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Normal Sweating and Tear Production in Congenital Ichthyosiform Erythroderma with Deafness and Keratitis

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Abstract. A 24-year-old female patient with congenital ichthyosiform erythroderma, deafness and vascularizing keratitis yet also exhibiting normal sweating and tear production when tested is reported. This unique finding indicates that a certain subgroup of patients with this generalized ectodermal disturbance may benefit from treatment with the aromatic retinoids.

Key words: Ichthyosiform erythroderma; Sweating; Tear production; Aromatic retinoids

The group of ichthyosiform dermatoses comprises the normokinetic ichthyoses (ichthyosis vulgaris and X-linked ichthyosis), the hyperkinetic forms (epidermolytic hyperkeratosis and the lamellar ichthyoses), and a complex category of congenital ichthyosiform syndromes associated with other developmental defects (1). The latter group harbours a rare disorder of ichthyosiform erythroderma with deafness and vascularizing keratitis, which was probably first described by Burns in 1915 (2). A consistent finding in this syndrome is the dry skin, referred to as "anhidrosis", reported by several authors (2-7). Recently, however, we investigated sweat and tear production in a patient with this syndrome and discovered that they both appeared normal.

CASE REPORT

A 24-year-old woman was referred to our Department because of a generalized eruption of the skin which had been present since birth. She was the youngest of nine siblings. There was no family history of this disorder and no consanguinity. Soon after birth the skin became thickened, scaly and reddish. Almost complete deafness was noted at the age of 4. From the age of 11 she had been photophobic and the right eye subsequently displayed marked visual loss. Dentition was incomplete and the teeth were malformed and showed carious signs at an early age. Genital development was uncomplicated and menarche occurred at the age of 16. Her psychosocial progress was considered satisfactory when compared with other deaf children.

On examination, her skin appeared universally ichthyotic with a red hue, especially on the face and limbs, and also showed some scaling. The palms and soles were hyperkeratotic. The scalp hair was rather scanty, eyebrows and eyelashes were sparse, and axillary and pubic hair were totally absent. The nails showed thickening and dystrophic changes.

Ophthalmological examination disclosed keratoconjunctivitis with extensive vascularization of the corneal epithelium with pannus formation of both eyes. Bullous dystrophia was seen in the lower segment of the right cornea but corneal ulcerations were not encountered. The visual acuity of the right eye was grossly diminished (1/6), whereas the left eye enjoyed normal vision. Tear production (Schirmer test) and composition, including IgA, were normal. Audiometric examination demonstrated profound perceptive (neurosensory) deafness in both ears.

Sweating on the forearm and the palm of the hand was investigated by the starch iodine test (Minor test). Sweating was normal on two occasions when compared with a control person. The more quantitative method as described by Thomson & Sutarman (8) was also performed. This test allows the recording of active palmar digital sweat glands. Numerous active sweat glands were encountered, indicating that our patient was not anhidrotic

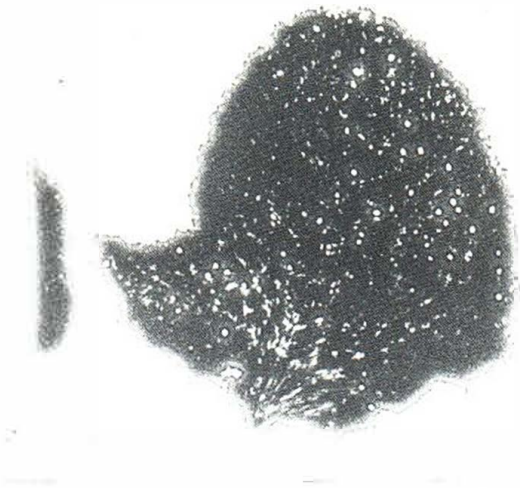


Fig. 1. Distribution of active sweat glands on the finger.

(Fig. 1). The number of active sweat glands seemed only slightly reduced, compared with a sex- and age-matched control.

Basic laboratory findings were either negative or within normal limits, including urine amino acid values. A biopsy specimen from affected skin showed a "basket weave" type of hyperkeratosis, focal parakeratosis and verrucose folding of the epidermis. There was minimal focal vacuolization. The capillaries in the papillary dermis were slightly dilated and small perivascular lymphocytic infiltrates were seen. The deep dermis indicated no abnormalities. Most importantly, the sweat glands appeared normal in distribution and appearance. The acrosyringium of the glands displayed no gross abnormalities. Histological examination of the hair was unrevealing; trichorrhexis nodosa was not demonstrated.

DISCUSSION

The present case was fully described elsewhere, when the patient was 18 years old (9). Allegedly she was anhidrotic, although sweat testing was not then performed. We too noticed that the skin had a dry appearance on physical examination. However, with the starch iodine test and with the method described by Thomson & Sutarman (8), sweating proved more or less undisturbed on two occasions. The number of functional sweat glands on the fingers was only slightly decreased according to the latter test. Intolerance to heat in the summer months, described by other authors (3, 4, 7) was not seen in our case. Finally, on histological examination of affected skin a regular distribution of sweat glands was encountered.

