

suggests the existence of a link between the 2. We think the connection could lie in the existence of an individual genetic predisposition to lichen sclerosus et atrophicus, where greater vulnerability to certain keratinocyte clones and the presence of environmental factors like viral herpes infections, could be responsible for the unusual clinical presentation of the lesions in our patient.

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Conflicts of Interest

The authors declare no conflicts of interest.

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Hyperkeratosis Lenticularis Perstans, or Flegel Disease, With Palmoplantar Involvement

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To the Editor:

Hyperkeratosis lenticularis perstans (HLP) is an autosomal dominant or sporadic keratinization disorder that occurs equally in men and women from 40-50 years old.¹ HLP has been described in association with endocrinial changes, and digestive and cutaneous tumors,^{2,3} and is characterized by the presence of small, asymptomatic erythematous papules that leave characteristic punctate bleeding when they become detached. The lesions generally occur symmetrically along the top of the foot and on the legs, appearing more rarely on the arms, forearms, palms, and soles, and even on the oral mucosa.^{4,5} Agreement has yet to be reached on the role of ultraviolet light in pathogenesis.⁶

Histologically, HLP is characterized by orthokeratotic, eosinophilic, and compact hyperkeratosis, hypogranulosis, thinning of the Malpighian layer, vascular dilation, and

band-like lymphocytic infiltrate in the papillary dermis. Immunohistochemical study shows a predominance of CD4+ T cells that is more evident in the early stages of the illness. Many treatment options have been described, although none has proved effective.^{7,8}

We present the case of a 64-year-old man with diabetes who consulted for asymptomatic, brown, hyperkeratotic papules that appeared progressively over the years with no relation to sun exposure. These were mainly located on the top of the feet, legs, arms, and forearms, leaving hemorrhagic pits on detachment (Figure 1). Pits or dimples could be seen on the palms and soles, reminiscent of the ungual pits of psoriasis (Figure 2). There was no family history of the disorder and tests including general biochemistry and thyroid profiling produced normal results. Biopsy of a papule from the top of the foot



Figure 1. Multiple erythematous and hyperkeratotic papules and characteristic punctate bleeding on the top of each foot.

displayed slight epidermal atrophy, compact orthokeratotic eosinophilic hyperkeratosis, and mild, predominantly leukocytic, lichenoid inflammatory infiltrate of the papillary dermis. Immunohistochemical studies for lymphoid markers did not establish a predominance of CD4+ over CD8+ T cells (Figure 3). The application of 30% urea every 12 hours, followed by topical calcipotriol, 2 times a day, led to a partial improvement of the lesions.

HLP, described by Flegel in 1958, is considered to be an autosomal dominant inherited keratinization disorder, although most cases are sporadic, affecting patients aged 40-50 years with no noted predominance in either sex.¹ The condition has been described in association with endocrine abnormalities including diabetes and hyperthyroidism, while a possible relationship with digestive and cutaneous tumors is more open to debate.^{2,3} There is controversy over the pathogenic role of ultraviolet light, with some authors differentiating between a classic, hereditary form of HLP and another sporadic or acquired form, related to chronic sun exposure.⁶

The condition is clinically characterized by asymptomatic erythematous or brown papules of 1 to 5 mm in diameter, which leave a characteristic punctate bleeding when removed. The lesions are generally located on the top of the feet and legs and, more rarely, on the arms, forearms, and the oral mucosa.⁴ Unilateral and localized forms have been described,^{9,10} as has possible palmoplantar involvement resulting in pitting—as was seen in our patient.⁵

In 1994, De Argila et al² published a series of 10 cases of HLP in patients aged from 41-80 years old. Of these, 3 were hereditary cases, 9 presented involvement of the lower limbs, and 5 of the upper limbs. Only 2 experienced any endocrine disorder (hyperthyroidism and type 2 diabetes) and none of them had the palmoplantar lesions. Our patient had suffered from diabetes for several years, but given the high prevalence of diabetes in the adult population and the lack of controlled studies we cannot rule out a causal association.²

Histologically, HLP is characterized by compact orthokeratotic hyperkeratosis, sometimes associated with parakeratosis, hypogranulosis, thinning of the Malpighian layer, and lichenoid lymphocytic infiltrate in the papillary dermis. The immunohistochemical study showed a predominance of CD4+ T cells, which is more marked in the early stages of the disease. Ultrastructural abnormalities were observed in the number and shape of the Odland bodies or keratinosomes.⁵

