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Plasma cell vulvitis: A case report

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ABSTRACT

Plasma cell vulvitis (PCV) is a rare inflammatory condition characterised by plasma cell infiltration in the vulva. A woman in her 80s was referred to a specialist gynaecology clinic with chronic, painful vulval ulcers that were non-responsive to topical betamethasone. Following a biopsy confirming PCV, combination therapy was initiated. This included non-pharmacological management, such as promoting aeration and using hypoallergenic clothing and washes, combined with the daily application of clobetasone cream 0.05% and clindamycin cream 0.1%. Additionally, topical estriol 1% was applied twice weekly. The patient experienced rapid symptom resolution, with the PCV lesion healing within six weeks of starting treatment. This case documents the rare occurrence of plasma cell vulvitis presenting as chronic vulval ulceration, and proposes a treatment regimen worth considering in instances where monotherapy has been ineffective.

1. Introduction

Plasma cell vulvitis (PCV) is a very rare cause of vulval ulceration, resulting from plasma cell infiltration within the vulva vestibule (1,2). First described in case reports in 1954, only a handful of cases have been reported in the literature until recently (3). Idiopathic PCV is hypothesised to be continuous with other plasma cell mucosities, including plasma cell balanitis, plasma cell orificial mucositis, atypical gingivostomatitis and plasmacytosis circumorificialis. While the aetiology of these lesions is unknown, factors such as viral infections, autoimmune conditions, hormonal changes and traumatic or irritant factors have been implicated in their development (3–5).

The mean age of diagnosis is 50 years; however, isolated cases have documented its occurrence in females as young as 8 years (5). Presentations may be asymptomatic or associated with burning, stinging sensations, pruritus, pain, dysuria, dyspareunia, bleeding, dryness and abnormal vaginal discharge (1–5). The literature describes a range of examination findings, including raised, sharply demarcated red-brown patches with shiny, lacquered paint appearances. Petechial haemorrhages may also be seen. Histopathology shows inflammatory infiltrates of polyclonal plasma cells, lozenge- or diamond-shaped keratinocytes, erythrocytes and hemosiderin deposition (5). Other histopathological findings include epidermal atrophy, spongiosis and Russell bodies (5). This constellation of symptoms often leads to frequent misdiagnosis, with an average 5-year delay in treatment (5). Aphthous ulcers, syphilis and infectious vulvitis are common clinical mimics that, especially those with infectious aetiology, can be easily excluded in histopathological analysis (5). Only a handful of cases have been reported in which PCV has preceded other diagnoses, including vulval cancer. Despite these instances, the prevailing opinion is that PCV remains grossly underreported (3,5).

Multiple treatment regimens are recommended in the literature. These include topical antibiotics and antifungal agents, topical steroids of varying duration and potency, topical calcineurin inhibitors and immune modulators (1–8). Only a handful of cohort studies and no randomised controlled trials exist for these treatment regimens (1–8). Here, a rare case is reported of plasma cell vulvitis presenting with vulval ulceration. The patient had a good response to a combination treatment regimen of topical clindamycin, estriol and clobetasone in conjunction with non-pharmacological therapies.

2. Case Presentation

A woman in her 80s presented with a shallow introital vaginal ulcer that had persisted for several years. She reported the ulcer as uncomfortable but denied any associated itching, abnormal vaginal discharge, or bleeding. Initially diagnosed and treated as an aphthous ulcer by her general practitioner, it was non-responsive to several courses of topical betamethasone. Consequently, she was referred to gynaecological services for further review. Her complex past medical history included

