Absence of Eccrine Sweat Glands without Other Ectodermal Defects

Lennart Juhlin

Department of Dermatology, University Hospital, Uppsala, Sweden

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Abstract. A unique case of total anhidrosis is reported, illustrating the problems of life without sweat glands.

Key words: Anhidrosis; Eccrine sweat glands; Sweating

Partial or complete absence of sweating is seen in patients with ectodermal dysplasia. They also have defects of hair, dentition and facial features. Anhidrosis due to the absence of eccrine sweat glands in a normal-looking person has to my knowledge not been described before. Such a patient and her problems are now reported.

CASE REPORT

The patient is a 26-year-old popular singer. Her mother also suffers from an inability to sweat, but one sister and two brothers are normal in this respect. The patient has been unable to sweat since birth, though a few minute beads of perspiration can occasionally be seen under her eyes. Hair, nails, teeth and facial features appear normal. Her feet in particular are dry and must have daily applications of cream to keep them soft. When she gets warm her skin turns reddish, she gets headache, feels sick and nauseated. Her body temperature rises to over 38°C. She becomes swollen, especially under the eyes and on the hands and feet. She must always try to keep herself cool. Since she was a child she has never been able to run fast, work hard physically, or take a sauna. The symptoms always arise in hot weather and when in her profession as a singer she is exposed to heat from the lamps used on stage and in television studios.

During recent years she tires more quickly and is unable to walk more than a few hundred metres in warm weather before getting short of breath and feeling that her heart is beating rapidly. In 1978 she appeared for medical consultation at a hospital. Her blood pressure was then 195/130 on both arms after rest. She had normal urinary sedimentation but slight proteinuria. Creatinine clearance rate was normal. Heart volume was 680 cc/m² of body surface, which is the upper normal limit. Echocardiography and electrocardiogram were normal. After treatment with thiazides and a betablocking drug her blood pressure became normal but her subjective symptoms remain unchanged. Serumalbumin, lipid and electrolytes tests proved normal. There were no signs of angiokeratoma corporis diffusum (Fabry). Tests for α-galactosidase and trihexosides in leukocytes from our patient and her mother were normal and eye examination showed no abnormality.

Tests for sweating were done on the hands, arms, feet, face, back and in the axillae, both after light exercise and also after intradermal injection of epinephrine 0.2 ml 1/10 000 and mecholyl 1/10 000. No eccrine sweating could be detected by the starch iodine method or after staining with orthoptalaldehyde (1). After injection of epinephrine, 5–8 functioning apocrine glands/cm² were registered in the axillae with the OPT method. Skin biopsies from the abdomen and arm revealed no ducts or glands. Nor could any sweat duct orifices be seen on replicas taken with cyanoacrylate (2) from various parts of the body.

Over the left clavicle there is a brachial cyst opening, a few mm² in area from which a viscous clear secretion exudes when she gets warm. Salivation and tears appear normal, as do the skin surface lipids (causal value over forehead 150 μg/cm²).

Various types of creams and water-based solutions have been tried, with limited success. A cold environment gives her the best relief.

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Necrobiosis Lipoidica in a Patient with Generalized Scleroderma: A Case Report

Gunhild Lange Wantzin and Takasi Kobayasi

Department of Dermatology, University of Copenhagen, Rigshospital, DK-2100 Copenhagen, Denmark

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Abstract. A 48-year-old woman with generalized scleroderma of the acrosclerosis type developed several lesions clinically, histologically and ultrastructurally, which were characteristic of necrobiosis lipoidica. The lesions were located to scar tissue. An increased collagen synthesis is suggested as a possible explanation.

