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PHOTOLETTER TO THE EDITOR

An eczema-like, pruritic, nonbullous form of bullous pemphigoid

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Abstract

We report a case with an unusual manifestation of bullous pemphigoid (BP) in a 57-year-old woman. She presented with a one-month history of pruritus and multiple annular patches with central regression on her trunk and extremities and there were no signs of corresponding internal malignancy. Eosinophilic spongiosis was observed in histopathologic examination and direct immunofluorescence studies revealed a strong linear staining pattern of the basement membrane zone with C3 and IgG. This unusual case indicates that a lack of blisters does not necessarily exclude a diagnosis of BP, and supports previous reports suggesting gyrate erythema can be an initial manifestation of bullous disease. (*J Dermatol Case Rep*. 2015; 9(2): 55-57)

Key words:

bullous pemphigoid, eczema, nonbullous, pruritus

Bullous pemphigoid (BP) is a chronic autoimmune bullous disorder most commonly seen in the elderly. Patients typically present with large, tense bullae in a generalized distribution favoring the flexural areas. However, there are various forms of BP, unlike the name of "bullous" pemphigoid (Table 1). Thus, clinical variants of BP were also various including localized, vegetating, dyshidrosiform, vesicular, erythrodermic BP, and pemphigoid nodularis, as well as childhood BP and pemphigoid sine bullae.[1]

A 57-year-old Korean woman presented with a one-month history of pruritus and multiple annular patches on her trunk and extremities. On physical examination, erythematous lesions with central regression were found covering her whole body except the head and neck area (Fig. 1). Two years prior she experienced a similar eruption on the skin of her abdomen and arms, which responded to topical corticosteroid treatment. She had no medical history, and there were no signs or symptoms of corresponding internal malignancy. Serum analysis revealed an elevated

Table 1. Various forms of bullous pemphigoid.

	Figure of lesion
Classic form	Erythema based large, tense blister
Non bullous forms	Eczematoid
	Serpiginous
	Targetoid
	Erythroderma
	Prurigo nodularis-like
	Vegetating
	Dyshidrotic dermatitis-like

WBC titer of 13.97 × 109/L (normal, 3.0-9.0 × 109/L) and eosinophil count of 830/mm^3 (normal, 0-300/mm^3). Histopathologic examination of a lesional skin biopsy specimen revealed mild parakeratosis and acanthosis of the epidermis. Perivascular lymphocytic and eosinophilic infiltrates with slight edema were observed in the upper dermis as were foci of eosinophils in the epidermis (eosinophilic spongiosis) (Fig. 2A). Direct immunofluorescence studies revealed a strong linear staining pattern of

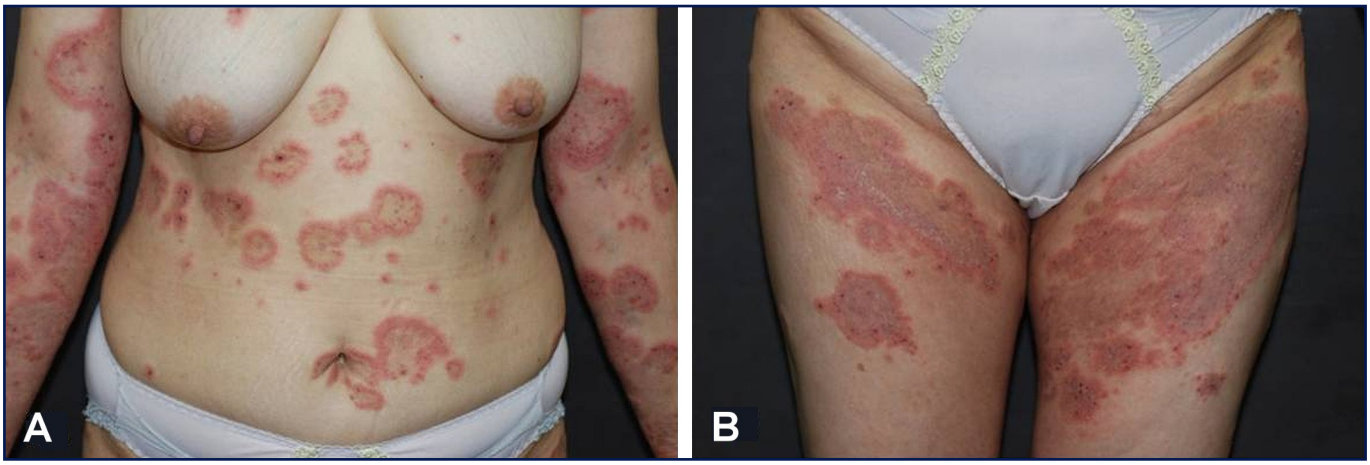

Figure 1

(A & B) Annular serpiginous migrating plaques with central clearing and a collarette-like scale on the trunk and extremities.

the basement membrane zone with IgG and C3 (Fig. 2B), however, the result of staining with IgA was negative. Immunoprecipitation revealed the presence of circulating autoantibodies against the 230 and 180 kDa antigens. So we finally diagnosed her as nonbullous form BP and treated her with 20 mg/d of methylprednisolone as starting dose, which was slowly tapered, and topical steroid for 3 weeks. Her skin lesions were resolved and no recurrence was observed during 1 year follow-up period.