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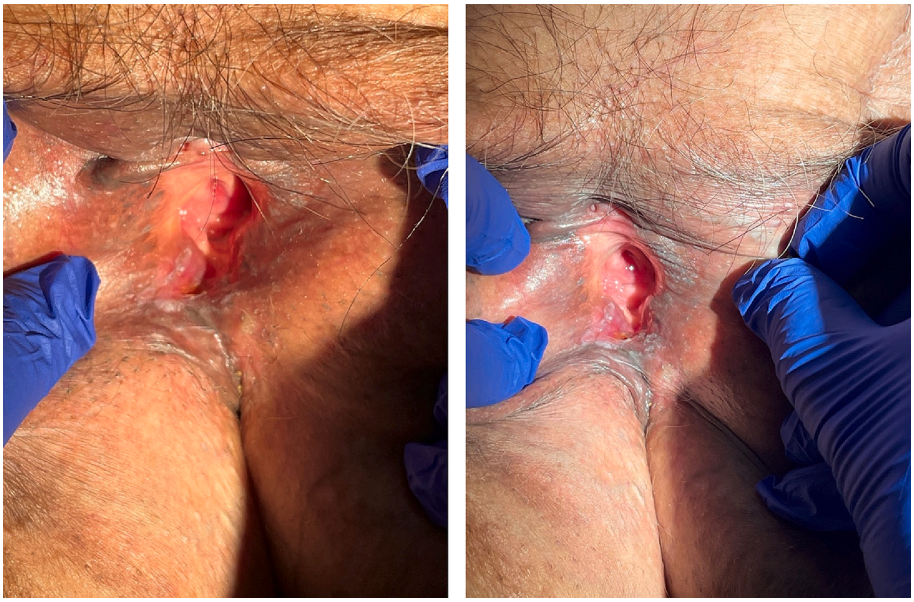

Fig. 1. Prior to the commencement of treatment, a 1 cm ulcerative lesion is observed on the right lower aspect of the vaginal introitus. It exhibits red-brown pigmentation and a shiny, lacquered appearance. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

hypertension, hypothyroidism, dyslipidaemia, deep-vein thrombosis, osteoporosis and lichen sclerosis.

Upon examination, a shallow ulcerative lesion, 1 cm in length, was located on the right lower aspect of the vaginal introitus. It was tender to touch and had classical red-brown pigmentation and a 'lacquered' appearances (Fig. 1). The remainder of her vagina appeared atrophic, which was normal for her age and otherwise healthy. Her cervix and cervical screening test completed during the initial review, returned negative results.

A biopsy of the lesion was taken, and histopathology revealed chronic inflammation with lymphocytes, histiocytes and a high density of plasma cells. The overlying squamous mucosa was atrophic but non-keratinising. Staining for infectious aetiologies returned negative results (Fig. 2). Following consultation with gynaecological pathology and dermatology specialists, the diagnosis of PCV was confirmed.

The patient commenced daily applications of clobetasone 0.05% and clindamycin 0.1% creams, in addition to topical estriol cream 1% applied twice weekly. Further non-pharmacological recommendations included using non-soapy water, undyed clothing and underwear and daily vulvar aeration.

At her routine six-week follow-up, she reported a complete resolution of symptoms. Examination revealed no sign of the previous PCV lesion (Fig. 3). She was advised to continue both topical estriol and clobetasone twice weekly, along with non-pharmacologic management strategies.

3. Discussion

Plasma cell vulvitis is a rare, chronic inflammatory condition of the vulva, often presenting with a protracted course and delays in both identification and the commencement of treatment. Numerous papers, including case reports, case series and systematic reviews, identify topical steroids as the most effective treatment (64% response rates), yet there are no standardised dosing regimens or recommendations regarding the duration of therapy. Clobetasol is the most frequently used of the available topical steroids, followed by hydrocortisone in terms of efficacy (1,5).

Virgil et al. conducted a retrospective cohort study comparing the efficacy of fusidic acid and betamethasone against clobetasol and tacrolimus. Although the results were equivocal, with all patients showing significant improvement, clobetasol and tacrolimus provided the greatest symptomatic relief (1). Consequently, clobetasol, one of the most potent topical steroids, has been recognised as the most efficacious treatment (1,3–5).

A limited number of case studies have reported effective treatment when steroids are combined with topical estrogen. The works of Scurrey et al., along with a few early studies, established that topical estrogen alone is unlikely to provide successful treatment of PCV (8,9). Preswood et al. documented the successful use of 0.01% estrogen in combination with hydrocortisone 100 mg suppositories for cases that had failed topical steroid monotherapy (7). The current case involves a less potent topical steroid, clobetasone, used in combination with estrogen, to successfully treat lesions for which betamethasone alone had failed.

