SNEDDON-WILKINSON DISEASE AND ARTHRITIS

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Sneddon-Wilkinson disease (subcorneal pustular dermatosis) is an uncommon disorder. An unusual association with seronegative arthritis is reported with review of literature.

Key Words : Sneddon-Wilkinson disease, Subcorneal pustular dermatosis, Arthritis

Introduction

Sneddon-Wilkinson disease (SWD) is a rare pustular disease often associated with other systemic illness. Several associations with paraproteinaemias have been observed. Association with IgA of the M component and inflammatory diseases such as rheumatoid arthritis, systemic lupus erythematosus, and unclassified arthritis has been reported.[1] A case of SWD with arthritis is described.

Case Report

A-26-year-old woman presented with a 12 months history of an itchy rash. On careful examination a pustular eruption was observed on the buttocks, flanks and trunk. The lesions spread centrifugally in a gyrate configuration with flaccid pustules of 2-5 mm diameter and a peripheral distribution (Fig. 1). She also developed recurrent episodes of joint pain at about the same time as these lesions for which she was prescribed oral ibuprofen with temporary relief. No oral lesions were observed and her tongue was normal. Clinical differential diagnosis included, SWD, superficial pemphigus (IgA or pemphigus foliaceus), and bullous impetigo. Gram and lactophenol staining of swabs obtained from the puatules for bacteria and fungus were negative.

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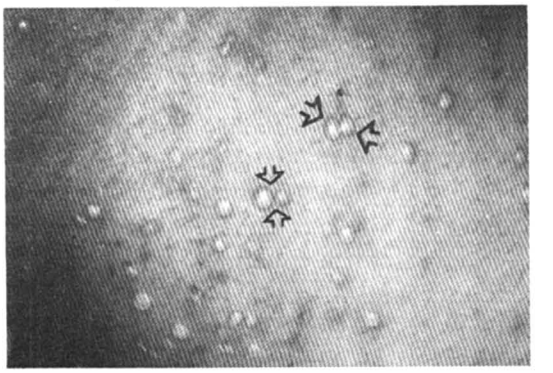

Fig. 1. Flaccid yellow pustules extending centrifugally (buttocks).

Laboratory examinations including complete blood count, ANA titre, ELISA for HIV 1 & 2, serum creatinine, liver function tests and assay of glucose-6-phosphate dehydrogenase activity were within normal limits. IgA rheumatoid factor was consistently negative. Erythrocyte sedimentation rate was 70mm/1hour.

Histology of a pustular lesion revealed a flaccid subcorneal pustule with numerous neutrophils and occasional eosinophils (Fig. 2).

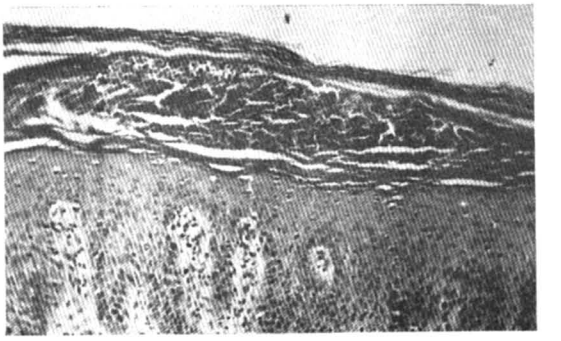

Fig. 2. A pustule is present below the horny layer without acantholysis (H&E x40).

No obvious acantholysis was observed. A diagnosis of SWD was made on the basis of typical clinical and histological features. Patient was given 100mg of dapsone per day. Her itching abated within 2 days and skin lesions markedly improved within 3 weeks. After 2 months of treatment her joint pain also subsided.

Discussion

Sneddon and Wilkinson first described subcorneal pustular dermatosis and separated this entity from other unclassified pustular eruptions.[2] This rare disease is characterized by flaccid sterile pustules which have a tendency to coalesce forming annular and circinate patterns.[1] The pustule of SWD is subcorneal filled with polymorphonuclear leucocytes.[1,2] Although immunofluorescence finding are usually nagative,[1] rarely intercellular, intrapustular or subcorneal epithelial deposition of IgA has been reported. This group of cases may be put in the group of "Intercellular IgA vesiculo-pustular dermatosis" [3,4] The characteristic features of this group are vesiculopustular eruption, intraepidermal pustule formation, and in vivo bound and/or circulating IgA anti-intercellular antibodies.[3,4] Indirect immunofluorescence studies have indicated specific reactivity patterns of the IgA antibodies with epidermis.[3,4] Now it is clear that at least two distinct disorders exists: a subcorneal pustular dermatosis type and an intra-epidermal neutrophilic type. Sera obtained from the patients of the former reacted with 115 and 105 kilodalton (KD) epidermal antigens suggestive of desmocollin I & II. In contrast, one serum from the latter case got attached to an indeterminate 120 KD epidermal protein.

Although dapsone remains the drug of choice of SWD therapy, several other medicines have been used singly or in combination.[5] These include: sulphamethoxypyridazine, minocycline, PUVA, systemic steroid and/or azathioprine, clobetasol propionate and colchicine.

Joint diseases such as rheumatoid and unclassified arthritis have rarely been reported with SWD.[1] Previous reports also indicate that dapsone is able to improve both skin and joint disease.[1] Improvement in this patient with only dapsone may suggest that similar mechanisms are involved in these conditions. Dapsone probably acts by inhibition of neutrophilic migration and chemoattractants, suppression of circulating immune complexes and inflammatory mediators such as prostaglandins and cytokines resulting in resolution of both skin and joint disease.[6]

References

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