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LETTER TO THE EDITOR

Case of papuloerythroderma of Ofuji-like eruption during the course of bullous pemphigoid

Dear Editor,

Deck-chair sign indicates a selective sparing of skin folds observed characteristically in papuloerythroderma of Ofuji (PEO).[1,2] This sign can also be observed in acanthosis nigricans, erysipelas, discoid lupus erythematosus, acute contact dermatitis, angioimmunoblastic lymphoma, cutaneous Waldenström's macroglobulinemia and leprosy.[2,3] However, this sign associated with bullous pemphigoid (BP) has not been reported. We encountered a case with erythroderma sparing skin folds during the course of BP.

A 75-year-old Japanese man was referred to us with a 2-year history of pruritus, edematous erythema and vesicular papules on the trunk, extremities and palmar/plantar surfaces except for the skin folds (Fig. 1a). He had undergone gastric resection for gastric cancer 6 years earlier. Eosinophil percentage was 8.6% in the peripheral blood of which white blood cell count was 7600/μL. Chemiluminescent enzyme immunoassay revealed the titer of serum anti-BP180-NC16a-domain antibodies was more than 1000 U/mL (normal range, <9). Histopathological examination showed subepidermal blistering and eosinophil infiltration into the dermoepidermal junction (Fig. 1b). Direct immunofluorescent assay revealed linear deposition of immunoglobulin G and M along the dermoepidermal junction (Fig. 1c,d). BP was diagnosed. Administration of dipeptidyl peptidase-4 inhibitor that he had been taking for type 2 diabetes mellitus during those 2 years was

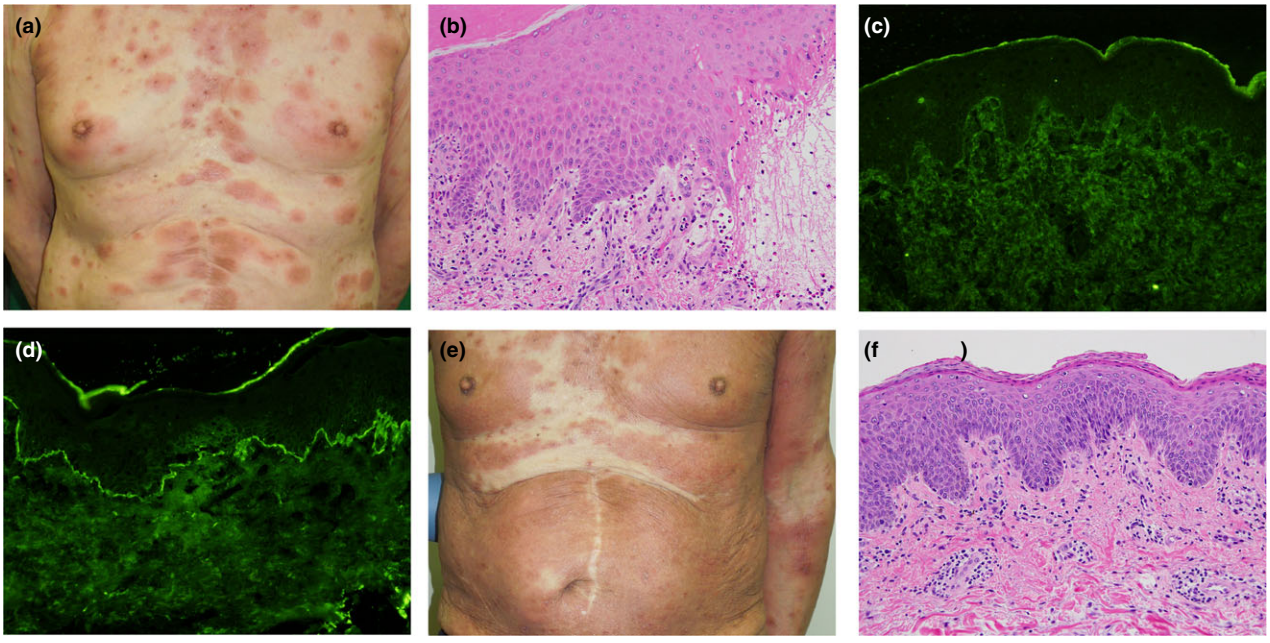

Figure 1. (a) Clinical findings at first visit. Edematous erythema and vesicular papules are evident on the trunk and upper extremities. (b) Histopathological findings at first visit. Subepidermal blistering and infiltration of eosinophils into the dermoepidermal junction are observed (hematoxylin–eosin [HE], original magnification ×200). (c) Findings of direct immunofluorescent assay using fluorescein isothiocyanate-conjugated anti-immunoglobulin G antibodies (Medical & Biological Laboratories, Nagoya, Japan). Linear deposition of immunoglobulin G in the dermoepidermal junction is seen (×200). (d) Findings of direct immunofluorescent assay using fluorescein isothiocyanate-conjugated anti-complement component 3 antibodies (Medical & Biological Laboratories). Linear deposition of complement component 3 is seen (×200). (e) Clinical findings after beginning treatments. Erythroderma sparing skin folds is apparent. (f) Histopathological findings of erythroderma with sparing of skin folds. Acanthosis, parakeratosis, spongiosis, vacuolar degeneration, incontinentia pigmenti histologica, individual cell keratinization and lymphocyte infiltration in the upper dermis are evident (HE, ×200).

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discontinued. Then, diabetes mellitus was controlled within a range of 5.9–7.5% of hemoglobin A1c by insulin preparation.

One course of steroid pulse therapy following daily systemic corticosteroid therapy of betamethasone at 0.05 mg/kg per day (2.5 mg/body per day) and topical steroid therapy using clobetasol propionate was performed, then one course of i.v. immunoglobulin therapy at 400 mg/kg per day for 5 days was added. After these, pruritic fresh erythema and vesicular papules subsided and the titer of anti-BP180-NC16a-domain antibodies decreased to 584 U/mL. The clinical manifestations changed to erythroderma sparing skin folds with mild pruritus (Fig. 1e). Eosinophil percentage was 4.0% in the peripheral blood of which white blood cell count was 9100/μL. Histopathological examination revealed acanthosis, parakeratosis, spongiosis, vacuolar degeneration, incontinentia pigmenti histologica, individual cell keratinization and lymphocyte infiltration in the upper dermis with no deposition of immunoglobulin (Fig. 1f). Two months later, the titer of anti-BP180-NC16a-domain antibodies decreased to 35 U/mL, at which point erythema had mostly resolved. Between onset and remission of erythroderma, the administrated drugs were not changed.

The major criteria for PEO are as follows: (i) erythroderma-like eruptions formed by coalescence of flat-topped, red-to-brown papules with cobblestone-like appearance; (ii) itch; (iii) deck-chair sign; (iv) histopathological exclusion of other skin diseases; and (v) absence of causative factors.[4] Because the present case showed no characteristic papular eruptions, the criteria were not fulfilled.

Because immunoglobulin in the part of the erythroderma did not deposit, this case may not match erythrodermic BP. Histopathological eczematous changes suggest that scratch behavior might have been associated with the erythroderma. To detect the histopathological findings specific in erythroderma sparing skin folds associated with BP, such cases should be accumulated. Dermatologists should keep in mind that erythroderma sparing skin folds can occur during the course of BP to avoid mistaking BP for PEO.

CONFLICT OF INTEREST: None declared.

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