Several authors have emphasized that dryness of the skin or anhidrosis is a consistent feature of the syndrome (2-7). However, very few cases have been tested for sweat production and all showed marked hypofunction of the sweat glands (3, 6, 7). Furthermore, it has been suggested that tear production may also be diminished in these patients (4), although other authors have stressed the normal lacrimation in their cases (6, 7, 10). Thus, there may be a subgroup of patients who do have normal sweat and tear production.

Our observation may have important implications. The therapeutic resorts for this ailment remain unsatisfactory. In theory, the aromatic retinoids may constitute a promising approach for this ichthyosiform skin condition (11). However, if basal tear production is reduced, the side effects of the aromatic retinoids such as xerophthalmia render this treatment a risky procedure. The vascularizing keratitis, often complicated by pannus formation, corneal ulceration and serious impairment of vision may thus be a limiting factor for the institution of this therapy modality, particularly if basal tear production is diminished. It is felt that patients with the syndrome should not be subjected to additional, iatrogenic, risk factors. Therefore, functional tests on the sweat glands—and especially the lacrimal glands—constitute a pre-requisite for treatment with the aromatic retinoids. Unfortunately, informed consent for therapy with the oral retinoid Ro 10-9359 (Tigason) was not obtained in the present case, mainly because of the fear of further loss of visual acuity and the impending hazard of further hair loss due to this modality.

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Loiasis: A Case Report

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Loiasis is one of the filaria infections which affect man, caused by a nematode worm, *loa loa*, endemic in the West and Central African rain forest area up to the Sudan (7). Humans are infected during the bloodsucking phase of the infectious mangrove fly (*Chrysops* spp.) (3). Infective larvae pierce the proboscis sheath of the fly and subsequently burrow through the skin of the mammal host, i.e. man. Development into an adult worm takes 3–18 months. However, already several months after the infection, the typically subcutaneous swellings may appear, most likely an allergic tissue response of the host to toxins from migrating adult worms. These swellings are well known as Calabar swellings and usually disappear within 2–3 days. The swellings, which can be located near or around joints of wrist, knee or ankle can give rise to local pain and restriction of mobility, suggestive of an acute arthritis. The passage of a worm beneath the conjunctiva causes swellings of the eyelids and the worm can even be observed by the patient (1, 6).

Besides these symptoms produced by the adult worm, a *loa loa* infection can go unnoticed, except for a chance laboratory finding indicating the presence of a (hyper)eosinophily. Loiasis imported

into the USA and Europe has been reported several times (2, 4, 8, 9, 10, 11).

We report here a patient with loiasis imported into the Netherlands and from which 2 adult worms could be surgically removed from the skin during therapy.

CASE REPORT

A female patient, 27 years old, had returned to the Netherlands over six months ago after a one year stay in Nigeria. Shortly after her return she started to complain about fugitive swellings around her wrists and lower part of the arm. During the previous few weeks swellings of both eyelids also occurred. The swellings lasted several days. Blood examination revealed an eosinophily of $2400 \times 10^6/l$, no microfilariae were detectable. By using an antibody against microfilariae, a positive serum test was found. IFA 1:64. Diethylcarbamazine citrate (DEC) treatment was started, with a dosage gradually increasing from 1.0 mg to 6.0 mg/kg daily, which is equivalent to 0.5–3.0 mg DEC base/kg. Two days after the treatment was begun, two reddish oedematous swellings appeared, one on the left upper arm and one on the left upper leg. Both caused itching and, on inspection, indicated a worm-like swelling in the centre (Figs. 1, 2).

Through a small incision, the worms could be removed (Figs. 1–4). The treatment was completed in 3 weeks. The patient has since had no more complaints and can be considered cured.

DISCUSSION

If a diagnosis of *loa-loa* infection is expected, the case history becomes an important piece of information. The endemic area is limited, and it is known that, while the infective larvae take many months to develop into adult worms, the latter can survive for many years, and cause the Calabar swellings. A presumptive diagnosis can be confirmed by the detection of microfilariae in peripheral blood, collected during the daytime. However, in a primary infection it may take several years before microfilariae can be found in the circulating blood. In patients without detectable microfilariae, one of the serological tests can be used (1).

When chemotherapy is indicated, diethylcarbamazine (Hetrazan[®]) is administered for 2–3 weeks. Therapy with DEC is started with an initial dose of 50 mg three times daily and each dose thereafter is increased by 50 mg until a total daily dose of 6 mg/kg of body weight is reached (5).

During the initial treatment with DEC one has to be alert for allergic reactions. To counteract these, it is advisable to give antihistamine or corticoster-