Key words: Scleroderma; Necrobiosis lipoidica; Scar tissue

The clinical and histological picture of necrobiosis lipoidica (N.l.) has been known since 1930 (7). N.l. is a rare condition occurring in about 3 per thousand

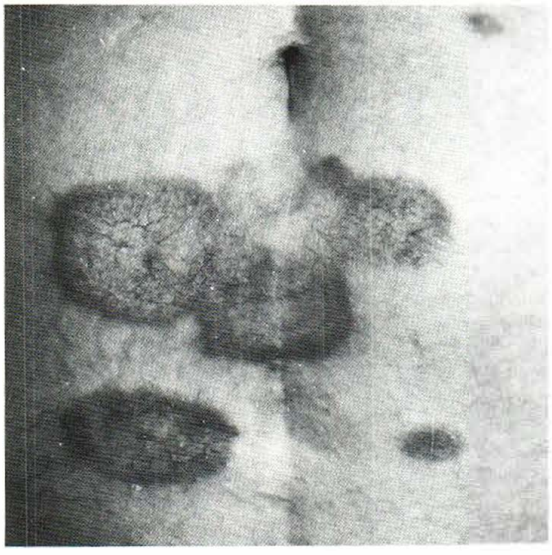

Fig. 1. Typical necrobiosis lipoidica lesion on the abdomen, with central atrophy and yellowish discolouration, surrounded by a reddish-brown margin. Numerous teleangiectases appear over the lesion.

diabetics but is not related to the duration or severity of this disease (10). Even non-diabetic patients have been known to develop N.l. (2). About 70% of patients with N.l. have diabetes mellitus (2). Necrobiosis may involve most regions of the body but is mainly seen on the lower legs and, in that case, is mostly related to diabetes (10). Recently, localization of N.l. to scar tissue has been reported (9). The present report concerns a patient with generalized scleroderma with clinical, histological and ultrastructural necrobiosis lipoidica on the abdomen, lower right arm and left thigh.

CASE REPORT

This 48-year-old woman had always had cold hands and feet. Since 1960, following a gynecological operation, she developed Raynaud's phenomenon. In 1963 thoracal sympathectomy was carried out, with no beneficial result, and in 1964 prednisone treatment was started, also without any effect. Early in 1965 she was referred to the Dermatological department where active, progressive generalized scleroderma of the acrosclerosis type was diagnosed. The sclerosis involved arms, face, upper part of the chest and lower part of the lower legs. Flexion contractures of fingers and elbows were observed.

Initially the patient was treated with benzylpenicillin diethylamino hydro-iodide-ethyl-ester (Leocillin®) with some success until 1966, when treatment with penicillamine, 900 mg daily, was started and maintained up to the time of reporting. Three times the treatment with penicillamine had to be discontinued because of mild side effects. Treatment with glutamine, 300 mg daily, was started in 1977. In 1971, treatment with apresoline was given for 11 months. After 7 years, the sclerosis regressed—except on the hands, where the contractures of the fingers persisted.

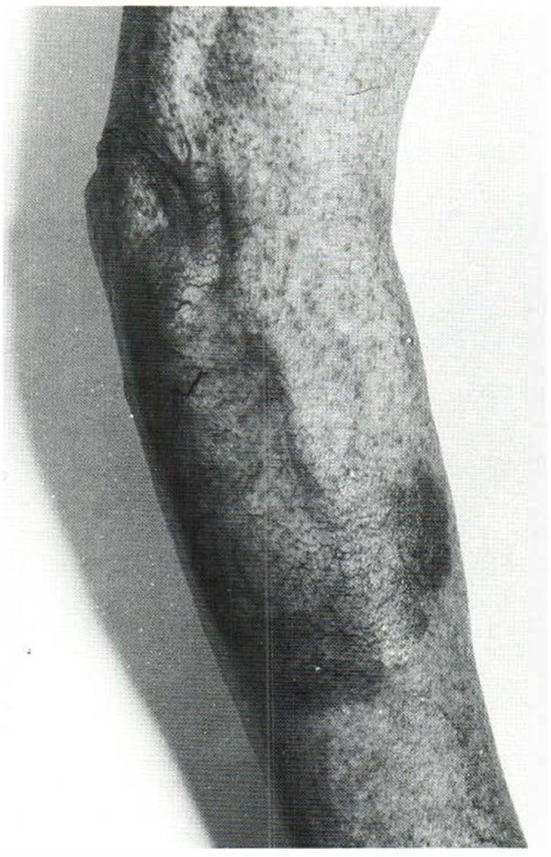

Fig. 2. Typical necrobiosis lipoidica lesions on the right lower arm.