In 2006, Ando et al¹¹ provided more detailed descriptions of the histological, immunohistochemical, and ultrastructural differences between early and late HLP, concluding that the characteristics of the early stage were compact hyperkeratosis with focal parakeratosis, hypogranulosis, epidermal atrophy, marked mononuclear infiltrate of a lichenoid type with a predominance of CD4+ T cells and superficial vascular dilation. Electron microscopy reveals the presence of lymphocytes with cerebriform nuclei, clusters of filaments and amorphous substance in corneocytes, and changes in the morphology



Figure 2. Punctate plantar pits.

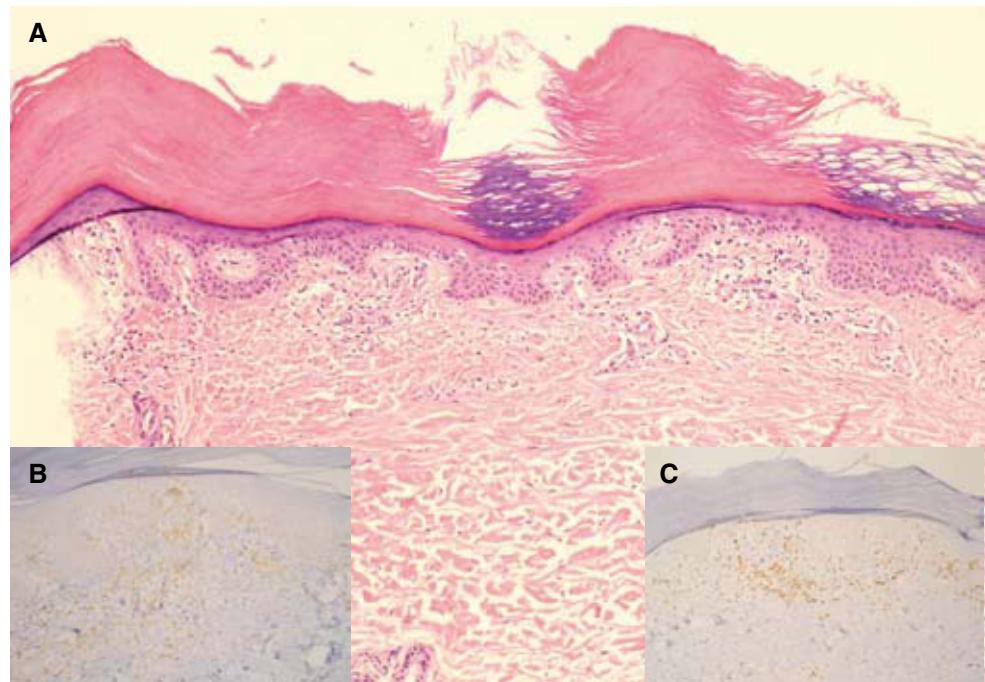


Figure 3. Papule on the top of the foot (hematoxylin-eosin, $\times 10$). A. Compact orthokeratotic hyperkeratosis, slight epidermal atrophy, flattening of the interpapillary ridges, and scant lichenoid lymphocytic inflammatory infiltrate in the papillary dermis. B. Immunostaining for CD4 (PAP, $\times 20$). C. Immunostaining for CD8 (PAP, $\times 20$).

and number of Odland bodies. They suggested that the absence or less marked presence of these characteristics in the later stages was due to the lesions reaching a more advanced stage of development.¹¹

Differential diagnosis should include disseminated superficial actinic porokeratosis, Kyriele disease, stucco keratosis, acrokeratosis verruciformis of Hopf, and lichen nitidus.

Many treatment options have been discussed including topical and systemic retinoids, 5-fluorouracil, vitamin D derivatives, psoralen UV-A therapy, excision, and dermabrasion of the lesions. At present all of these are considered unsatisfactory due to high rates of recurrence.^{7,8}

We draw attention to the uncommon presence of punctate palmoplantar pits in HLP and in our case the controversial association with diabetes and sun exposure.

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Conflicts of Interest

The authors declare no conflicts of interest.

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