Atypical Variant of Bullous Pemphigoid
—Prolonged Eruptions of Papulourticarial Lesions—

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A 63-year-old woman had a large number of pruritic papulourticarial eruptions on her abdomen and extremities for 6 months, with only a single tense vesicle on her arm. The biopsy specimen from a papular lesion showed foci of microvesicles at the dermoepidermal junction, and an abundance of eosinophilic infiltrate. Immunofluorescent studies showed linear deposits of IgG, IgE, and C3 at the basement membrane zone (BMZ) and circulating anti-BMZ antibodies in her serum, consistent with bullous pemphigoid.

This case can be distinguished from most of the other variant of bullous pemphigoid, particularly in the aspect of clinical presentation. It further emphasizes the concept that bullous pemphigoid represents a spectrum of bullous disease with heterogeneous clinical features. (Ann Dermatol 1:33—36, 1989)

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Bullous pemphigoid (BP) is a wide spectrum disease that can affect the skin and mucous membranes. Clinically, most patients with BP present with large tense bullae. However, clinical variants of vesicular, erythematous, localized, erythrodemic, nodular, seborrheic, and vegetans forms may occur. The degree of pruritus is variable in this disorder and ranges from severe to nonexistent. The patient reported herein has an unusual variant of immunopathologically confirmed BP who presented long lasting pruritic papulourticarial lesions which were not considered to be a prodromal symptom.

REPORT OF A CASE

A 63-year-old woman visited the dermatology clinic at Hanyang University Hospital in November 1987, complaining of a pruritic papular dermatosis on her abdomen and arms. She first noted several erythematous papular eruptions on the periumbilical area 6 months before her visit. The lesions had gradually spread on her abdomen and arms over the ensuing weeks. She had noted only a few vesicular lesions developing sporadically. The patient had taken antihistamines and applied topical corticosteroids for the pruritic symptoms with some benefit. She was in good health prior to developing the skin lesions. She had no drug history relevant to her skin disease, nor did she have any systemic disease or infection at the time of her visit. Her family history and past history revealed nothing of significance.

Physical examination of this normotensive, thin woman, weighing 40 kg, was essentially unremarkable except for the pruritic skin condition. On her abdomen, inner and extensor arms, and parts of her inner thighs a large number of erythematous, relatively flat papules with a somewhat urticarial nature were dispersed (Fig.1). Each papular lesion was rather firm and measured 2 to 6 mm in diameter. Nikolsky's sign was not seen on these papular lesions. The patient had only one tense vesicle measuring 4 mm on her left forearm which initially erupted as a tiny clear vesicle on an erythematous papule. The mucous membranes were not involved, and there were no lymphadenopathies.

Histologic examination of a skin specimen obtained from a papular lesion on the abdomen showed irregular acanthosis and an occasional spongiosis of
subepidermal blister containing many eosinophils and neutrophils.

Laboratory studies, including a complete blood cell count with differential, were within the normal ranges except for a 21% reading of eosinophils. The erythrocyte sedimentation rate was elevated to 44 mm/hr. A urinalysis, stool examination, and roentgenogram of the chest were normal. The pattern of serum protein electrophoresis, the values of liver, thyroid, and kidney function tests, fasting blood sugar level, serum concentration of immunoglobulins (C3, C4, and MCH50), C3 and C4 concentrations, ASLO, VDRL, antinuclear antibody, alpha-1 antitrypsin level, and C-reactive protein were all within the normal limits or negative. The serum examination for cryoglobulin was positive, and the serum IgE level was slightly elevated to 620 IU/ml. Intradermal skin tests with recall antigens of PPD and Trichophyton showed positive reactivities.

A preliminary diagnosis of bullous pemphigoid was made, and confirmed by immunofluorescence (IF) studies. The patient was treated with a regimen of systemic prednisolone (25mg/day) and azathioprine (75mg/day), as well as antipruritic topical preparations. The skin lesions soon subsided without scarring and she became symptom free in 3 weeks. Three months later, when clinical remission of the skin lesions was evident, she could manage with only a minimum dose of prednisolone and 50 mg of azathioprine a day.

### IF Studies

Direct and indirect IF studies were performed according to the established procedures, using commercially obtained monospecific fluorescein-conjugated antisera (Meloy Laboratories, Inc., Springfield, VA, U.S.A.). Direct IF studies of papular and peribullous specimens demonstrated linear deposits of IgG, IgE, and C3 along the basement membrane zone (BMZ) (Fig. 2). The intensity of IgE staining was weaker than IgG, but recognizable. Indirect IF studies of the serum, using normal human flank skin as the substrate, detected anti-BMZ IgG and IgE antibodies, at a titer of 1:40 and 1:10 respectively. The patient did not have IgA class autoantibody to the BMZ.

Another system of indirect IF studies, using 1.0 mol/L sodium chloride split intact skin preparations...