Plasma Cell Vulvitis (Zoon's Vulvitis): A Rare Case Report with Emphasis on Diagnostic Challenge

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Abstract: Plasma cell vulvitis (PCV), also referred to as Zoon's vulvitis, is a rare and chronic inflammatory condition of the vulva characterized by distinctive red, glistening patches with a subtle red-orange hue. The condition can be asymptomatic or present with symptoms such as discomfort, dyspareunia, and pruritus, often mimicking other vulvar mucosal disorders like lichen planus. Due to its rarity and the overlap of symptoms with more common vulvovaginal conditions, PCV is frequently underreported and misdiagnosed. The exact etiology of PCV remains unclear, with possible associations to herpes simplex virus (HSV) and abnormal immune responses being hypothesized. This manuscript presents a case of PCV in a 46-year-old female who presented with a red, glistening, focally ulcerated patch on the vulva.

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Introduction

Plasma cell vulvitis (PCV), also called Zoon's vulvitis, is an uncommon, chronic inflammatory condition of the vulva, marked by distinct red, glistening patches with a subtle red-orange tint (Goldstein et al., 2005; Damiani et al., 2017). The prevalence of PCV is not well-documented due to its rarity and likely under reporting, which can be attributed to the overlap of its symptoms with more common vulvo-vaginal disorders (Damiani et al., 2017). PCV can present as an asymptomatic lesion or cause symptoms such as discomfort, dyspareunia (pain during intercourse), and pruritus (itching), often resembling other vulvar mucosal conditions like lichen planus (Virgili et al., 2015).

The exact pathophysiology of PCV is not well understood. Some hypotheses suggest a possible link to herpes simplex virus (HSV), while others propose that it may result from an abnormal immune response (Morioka et al., 1988). This case report describes a rare occurrence of plasma cell vulvitis in a middle-aged woman, emphasizing the importance of biopsy for timely diagnosis and treatment. It highlights how patients presenting with well-defined red plaques and persistent vulvar discomfort should prompt consideration of PCV in the differential diagnosis.

