An Unusual Form of Pemphigus Vegetans

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Abstract. Numerous vegetating plaques developed in a 63-year-old man followed up for the management of pemphigus vulgaris of 5 years' duration. These lesions occurred over the trunk and right upper arm with no involvement in the intertriginous area, and were controlled with systemic steroid and ODT. Direct immunofluorescence test showed heavy depostion of fibrinogen in the papillary dermis. To our knowledge, the occurrence of pemphigus vegetans in the non-intertriginous area is rare.

Key words: Pemphigus vegetans; Fibrinogen; Steroid

CASE REPORT

A 63-year-old man first came to our clinic in December 1974 with multiple bullous lesions involving almost the entire body surface. He had had a 6-month history of enanthema in the oral mucosa. Skin biopsy specimen confirmed pemphigus vulgaris, and direct immunofluorescence microscopy showed IgG and C_3 deposits in the intercellular epidermal space. The titer of pemphigus antibody was X640. The patient was treated first with 80 mg of betamethasone daily and was maintained on 5–10 mg of prednisolone after his first admission. In December 1979, the patient developed relapsing vesicle formation with thickening of vegetating plaques (Fig. 1) on the trunk and right upper arm (Fig. 2). At that time there was no vegetating enanthema. He was rehospitalized. Biopsy of the plaque on the back revealed hyperkeratosis, papillomatosis (Fig. 3), suprabasal acantholytic bulla (Fig. 4) and abscess formation similar to the Neumann type (Fig. 5), but no eosinophilic spongiosis was observed. Direct immunofluorescence test demonstrated IgG and C_3 deposits at the bottom of histological bulla (Fig. 6) and heavy depostion of fibrinogen in the papillary dermis (Fig. 7). The vesicle adjacent to plaques was found to be acantholytic, and erythema near the vesicle exhibited IgG and C_3 deposits in the interepidermal space. Many plaques were reduced in size after oral administration of 3 mg of betamethasone, but the lesion at the sacral region was

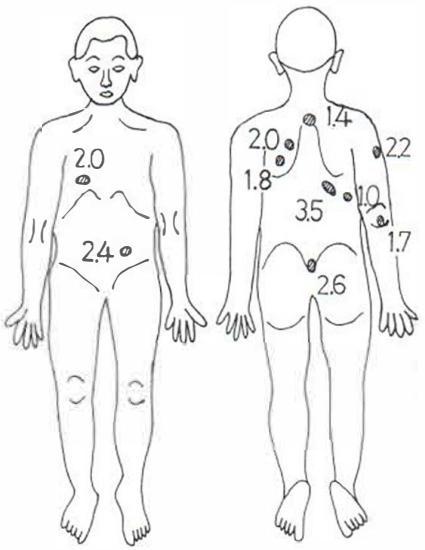

Fig. 2. Scheme showing distribution of the lesions. The numeral (cm) shows maximum diameter of plaque (hematoxylin-eosin, ×200).

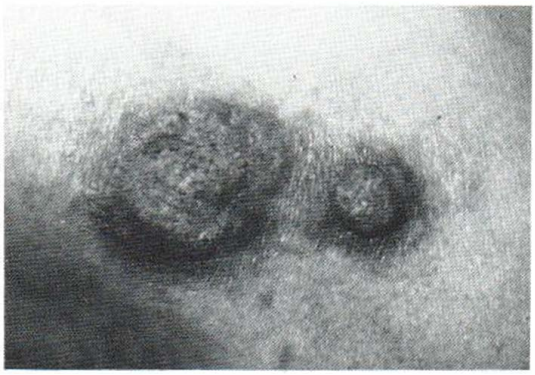

Fig. 1. Vegetating plaques on middle part of back.

Fig. 3. Skin biopsy specimen from back shows hyperkeratosis, and papillomatosis (hematoxylin-eosin, ×20).

