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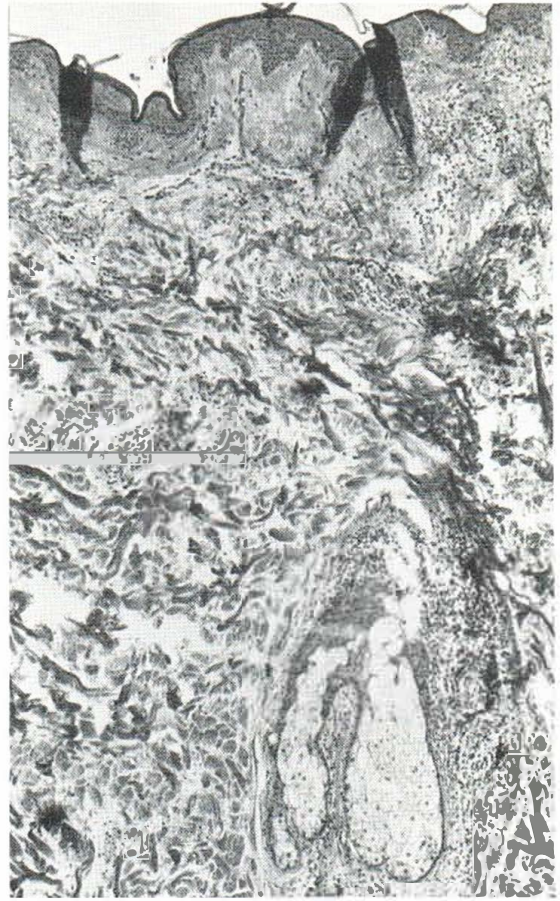


Fig. 1. A perivascular lymphocytic infiltration, especially marked around a hair follicle with its sebaceous gland. No mucinous deposition in this area. (Original magnification,  $\times 11$ .)

## Reticular Erythematous Mucinosis Syndrome: Report of a Case

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**Abstract.** A 47-year-old man with erythematous, maculopapular rash on the chest and the back. Histology showed perivascular infiltration of lymphocytes, especially around hair follicles, and alcian green positive deposits in the dermis. Some improvement was achieved by treatment with 200 mg oxychloroquine sulphate (Plaquenil, Winthrop) twice daily.

**Key words:** REM syndrome; Reticular erythematous mucinosis syndrome; Mucinoses

The reticular erythematous mucinosis syndrome is a rare disease. It was first described in 1974 by Steigleder et al. (3, 4). Their 4 patients (3 men and 1 woman) had all for some years been suffering from a then unknown dermatosis. As a preliminary diagnosis, Steigleder coined the term "round-cell-erythematosis". The rash affected the skin on the chest and upper part of the back. It was a more or less sharply outlined sheet or net-like erythema, with a slightly infiltrated centre. There were some complaints of itching and also of aggravation upon sun-exposure. In each case, histology showed pronounced perivascular infiltrates of round cells—predominantly lymphocytes—and an alcian blue-staining substance in the dermis. The material was not metachromatic and did not stain with



Fig. 2. Thread-like deposits of mucin between collagen fibres in the reticular dermis. (Original magnification,  $\times 114$ .)

mucicarmine. The epidermis showed no atrophy and there were no changes in the PAS-positive membrane.

In 1975, Smith et al. (2) described a case of REM syndrome. Also in 1975, Steigleder (5) reported on a patient with features of both reticular erythematous mucinosis and a plaque-like form of cutaneous mucinosis. In 1960, Perry et al. (1) also described patients with a plaque-like form of cutaneous mucinosis.

#### CASE REPORT

Our patient is a 47-year-old male with no previous illness, who in 1959 developed an erythematous, maculopapular rash in the sternal area and between the scapulae. The borders of the erythema were rather indistinct in some places and in others formed small, tongue-like spurs into the neighbouring, normal skin. During the following years

there was a slow, symmetrical progression of the skin manifestations. The patient was in good general health and had no itching or complaints connected with internal organs. There was no progression after sun exposure and no drugs were used.

*Laboratory findings:* ESR 3 mm/hr. Treponemal serological tests were negative. No deposition of immunoglobulins and complements could be detected by direct immunofluorescence studies on the affected skin. The serum levels of immunoglobulins were normal. Serum antibodies against thyroglobulin were absent. Thyroid function tests were normal.

Xenon lamps using various Schott filters were used to investigate the sensitivity to UVA and UVB and invisible wavelengths showed normal reactions.

*Histology.* From 1960 to 1976 seven biopsies from this patient have been examined. The histologic picture has shown some variation. However, most of the biopsies demonstrated deposition of a fibrillar (sometimes granular) material between the collagen fibres in the upper part of dermis. This material stains green with alcian green and weak red with mucicarmine. It does not (or at least only very weakly) stain metachromatically with toluidine blue. The material is found not only in the neighbourhood of the adnexal structures, but also at some distance from these, and it does not seem to be especially connected with the blood vessels. In addition, most of the biopsies showed a fairly dense perivascular infiltration of lymphocytes. This infiltration was found throughout the dermis, especially around the hair follicles. There was no basal cell degeneration of the epidermis or the hair follicles. In the upper part of the dermis there was slight edema.

#### Staining methods

- (i) Alcian green: 1% aqueous solution in 1% acetic acid;
- (ii) Toluidine blue: 1% aqueous solution using standard method;
- (iii) Mucicarmine: Southgate's method.

#### Therapy

The patient was treated with 200 mg oxychloroquine sulphate (Plaquenil, Winthrop) twice daily, and some improvement was observed after 9 weeks.

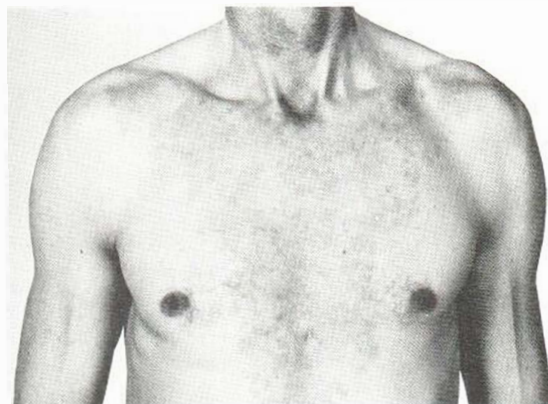


Fig. 3. Clinical lesion over the skin of the chest.

## DISCUSSION

In the present case report, the patient had been observed at our department repeatedly during several years before the actual diagnosis was established. We have found the clinical and histological picture described earlier to be characteristic of the syndrome. Histologically, the often dense lymphocytic infiltrates around the blood vessels and especially around the hair follicles seem to be just as characteristic as the mucinous deposits. These cellular infiltrates look like those found in lupus erythematosus. However, the lack of basal cell degeneration in the epidermis and in the hair follicles excludes this diagnosis. This fits well with the negative result of immunofluorescence studies, and the normal serum level of immunoglobulins.

Steigleder et al. (3) have described some relation to sun exposure, but our patient had no such symptoms, and the light sensitivity tests were negative. Steigleder et al. (3) treated their patients with an oral antimalarial drug (Rhetis, Winthrop) chloroquine diphosphate 65 mg, hydroxychloroquine sulphate 50 mg, mepacrine hydrochloride 25 mg.

After some weeks improvement was noted. We used 200 mg oxychloroquine sulphate (Plaquenil, Winthrop) twice a day for our patient, and after 9 weeks of treatment, partial regression was seen. Future controls will show if the improvement is continuing, or if there will be relapse in spite of continued treatment.

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