Our Dermatology Online

Case Letter

Amicrobial pustulosis of the folds mimicking Hailey–Hailey disease in a patient with systemic lupus erythematosus and Sjögren's syndrome

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Sir,

Amicrobial pustulosis of the folds (APF) is a rare disease of neutrophilic dermatosis involving the scalp, ears, axillae, umbilicus, groin, and buttocks [1-3]. Herein, we describe a case of APF involving the bilateral axillae mimicking Hailey–Hailey disease in association with systemic lupus erythematosus (SLE) and Sjögren's syndrome (SjS).

A 63-year-old female had been undergoing treatment for SLE and SjS for six years. She had been taking prednisolone at a dose of 4 mg per day on the first visit to us. She also had a history of Crohn's disease in remission. She complained of recurrent, erosive erythema on the axillae and beneath the breasts, which worsened in summer. A physical examination revealed infiltrative erythema with superficial erosions on the bilateral axillae (Figs. 1a and 1b). A laboratory examination revealed normal liver and kidney function, yet positive results for serum antinuclear antibodies (1:1280), anti-double strand DNA antibody (128 IU/mL), anti-Sm antibody (index: 6.7), anti-SS-A antibody (index: 131.5), and anti-SS-B antibody (index: 93.7). The erythrocyte sedimentation rate was high (32 mm in the first hour), and the CRP level was slightly increased (1.19 mg/dL). Bacterial cultures were positive for group G streptococci and the *Corynebacterium* species. A biopsy specimen did not reveal acantholysis of the epidermis, yet mild epidermal acanthosis without intense infiltration of inflammatory cells. Hailey–Hailey disease was excluded, and the patient was followed under treatment with topical corticosteroid ointment. During the course, the skin lesions exacerbated, and new pustules and erosive erythemas appeared on the groin and the anogenital areas (Fig. 1c). A histopathological examination revealed subcorneal neutrophilic abscesses in the epidermal and dermal perifollicular areas (Figs. 2a – 2d).

APF is a rare condition included as one of the neutrophilic dermatoses. It mainly affects the scalp and ear canals, as well as the intertriginous areas such as the axillary, groin, and perianal regions. If it involves the scalp, it typically results in alopecia [4]. APF is characterized by small, sterile pustules and erythemas and, histopathologically, subcorneal or intraepidermal neutrophil infiltration sometimes forming spongiform pustules, parakeratosis, and cellular infiltrates in the upper dermis. Pustules are both follicular and non-follicular. In the present case, we initially suspected Hailey–Hailey disease, because wet erythemas with superficial tiny pustules were observed on the skin folds such as the bilateral axilla and anogenital areas. However, the initial biopsy did not show acantholysis of the epidermal cells. A second biopsy revealed subcorneal neutrophil infiltration, follicular epithelial infiltration forming neutrophilic abscesses, as well as neutrophil infiltration around the follicles in the dermis.

APF is known to occur in patients with autoimmune diseases such as SLE. Marzano et al. proposed the diagnostic criteria for APF, which require association with one or more autoimmune disorders, positive antinuclear antibody at more than 1:160, and the presence of one or more serum autoantibodies [5]. In the present case, although the activity of APF and

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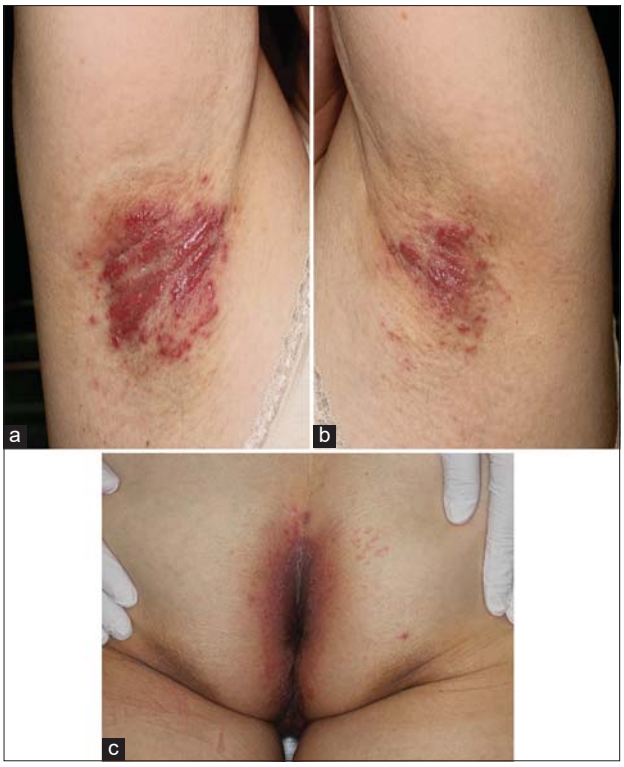

Figure 1: (a and b) Infiltrative erosive erythema on the bilateral axillae. (c) Erosive erythema and scattered papules in the intergluteal region.

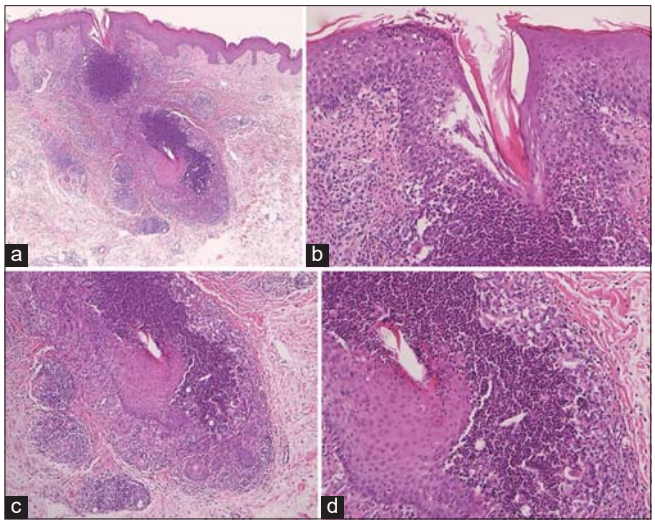

Figure 2: (a) Neutrophil infiltration in the follicular epidermis (40×). (b) Subcorneal neutrophil infiltration and follicular pustule (200×). (c) Prominent neutrophilic abscess in the dermis (100×). (d) Higher magnification showing giant cells outside the neutrophilic abscess (200×).

SLE/SjS was not parallel in behavior, the patient's previous history of Crohn's disease, as well as the presence of concurrent SLE and SjS, may have suggested the risk of developing APF. In addition, APF has recently been suggested to be a spectrum of autoinflammatory disorders, and several cases of APF in association with neutrophilic dermatoses, such as palmoplantar pustulosis and pyoderma gangrenosum, have been reported [6,7]. APF is a type of neutrophilic dermatosis and needs differentiation from Hailey–Hailey disease.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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