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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, anogenital 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.

MOTS CLÉS
Pustulose amicrobienne des plis ;
Pustulose aseptique ;
Lupus érythémateux ;
Dermatose neutrophilique ;
Maladie auto-inflammatoire

Résumé
Introduction. — Il y a 25 ans, le Professeur Béatrice Crickx rapportait aux Journées Dermatologiques de Paris une nouvelle association : « pustulose amicrobienne et lupus érythémateux systémique », aujourd'hui nommée pustulose amicrobienne des plis (PAP). L'objectif de ce travail est d'analyser les caractéristiques cliniques et paracliniques de la PAP et de montrer l'évolution progressive des connaissances de cette affection.
Matériel et méthode. — À partir d'un cas de PAP constaté dans notre service, nous avons effectué une revue de la littérature depuis 1991. Une recherche d'articles scientifiques a été réalisée en interrogeant la base de données MEDLINE avec les mots clés « Amicrobial Pustulosis » ou « Pustular Dermatosis » et « Folds ».
Résultats. — Nous avons rassemblé 63 cas de PAP. Au total, 90 % étaient des femmes, l'âge moyen de 30 ans. L'atteinte cutanée correspondait à des pustules sur fond érythémateux, souvent érosif touchant les plis cutanés, la région génitale, le cuir chevelu, le conduit auditif externe, l'ombilic. L'examen histologique montrait une pustule spongiforme sous-cornée associée à un infiltrat inflammatoire du derme.
Discussion. — La PAP appartient au groupe des dermatoses neutrophiliques. Sa physiopathologie complexe est principalement d'origine auto-inflammatoire. Elle est associée à des maladies auto-immunes variées, le plus souvent au lupus érythémateux systémique. Son diagnostic est difficile et repose sur une corrélation anatomoclinique et un examen microbiologique négatif. Le traitement le plus efficace à ce jour reste la corticothérapie par voie générale, mais de nouvelles perspectives thérapeutiques existent comme l'anakinra et les anti-TNF-alpha.

Twenty-five years ago at the Journées Dermatologiques de Paris, Prof. Béatrice Crickx described a new association of microbial pustules of the skinfolds of sudden onset in two female patients presenting systemic lupus erythematosus (SLE): ''amicrobial pustulosis and systemic lupus erythematosus'' [1].

Two further similar cases ensued [2—6], followed by cases of amicrobial pustulosis associated with other autoimmune diseases, dubbed amicrobial pustulosis associated with autoimmune disease (APAD) [7]. Precise diagnostic criteria were suggested in 2008 (Table 1), with the entity being renamed ''amicrobial pustulosis of the folds'' (APF) [8]. It currently forms part of the group of neutrophilic dermatoses [9,10].

The aim of this study was to analyse the main clinical and laboratory characteristics as well as the treatment of APF based on a new case and on a literature review, and also to describe the progressive advancement of knowledge that has resulted in the classification of this disease as an independent entity within the neutrophilic dermatoses group.

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 (Dermoval® 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-auricular 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