BP presenting as an eczematous or urticarial eruption without blisters or with a lengthy prodromal period has been reported.[2,3] Nakatani *et al.*[4] reported a case of bullous pemphigoid preceded by prodromal eczematous eruptions that lasted an unusually long time of 11 years. The diagnosis of all BP subtypes is confirmed by immunofluorescence findings.[1] Here we presented a case with an unusual manifestation of BP. Clinically, the patient presented with generalized, sharply marginated, erythematous scaly patches and plaques. The clinical features mimicked lesions found in erythema annulare centrifugum or erythema gyratum repens. However, direct immunofluorescence microscopy findings were characteristic of BP, and circulating autoantibodies were found to recognize both BP180 and BP230. This case indicates that a lack of blisters does not necessarily exclude a diagnosis of BP, and supports previous reports suggesting gyrate

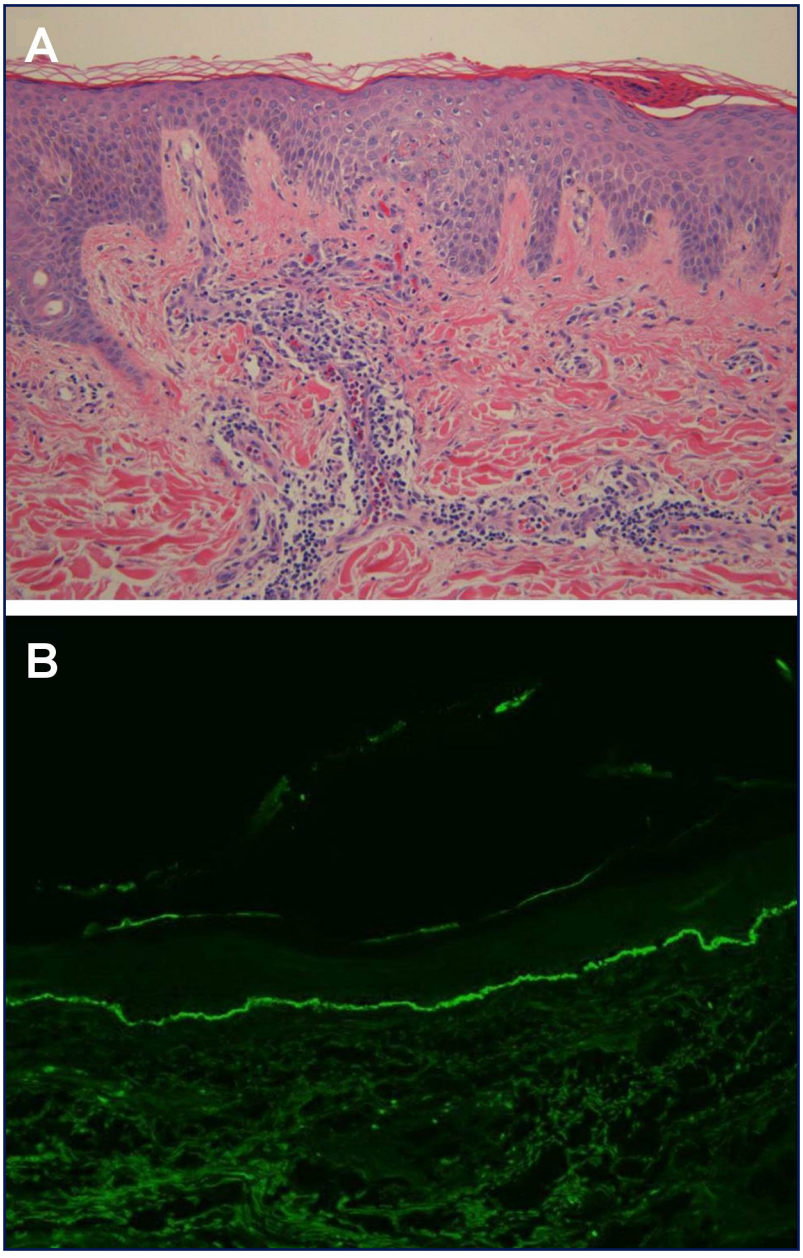

Figure 2

(A) Lesion histology showing foci of eosinophilic spongiosis and perivascular lymphocytic and eosinophilic infiltrates. (B) Direct immunofluorescence of a perilesional skin biopsy showing linear deposits of IgG along the dermoepidermal junction and on the surface of basal keratinocytes.

erythema can be an initial manifestation of bullous disease.[5] BP can also present as gyrate erythema with or without malignancy, and it should not be ignored by physicians, and further studies are needed to elucidate the immunopathologic mechanisms that determine various clinical manifestations of BP. BP should be kept in mind as differential diagnosis in atypical, long-lasting nonbullous pruritic lesions. That in elderly or middle aged patients any pruritic rash persisting for few months should be investigated by immunofluorescence and / or ELISA for BP antigens to rule out pemphigoid even if clinically or histologically no blisters are seen.

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