0.95 and 8.6% with 0.99 confidence probability, respectively), it does not suggest either that routine ophthalmological screening of all patients with AD is indicated.

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Pyoderma Gangrenosum Associated with Transient Acantholytic Dermatosis (Pemphigus Erythematosus-like) and Paraproteinemia

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Abstract. A 69-year-old man with recurrent eruptions of pyoderma gangrenosum for 4 years is described. The patient also suffered from paraproteinemia (increased IgA with M-component) and transient acantholytic dermatosis resembling pemphigus erythematosus. He had no intestinal symptoms.

Key words: Pyoderma gangrenosum; Paraproteinemia; Subcorneal acantholytic bulla formation; Pemphigus erythematosus-like eruption; Transient acantholytic dermatosis

Pyoderma gangrenosum is an ulcerating inflammatory skin condition of unknown origin, but with typical clinical characteristics. Pyoderma gangrenosum is considered to be an auto-immune disease and is often accompanied by chronic ulcerative colitis, rheumatoid arthritis and paraproteinemia.

Pemphigus erythematosus is also regarded as an auto-immune disorder. It is extremely rare in Denmark.

This case report presents a patient suffering from pyoderma gangrenosum, paraproteinemia, and transient acantholytic dermatosis resembling pemphigus erythematosus.

CASE REPORT

A 69-year-old man with no family history of skin disorders. At the age of 65 years the patient developed a painful bluish discoloration on his left lower leg without any preceding trauma. Treatment with systemic and topical antibiotics was instituted, but he developed a putrid, necrotic ulceration measuring 14×16 cm. The edges were violaceous, elevated and undermined (Fig. 1). The patient had no intestinal symptoms.

Histopathological examination showed pseudoepithelial hyperplasia and dermal abscesses with inflammatory infiltrate of neutrophil granulocytes and lymphocytes. Many fibroblasts and blood vessels were present. Fibrinoid necrosis was present in the vascular wall.

The patient was treated with salicylazosulphapyridine 3 grams daily and topical antibiotics. As this treatment was without effect, a split-skin transplantation was performed, and the ulcer healed.

One year later the patient developed non-pruritic erythematous crusted skin lesions, mainly on the trunk, and for a short time there were facial lesions too. In some places small bullae were present, but no papules. The lesions healed spontaneously, forming brownish crusts.

Histopathological examination revealed an acanthotic epidermis, subcorneal bulla formation with neutrophil granulocytes and acantholytic cells, and in the dermis perivascular infiltration with lymphocytes and granulocytes was found. Direct immunofluorescence microscopy (from several lesions on the trunk) demonstrated only fluorescence of complement C3 at the dermo-epidermal junction. Indirect immunofluorescence microscopy was negative. Fluorescence could not be demonstrated in skin biopsy specimens from normal skin.

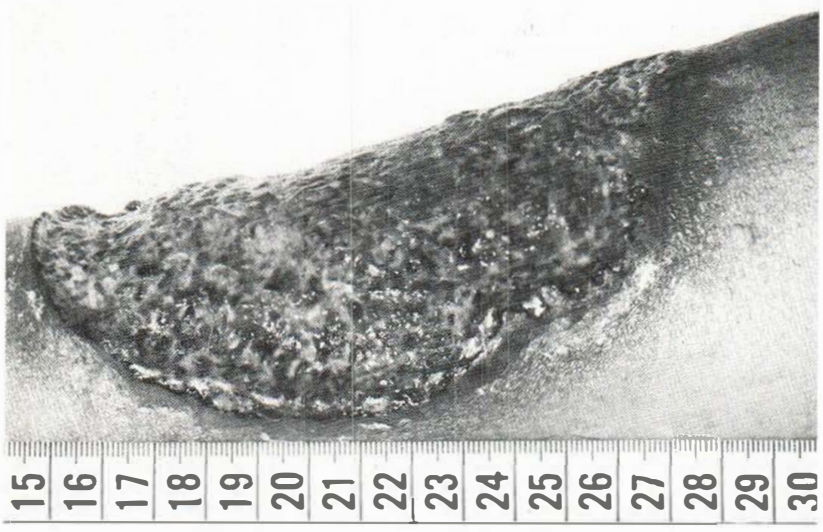

Fig. 1

At the same time the patient developed a 20×20 cm pyoderma gangrenosum lesion in the lumbar region.

Again the patient was treated with salicylazosulphapyridine and topical antibiotics, and the pemphigus-like lesions with topical corticosteroids. The ulcer healed as also did the blister lesions.

After a period of 2 years the patient again developed a pyoderma gangrenosum lesion, now in the groin and erythematous bullous skin lesions on the trunk (Fig. 2). Once more these pemphigus erythematosus-like lesions were treated with topical steroid and the pyoderma gangrenosum ulcer with clofazimine (Lampren®) 300 mg daily for 9 weeks. The ulcer healed, but the bullous eruption remained unchanged. The only side effect of the treatment with clofazimine was a pronounced, diffuse redness of the skin and a reversible corneal epithelial dystrophy.

Laboratory data showed an increased sedimentation rate (45 mm/hour), increased complement C3 and C4, increased IgA: 14.2 (normal range 0.6–3.4 g/l) with a M-component, while IgG and IgM were normal; antinuclear antibodies (ANA), rheumatoid factor and LE cells could not be demonstrated. Other routine hematological examinations were normal. X-ray examination of lungs and colon showed no abnormalities. Biopsy material taken from the mucous membrane of the rectum showed no pathological changes.

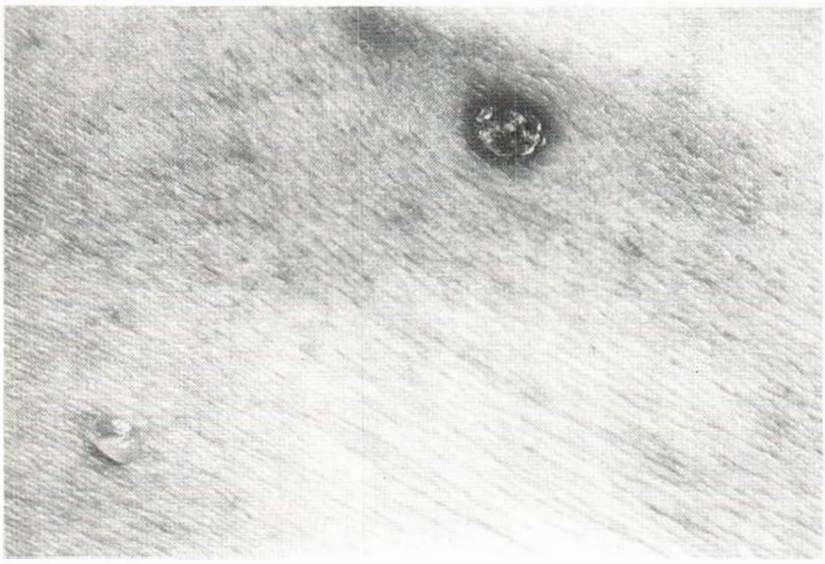

Fig. 2

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