounced edema and homogenization of collagen in the upper dermis. Hyperkeratosis and follicular plugging in the epidermis and inflammatory infiltrate in the mid-dermis are observed [hematoxylin and eosin, 40×].)

![Figure 3](Rebiopsy specimen from the eroded patch of the labia minora shows epidermal thinning, absence of the stratum corneum, irregular acanthosis, and spongiosis in the epidermis. A dense dermal lichenoid infiltrate with many plasma cells and lymphocytes is observed [hematoxylin and eosin: (A) 100×; (B) 400×].)
lesion. However, we note that similar lesions may be exhibited by other diseases, such as VCP. Herein, we described an interesting and rare case of VCP in pre-existing lichen sclerosus. Dermatologists should understand that VCP can result from lichen sclerosus and that it resembles squamous cell carcinoma.

References