In 1971, when the patient was 40 years old, five 1–5 cm lesions were observed on the abdomen (Fig. 1). Clinical examination revealed round indurated lesions with central atrophy and yellowish discolouration surrounded by a reddish-brown margin. Numerous teleangiectases appeared over the lesions. The clinical diagnosis necrobiosis lipoidica was supported by histology. The histopathological examination showed dermal changes with perivascular infiltration of lymphocytes, plasma cells, histiocytes, mast cells, fibroblasts and giant cells. The collagen was hyalinized and necrobiotic. Nuclei were missing from a large area. In some parts, the infiltrate was granulomatous.

In 1972, a 12×5 cm lesion developed on the right lower arm (Fig. 2). The clinical appearance was identical with the alterations on the abdomen described one year earlier. The lesion appeared in an area where a muscle biopsy had been taken one year previously.

In 1978, a 2×2 cm indurated, yellowish lesion surrounded by a reddish-brown margin and with teleangiec-

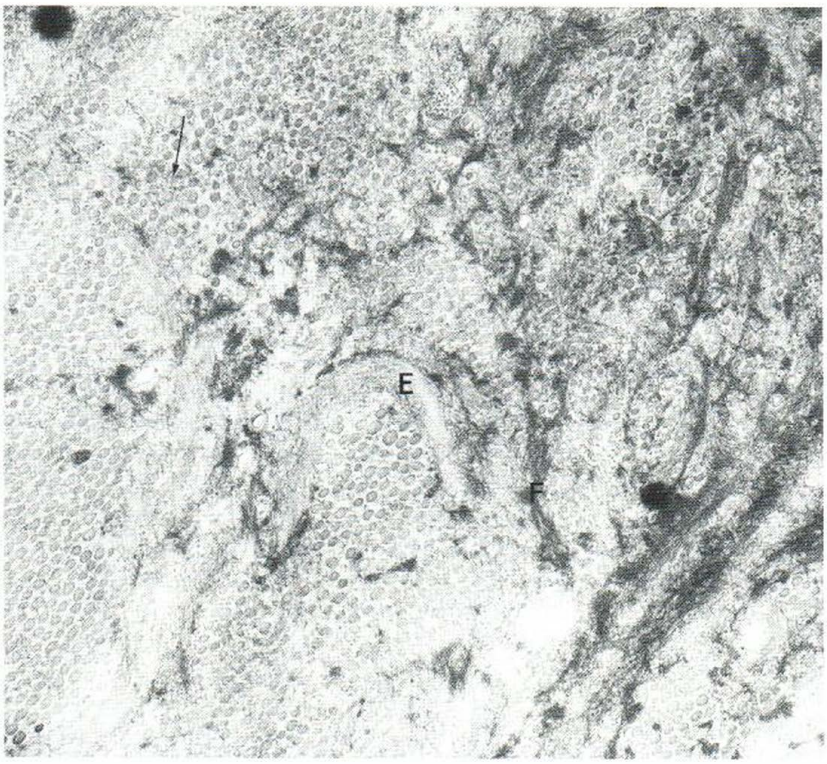

Fig. 3. Dermal fibrils in the palisade-like cell infiltrate. E=Elastic fibre. F=Fibrin. Arrow, granular material. ×30 000.

