Hyperkeratosis lenticularis perstans: Report on Three Cases

INGER ROSDAHL and KARIN ROSÉN

Department of Dermatology, Sahlgrenska Hospital, Gothenburg, Sweden

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Three women with hyperkeratosis lenticularis perstans (HLP) are described. The lesions developed after the age of 45 and there was no family history of this disease. The possible role of UV-light in the pathogenesis of this disease is discussed. *Key words: Electron microscopy; Membrane-coating granules; UV-light* (Received January 28, 1985.)

K. Rosén, Department of Dermatology, Sahlgrenska Hospital, S-413 45 Gothenburg, Sweden.

Hyperkeratosis lenticularis perstans (HLP) is a rare disease of keratinization (1). It is characterised by asymptomatic horny papules on the extensor surfaces of the extremities. The microscopic picture, including a reduced number of membrane-coating granules (MCG), separates this disorder from other focal kertinization disorders, such as stuccokeratosis, Kyrle's disease, porokeratosis of Mibelli and disseminated superficial actinic porokeratosis (DSAP) (2, 3, 4). Three patients with HLP are described in this report.

CASE REPORTS

Case 1

A 70-year-old fair-skinned women with a negative family history started to develop asymptomatic lesions on the extensor surfaces of her feet and lower legs at the age of 45. The lesions spread during 10 years to her thighs and the extensor surfaces of her arms and hands. Her palms and soles were not affected. Today the patient presents multiple, asymptomatic, rough, horny papules, 1–5 mm in diameter, on uninflamed skin. The diagnosis HLP was confirmed by light and electron microscopy (see below). Topical treatment with sunlight, keratolytics, tar preparations and clobetasol propionate gave no benefit. Some lesions have been curetted and have not recurred. This treatment has left atrophic scars.

Case 2

A 76-year-old woman developed asymptomatic warty classical HLP papules 1–10 mm in diameter on the extensor surfaces of her upper and lower extremities (Figs. 1, 2). Occasional lesions were found on her buttocks. Some larger disciform lesions were treated with curettage and did not reappear. No one in her family is known to have a similar skin condition. This patient had skin type I and was earlier treated for several actinic keratoses and basal cell carcinoma of the face.

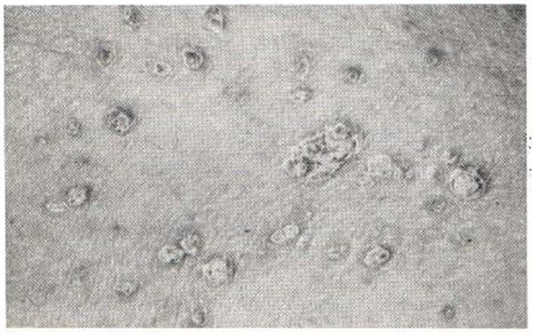
Case 3

A 51-year-old woman developed palmoplantar pustulosis (PPP) at the age of 45. Except for a grandfather with psoriasis, her family history for skin diseases was negative. Her PPP was treated with PUVA (8-MOP+UVA). After 12 months and an approximate UVA dose of 900 J/cm^2 to her feet, about 50 small, reddish-brown keratotic papules developed on the back of her legs. Light and electron microscopy of these papules verified the clinical diagnosis of HLP. During etretinate treatment for her PPP, the HLP lesions on her legs vanished, only to reappear after this medication was stopped.

Histology

Microscopic examination was performed on several skin biopsies from all patients. Paraffin sections showed an orthokeratotic scale with patchy parakeratosis overlying an area of atrophic epidermis (Fig. 3). In this area, the granular cell layer was reduced or absent. The lesions had sharp borders, sometimes demonstrating a church-spire like elevation at the edge. No cornoid lamella was found. A superficial band-like lymphocytic infiltrate was noticed close to the epidermis strictly under the lesion.

The biopsies for electron microscopy were processed according to standard procedures i.e. fixed in glutaraldehyde and osmium tetroxide, embedded in Epon, sectioned and stained with uranyl acetate



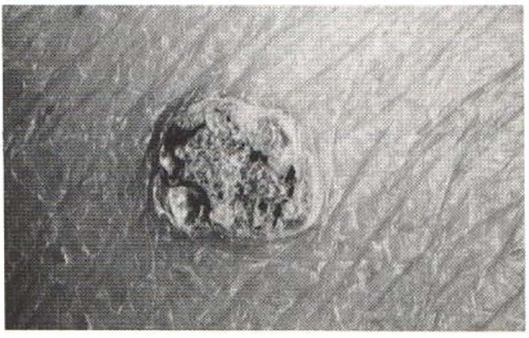


Fig. 1. Close-up of papules on the lower leg (case 2).

Fig. 2. 10 mm large keratotic disc on the lower leg (case 2).

and lead citrate. The sections were viewed in a Philips EM 300 Microscope. In a scale from the lesions, the compact corneocytes contained a dense matrix with no obvious keratin pattern. The desmosomal discs most often persisted up through the stratum corneum of the scale. In the underlying atrophic epidermis, no membrane-coating granules could be found, while they were present in the epidermis just outside the lesions.

DISCUSSION

Hyperkeratosis lenticularis perstans is considered a rare disorder of keratinization. Only about 50 cases have been described. As we have seen 3 cases in a short period of time, this disease may not be so uncommon in its more discrete forms. In earlier publications, HLP is noted to be an autosomal dominant disease (5). Sporadic cases have also been reported in the last few years (6, 7). It is notable that none of our 3 patients had a positive family history.

Among other focal keratinization disorders, DSAP must especially be considered in the differential diagnosis of these cases. However, the individual lesion was clinically different from DSAP and the cornoid lamella typical of porokeratosis was missing. Furthermore, the EM picture was not consistent with DSAP and the lack of MCG observed in our biopsies is considered to be characteristic of HLP (4).

Many patients with HLP are light sensitive; some have skin tumours and solar keratoses (2, 5, 8, 9). The possibility that external factors, for example UV light, may play a role in the pathogenesis of HLP has been discussed. Our observations support this hypothesis.

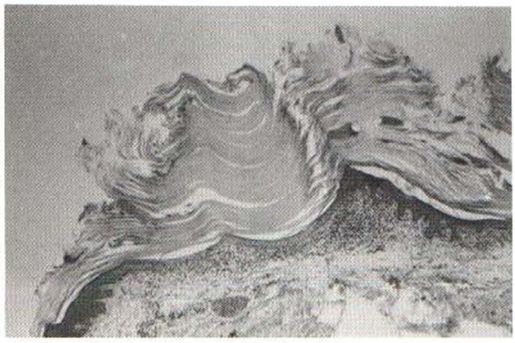


Fig. 3. Light microscopy of paraffin section (case 1). For further details, see text.

All our patients are blond and one has multiple solar lentigenes and was treated for actinic keratosis and basal cell carcinoma of the face. An even stronger case is offered by patient 3, in whom the time of onset of the lesions and the atypical localization to the calves indicate an association with her PUVA treatment. The distribution of these lesions corresponded to the extension of the PUVA light bow. Against the hypothesis of light provocation, it may be argued that HLP has also been described in women of darker complexion (10). It is possible that there are two types of HLP—one classical, with an autosomal dominant transmission, and one sporadic, perhaps more discrete, type, where UV light might be a provoking factor.

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