DISCUSSION

The case presented here showed typical features, clinically as well as histologically, of pyoderma gangrenosum.

Pyoderma gangrenosum is often accompanied by ulcerative colitis, regional enteritis, rheumatoid arthritis, and paraproteinemia (1), and in the present case a paraproteinemia of the IgA was found.

Salicylazosulphapyridine (8) and clofazimine (Lampren®) (7, 9) are effective in the treatment of pyoderma gangrenosum. In our patient, treatment with salicylazosulphapyridine was only partially successful, but clofazimine healed the ulcer completely.

The origin of transient acantholytic dermatosis is not known. It belongs to the group of primary acantholytic diseases (3). This syndrome is not uncommon in men over 40. The lesions are mostly papules or papulovesicles, which may be oedematous and crusted. Severe pruritus is often present (2).

Transient acantholytic dermatosis may mimic pemphigus erythematosus, which usually has a benign course compared with other forms of pemphigus and may persist indefinitely as a localized disease without ever becoming generalized (6). The two diseases can be differentiated from each other by immunofluorescence studies.

In pemphigus erythematosus, direct immunofluorescence microscopy reveals intercellular linear fluorescence of IgG and complement as well as fluorescence at the dermo-epidermal junction (LE bands) (4, 6). This fluorescence can be demonstrated in almost all patients with pemphigus erythematosus, especially in lesions on the face, but occurs less frequently in lesions localized to the trunk (4).

Among the group of pemphigus diseases, pemphigus erythematosus is the one most commonly found in association with other auto-immune diseases (6). A few cases have been reported where pemphigus erythematosus was accompanied by systemic lupus erythematosus, thymoma, myasthenia gravis or rheumatoid arthritis (5).

Reports of the simultaneous appearance of transient acantholytic dermatosis, pyoderma gangrenosum and paraproteinemia have, to the best of our knowledge, not been published before. At present we are not able to interpret this odd combination of disorders, but in the hope that others have been confronted with the same problem, we made this report.

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Acrokerato-Elastoidosis: A Case Report

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Abstract. Acrokerato-elastoidosis belongs to the group of diffuse palmo-plantar keratoses without associated symptoms. We report a 25-year-old man who has had typical skin lesions for 5 years. The histopathological changes are described. By electron microscopy, changes in dermal elastic fibres as previously reported in patients with acrokerato-elastoidosis were absent.

Key words: Acrokerato-elastoidosis; Palmo-plantar keratoses; Hyperhidrosis; Electron microscopy

A peculiar skin disorder localized to hands and feet was first described by Costa in 1952 (1). His patient

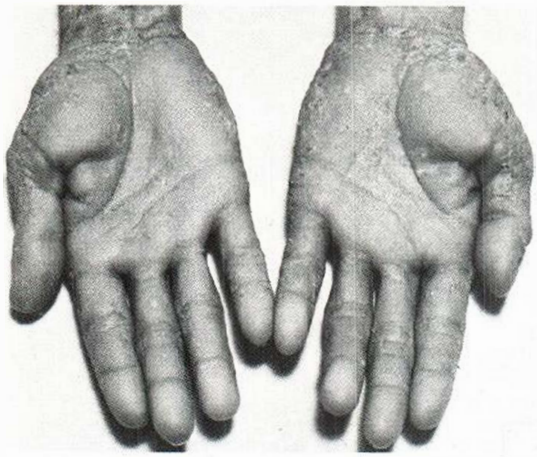

Fig. 1. Pronounced hyperkeratosis in the palms.

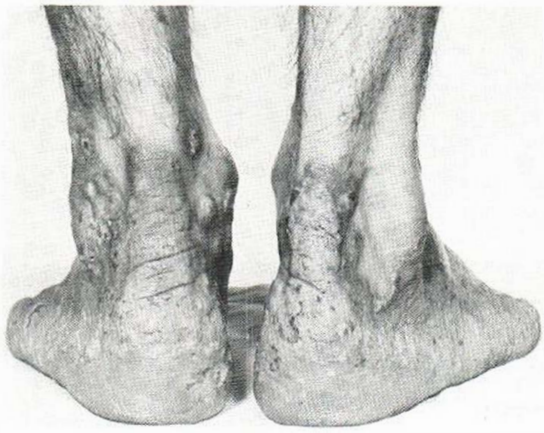

Fig. 2. Warty, slightly scaling skin-coloured keratosis in the Achilles tendon area.

suffered from hyperkeratosis accompanied by severe hyperhidrosis. Costa termed the disease 'acrokerato-elastoidosis'.

A case of acrokerato-elastoidosis is reported in the following.

CASE REPORT

A 25-year-old man with no family history of acrokerato-elastoidosis or other skin disorders. On his first hospital admission 5 years ago he presented severe hyperhidrosis localized to hands and feet. The onset was followed by bullous skin eruptions on the dorsal part of hands, on wrists and lower extremities, and increasing palmo-plantar hyperkeratosis. Three months following the onset the patient developed a severe infection of the affected areas and was feverish for some time. His general condition was seriously affected with peripheral polyneuropathia and paraesthesia of the three radial fingers of both hands. During this period the ESR was increased and there was pronounced leukocytosis with increased numbers of eosinophilocytes and increased serum IgE. All other laboratory findings were within normal ranges. Patch tests (European Standard Test Series) were negative and no fungoid growths were found.

