Acta Derm Venereol (Stockh) 1992; 72: 301–302

Subcorneal Pustular Dermatosis in a Patient with Crohn's Disease

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A case of subcorneal pustular dermatosis (Sneddon–Wilkinson disease) is reported in a patient with a one-year history of Crohn's disease. Subcorneal pustular dermatosis has been described in association with monoclonal gammopathy, but to our knowledge it has not been associated with Crohn's disease. This new association reinforces the hypothesis of a possible common pathogenesis for neutrophilic dermatoses and inflammatory bowel diseases. ***Key words: Neutrophilic dermatoses; Ulcerative colitis; Pyoderma gangrenosum; Sweet's syndrome.***

(Accepted December 16, 1991.)

Acta Derm Venereol (Stockh) 1992; 72: 301–302.

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Subcorneal pustular dermatosis (SPD), also referred to as Sneddon–Wilkinson disease, is a chronic relapsing vesiculopustular eruption, mainly involving the trunk and intertriginous areas, usually seen in women past the age of forty (1). Its association with a monoclonal gammopathy, most commonly an immunoglobulin A-paraproteinemia, is well recognized (2). SPD has also been occasionally described in patients with rheumatoid arthritis (3) and ulcerative colitis (4), but the association with Crohn's disease has not previously been reported.

CASE REPORT

A 37-year-old man was admitted to our department with a one-month history of a pustular eruption. One year before admittance, a diagnosis of ileocolonic Crohn's disease had been made on the usual clinical, morphological and histological criteria (5). A first cousin of him was also known to suffer from Crohn's disease. At his admission, Crohn's disease was well controlled, i.e. Crohn's disease activity index < 150 (6), by mesalazine (1.5 g per day).

On physical examination, he presented with flaccid pustules with hypopyon on erythematous bases involving the trunk, axillae, groin folds and arms. Recurrent waves of lesions coalescing led to circinate and polycyclic patterns. There was no fever or diarrhea. Histology of a pustular lesion showed a subcorneal pustule filled with polymorphonuclear leucocytes, mainly neutrophils (Fig. 1). A perivascular dermal infiltrate was present, consisting predominantly of neutrophils and rarely eosinophils. There was no spongiosis or acantholysis. Direct and indirect immunofluorescence were negative. Haematological investigations and a full biochemical screen were normal and no paraprotein was detected.

A diagnosis of SPD was made on the basis of the clinical picture together with the histopathological findings. The patient was given dapsone 100 mg daily which led to control of cutaneous lesions within 6 weeks. At a follow-up six months later, the skin lesions had not recurred.

DISCUSSION

Crohn's disease may have distinctive mucocutaneous manifestations which may be related to the primary granulomatous process, to nutritional deficiencies, to therapy or may be idiopathic in nature. Idiopathic conditions include erythema nodosum, epidermolysis bullosa acquisita, necrotizing vasculitis, finger clubbing and neutrophilic dermatoses (7).

Neutrophilic dermatoses, e.g. SPD, intra-epidermal neutrophilic immunoglobulin A dermatosis, Sweet's syndrome, pyoderma gangrenosum and erythema elevatum diutinum, are non-infective skin diseases without any known etiology, characterized by a cutaneous neutrophilic infiltrate (8). Reports of overlap syndromes between those conditions (9, 10), together with the efficacy of drugs acting on neutrophils, have led to the concept of the neutrophilic dermatoses clinicopathological spectrum. Furthermore, these dermatoses share possible association with a similar range of diseases including myeloproliferative disorders (11) and evolutive inflammatory bowel diseases, i.e. Crohn's disease and ulcerative colitis. Pyoderma gangrenosum is by far the most frequent neutrophilic dermatosis occurring in inflammatory bowel diseases, especially ulcerative colitis (9). Sweet's syndrome has been reported in association with Crohn's disease in four patients (12, 13, 14) and erythema elevatum diutinum has been mentioned in one case (15). Up to now, SPD has only been recorded in one patient with a non-evolutive ulcerative colitis (4). Interestingly, in our case, the bowel disease was also quiescent. Although concurrence of SPD and Crohn's disease may be coincidental, the probability of two rare conditions occurring together seems low. This observation enlarges the spectrum of neutrophilic dermatoses associated with inflammatory bowel diseases and raises the question of a possible common pathogenesis for these two groups of diseases. Neutrophils could be the causal link.

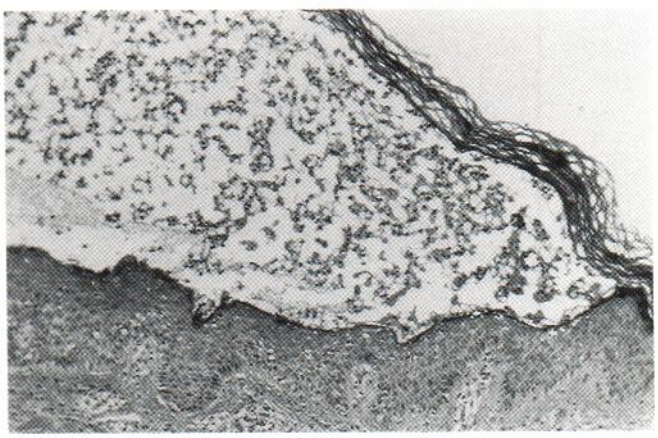

Fig. 1. Subcorneal pustule containing numerous neutrophils (H&E; ×100).

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