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LETTER TO THE EDITOR

Journal of Cutaneous Immunology and Allergy
WILEY

Anti-BP230 antibody–positive bullous pemphigoid complicated by ulcerative colitis

Dear Editor,

Bullous pemphigoid (BP) is characterized by tense blisters and erythema; BP autoantibodies react with the BP180 and/or BP230 antigens. Anti-BP230 antibody–positive BP is an autoimmune disease that causes subepidermal blistering and is associated with the production of autoantibodies against the intracellular plaque protein, BP230, which accounts for 5%–8% of all BP.[1] Here, we have recorded a rare case of anti-BP230 antibody–positive BP, which developed subsequently to ulcerative colitis (UC).

A 12-year-old boy with UC was treated with 3000 mg of mesalazine daily for two months, followed by 30 mg of prednisolone daily. When the prednisolone was tapered to 5 mg daily, pruritic edematous erythema appeared over the entire body surface. Despite being treated with a topical steroid for one month, erythemas were further distributed across the patient's body. Upon examination, tense blisters of 1–3 cm in diameter were found on his right upper arm and right foot (Figure 1A,B). He had no past history. The patient's mother had also been diagnosed with UC.

A biopsy was taken from the erythema on his left thigh, and direct immunofluorescence (IMF) revealed liner deposition of C3 on the basement membrane zone (BMZ) (Figure 1C). BP180 NC16a was not detectable through chemiluminescent enzyme immunoassay (MBL, Nagoya, Japan), but an IgG enzyme-linked immunosorbent assay (ELISA) for BP230, which was examined in the manner described by Hashimoto et al,[2] was positive (index value 26.5, cutoff 9.0). Based on these findings, a diagnosis of BP230-type BP associated with UC was made. Daily administration of 10 mg (0.25 mg/kg) of oral prednisolone improved his skin lesions, with the prednisolone dosage gradually tapered to 1 mg daily. He has had no recurrence for over 2 years.

To our knowledge, 28 cases of linear IgA bullous dermatosis (LAD) and 24 cases of BP180-type BP associated with UC have been reported.[3–9] Therefore, this is the first case of BP230-type BP associated with UC. According to previous reports, the patients all developed UC before BP, with a range of 6 months to 23 years between diagnoses. In contrast to BP180, the pathogenic relevance of autoantibodies against BP230 remains elusive,[1] and it has been reported that BP230-type BP tends to present with milder clinical phenotypes than does BP180-type BP. Additionally, blisters are not consistently present with BP230-type BP. In this case, edematous erythemas, rather than tense bullae, were the primary symptom. It is unclear whether the association between BP230-type BP and UC is significant or coincidental. BP230, as well as BP180, is expressed in intestinal epithelial cells[10]; therefore, we speculate that sensitization to those proteins in the inflamed intestinal epithelia might lead to a production of anti-BP180/230 autoantibodies, though further studies are needed to elucidate the pathophysiology of BP associated with UC.

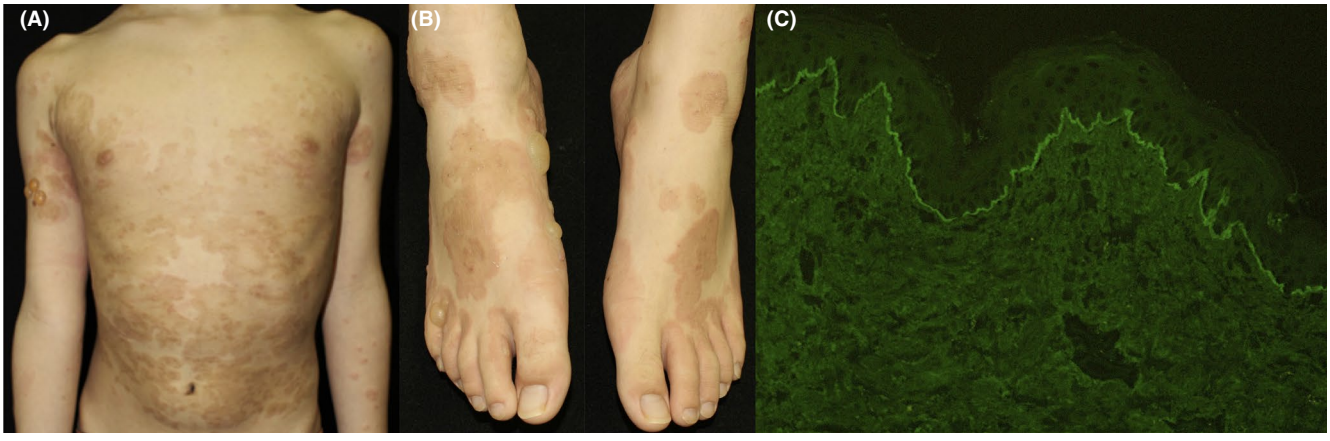

FIGURE 1 Edematous erythema distributed over the entire body surface, with tense bullas on the patient's right upper arm and right foot (A, B). Linear deposition of C3 on the basement membrane zone (C)

CONFLICT OF INTEREST

Dr Daisuke Tsuruta is a member of the Journal of Cutaneous Immunology and Allergy Editorial Board. Management of the peer review process, and all editorial decision-making, for this article was undertaken by Editor in Chief, Yoshiki Tokura who managed this article.

[Correction added on 10 September 2019, after first online publication: Conflict of Interest statement has been updated.]

APPROVAL OF THE RESEARCH PROTOCOL

N/A.

INFORMED CONSENT

Written informed consent was obtained from the patient.

REGISTRY AND THE REGISTRATION NO. OF THE STUDY/TRIAL

N/A.

ANIMAL STUDIES

N/A.

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