Images in Women's Dermatology

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Vulvar burning with erythematous macules of the vestibule

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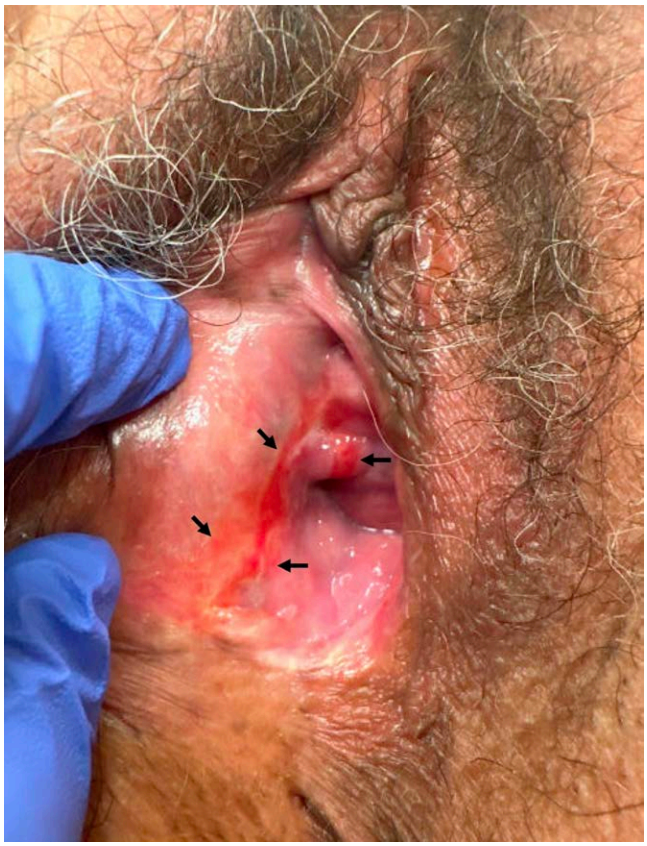

Fig. 1. Bright red to brown erythematous macules coalescing into a patch on the vestibule bilaterally.

Case summary

A 55-year-old woman presented with a 1-year history of vulvar pain and burning. She denied vaginal discharge or oral lesions. Previous treatments included short courses of individual topical agents: tacrolimus ointment, estrogen cream, topical testosterone, as well as topical baclofen for presumed vulvodynia, without improvement. She was not applying topicals or using premoistened wipes. She had a history of Hashimoto's thyroiditis, on levothyroxine. Her past medical/surgical history included nephrolithiasis and urinary stress incontinence status post-transobturator urethral suspension 12 years prior. She denied trauma to the area. On physical examination, the patient appeared healthy, alert, and in no acute distress. No nail changes or oral lesions were observed. Vulvar examination revealed bright red to brown erythematous macules on the vestibule bilaterally (Fig. 1). There was no evidence of scarring, erosions, ulcerations, or fissures. Punch biopsy revealed epidermal thinning and a diffuse plasma cell infiltrate (CD138 positive) in the dermis.

Question 1

What is the most likely diagnosis?

A. Fixed drug eruption
B. Lichen planus
C. Mucous membrane pemphigoid
D. Pemphigus vulgaris
E. Plasma cell vulvitis

Correct answer: E. Plasma cell vulvitis. This diagnosis is supported by physical exam findings of bright red to brown erythematous macules on the vestibule bilaterally and biopsy revealing diffuse CD138 positive plasma cell infiltrate in the

What is known about this subject in regard to women and their families?

- Plasma cell vulvitis is an uncommon vulvar condition that can lead to burning, itch, and pain, and diagnosis is often delayed.

What is new from this article as messages for women and their families?

- This article is a case highlighting the classic morphologic findings of plasma cell vulvitis, which clinicians should consider in the differential diagnosis for vulvar mucosal dermatoses.

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dermis. The other answer choices would not have biopsy findings with a dense infiltrate of plasma cells.

Discussion

Plasma cell vulvitis (PCV) is a rare inflammatory vulvar dermatosis with under 250 cases reported in the literature. It typically occurs between the fifth to eighth decade.[1] While symptoms of pain, burning, dyspareunia, stinging, and pruritus are common, this condition may also be asymptomatic.[1] PCV is thought to occur on a spectrum of plasma cell mucositis that can also involve the glans penis and oral mucosa.[1] Some consider PCV to be a variant of mucosal lichen planus. Regardless, PCV has a unique clinical morphology that is distinct from vulvovaginal lichen planus. The clinical presentation consists of well-demarcated copper colored to bright red or brown macules or patches, as seen in our patient. Findings may mimic erosions but upon further inspection and palpation, the epidermis is found to be intact. The absence of Wickham's striae and absence of architectural changes are integral to the diagnosis and helpful for ruling out lichen planus or other prescarring inflammatory conditions. Lesions are mostly commonly visualized on the vestibule, labia minora, and periurethral area.[1]

While the etiology of PCV is unclear, an autoimmune pathogenesis is speculated, and viral, irritant, and hormonal factors have also been implicated.[1]

Diagnosis is often delayed, with one study finding a diagnostic delay of almost 5 years.[2] The differential for PCV is broad and includes other inflammatory vulvar dermatoses such as vulvovaginal lichen planus, lichen sclerosus, mucous membrane pemphigoid, and pemphigus vulgaris. Additionally, neoplastic conditions (differentiated vulvar intraepithelial neoplasia or extramammary Paget disease) and infectious conditions (syphilis) should be considered in the differential.[1]

PCV is often refractory to treatment and treatment guidelines do not exist. Modalities frequently reported include topical corticosteroids, topical immunomodulators, and topical calcineurin inhibitors, with a recent systematic review finding the most evidence for topical clobetasol 0.05% and topical tacrolimus 0.1% ointment.[1] Other therapeutic options that have been reported include surgical excision, cryotherapy, and carbon dioxide laser ablation.[1] A case series reported successful treatment with hydrocortisone suppositories alternating with estradiol vaginal cream every night for 4–8 weeks.[3] Our patient was treated with intravaginal 25 mg hydrocortisone suppositories alternating with estradiol 0.01% cream every other night for 8 weeks with improvement clinically and symptomatically.

PCV is an underrecognized entity and should be considered in patients with characteristic morphologic findings, particularly if lesions are localized to the vestibule or labia minora. Histopathology will reveal a predominant plasma cell infiltrate and can confirm the diagnosis. Early recognition and initiation of multimodal therapy can improve clinical outcomes.

Question 2

Which of the following is most likely to be seen in the clinical course of PCV?

A. Architectural changes (scarring)
B. Development of oral lesions
C. Dyspareunia
D. Progression to squamous cell carcinoma
E. Recurrent urinary tract infections

Correct answer: C. Dyspareunia. While architectural changes and squamous cell carcinoma can develop in other vulvar conditions like lichen planus and lichen sclerosus, they do not develop in PCV. It is important to remember that PCV can occur as a concomitant condition with lichen sclerosus, but in this case, the patient only has PCV. Oral lesions can be seen in lichen planus but are not found in PCV. Urinary tract infections may develop due to changes in urethral anatomy with scarring conditions that can affect the urethra, however, PCV is not a prescarring condition and thus, recurrent UTIs should not be seen in PCV.

Conflicts of interest

None.

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Study approval

N/A

Author contributions

CK and AK: Participated in research design and writing of the paper.

Patient consent

Informed, written consent was received from all patients for whom photographs are present in the manuscript.

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