Plasma cell vulvitis: a review of two cases

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Summary

Zoon first described plasma cell balanitis in 1950 and since then there have been many cases described. The equivalent condition in women is plasma cell vulvitis (PCV), which is less frequently reported in the literature (1, 2). It provides a diagnostic and therapeutic challenge, with large numbers of women receiving a delayed diagnosis and inappropriate treatments (3). The condition presents with pruritus, pain, dyspareunia and burning in the vulval area (3-5). Examination may reveal erythematous patches, erosive lesions, pinpoint petechial and purpuric spots (4, 5). Histological features of the condition include the predominant feature of subepidermal plasma cell infiltrates in a band-like pattern, epidermal thinning which is thought to be a result of reduced size and quantity of keratinocytes and reduced size of the horny and granular cell layers. Keratinocytes become diamond or lozenge shaped (5, 6). Plasma cells usually predominate in the biopsies taken (5, 6). Further non-specific features include spongiosis, erythrocyte extravasation, and vascular dilatation and haemosiderin deposits. Haemosiderin gives the lesions their characteristic red/brown appearance. Topical steroids have been shown to be effective in many cases (4-6).

Case 1

A 51-year-old peri-menopausal female presented to the Sexual Health Clinic at Royal Perth Hospital, with a one-year history of ulceration, pruritus and pain in the vaginal introitus. She also complained of dysuria and urinary frequency. She had not been sexually active for nine years. Examination confirmed an inflamed, 4 x 5 cm area at the vaginal introitus extending from the 5 o’clock to the 8 o’clock position. After an initial biopsy which showed non-specific inflammatory changes, a further biopsy showed squamous mucosa with a moderately dense inflammatory infiltrate in the stroma (Figure 1). The inflammatory infiltrate included relatively abundant plasma cells as well as lymphocytes and scattered eosinophils. Lichenoid inflammation was present, with infiltration of the basal layer of the epithelium by inflammatory cells associated with vacuolar change in some keratinocytes and spongiosis (Figure 2). Focally there was haemosiderin deposition in the stroma (not shown). There was no vasculitis or malignancy. Findings were consistent with a diagnosis of PCV. The patient was commenced on betamethasone dipropionate (0.05%) cream twice a day for three weeks and her symptoms improved significantly. Some five months later there was a small area of clinical recurrence in the vaginal introitus (Figure 3), which was asymptomatic. She was again commenced on betamethasone dipropionate (0.05%) twice daily and has not returned for follow up.

Introduction

Zoon first described plasma cell balanitis in 1950. Plasma cell vulvitis (PCV) is the equivalent condition in women, which is less frequently reported in the literature (1, 2). It provides a diagnostic and therapeutic
Case 2

A 79-year-old female presented to a private Dermatologist, with pain in the vulval area with severe dysuria, pruritis, frequency and urinary incontinence. She had attributed her symptoms to a course of antibiotics for a respiratory infection. Previous treatment included ketoconazole cream, sodium bicarbonate douches, vitamin A cream, nappy cream and hydrocortisone 1% cream (Figure 4).

Examination confirmed a marked inflammatory eruption on the labia majora and minora. Initially treatment involved betamethasone dipropionate (0.05%) ointment twice daily. Within one week there had been partial resolution but the labia minora, especially the lateral aspect, remained inflamed.

Vulval biopsies showed mild epithelial spongiosis and a sub-epithelial infiltrate of plasma cells (4). Stains for pathogens including treponema were negative.

After two months of treatment with various strengths of topical corticosteroids, including betamethasone dipropionate (0.05%) ointment, methylprednisolone aceponate 0.1% ointment and finally betamethasone dipropionate (0.05%) OV ointment there was near-complete improvement of signs and symptoms with only a small residual area at the right labia minora. A year later the patient had a recurrence of severe symptoms that failed to improve with betamethasone dipropionate OV ointment application. She was hospitalised for four times daily wet dressings with betamethasone dipropionate 0.05% ointment and emollient. After 11 days of treatment there was resolution and she was discharged. However the symptoms recurred despite ongoing topical corticosteroid therapy. She was commenced on tacrolimus 0.1% ointment and had significant improvement within two weeks of treatment. She continued on treatment for approximately two years and remained asymptomatic. She now uses intermittent tacrolimus ointment for occasional flares which settle promptly.
Discussion

PCV is a rare condition, representing less than 2% of chronic vulval symptoms, with unknown incidence and aetiology, occurring in cases aged between 8-80 yrs (6). Several triggers have been hypothesized, including friction, warmth, poor hygiene and herpes simplex infection (2, 4-6). Differential diagnoses include eczema, lichen planus, immunobullous disease, fixed drug eruption, Paget’s disease, squamous cell carcinoma, herpes simplex virus infection and candidiasis (4).

Many different treatments have been reported but they are limited to individual cases and case series leading to difficulty in optimising therapy (5-8). Topical corticosteroids, vaginal oestrogen, oral conjugated estrogens, medroxyprogesterone acetate and local excision showed some promise in four cases (9). PCV responded well to topical corticosteroids in a further four cases (10). Tacrolimus 0.1% ointment treatment was associated with disappointing clinical and histological improvement, in contrast to that shown in Zoon’s balanitis (5). Three cases have been successfully treated with misoprostol (11). Two cases were managed surgically after being refractory to all treatment modalities (12). Combination treatment using antibiotics and topical steroids achieved resolution of two cases and a further case after surgical excision (13). In another case series, no significant difference was found between various topical interventions (fusidic acid and topical corticosteroids, topical corticosteroids alone and tacrolimus) (14). Other treatments described include systemic immunomodulating drugs, and physical therapies such as cryotherapy, intralesional injections and CO2 laser ablation. Generally treatments were more effective in reducing symptoms than signs in patient with PCV (14).

PCV should be considered in patients with vulval symptoms who fail initial therapy and continue to have pruritus, pain or burning (4). They may have non-specific initial biopsy results and swabs for pathogens will be negative. The mainstay of treatment is topical steroids, but relapse or non response poses a clinical dilemma for ongoing treatment (4, 7). An appropriate therapeutic pathway has not yet been developed for PCV, as well as agreed end-points for treatment cessation. In our cases one patient responded more typically to conventional treatment and the second patient was more refractory but responded well to tacrolimus which has had variable results in the literature. More research is warranted to further define this condition and its optimal treatment.

References