



ZOON'S VULVITIS: AN UNCOMMON IDIOPATHIC DERMATOSIS SUCCESSFULLY TREATED WITH TOPICAL ESTROGEN

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Background/Objective: Zoon's vulvitis, also known as plasma cell vulvitis or circumscribed plasma cell vulvitis, is a rare, benign vulvar dermatosis of unknown etiology, possibly associated with autoimmune conditions. It predominantly affects postmenopausal women and due to its nonspecific clinical presentation and overlap with other vulvar dermatoses, the condition is frequently underrecognized, leading to delayed diagnosis. Histopathological evaluation is essential, typically revealing a dense inflammatory infiltrate rich in plasma cells and lymphocytes [1,2]. This report aims to present a case of Zoon's vulvitis with persistent clinical presentation and initial therapeutic failure, emphasizing diagnostic confirmation and therapeutic alternatives.

Methods: This is a case report based on data extracted from the patient's medical records. The patient provided written informed consent for publication, and the study was approved by the Research Ethics Committee of Irmandade da Santa Casa de Misericórdia de São Paulo.

Case Report: A 47-year-old woman was referred to a Lower Genital Tract Pathology clinic with a two-month history of painful erythematous vulvar lesions associated with persistent burning. She had previously been evaluated in primary care settings and treated with topical healing agents, without clinical improvement. She denied associated symptoms, comorbidities, regular medication use, or substance use. Her history was notable for menopause at age 40, followed by four years of oral hormone therapy, and sexual abstinence for the past seven years. Physical examination revealed atrophic vulvar mucosa with erythematous macules located in the fossa navicularis, extending to the vaginal introitus, associated with significant local tenderness (Figure 1). The cervix and vagina were atrophic but otherwise normal, with no lesion extension.

Initial diagnostic hypotheses included vulvar hematoma secondary to atrophy, Zoon's vulvitis, and erosive lichen planus. A biopsy was performed, and empirical treatment with cold boric acid solution and topical dexamethasone was initiated. After 15 days, there was no clinical improvement. Histopathological analysis demonstrated chronic vulvitis with epidermal leukocytic infiltration, hemorrhagic foci, and vascular proliferation, without evidence of malignancy. Immunohistochemistry revealed CD138 positivity in plasma cells, CD3 positivity in numerous T lymphocytes, CD20 positivity in rare B lymphocytes, CD34 positivity in vascular structures, and negative p16 and p53 expression, confirming the diagnosis of plasma cell vulvitis (Figure 2). Topical corticosteroid therapy was escalated to clobetasol propionate 0.05% once daily for 30 days, without clinical response. The treatment was then switched to topical conjugated estrogen (0.625 mg/g) for 21 days, resulting in complete resolution of both symptoms and lesions (Figure 3). Maintenance therapy with topical estrogen twice weekly was instituted, and the patient was subsequently discharged with outpatient follow-up.

Conclusions: Zoon's vulvitis is a challenging condition, often misdiagnosed as other inflammatory or infectious vulvar dermatoses. Histopathological evaluation with immunohistochemical analysis is essential for diagnostic confirmation. Although high-potency topical corticosteroids are considered first-line therapy, the therapeutic response is variable. This case highlights the potential role of topical estrogen therapy as an effective alternative in postmenopausal patients who do not respond to conventional treatment, contributing to improved clinical management of this underrecognized condition.



Figure 1. Assessment of the vulva showing reddish macules in the region of the fossa navicularis, extending to the vaginal introitus.

