

HYPERKERATOSIS LENTICULARIS PERSTANS (FLEGEL)

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Abstract. A case of hyperkeratosis lenticularis perstans (Flegel) is presented. The clinical and histological observations are essentially in accordance with earlier findings. On the basis of morphological and histochemical findings a possible origin of the disease arising in a sweat duct anomaly is suggested.

During the 15 years of its existence as a well defined distinctive dermatological entity (1, 2, 4, 6-14), only a few cases of hyperkeratosis lenticularis perstans (Flegel) (HLP) have been described. It has been possible to distinguish HLP from other papular keratoses such as Porokeratosis actinica, elastoma interpapillare perstans, stucco-keratosis, tar warts and arsenical keratosis, on the basis of somewhat divergent clinical features and, above all, upon certain histological peculiarities. It has been suggested that HLP may be identifiable with Kyrle's disease (10), on the evidence of coexistence in one patient and similarities in histological appearance. It has not been possible to reach any definite conclusions regarding aetiological and nosological classification.

CASE REPORT

The patient is a 66-year-old woman who was operated on in 1963 for mammary cancer. No signs of recurrence were seen. She was treated for a gastric ulcer in 1971. The patient noticed a reduced tolerance to sun exposure, apparently related to a poorly pigmented skin. Light tests with xenon lamp radiation and subsequent biopsy studies proved negative.

In 1971 skin lesions appeared on the patient's legs and during the following year lesions appeared on her shoulders and arms. There was no involvement of the palms of the hands and soles of the feet. At the onset of the eruptions the patient felt them to be somewhat tender together with some prickling sensations, which later disappeared. When examined in 1972 the lesions consisted of irregularly shaped harsh, roughly scaling papules varying in size from a pinhead to a peanut (Fig. 1). In some places the papules showed a tendency to form groups, merging into slightly elevated

plaques with a diameter of 1-2 cm. The lesions were greyish brown in colour and there were no conspicuous signs of inflammation of the surrounding skin. Teleangiectatic capillaries could be seen in most of the lesions. No spontaneous involution was noted. There was no family history of similar skin changes. In addition the tongue was diffusely coated with a rough yellowish-grey hyperkeratotic layer. The fingernails showed longitudinal striation.

HISTOPATHOLOGY AND HISTOCHEMISTRY

Two biopsies of the skin were taken from different representative areas. A separate excision was made for phosphorylase staining according to Eränkö & Palkama (5).

The histological picture (Fig. 2) was dominated by a massive hyper-parakeratotic layer contrasting with an underlying thin epidermis, which in some places was reduced to one single cell-layer. In one place a parakeratotic plug came into direct contact with the dermis (Fig. 3). Tent-like prominences alternated with deep invaginations. The granular layer was almost completely absent and the basal layer was partly eliminated by colligation due to heavy lympho-histiocytic band-like infiltrations into the adjacent corium. The lower border of this infiltration was sharply demarcated, forming a horizontal line (Fig. 4). An almost total absence of elastic fibres contrasted with the abundance of reticulum fibres in the area of the infiltration. The blood vessels were dilated. Irregularly shaped, anuclear, PAS-positive bodies (colloid bodies) were found in some places beneath the epidermis, and even somewhat deeper, intermingled with the inflammatory cells. In other places the basal layer remained intact (Figs 5, 6). Fig. 5 shows a funnel-like, wide epidermal invagination containing lamellar, horny masses and the horny part of the sweat duct. In an adjacent section, beneath the epidermal invagination, the intra-epidermal sweat duct was found to be in direct connection with an irregularly

