PEMPHIGUS HERPETIFORMIS

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A 70-year-old Indian lady with a two year history of a blistering eruption is described. This eruption clinically resembled dermatitis herepetiformis and responded to dapsone, however, histological and immunological investigations showed features of pemphigus herepetiformis.

Introduction

A 70-year-old woman presented with a two year history of itching and redness of the skin followed by blister formation. The blisters initially were single but later became grouped. There was no history of worsening with gluten containing foods.

At initial presentation grouped tense vesicles on an erythematous base were seen distributed extensively over the forearms, arms, anterior abdominal wall, thighs, legs and feet (Fig. 1). Nikolsky sign was negative. The differential diagnoses considered were dermatitis herpetiformis (DH) and vesicular pemphigoid.

Case Report

A skin biopsy showed a subcorneal bulla containing acantholytic cells, eosinophils and neutrophils (Fig. 2). It was reported as consistent with pemphigus foliaceous. Direct immunofluorescence revealed the presence of IgG in the intercellular spaces of the epidermis. C3 was also deposited at the dermoepidermal junction like a "lupus hand". Therefore, a diagnosis of 'dermatitis herpetiformis like pemphigus' or, pemphigus herpetiformis was considered. A Tzanck smear was negative for acantholytic cells. There was

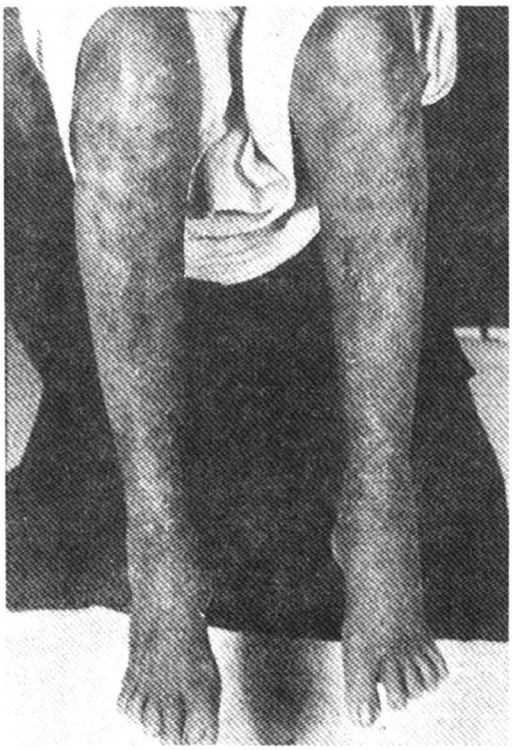

Fig. 1. Grouped vesicles and crusting on the legs.

no evidence of malabsorption; 72 hour faecal fat excretion was normal. The patient refused a jejunal biopsy.

The patient was given dapsone 100 mg orally per day, following which the blisters subsided in 4 days. Dapsone was then stopeed as a provocative test and the skin lesions reappeared within 48 hours. Dapsone 100 mg daily orally was resumed, but as the blisters were not controlled this time with dapsone alone, prednisolone 40

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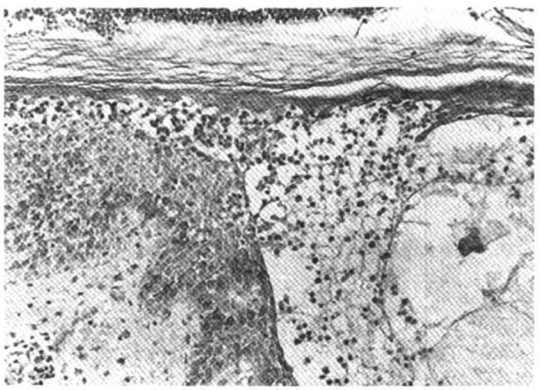

Fig. 2. A subcorneal bulla containing acantholytic cells and several neutrophils (H&E x 160).

mg/day orally was added. Blistering now subsided in a week. The dose of prednisolone was then gradually tapered over several weeks to a maintenance dose of 5 mg every successive alternate day. The patient has remained blister free for the 15 months she has been followed up.

Discussion

'DH like pemphigus' was first described by Brocq in 1892. In 1943 Civatte observed occasional subconrneal vesicles in patients with dermatitis herepetiformis.[1] Others felt that this may be a unique new disease and suggested the term acantholytic herpetiform dermatitis.[2] Yet other reports suggest that this is only a subset of pemphigus. It was Jablonska who first coined the term herpetiform pemphigus.[3]

Clinically, the lesions may resemble those of dermatitis herpetiformis, bullous pemphigoid or linear IgA dermatosis. In pemphigus herpetiformis, the lesions may be urticarial plaques with central healing and peripheral papulovesicles, or grouped papules, vesicles or bullae in a herpetiform pattern as in our patients. The locations of the blister has varied in various case reports from subcorneal[4] as in our patient, to suprabasal.[5]

Pemphigus herpetiformis has been treated employing either dapsone, oral steroids or a combination of both. Although our patient initially responded to dapsone alone, later, a low dose of systemic steroids was required to control the eruption.

This case is being reported because pemphigus herpetiformis is rare in India. The direct immunofluorescence test on the skin biopsy of our patients, in addition to staining the epidermal intercellular spaces for IgG, also showed C# deposition at the dermoepidermal junction, a feature usually seen in pemphigus erythematosus but not so far reported in pemphigus herpetiformis.

References

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