Brunsting-Perry pemphigoid transitioning from previous bullous pemphigoid

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Key words: Brunsting-Perry pemphigoid; bullous pemphigoid; epitope spreading.

INTRODUCTION

Brunsting-Perry pemphigoid is a variant of cicatricial pemphigoid, bullous pemphigoid, or epidermolysis bullosa acquisita that is commonly characterized by the presence of atrophic scars, with little or no mucosal involvement, and affects the head, face, neck, and upper trunk.[1] Here, we report a case of Brunsting-Perry pemphigoid after remission of previous bullous pemphigoid.

CASE REPORT

A 63-year-old Chinese man was referred to our hospital in August 2013 with a 3-month history of pruritic lesions. Scattered tense vesicles and bullae appeared on his hands (Fig 1, *A*), upper limbs, and trunk. The patient received a skin biopsy, which showed a subepidermal bulla as well as superficial inflammation in the upper dermis, with eosinophil infiltration. Direct immunofluorescence studies showed linear deposition of IgG and C3 along the basement membrane zone of the peribullous area. An enzyme-linked immunosorbent assay for IgG antibodies against the BP180 NC16A domain resulted in a value of 90.44 (index >15: positive) and of 33.75 (index >9: positive) for BP230 (Fig 1, *B*). In accordance with the typical clinical features, immunopathologic findings, and specific antibodies, he received a diagnosis of bullous pemphigoid and was treated with 30 mg prednisone per day. The skin lesions gradually subsided, and prednisone was tapered and finally stopped after 2 years. After the patient stopped receiving prednisone, he experienced occasional recurrences that were quickly relieved with topical steroids.

In July 2017, he revisited our department, with a 7-month history of lesions. Clinical examination revealed crusts and ulcers on the parietal scalp, with scarring alopecia (Fig 2, *A*). Ulcers were occasionally observed on the oral mucosa. Indirect immunofluorescence using sodium chloride−split (at 1 mol/L) healthy human skin sections revealed that there were anti−basement membrane zone antibodies bound to the roof of the split (Fig 2, *B*). The enzyme-linked immunosorbent assay index value for BP180 NC16A was 25.45 and was 110.82 for BP230 (Fig 1, *B*). The enzyme-linked immunosorbent assay results for type VII collagen were negative. Direct immunofluorescence showed no IgG, C3, immunoglobulin M, or IgA deposition at the basement membrane zone, which may be because of the repeated erosion and scar formation of the scalp. In accordance with the clinical features and immunopathologic findings, the patient received a diagnosis of Brunsting-Perry pemphigoid. He received oral prednisone (10 mg/d), debridement (Fig 3, *A*), and wet packing with dexamethasone at 0.1 mg/mL, which adequately controlled the lesions (Fig 3, *B*). A dosage of prednisone of 5 mg/d was maintained by the patient for 1 year; the antibody titer was 5.89 for BP180 NC16A and 28.39 for BP230.

DISCUSSION

To date, the pathogenesis of Brunsting-Perry pemphigoid remains to be elucidated. Traditionally, the involvement of the BP180 C-terminal domain in the formation of the scarring phenotype of cicatricial pemphigoid has been suspected.[2] However, a recent review on Brunsting-Perry pemphigoid suggests

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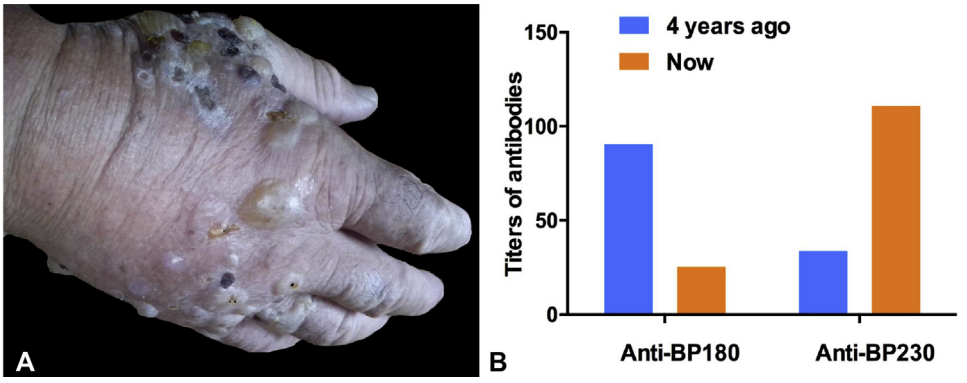

Fig 1. A, Appearance of vesicles, bullae, and crusts on the right hand at the first visit. **B**, Autoantibody against BP180 NC16A was dominant 4 years ago, but 4 years later, BP230 was dominant according to the enzyme-linked immunosorbent assay.

