MIXED FORM OF PEMPHIGUS VULGARIS AND DERMATITIS HERPETIFORMIS

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A 45-year-old female had grouped pruritic papules and vesicles with a few scattered tense bullae, symmetrically distributed on the extensor surfaces of limbs, back and buttocks. Nikolsky's sign was negative. Histopathology revealed intra-epidermal suprabasal cleft along with acantholysis. Eosinophilic spongiosis was also present. The patient recovered with dapsone.

Key words : Mixed bullous disease, Pemphigus vulgaris, Dermatitis herpetiformis.

Pemphigus vulgaris may present as dermatitis herpetiformis.[1,2] Montgomery[3] stated that occasionally acantholytic cells would be present in dermatitis herpetiformis. A variant of pemphigus foliaceous clinically mimicking dermatitis herpetiformis has already been recognised as a separate entity under the name pemphigus herpetiformis.[4,5]

Case Report

A 45-year-old lady was having pruritic skin lesions for the last 7 years. The disease aggravated every summer and remained somewhat subsided during the rainy season and winter. She had had her last child-birth 16 years ago. She had no such lesions either during pregnancy or puerperium. There was no family history of a similar disease.

The lesions consisted of groups of papules and vesicles surrounded by erythema, symmetrically distributed on the extensor surfaces of the extremities, the back, the shoulders and the buttocks. A few tense bullae were also present on the hands, the legs and the back. The bullae did not break easily. Oral mucosa was not involved. Nikolsky's sign was negative. There was no associated diarrhoea. She had mild anemia. Liver, spleen and superficial lymph nodes were not palpable. No systemic abnormality was detected.

Fig. 1. A few grouped papules and vesicles and a tense bulla on the lower leg.

Histopathological studies from an early vesicle showed a suprabasal cleft within the epidermis with many acantholytic cells. A superficial old healing split was also visible. Eosinophilic spongiosis was seen in some areas of epidermis. Dyskeratosis was absent in the granular layer. The dermis had a moderate number of inflammatory cells, among which eosinophils were prominent. Other investigations such as complete hemogram, routine urine and stools examination, blood sugar, blood urea and creatinine, chest X-ray, barium X-ray of gastro-intestinal tract, Mantoux test and VDRL test did not show any abnormality except

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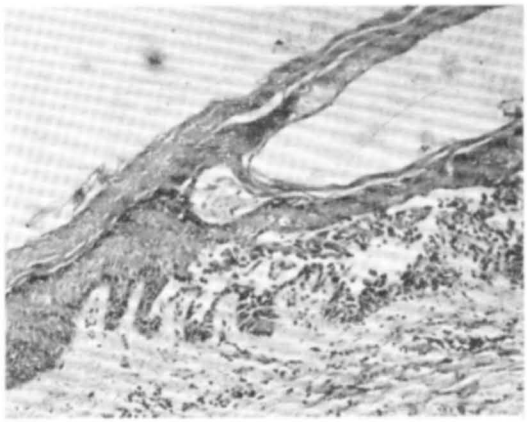

Fig. 2. Suprabasal cleft within the epidermis along with acantholytic cells. Eosinophilic spongiosis is present. A superficial old healing split is also visible (X80).

hemoglobin level of 10.5%. There was no blood eosinophilia.

The patient responded to dapsone, 100 mg twice a day in the first week, 100 mg thrice daily for the next two weeks when all the lesions disappeared. The same dose was maintained for 4 more weeks and then gradually reduced to 100 mg daily. For the last 6 months, the patient had no lesion.

Comments

Singh et al[1] reported a case of pemphigus vulgaris presenting as dermatitis herpetiformis but eosinophilic spongiosis was not seen. The case reported by Singh et al[2] was a treated case of classical pemphigus vulgaris, developing dermatitis herpetiformis like lesions during the remission period. Eosinophilic spongiosis was evident histopathologically. Administration of dapsone and prednisolone controlled the lesions.

The present case having a clinical picture of dermatitis herpetiformis resembled histopathologically pemphigus vulgaris. Eosinophilic spongiosis was also associated. The ailment was controlled well with dapsone. Thus, the case represents a mixed bullous disease having both the features of dermatitis herpetiformis and pemphigus vulgaris.

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