Due to frequent misdiagnosis, most cases of PCV have been exposed to and failed several antimicrobial regimens prior to the commencement of topical steroids (3). Therefore, antibiotic treatment alone is not considered an appropriate monotherapy for PCV (5,8). However, antibiotics have proven useful when combined with topical estrogen and steroids, as demonstrated in both the current study and the work of Nyugen et al. In their 9-year cohort study, Nyugen et al. reported on 11 women diagnosed with PCV, 5 of whom experienced resolution of symptoms. Of those who experienced symptom resolution, all but one were treated with a combination of clobetasol, topical estrogen and topical clindamycin (3). One woman received systemic antibiotic therapy in addition to topical treatment, and one woman reported symptom resolution without treatment (3).

Lastly, there is sparse literature on the use of non-pharmacological management alongside pharmacological treatment for PCV. Our case suggests that such an integrated approach might be significant, reflecting standard treatment for related conditions such as plasma cell balanitis, which emphasises cleanliness and dryness as first-line care.

The potential overlap of PCV with other chronic dermatological conditions, such as concomitant vulvar lichen sclerosis reported by Yun et al., and its implications for treatment response are also noteworthy (1–3,5,8). Our case, featuring a previous history of lichen sclerosis, contributes to the limited literature on this subject, providing insight into the management of the condition when coexisting with other dermatological conditions (3,8,9).

