A rare cause of vulval swelling

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DESCRIPTION

A 46-year-old woman presented to the emergency department with a 6-month history of gradually worsening vulval swelling, yellow discharge and excruciating pain on movement. She had previously attended her general practice, as well as her local urgent care centre, where she received multiple courses of oral antibiotics for presumed folliculitis and a course of oral anti-fungal for presumed thrush without improvement. On her latest presentation, the patient was assessed by a gynaecologist in the emergency department. After review, an urgent referral was made to dermatology.

On further assessment, the patient did not report any systemic symptoms. Medical history was significant for polycystic ovary syndrome and she did not take any regular medications. Family history was unremarkable for autoimmune conditions. Clinical examination revealed gross vulval oedema with erythema and multiple superficial erosions (figure 1). Oral ulcers, gum-line erythema and hard palate erosions were noted, as well as small, discrete, crusted ulcerations on the abdomen. Laboratory investigations revealed a raised erythrocyte sedimentation rate. A sexually transmitted disease screen, viral swabs and bacterial cultures were negative. Histological analysis showed an intraepidermal split at a suprabasal level with acantholytic keratinocytes (figure 2). Intracellular epidermal deposits of IgG and C3 were also shown on a direct immunofluorescence assay, confirming a diagnosis of pemphigus vulgaris.

The patient was initially treated with high-dose prednisolone and mycophenolate mofetil. Despite this, the disease progressed. Rituximab, a chimeric monoclonal anti-CD20 antibody that induces depletion of B cells in vivo was added 3 months later. Six months post-therapy-initiation, the patient remains in remission.

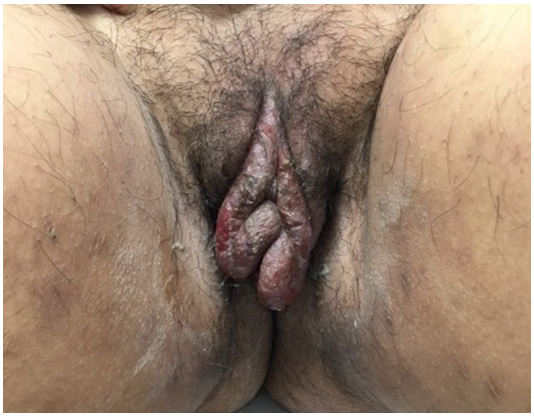

Figure 1 Severe vulval oedema, swelling and erythema with multiple superficial erosions.

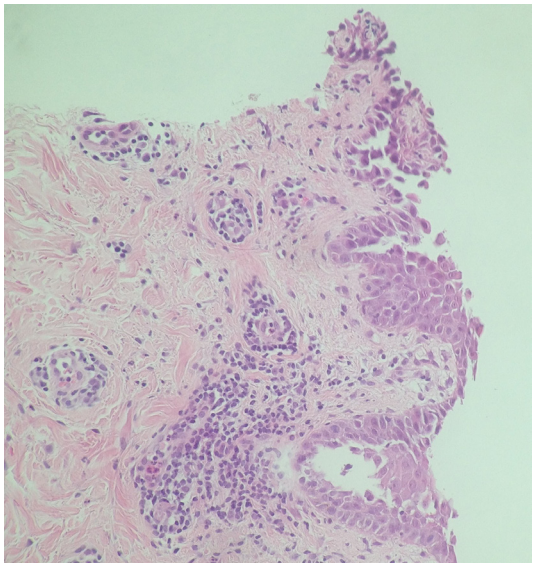

Figure 2 Skin biopsy showing intraepidermal and suprabasal split of the epidermis, acantholytic keratinocytes and perivascular infiltrate of inflammatory cells.

Pemphigus vulgaris is a rare, autoimmune blistering condition. It is characterised by the loss of cellular adhesion in the outer layer of skin and mucosa. This is caused by the development of IgG autoantibodies against desmoglein 1 and/or 3.[1] Varying frequencies of genital involvement in pemphigus vulgaris have been reported.[2] Although genital involvement usually occurs when other sites are involved, there have been reports of isolated genital involvement with pemphigus vulgaris.[2 3] Differential diagnoses for vulval pemphigus vulgaris includes bullous pemphigoid, bullous tinea, Behcet's disease, herpetic stomatitis and extra-cutaneous manifestations of granulomatous diseases, such as Crohn's disease. Pemphigus vulgaris involving the vulva is rare and requires a high index of suspicion with prompt specialist referral.

Learning points

- Clinicians should consider immunobullous diseases, such as bullous pemphigoid and pemphigus vulgaris as a cause for chronic mucosal lesions at genital sites.
- Clinicians should consider urgent referral to dermatology in cases of unresolving vulval erosions.

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