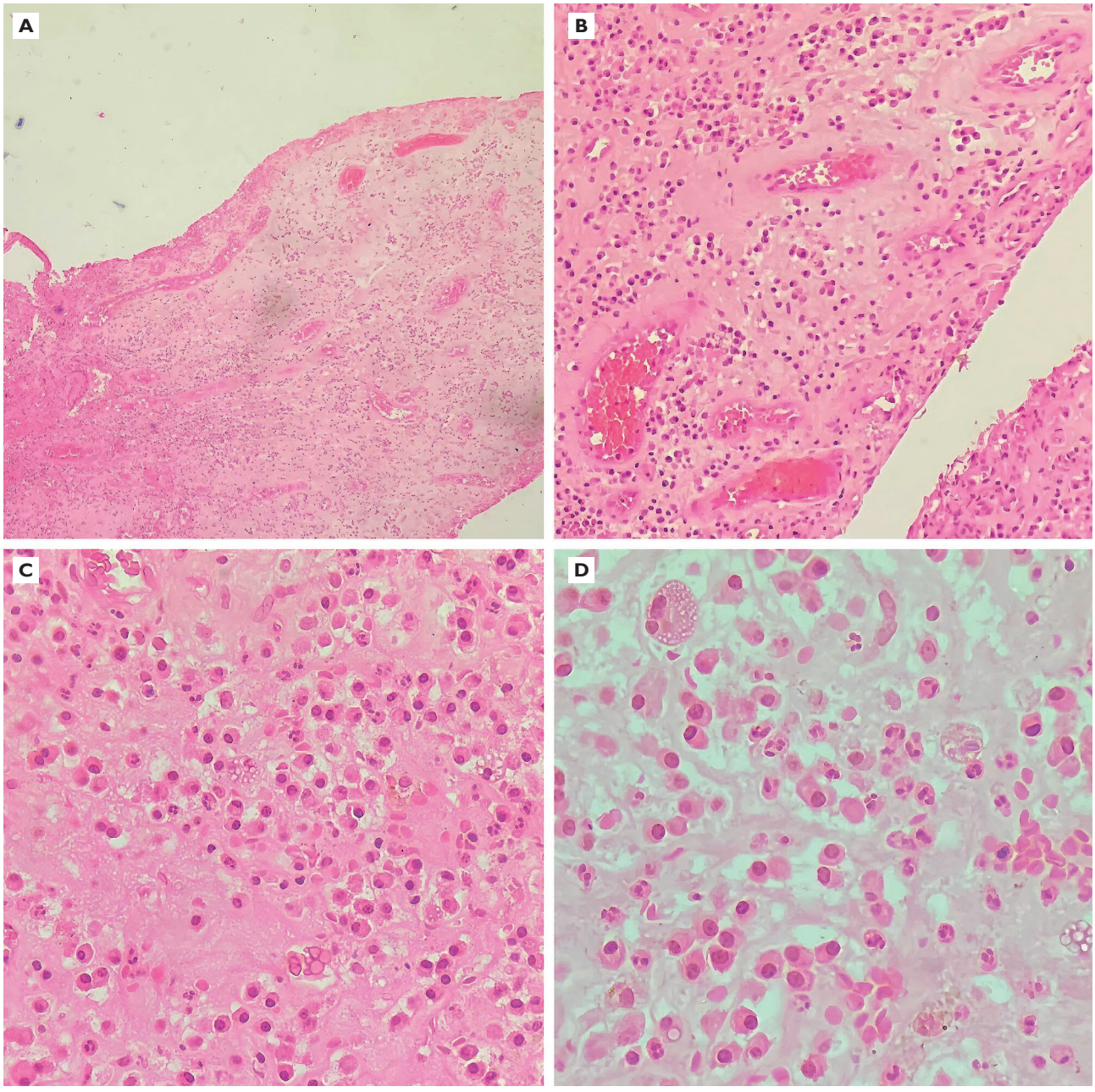

Figure 1: Histopathological features of plasma cell vulvitis. (A) Atrophic epidermis with focal ulceration (haematoxylin and eosin [H and E], 10×). (B) Subepithelial dense plasma cell infiltration with ectatic, congested vessels (H and E, 20×). (C and D) High-power view showing numerous plasma cells with eccentric nuclei and interspersed macrophages (H and E, 40×).

Case report

A 46-year-old female patient presented to the gynecology outpatient clinic with complaints of vulvar irritation, lower abdominal pain, and urinary incontinence for the past five months. She had a history of hysterectomy two years prior. There was no history of vaginal discharge, vulvar itching, allergic reactions, or medication use. Physical examination revealed a red, glistening, focally ulcerated patch on the vulva, measuring 1.5×2 cm in diameter. A biopsy was performed, and the specimen was sent for histopathological examination.

Histopathology revealed fragmented tissue lined by thin, atrophic epidermis with focal areas of ulceration. The underlying sub-epithelial stroma showed dense infiltration of plasma cells along with lymphocytes and histiocytes. Ectatic and congested small-caliber blood vessels were also noted (Figure 1). Based on these findings, a diagnosis of plasma cell vulvitis was made. The patient was prescribed topical steroids (2% hydrocortisone) and scheduled for follow-up.

Discussion

Zoon originally identified a chronic, benign inflammatory disorder of the penis and prepuce, histologically characterized by plasmacytic infiltration, which he termed balanitis plasmacellularis (Neri et al., 1995). In 1954, Garnier first described similar lesions in women as Zoon's vulvitis (Virgili et al., 2015).

PCV is indeed a rare condition, with an underreported prevalence likely due to its overlap with more common vulvovaginal disorders such as lichen planus, lichen simplex chronicus, and psoriasis, all contributing to its perceived rarity. Proper diagnosis typically requires a thorough medical history, physical examination, and often a biopsy to distinguish it from other similar conditions (Damiani et al., 2017).

The causes of PCV are still unclear. Due to its association with desquamative gingivitis, autoimmune polyglandular endocrine failure, and circulating antibodies, some researchers believe PCV may be linked to an autoimmune disorder (Doherty et al., 1993). Histopathologically, the hallmark of plasma cell vulvitis is a dense infiltrate of plasma cells in the subepithelial region. The overlying epithelium may show atrophy, hyperkeratosis, or parakeratosis. In addition to plasma cells, a mixed inflammatory infiltrate of lymphocytes and histiocytes may also be observed (Joshi, 1999).

Given the rarity of PCV, treatment strategies are often based on case reports, as incidence and management guidelines are poorly documented. Most case studies recommend topical steroids, although some describe the use of tacrolimus and other immunosuppressive therapies (Botros et al., 2006; Birx et al., 2010).

Conclusion

Owing mainly to its clinical similarity to more prevalent vulvovaginal dermatoses, PCV is still an uncommon, underdiagnosed, and sometimes misunderstood chronic inflammatory condition. This example emphasizes how crucial it is to take PCV into account while making a differential diagnosis for persistent vulvar lesions. Histopathological analysis is essential for making a conclusive diagnosis since it shows distinctive infiltrates that are rich in plasma cells. While the precise cause is still unknown, new data points to potential viral or autoimmune connections. Increased knowledge among pathologists and clinicians, along with timely biopsy and suitable therapy, can greatly enhance patient outcomes and avoid needless morbidity brought on by an inaccurate or delayed diagnosis.

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