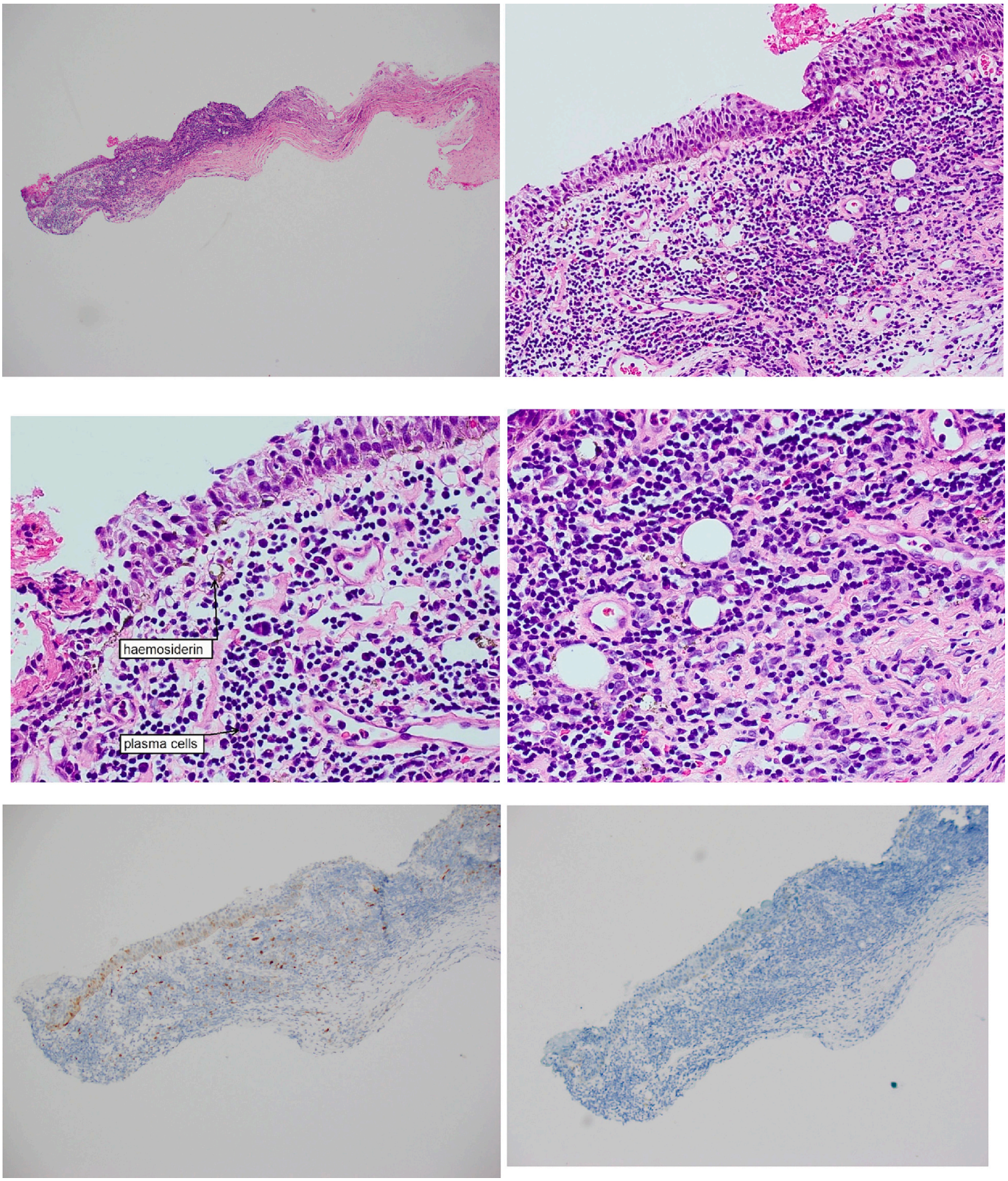

Fig. 2. Vulval biopsy histopathology, with images arranged clockwise from the top left: low-power x40, medium-power x200 and high-power magnification x400. Lymphocytes, histiocytes and occasional eosinophils are also present. Bottom right, p16 immunohistochemistry (ICH) negative. Bottom left; spirochete ICH and PAS-D staining for fungal organisms are all negative. Middle left; high-power magnification with arrows on the high-power image reveal slight atrophy of the squamous mucosa, which is non-keratinising. Heavy chronic inflammation characterises the underlying stroma, featuring a large number of plasma cells identifiable by 'pushed aside' nuclei and clockface chromatin. Hemosiderin deposits are indicated by brown pigmentation (arrows). Finally no HPV dysplasia is evident in any of the slides. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

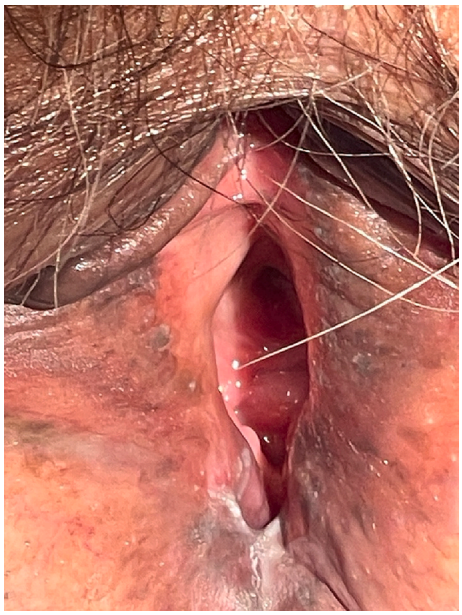

Fig. 3. Six weeks after the start of treatment, the complete resolution of the lesion depicted in Fig. 1.

4. Conclusion

Plasma cell vulvitis remains a rare and underreported cause of vulval ulceration, presenting significant challenges in both diagnosis and treatment. This case adds valuable insight to the limited literature on the condition, not only demonstrating the rare occurrence of PCV presenting as chronic vulval ulceration, but also illustrating how it can be successfully managed with a combination of topical antibiotics, estrogen and a mild steroid supplemented by non-pharmacological management stratergies. Comparison with existing case studies indicates a consensus on the efficacy of topical combination regimens in treatment, suggesting the potential for less potent steroids in combination therapies. There is an ongoing need for further research to establish standardised treatment guidelines, particularly for cases with atypical presentations and those that are refractory to monotherapy. Ultimately, enhancing awareness and documentation of such cases is crucial for improving the understanding and management of PCV.

Contributors

Lauren Fisher was involved in acquisition and interpretation of case information, undertaking literature review, drafting and revision of article.

Eman Alnaggar was involved in patient care, conception of the case report, drafting and revision of article for critical appraisal and providing important intellectual content.

Both authors approved the final submitted manuscript.

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Patient consent

Informed consent was obtained from the patient, and confirmed by her legal representatives, for the publication of this case report.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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