A case of Bullous Pemphigoid Associated with Lichen Sclerosus et Atrophicus

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Sir,

We encountered a 66-year-old woman with bullous pemphigoid (BP) that developed on lichen sclerosus et atrophicus (LSA) lesions.

The patient suffered from LSA for 12 years. While LSA most commonly affects the vulval area, her LSA involved the trunk, extremities, and the anogenital skin.[1] Topical treatment with corticosteroids and tacrolimus yielded limited success.

Tense bullae appeared on the sclerotic LSA lesions in the genital region 12 years after the diagnosis of LSA [Figure 1a and b]. The bullous lesions spread to the skin on the abdomen and chest affected by LSA. Her serum BP antibody, BP180, was 46.9 U/ml (chemiluminescence enzyme immunoassay). Skin biopsies from her abdomen and genitalia revealed subepidermal cleft with predominant eosinophilic infiltration [Figure 1c and d]. In the upper dermis, we noted pronounced edema and collagen homogenization [Figure 1c and e]. Direct immunofluorescence study of the same specimens revealed linear deposits of immunoglobulin G in the basement membrane zone [Figure 1f]. The above findings led to the diagnosis of BP arising on LSA. Oral prednisolone (PSL, 30 mg/day) was started. The bullous lesions improved quickly, and her autoantibodies became negative. The PSL dose was gradually tapered to 10 mg, and she suffered no relapse in 1 year.

Although the etiology and pathogenesis of LSA are not fully understood, they are thought to be multifactorial with the involvement of autoimmune-related factors. [2-4] Our review of the English literature found only three cases of BP associated with LSA. [2-4] Meyrick Thomas et al.[5] who investigated autoimmune-related phenomena in 350 women with histologically confirmed LSA demonstrated that 21.5% had one or more autoimmune diseases including alopecia areata (9%), vitiligo (6%), hypothyroidism (6%), and BP (1%); 42% presented with an autoantibody. With respect to autoantibodies against BP antigens, Baldo et al.[1] reported that the noncollagenous 16A domain of BP180 was a target for circulating T-cells in over 40% of their patients with vulval lichen sclerosus or lichen planus; 4 of 19 manifested associated autoantibodies to BP180. A lichen sclerosus cohort study of 149 patients revealed elevated anti-BP antibodies in 5 (3.4%), BP180 in 4, and BP230 in one patient.[6] These studies suggested an immunogenetic connection between LSA and anti-BP autoantibodies. Ours was a rare case of BP with LSA and further studies to examine the pathological role of anti-BP autoantibodies produced in LSA patients who develop associated BP are needed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and anonymity cannot be guaranteed.

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Nil
Conflicts of interest

There are no conflicts of interest.

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