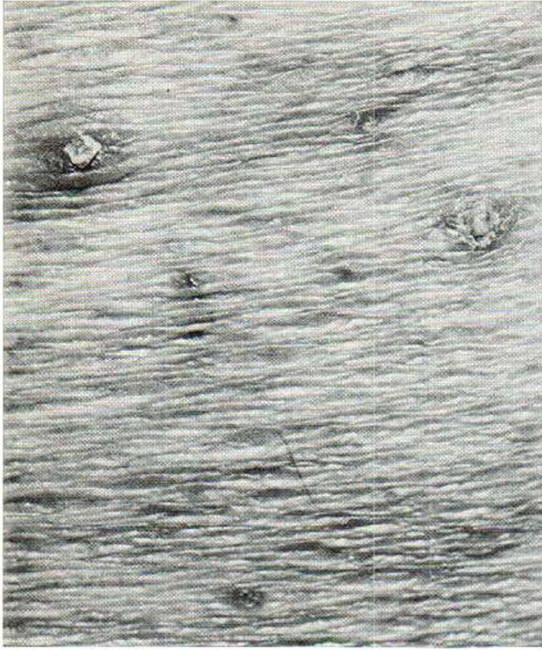


Fig. 1. Clinical manifestations.

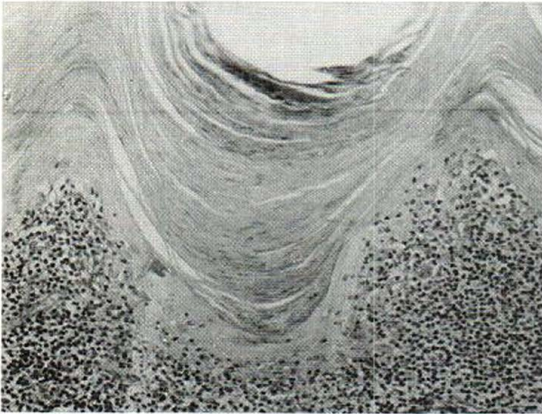


Fig. 2. Funnel-like epidermal invagination filled with hyperkeratotic material. On both sides, tent like prominences are seen. Heavy, lympho-histiocytic infiltration invading the epidermis. Liquefaction degeneration of the basal layer.

shaped intradermal sweat duct proliferation (Fig. 6), a part of which is also seen in Fig. 5. In the same zone the atrophied sweat coil is found, located unusually high in the corium. Phosphorylase staining showed a positive reaction at the bottom of the epidermal invagination and in the walls of the sweat ducts (Fig. 7).

DISCUSSION

The clinical appearance, distribution and permanence of the lesions correspond well to the hitherto

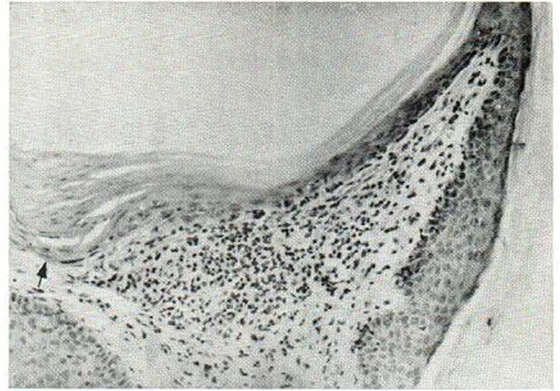


Fig. 3. A parakeratotic plug in direct contact with the corium (arrow).

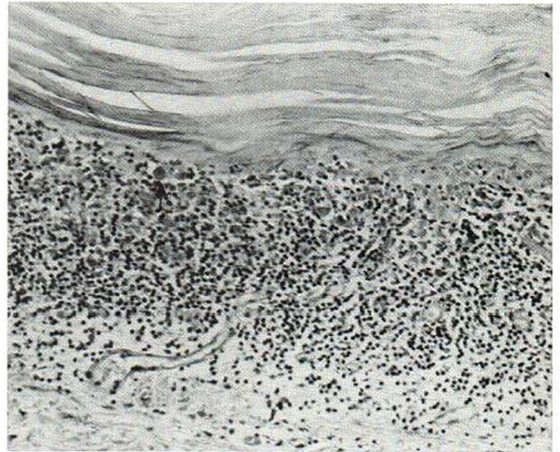


Fig. 4. Band-like lympho-histiocytic infiltration beneath the epidermis with teleangiectasiae. The lower border forms a sharp line. Liquefaction degeneration of the basal cells and colloid bodies (arrows).



Fig. 5. Funnel-like epidermal invagination in direct connection with the epidermal sweat duct (thin arrow) and an irregularly shaped dermal sweat duct proliferation (thick arrow).