The patient's general condition improved slowly, whereas the skin lesions remained unchanged. The eruptions were sharply demarcated and pronounced hyperkeratosis was found symmetrically on palms and soles (Fig. 1). The skin lesions on the dorsum of fingers and feet displayed papulous, partly confluent, skin-coloured keratosis with a warty, slightly scaling surface. The same changes were present in the Achilles tendon area, on the lower legs and wrists (Fig. 2). The severe hyperhidrosis persisted. The pathological laboratory results normalized simultaneously with the improving general condition.

Histology

Biopsies were taken from hands, feet, and lower legs for light microscopy, and from the lower legs for electron microscopy.

Specimens for light microscopy were fixed in formalin. Paraffin sections were stained with hematoxylin and eosin, Verhoeff's elastic stain, Foot's silver impregnation for the detection of reticulin fibres, periodic acid-Schiff (PAS) technique and with alizarin red S for demonstration of calcium. Sections were further stained with alcian blue, pH 1.0 and 2.5, for the detection of acid glycosaminoglycans.

Specimens for electron microscopy were fixed in 2.5 % glutaraldehyde in 0.1 M cacodylate buffer, pH 7.2, post-fixed in 1 % osmium tetroxide in 0.1 M cacodylate buffer, pH 7.2, dehydrated in a graded series of acetone, and embedded in Araldite. For orientation purposes semi-thin sections were stained with toluidine blue. Ultra-thin sections were stained with uranyl acetate and lead citrate and studied in a Jeol 100CX electron microscope.

By light microscopy all biopsies revealed a variable and locally pronounced hyperkeratosis, acanthosis and slight spongiosis. Cellular layers of stratum granulosum were present and in some places thickened. The upper dermis showed moderate discrete mononuclear cell infiltrates with occasional mast cells. Elastic fibres of the papillary layer as well as the coarser elastic fibres of the reticular layer showed only slight disarrangement and fragmentation. In all other respects the structure of the dermis appeared normal. Calcium deposits were not demonstrated.

By electron microscopy, elastic fibres in all parts of the dermis appeared normal. In the middle and lower parts of the dermis they were dispersed between tightly packed bundles of collagen fibrils (Fig. 3). Elastic fibrils were seen on the surface of the homogeneous matrix and inside the matrix they formed thin parallel lines of moderate electron density arranged along the axes of the fibres. The fibrillar component of the fibres was not reduced. In some of the larger fibres from the deep part of the dermis the matrix lines might show a slight increase in width (Fig. 4). However, accumulations of electron-dense material, holes in the matrix, or disaggregated zones of matrix were not found. Dense material suggesting deposition of calcium salts was not observed. The distribution of collagen fibres

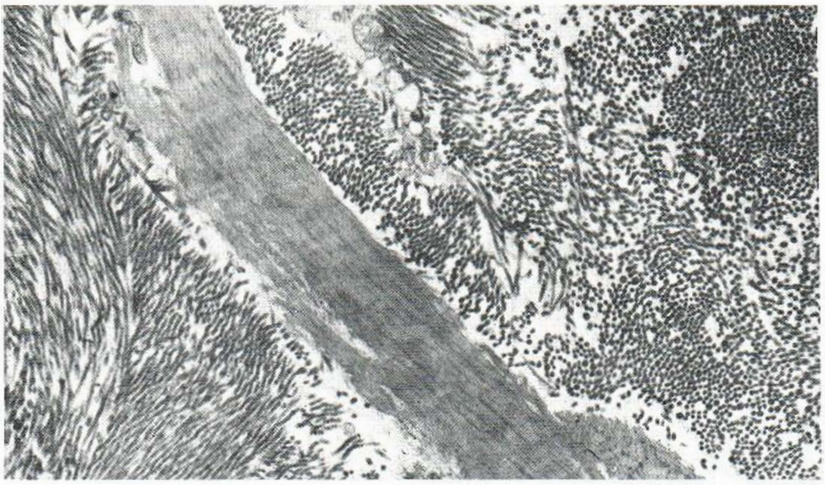

Fig. 3. Elastic fibre from the upper part of the dermis. Original magnification ×12 000.

did not differ from that of normal skin and the fine structure of collagen fibrils was normal.

The sparse perivascular mononuclear cell infiltrates found in the superficial dermis contained mast cells, lymphocytes, plasma cells and histiocytes. Fibroblasts were normal.

Therapy

Treatment with systemic prednisone, methotrexate, dapsone, various antibiotics, tar, silver nitrate and Grenz-X-Rays was of no effect.

Local treatment with potent steroids holds the skin lesions in check. Treatment with retinoid acid 50 mg daily has afforded some relief.