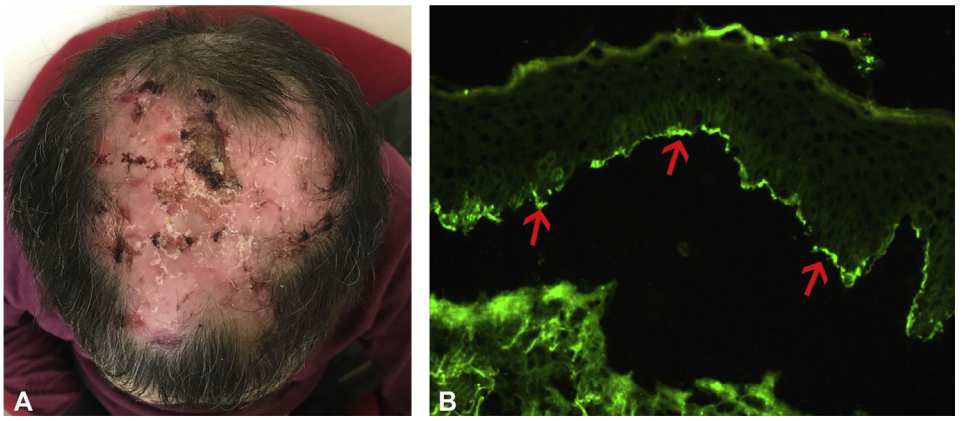

Fig 2. A, Crusts and trophic scars on the parietal region, with hair loss. **B**, Indirect immunofluorescence on sodium chloride–split (at 1 mol/L) skin revealed that IgG antibodies were bound to the roof of the split.

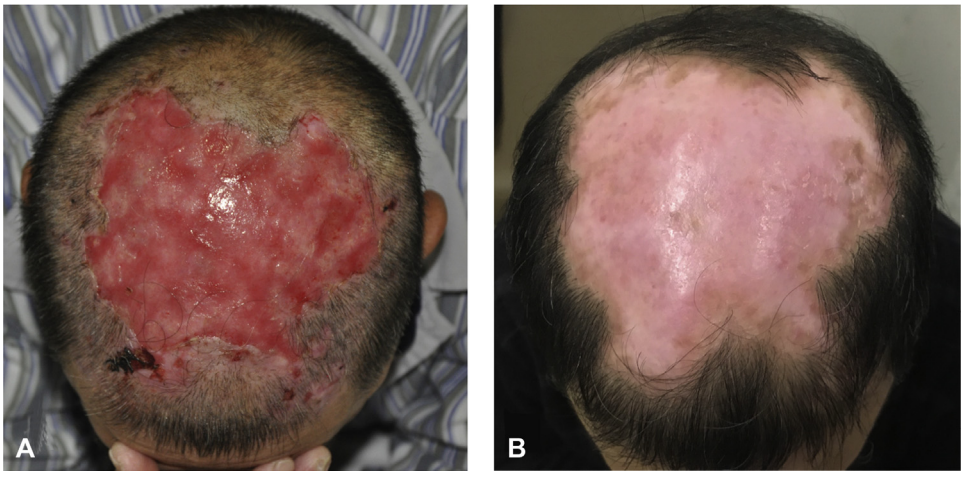

Fig 3. A, Necrotic crusts were removed before topical treatment. **B**, Lesions were relieved, leaving trophic scars.

otherwise; autoantigens such as BP180 (both NC16A and C-terminal domains), type VII collagen, laminin 332, BP230, and LAD-1 have been reported to be associated with Brunsting-Perry pemphigoid. The BP180 C-terminal domain is not essential in the pathogenesis of Brunsting-Perry pemphigoid.[3,4]

We report an unusual case of bullous pemphigoid that transformed into a typical Brunsting-Perry pemphigoid in a 63-year-old man. Analysis indicated a switch in the dominant IgG anti—basement membrane zone antibody from anti-BP180 NC16A to anti-BP230. To the best of our knowledge, this is the first account of this phenomenon. With respect to previous reports, this rare transition from bullous pemphigoid to Brunsting-Perry pemphigoid may be due to the spreading of the intermolecular dominant epitope from BP180 NC16A to BP230.[5,6] Further investigation is needed to discern the mechanism.

In terms of clinical treatment, topical steroids and low-dose oral prednisone were effective in this patient. Topical and oral steroids are traditional methods for treating Brunsting-Perry pemphigoid; other therapies include azathioprine, intralesional steroids, dapsone, and doxycycline/niceritrol.[3]

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