Absence of Eccrine Sweat Glands without Other Ectodermal Defects

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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

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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