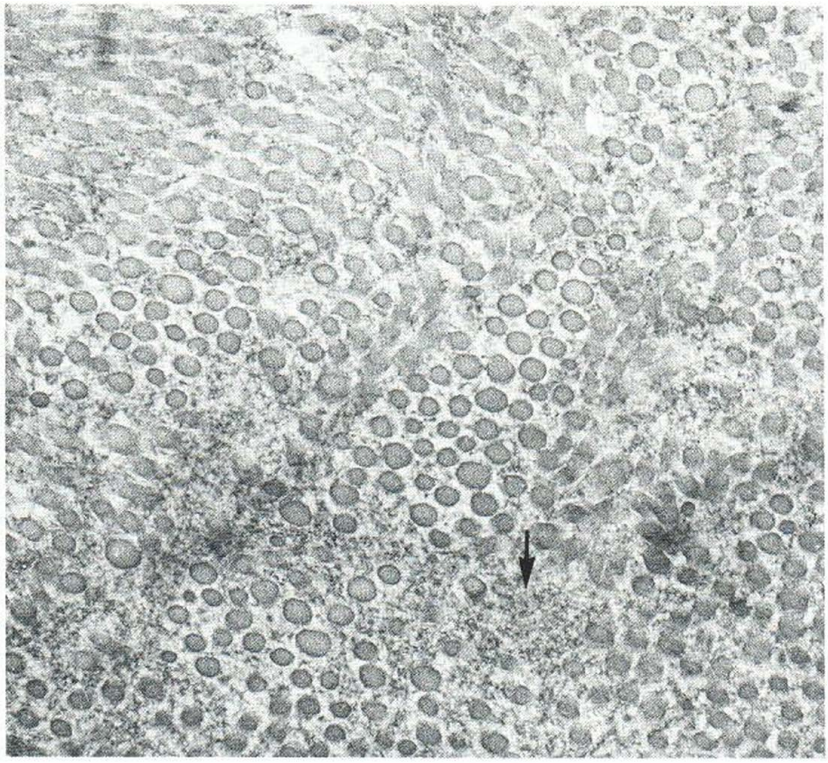

Fig. 4. Collagen fibrils with various diameters. Some fibrils show an irregular arrangement. Dense granular material is indicated by an arrow. ×60 000.

tases was observed on the left thigh. The lesion had developed on the same spot where a skin biopsy had been taken one year before.

All the described lesions (i.e. on the abdomen, right lower arm and left thigh) were identical and clinically diagnosed as necrobiosis lipoidica.

Electron microscopy

The dermis showed histiocytic cell infiltration with a tendency to palisade arrangement. In the indistinct palisade, collagen fibrils, a few elastic fibres, fibrin and dense granular material were found (Fig. 3). The diameters of the collagen fibrils varied between 33 and 100 nm. Some collagen fibrils were arranged unparallel in the bundle, while the others appeared bent and wavy (Fig. 4). Elastic fibres showed blurred contours and there were no tubular fibrils.

Laboratory investigation

Hemoglobin, ESR, leukocyte and thrombocyte counts, immunoglobulins, liver function tests, se-calcium, se-phosphorus, LE cells, standard test for syphilis and urinalysis were all within normal limits. Fasting blood sugar and glucose tolerance tests were normal.

DISCUSSION

The diagnosis necrobiosis lipoidica (N.l.) is not always clear-cut, as the lesions may resemble granuloma anulare, rheumatoid nodule, xanthelasma and Miescher's granuloma disciformis (2, 6). Clinically, however, this patient demonstrated a clear case of N.l., the diagnosis being supported by light- and electronmicroscopical examination. As reported by Muller & Winkelmann (6), N.l. may be subdivided into two groups, one mainly occurring in diabetic patients, another in non-diabetics, showing more prominent multinucleate giant cells, histiocytes and epithelioid cells. Non-diabetic N.l. lesions generally appear on the face and scalp (10). In the present case, the lesions were found on the abdomen, the lower right arm and the left thigh—but not on the lower legs, which is the main location when combined with diabetes.

The first lesions on the abdomen appeared in the area of a scar, and the other two lesions in old scars left after biopsies. In sclerodermal skin, the epidermis is thin and atrophic, while the dermis is thickened and hypertrophic. Both light- and electronmicroscopy demonstrate the dermal thickening to be caused mainly by increased quantities of collagen fibres (4). Ultrastructural studies on N.l. have shown rather compact collagen fibril bundles and few disintegrated elastic fibres in the necrobiotic areas, besides numerous acid mucopolysaccharide filaments and thickened vascular walls. The infiltrating cells were the ones found by light microscopy (5).

In the present study, the findings are suggestive of necrobiotic changes. Small arterial and arteriolar lesions were found simulating those described in scleroderma, i.e. consisting of concentric acellular thickening of the intima, with narrowing or occlusion of the lumen (3). Diabetic 'micro-angiopathy' has been suggested to underlie the pathogenesis of N.l. (1, 8). In non-diabetic N.l., vascular changes are less marked, which means that some other explanation of the aetiology is needed. Recently, Sahl reported N.l. lesions in scar tissue and suggested that immunological factors may determine the localization of the lesions (9). Both in active scleroderma and in scar tissue an increase of collagen synthesis takes place. This added synthesis of collagen may possibly have triggered off the necrobiosis lipoidica lesions.

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