Bullous Pemphigoid of the Perineum and Perianal Area: A Rare Localized Form in Adults

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Sir,

A 72-year-old woman presented with a persistent eruption of the perineum and perianal area, which consisted of pruritic erythematous plaques and erosions with purulent discharge. The patient was treated with systemic and topical antifungal agents for 2 months, with no signs of improvement. She was obese and reported itch in this area for more than a year, as well as intertriginous lesions during the summer.

Histological examination revealed a subepidermal cleft filled with numerous eosinophils and few neutrophils. Perilesioual direct immunofluorescence (DIF) was typical of bullous pemphigoid (BP) with linear deposits of C3 and IgG at the basement membrane zone (BMZ). By indirect immunofluorescence (IIF) a linear fluorescence of C3 was also observed and with salt-split skin test on monkey oesophagus substrate, there was a clear fluorescence on the epidermal side of the artificial blister. A BP180NC16A enzyme-linked immunosorbent assay (ELISA) test was positive with a value of 14.7 U/ml (cut-off 9 U/ml). A BP230 ELISA test, however, was negative. Direct examination and culture of purulent discharge was negative for bacteria or fungi. Histology, DIF, IIF and BP180ELISA findings established a diagnosis of BP.

The patient was treated with oral prednisolone 0.5 mg/kg/day for 2 weeks with gradual tapering of dose and tacrolimus ointment applied twice a day for a period of one month and once afterwards. No rebound after one year on maintenance therapy and no residual scarring have occurred. Itch is rarely reported.

Localized bullous pemphigoid (LBP) is a form of BP that requires a high index of clinical suspicion, as it presents with quite atypical clinical pictures. It accounts for 16–29% of all cases of BP, but the true incidence may be greater as it is often misdiagnosed (1).

In children it presents on the palms and soles or on the genitalia (childhood vulvar pemphigoid) (2). In adults, LBP occurs predominantly on the shins of elderly women, but it has been reported to the soles (3), around sites of trauma, irradiation (3) or psoralen + ultraviolet A (PUVA) therapy, on peristomal areas or fistulas (4).

As a possible pathogenetic mechanism for LBP, in most studies, an epitope spreading phenomenon on the BMZ is thought to be induced by trauma, thermal burn or chronic irritation. Proteins, components of BMZ, are exposed to an autoimmune response, leading to the formation of blisters. In our case chronic itch, caused by chronic intertrigo or lichen sclerosus and atrophicus in early stages, could probably induce an antiBP180NC16A response. In a recent study, weak circulating BMZ antibodies were detected by immunoblotting in 61% of patients with erosive lichen planus of the vulva, providing evidence for an autoimmune response in this disorder as well (5).

Differential diagnosis in LBP of the perineum and perianal area includes lichen sclerosus and atrophicus, pemphigus vulgaris, irritant dermatitis or intertrigo. The clinical course, histological and immunofluorescence findings usually help to distinguish LBP from the above clinical entities.

First-line therapy in localized forms of BP includes steroids, systemic or very potent topical ones. Tacrolimus ointment has been reported to be an effective alternative in the management of BP (6). This topical immunomodulator is known to inhibit T-cell activation, and, as T cells have a determining role in the pathogenesis of BP, it is an efficacious regimen without the adverse effects of topical steroids. It is also important that tacrolimus does not induce Candida infections in the genital area. In our patient, a combination of systemic prednisolone for a short period and maintenance treatment with topical tacrolimus proved beneficial.

Prevalence of localized forms of BP would probably increase as the mean survival rate increases. Every case of itch in elderly patients must be examined fully and diagnosed early, in order to avoid prolonged and inappropriate therapeutic modalities.

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