

Systemic Sarcoidosis with Necrobiosis Lipoidica-like Scalp Lesions

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Abstract. A 78-year-old woman is described, having systemic sarcoidosis for 15 years, involving the lungs, lymph nodes and the skin. Over the last 6 years she developed a progressive cicatricial alopecia with histologic changes of the granulomatous type of necrobiosis lipoidica surrounded by sarcoid granulomas.

Key words: Sarcoidosis; Necrobiosis lipoidica

The clinical and histological distinction between the granulomatous type of necrobiosis lipoidica and cutaneous sarcoidosis presents difficulties (3). Both are capable of provoking scalp involvement (5). This report presents an unusual case of systemic sarcoidosis with necrobiosis lipoidica-like changes developing in scalp lesions.



Fig. 1. Cicatricial alopecia with atrophic centre containing superficial ulcerations, teleangiectasia and yellowish deposits, surrounded by a reddish brown, infiltrated and demarcated border.

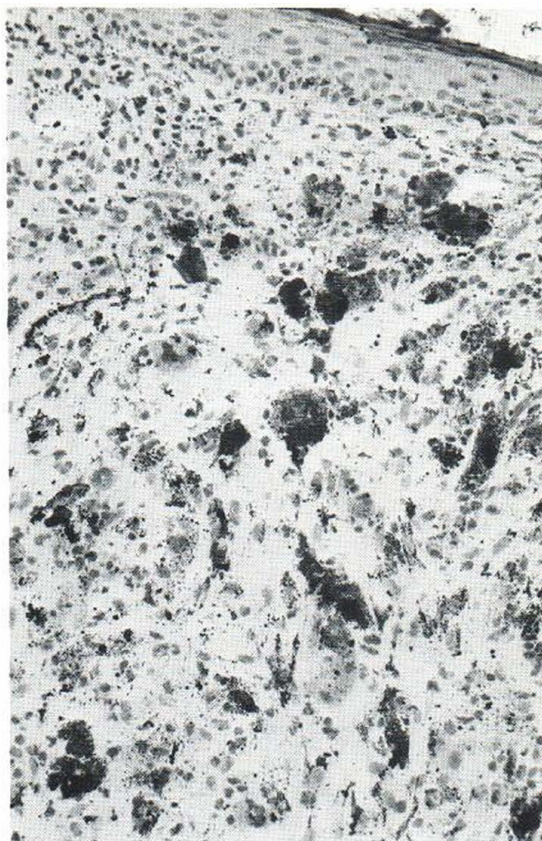


Fig. 2. Lipid deposits extracellularly and intracellularly in lipophages and giant cells. Biopsy taken from the atrophic centre. (Oil Red O, $\times 165$.)

CASE REPORT

A 78-year-old woman was seen at our clinic for the first time in 1975. Since 1961 she had had systemic sarcoidosis involving the lungs, lymph nodes and the skin of the face and scalp. The diagnosis had been verified by mediastinal lymph node biopsy and skin biopsies from lesions on the face and scalp. X-rays of the lungs showed diffuse fibrosis though without progression over the years of observation. The lesions on the face and scalp remained unchanged up to 1969, but over the last 6 years they progressed slowly.

Clinical examination revealed a cicatricial alopecia covering an area 20×10 cm in the frontoparietal region (Fig. 1). Most of the lesion was atrophic, with superficial ulcerations, teleangiectasia and sprinkled with yellowish deposits. The border of the lesion was reddish brown, infiltrated and sharply demarcated. Three smaller elements were seen on the face.

Laboratory investigation. Hemoglobin, ESR, leukocyte count, liver function tests, serum calcium, and urine analysis were all within normal limits. Wassermann reaction was negative. Mantoux was negative. Kveims test was negative (Finsen Institute, 1976). X-rays of hands,

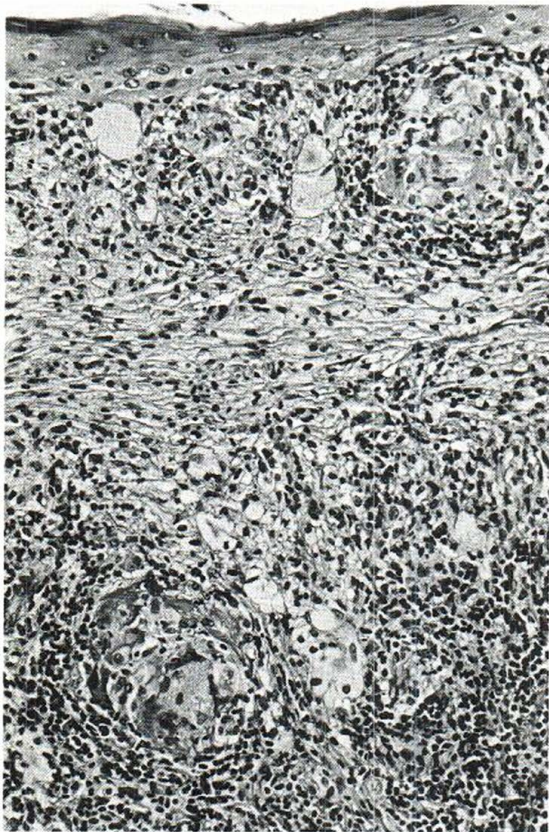


Fig. 3. Granulomatous infiltrate containing epithelioid cells, lymphocytes and giant cells of foreign body type. Biopsy taken from the border. (H. + E., $\times 165$.)

feet and skull were normal. Ophthalmological examination was normal. Fasting blood sugar was elevated to 12.2 mmol/l. Glucose tolerance test with 50 g per os showed a diabetic curve with a maximal plasma glucose level of 15.9 mmol/l and a 2½-hour value of 12.2 mmol/l. Total plasma lipid (10.8 g/l) and triglyceride (2.74 mmol/l) levels were slightly elevated, showing a hyperlipemia of Friedrichson type IV. X-ray of the lungs showed diffuse fibrosis.

