REVIEW

Amicrobial pustulosis of the folds: Where have we gone 25 years after its original description?

Pustulose amicrobienne des plis : état des lieux 25 ans après la description princeps

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Amicrobial pustulosis of the folds; Aseptic pustulosis; Lupus erythematosus; Neutrophil dermatosis; Auto-inflammatory disease

Summary
Background. — Twenty-five years ago at the Journées Dermatologiques de Paris, Prof. Béatrice Crickx described a new association, "antimicrobial pustulosis and systemic lupus erythematosus", a condition now known as amicrobial pustulosis of the folds (APF). The aim of this study is to analyse the clinical and laboratory characteristics of APF and to outline the gradual advancement of knowledge regarding this disease.

Materials and methods. — Based on a case of APF seen in our department, we carried out a review of the literature since 1991 by searching the Medline database for scientific articles using the following keywords: "Amicrobial Pustulosis" or "Pustular Dermatosis" and "Folds".

Results. — We collated 63 cases of APF. In total, 90% involved women and the mean age was 30 years. Cutaneous lesions consisted of pustules in an erythematous setting, frequently erosive and affecting the skinfolds, ano-genital region, scalp, external auditory meatus and umbilicus. Histological examination revealed spongiform subcorneal pustulosis associated with mixed inflammatory infiltrate in the dermis.

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Discussion. — APF belongs to the spectrum of neutrophilic dermatoses. Its complex physiopathology involves an auto-inflammatory mechanism. It is associated with a variety of autoimmune diseases, in most cases systemic lupus erythematosus. Diagnosis of the condition is difficult and is based on histological correlation and negative microbiological culture. The most effective treatment is still systemic corticosteroids, although new therapeutic alternatives are emerging such as anakinra and anti-TNF-alpha drugs.

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Patients and methods

A 30-year-old female patient consulted for weeping pustular dermatosis present for two years. The rash began on her back, before rapidly appearing on the scalp, pubis, buttocks and thighs. An initial diagnosis was made of pustular psoriasis and the patient was treated with clobetasol propionate (Dermovate® cream) and then oral cyclosporine 150 mg b.i.d. for 3 months. The outcome was initially favourable but the dermatosis returned one year later with extensive pustules and erosion of the scalp with alopecia (Fig. 1) and of the retro-aural folds (Fig. 2), and erythematous and pustular plaques on the inguinal folds (Fig. 3), pubis, and umbilicus, as well as intergluteal intertrigo. Bacteriological and mycological cultures were negative. The picture was at this point highly evocative of APF, and screening for antinuclear antibodies was positive at 1/320 with the presence of anti-Ro/SSA antibodies. The skin biopsy exhibited spongiosis of the epidermis with multilocular subcorneal pustules and inflammatory infiltrate in the dermis (Fig. 4). Treatment was initiated with oral corticosteroids.

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