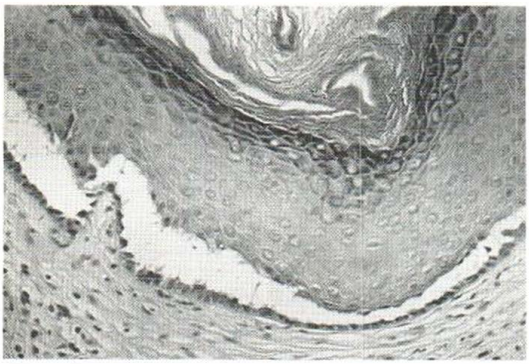

Fig. 4. Same specimen shows suprabasal bullae formation in epidermis (hematoxylin-eosin, ×100).

enlarged in spite of the low titer of pemphigus antibody (X10), even after an increased dose of oral betamethasone. This plaque regressed after topical application of clobetasol propionate under ODT.

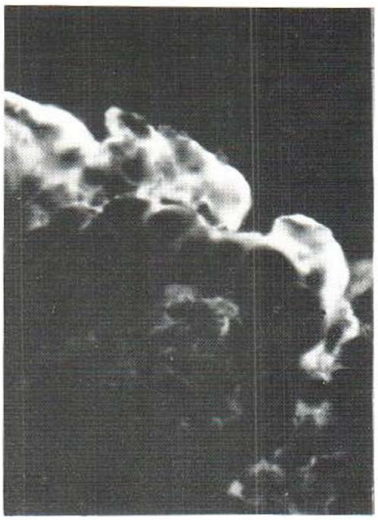

Fig. 6. Direct immunofluorescence microscopy of involved nodule. Intercellular deposition of IgG at bottom of acantholytic bulla (×400).

DISCUSSION

Pemphigus vegetans is composed of the Neumann and Hallopeau types, and is thought to be more benign than pemphigus vulgaris (1). The common Neumann type begins and ends as pemphigus vulgaris—before the advent of steroids. Vegetative forms may occur in a patient with pemphigus vulgaris as the condition remits under steroid control (2). Our case seemed to be clinically consistent with the Neumann type.

The characteristic findings in our case include the following: 1) pemphigus vegetans arose on the non-intertriginous area in the course of pemphigus vulgaris of 5 years' duration, and the unusual clinical feature might be partly due to the prolonged administration of steroids; 2) most vegetating lesions, which occurred with flare, were cured after treatment with oral steroid in a higher dose, and it may be said that the pathogenesis of pemphigus vegetans is related to the increase in pemphigus antibody; 3) one lesion in the sacral area, which was found to respond favourably to topical steroid under ODT, grew larger despite the low titer of pemphigus antibody and increased dose of oral steroid, and this might be associated with mechanical stimulation, taking into consideration that decubitus often arises in the same area; 4) fibrinogen deposit in the papil-

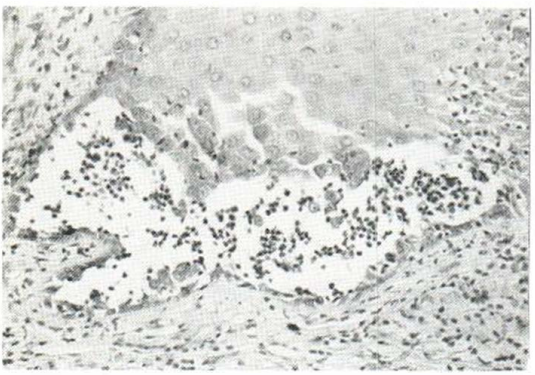

Fig. 5. Specimen shows features of abscess formation (hematoxylin-eosin, ×200).

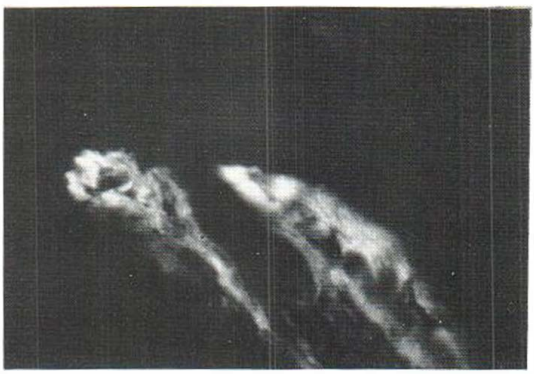

Fig. 7. Diffuse deposition of fibrinogen in papillary dermis (×200).

lary dermis was observed in biopsy specimens from nodular lesions (biopsy was done twice), and it would appear that a disturbance of the local fibrinolytic system is one of the causative factors in pemphigus vegetans.