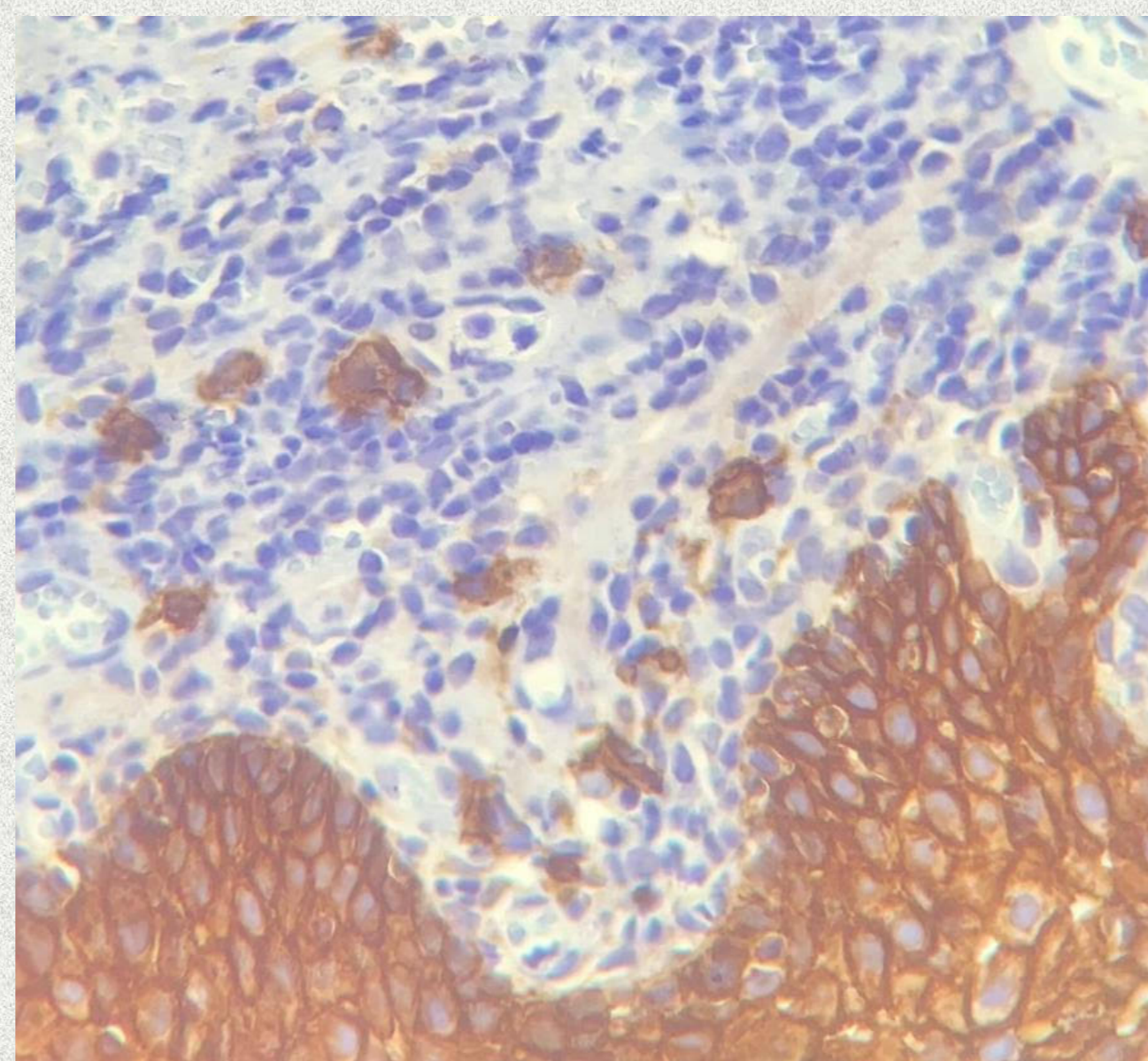


Figure 2. Immunohistochemical staining (CD138), 40× magnification, demonstrating positivity with abundant plasma cells within the inflammatory infiltrate in the vulvar squamous epithelium.



Figure 3. Assessment of the vulva after topical estrogen therapy demonstrates complete resolution of the lesion.

References:

1. Salopek TG, Siminoski K. Vulvitis circumscripta plasmacellularis (Zoon's vulvitis) associated with autoimmune polyglandular endocrine failure. *Br J Dermatol.* 1996 Dec;135(6):991-4.
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