DISCUSSION

Acrokerato-elastoidosis Costa belongs to a group of diffuse palmo-plantar keratoses (5, 6, 7). The disease is regarded as an autosomal dominant hereditary disorder (7), although cases without hereditary predisposition have been described (2, 3, 6).

The skin manifestations usally occure before the age of 20 years (7), but later onset has been described (6). However, in our patient the onset was more violent and the general condition more seriously affected than in previous reported cases.

Localized hyperhidrosis is now regarded as belonging to the clinical picture of acrokerato-elastoidosis (7).

The most important differential diagnoses are: Kerato-elastoidosis marginalis of the hands (Kocsard), acrokeratosis verruciformis (Hopf), diffuse palmo-plantar keratoderma (Thost-Unna), progressive palmo-plantar keratoderma (Greither), and

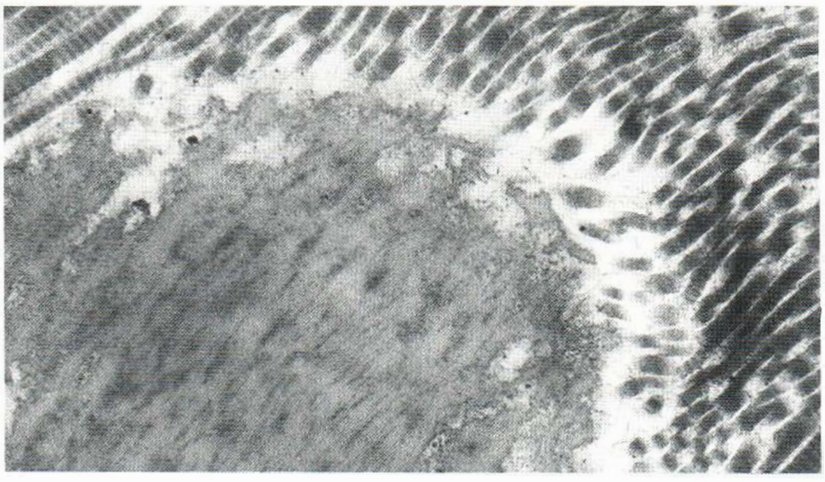

Fig. 4. Elastic fibre from the deep part of the dermis. Original magnification ×48 000.

mutilating keratoderma (Vohwinkel). Only the latter three diseases are frequently accompanied by hyperhidrosis (5, 7). In kerato-elastoidosis marginalis of the hands (8) the keratosis is linear and localized to the radial margin of the index finger and to the ulnar margin of the thumb, and hyperhidrosis is not present. In acrokeratosis verruciformis the warty papules are flat or convex and isolated in small groups. This condition is often present at birth or has its onset during early childhood, similar to diffuse palmo-plantar keratoderma, whereas acrokerato-elastoidosis has its onset later in life.

In diffuse palmo-plantar keratoderma the hyperkeratosis is surrounded by a band of erythema. In mutilating keratoderma the keratosis are starfish-shaped and cicatricial alopecia can be seen. In progressive palmo-plantar keratoderma, hyperhidrosis is not always present, and if so only localized to the palms (5).

Electron microscopy of biopsies taken from the skin of the hands of 2 patients, 47 and 59 years of age, suffering from acrokerato-elastoidosis, revealed pronounced changes of the dermal elastic tissue in the form of granular disaggregation (7). Similar changes were absent in the present case. In our patient, however, the biopsies were taken from non-sun-exposed areas on the lower legs. The elastic fibre changes reported by Jung et al. (7) apparently do not differ morphologically from the changes found in the skin of older people and in senile skin, especially in chronically sun-exposed skin (4). We therefore suggest that the ultrastructural changes found by Jung et al. (7) are probably non-specific.

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Granulomatous Secondary Syphilis Coinciding with PUVA Treatment in a Patient with Psoriasis: An Apparent Failure of PUVA Therapy

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Abstract. A 39-year-old man is described who has been suffering from psoriasis for many years. Because of the extensive involvement of the skin he was treated with PUVA therapy, initially with a good result. A second course of PUVA treatment was unsuccessful. Histopathological examination of the (persisting) skin lesions at this time showed clear granulomatous infiltration, in addition to psoriatic changes. Based on history, serological and dark-field examination, a secondary syphilis was diagnosed. The possible influence of PUVA therapy is discussed.

Key words: Psoriasis; Syphilis; Granulomatous reaction

It is generally accepted that skin lesions during secondary syphilis can show great variability in appearance and number (7). One of the clinical forms resembles psoriasis. Currently, PUVA therapy is widely used in the treatment of patients with psoriasis (13, 18, 19). The influence of PUVA treatment on other (skin) diseases is not well established. The development of systemic lupus erythematosus (3), parapemphigus (17) and possibly the development of skin cancer (16) have been described. In this paper we report a patient who was treated with PUVA therapy for psoriasis. During this treatment he developed a secondary syphilis.

REPORT OF A CASE

A 39-year-old man, skin type III (4) had been suffering from psoriasis vulgaris since he was 11 years old. For the