Amicrobial Pustulosis of the Folds: Report of Three New Cases

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Sir,

Amicrobial pustulosis of the folds (APF) is a rare, chronic and relapsing disease belonging to neutrophil dermatosis. APF involves major and small folds and is associated with autoimmune disorders and/or immunological abnormalities. Herein, we report the clinical, pathological, and laboratory findings of three new cases of APF.

The first case was a 46-year-old woman who presented with a 10-year history of recurrent pustular eruption on an erythematous base on the neck, axillae, submammary folds, lip commissures, pubis, and scalp [Figure 1a and b]. Histological examination showed hyperplasia with focal parakeratosis, mild exocytosis of neutrophils, spongiform pustules in the epidermis, and a scant, perivascular neutrophilic infiltrate in the superficial and mid dermis. The patient was treated with dapsone 75 mg/day and topical mometasone furoate, with marked improvement of the lesions after 12 weeks. The remission was obtained with a 7-day cycle of prednisone 0.5 mg/kg/day.

The second case was a 40-year-old woman who presented with a chronic pustular eruption with oozing and crusts of 4-month duration involving the inguinal folds, perineum, scalp, ears, and nose orifices. Histopathology showed parakeratosis, intraepidermal vesicles and pustules of neutrophils, and an infiltrate of lymphocytes and neutrophils in the superficial dermis. The patient was treated with oral corticosteroids (1 mg/kg/day) and oral clarithromycin 500 mg/bid for 5 days, with rapid improvement. In the following 3 years, the patient underwent four exacerbations, always responding to 0.5 mg/kg/day prednisone alone for 7 days.

The third case was a 44-year-old Hispanic woman affected by systemic lupus erythematosus (SLE) since the age of 18 years and with a 3-year history of recurrent erosions and nonfollicular pustules involving the intergluteal and submammary folds, axillae, genital area, retroauricular groove, and occipital region of the scalp [Figure 1c and d]. Histopathology showed hyperplastic epidermis with neutrophil exocytosis forming subcorneal aggregates. The upper dermis showed a mixed neutrophilic and lymphocytic infiltrate with perivascular and interstitial distribution [Figure 1e]. The patient was treated with oral corticosteroids (1 mg/kg/day) and topical mometasone furoate with clinical benefit after 2 weeks and total remission after 10 weeks. At 2 years of follow-up, the patient maintained complete remission.

To date, 69 cases of APF have been reported in the literature, with a 90% female prevalence. Marzano et al. coined the term APF and suggested diagnostic criteria. The list of differential diagnoses covers most subcorneal pustulosis as well as bacterial and candidal intertrigo. All our three cases showed an involvement...
of the major and minor folds, scalp, and genital area with erythematous plaques, erosions, and few nonfollicular pustules at the edge. Several cultures for bacteria and fungi collected from unopened pustules before antibiotic treatment were consistently negative. One patient had a previous diagnosis of SLE and one had autoimmune thyroiditis. ANA positivity was found in all our cases at a titer >1/640 with a homogeneous or speckled pattern. Histopathology showed spongiform subcorneal pustules and inflammatory dermal neutrophilic infiltrate with scattered lymphocytes. Given the rarity of APF, there are no standard therapeutic regimens. Systemic corticosteroids (0.5 mg/kg/day) represent the most effective treatment. Other therapies include dapsone 50–100 mg/day, cyclosporine 3–3.5 mg/kg/day, and steroid-sparing adjunctive therapies, such as colchicine 1 mg/day, hydroxychloroquine 400 mg/day, and methotrexate 15 mg/week. The clinical course of APF is chronic-relapsing, and in our series, only one patient showed no relapse.

Our cases confirmed that APF has a chronic course, but benign clinical behavior. Given the rarity of APF, there is no consensus on best treatment. We found a good response with low-dose systemic corticosteroids.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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