Histopathology. Hematoxylin and eosin stained preparations of skin biopsy material from the atrophic, ulcerated centre of the alopecia showed granulomatous accumulations containing giant cells of foreign body type, epithelioid cells and lymphocytes, as well as a large number of lipophages. Changes in vessel walls were not observed, however. In a few areas, thickened collagen fibres, occasionally of homogeneous appearance, were seen. Oil Red O stained preparations revealed large amounts of both extracellularly and intracellularly located lipid deposits (Fig. 2). Verhof's staining for elastic tissue showed an absence of elastic fibres in the granulomatous areas, whereas the reticulin staining of Gordon and Sweet in those areas revealed a network of reticulin fibres.

Another skin biopsy from the infiltrated, reddish brown border of the alopecia showed granulomatous accumulations similar to those described above, but with only a few lipophages and no extracellularly located lipid deposits. Thickened collagen fibres were not observed (Fig. 3).

DISCUSSION

Scalp lesions of necrobiosis lipoidica with sarcoid reaction have been described previously (2, 7). In the present case of systemic sarcoidosis, the clinical picture combined with the microscopical finding of large amounts of extracellular lipids and lipophages in the granulomas could indicate the presence of necrobiosis lipoidica. In sarcoid lesions, lipid deposits are seldom found and then only within epithelioid cells and in small amounts (4). The spotty appearance of thickened, homogeneous collagen fibres supports the diagnosis of necrobiosis lipoidica (3). Another possibility is that the histologic changes described in the centre represent an extreme development of the sarcoid process, which has not been reported hitherto. On the other hand, the surrounding sarcoid granulomas could be taken as secondary responses to local processes of a different character (1). Savin (6) has reported a case similar to the present one, though without histological confirmation. Wilson Jones (8) presented 29 cases of necrobiosis lipoidica of the granulomatous type localized to the face and scalp, and pointed out that facial necrobiosis was a distinctive condition. He excluded patients who had evidence of sarcoidosis in other areas. The present case is believed to represent a rare coincidence of two different diseases localized to the scalp: necrobiosis lipoidica of the granulomatous type centrally and sarcoid granulomas in the periphery. The granulomatous type of necrobiosis lipoidica is less often associated with diabetes than is the vascular type (3).

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Tranexamic Acid (Cyklokapron®) in Chronic Urticaria: A Double-blind Study

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Abstract. A double-blind study with tranexamic acid (Cyklokapron®) was carried out in 17 patients with chronic urticaria. All patients had slightly depressed C₁-esterase inhibitor value. No significant differences were found between TA and placebo treatment periods.

Key words: Chronic urticaria; Tranexamic acid

Within recent years several workers have reported good results with the plasmin inhibitor epsilon-aminocaproic acid or its analogue tranexamic acid (TA), in hereditary angioneurotic edema (HAE) (1, 3, 4, 5, 7).

HAE manifests itself by attacks of edema in subcutaneous as well as in submucous tissue. The disease in general is believed to result from an inborn defect in the synthesis of a serum α_2 -globulin that inhibits the first component of complement, and most patients with HAE have, besides a family history, very low values of C₁-esterase inhibitor (2).

Chronic urticaria (CU) is often followed by angio-edema, and in some cases patients with CU may be found to have slightly depressed C₁-esterase inhibitor values. In preliminary studies (6)

a number of these patients seemed to benefit from TA. The purpose of the present investigation was to test the possible effect of TA in patients with chronic urticaria combined with depressed C₁-esterase inhibitor value, in a double-blind study.

MATERIALS AND METHODS

The trial was carried out on 17 patients, 13 women and 4 men, aged 10–60 years (average 34.6 years). The mean C₁-esterase inhibitor value was 90 units, range 72–100 (normal: 101–172 units). The randomized double-blind study lasting 9 weeks was split up into a 4 week treatment period with TA or placebo, 1 week without treatment, followed by a 4 week cross-over period with placebo or TA. The dose of TA was 1 g three times daily.

The patients recorded daily the severity of urticaria, angioneurotic edema and itching. The physician's evaluation was performed once weekly, together with a laboratory investigation including a leukocyte and differential count, se-creatinine, GP-transaminases and a urine examination for albumen and sugar.

RESULTS

The results of the study can be seen in Table I. No statistically significant differences were recorded between treatment period for TA and placebo. All laboratory tests were normal throughout the study. The only side effect noted was diarrhoea (reported by one patient).

DISCUSSION

Although antihistaminics may be helpful in CU, their value is often limited, and alternative treatments have to be sought. Activation of plasminogen and formation of plasmin appears to be an important factor in HAE (5). Plasmin formation may also lead to formation of kinins, which can induce urticaria. It was therefore natural to try TA in CU, especially in patients with low C₁-esterase inhibitor values.

Table I. *Results of treatment expressed in average units for severity of disease and itching $\pm$ S.D.*

0=no reaction or itching, 1=minor reactions or itching, 2=moderate reactions or itching and 3=severe reactions and intense itching

Period	Urticaria	Angio-edema	Itching
TA	0.88 $\pm$ 0.8	0.45 $\pm$ 0.69	0.98 $\pm$ 0.92
Pause	1.03 $\pm$ 0.86	0.64 $\pm$ 0.85	1.09 $\pm$ 0.93
Placebo	0.92 $\pm$ 0.72	0.40 $\pm$ 0.64	1.07 $\pm$ 0.86