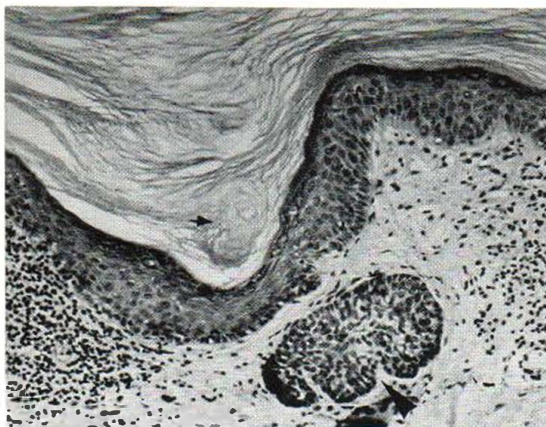


Fig. 6. Funnel-like epidermal invagination containing the horny part of a sweat duct (thin arrow). Distorted part of a sweat duct proliferation (thick arrow).

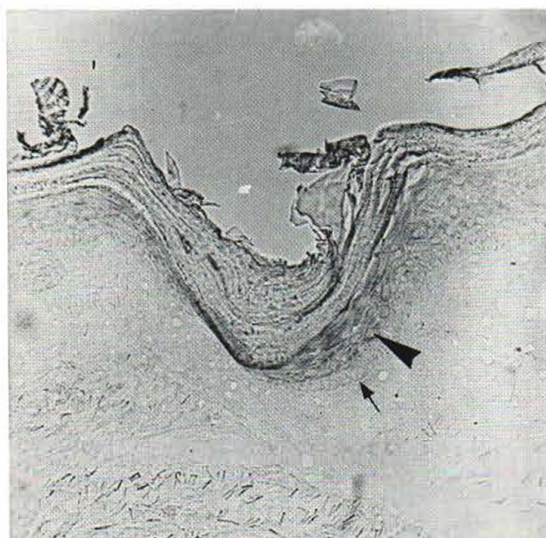


Fig. 7. Phosphorilylase staining. Positivity at the bottom of an epidermal invagination (thick arrow) contrasting with pale staining in the epidermal cells beneath (thin arrow) and on both sides.

published cases of HLP. The relation between cell infiltration beneath the epidermis and the basal layer varied in the previous descriptions. While neither cell invasion (10, 11) nor basal-cell colliquation were noted by some authors (11, 12), an extension of the infiltration into the epidermis in some areas has been described (1, 4, 8). However, absence of a basal layer under a massive parakeratotic plug has also been described (10). In addition, we found colloid bodies close to the basal parts of the epidermis. The possibility of HLP representing

an atypical form of lichen ruber planus has been discussed (11, 13), but rejected. The permanence of the lesions, absence of the granular layer facing a thick horny layer and, finally, an abundance of histiocytic elements in the infiltration, do not correspond with the findings in lichen ruber planus. In agreement with Kocsard et al. (10), direct contact between a parakeratotic plug and the corium, without penetration of kerato-parakeratotic material into the corium, was observed. The absence of cornoid lamellae and an otherwise quite different histologic picture, together with the negative response to xenon lamp radiation test, excludes the diagnosis of disseminated superficial actinic porokeratosis (3).

A peculiar finding in our case was an irregularly shaped sweat duct proliferation directly beneath the funnel-like epidermal invagination containing the horny part of the sweat duct (Fig. 5). Parallel with the progressive widening of epidermal invaginations, they became shallower by degrees. It seems that in the genesis of HLP a sweat duct anomaly might play a primary role. Flegel (6), however, did not find any convincing primary adnexal changes. Our findings point to a hyperplasia of the intradermal part of the sweat duct. Hyperkeratosis, epidermal atrophy and lympho-histiocytic infiltration in the adjacent corium consequently seem to be a secondary feature.

The permanence of the lesions, some reports of familial incidence (1, 2), and additional reports of HLP localized also to the palms and soles (7, 8), indicating a genodermatosis (1, 2, 14), tally with a possible nevoid hyperplasia of the intradermal sweat duct. However, a definite opinion regarding the aetiology of HLP can be established only through further experience and case reviews.

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