Drug: Betamethasone-Rinderon, Shionogi & Co. Ltd.; Prednisolone-Prednisolone, Shionogi & Co. Ltd.; Clobetasol propionate-Dermovate, Nippon Glaxo Ltd.

REFERENCES

1. Lever, W. F.: Pemphigus and Pemphigoid. pp. 40–51. Charles C. Thomas, Springfield, Ill., 1965.
2. Sneddon, I. B.: Bullous eruptions. *In* Textbook of Dermatology (ed. A. Rook, D. S. Wilkinson & F. J. G. Ebling), pp. 1441–1481. Blackwell Scientific Publications, Oxford, 1979.

White Fingernails Preceded by Multiple Transverse White Bands

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Abstract. In a patient with hepatocellular damage due to abuse of alcoholic liquors, multiple transverse white bands were found in all fingernails. On examination 3 months later the bands had disappeared, and the nails had developed into typical white nails. The changes were located in the nail bed, and were possibly caused by vascular alterations.

Key words: Multiple transverse white bands; White nails

In 1954 Terry described white nails in hepatic cirrhosis. In 82 of 100 consecutive patients with cirrhosis he found characteristic changes in the fingernails. The proximal part of the nails exhibited an opaque whiteness extending from the base of the nail, where the lunula was indistinguishable, to within 2 mm of the distal border, leaving a distinct distal zone of normal pink. No preceding abnormalities were noted (3).

In the following we report on a patient with hepatocellular damage due to abuse of alcoholic liquors, in whom multiple, transverse white bands preceded the development of white nails.

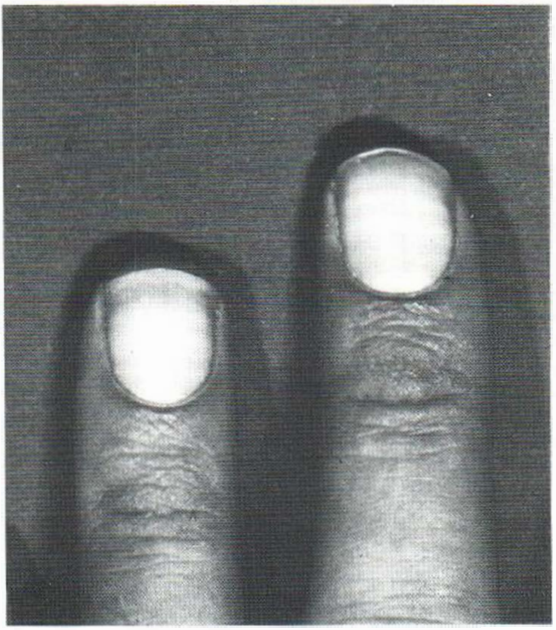

Fig. 1. Nails of 3rd and 4th left finger, showing multiple transverse white bands.

CASE REPORT

The subject is a 28-year-old male, a heavy abuser of alcohol for 10 years. In all nails 3–4 transverse, white bands ran parallel to the lunula, separated from each other by pink zones of normal appearing nail (Fig. 1). The bands were most pronounced in the fingernails. When blanching the fingertips, the bands disappeared. Otherwise the nails were normal. Laboratory evaluation showed an elevated serum-aspartate-amino-transferase (s-ASAT). The following tests were normal: Erythrocyte sedimentation rate, leukocyte count, serum alkaline phosphatase, prothrombin, serum bilirubin, serum creatinine, serum electrophoresis. When the patient was seen 3 months later the bands had disappeared, and the fingernails had assumed the typical picture of white nails (Fig. 2). Repeated laboratory examination showed only an elevated s-ASAT as 3 months earlier.

DISCUSSION

Nail dyschromias can be classified according to the site of alteration, i.e. the nail plate or nail bed. When blanching the fingertip, discolorations located to the nail bed tend to disappear, whereas changes located to the nail plate will remain grossly unaltered (1). In the patient described, the changes