JAMA Dermatology Clinicopathological Challenge

Unilateral Asymptomatic Progressive Eruption of Hyperkeratotic Papules

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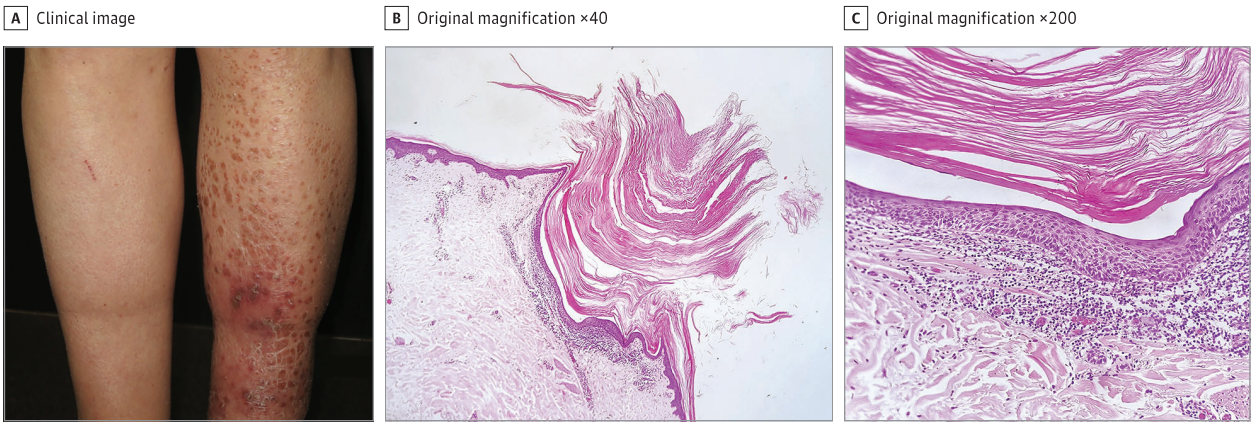

Figure 1. A, Clinical image of the reddish brown hyperkeratotic papules on the front of the left leg. B and C, Hematoxylin-eosin–stained lesional skin biopsy specimens revealing compact hyperkeratosis, epidermal atrophy, and bandlike lymphocyte infiltrate in the superficial dermis.

A man in his 60s presented with a 10-year history of a slowly progressive, asymptomatic cutaneous eruption on his left leg (**Figure** 1A). He had a 16-year history of diabetes mellitus that was being treated with metformin. Hematological and biochemical test results were unremarkable. There was no history of another endocrine disorder or malignant neoplasm. There was no family history of similar cutaneous findings. Physical examination revealed reddish brown hyperkeratotic papules 1 to 10 mm wide on the front and back of the left leg from knee to ankle. The rest of the physical examination findings were unremarkable. Removal of the scales caused slight bleeding. A lesional skin biopsy was performed (Figure 1B and C).

WHAT IS YOUR DIAGNOSIS?

A. Stucco keratoses

B. Hyperkeratosis lenticularis perstans

C. Disseminated superficial actinic porokeratosis

D. Kyrle disease

Diagnosis

B. Hyperkeratosis lenticularis perstans

Microscopic Findings and Clinical Course

Histopathologic examination revealed focal epidermal atrophy, orthokeratotic hyperkeratosis, and a bandlike infiltrate of lymphocytes in the superficial dermis compatible with unilateral hyperkeratosis lenticularis perstans (HLP) (Figure 1B and C). He was treated with acitretin and topical tacalcitol with substantial improvement of his cutaneous lesions (**Figure 2**).

Discussion

Described by Flegel[1] in 1958, HLP is a disease of uncertain origin, though it has been shown to have an autosomal dominant inheritance pattern with late onset.[2] This rare disorder is characterized by numerous symmetric hyperkeratotic papules. Lesions are most common on the distal portion of the legs and dorsal surface of the feet. Arms, palms, and soles can be affected. Other parts of the body, including the oral mucosa, are rarely affected, and the trunk is usually spared.[3] The patient has localized unilateral HLP, which is a rarely seen form of the disease[4,5] The lesions are usually

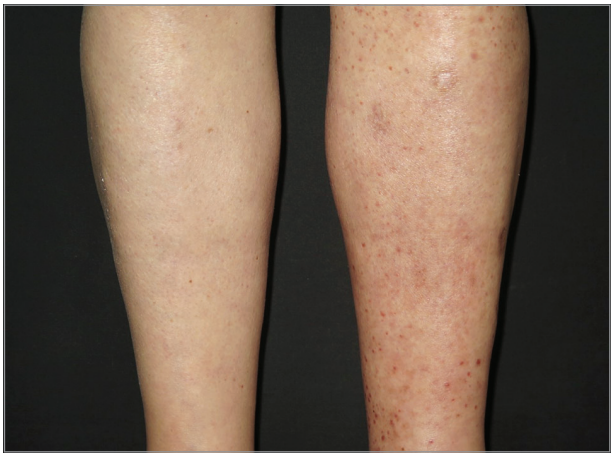

Figure 2. The clinical image after treatment with systemic retinoids and vitamin D derivates demonstrates improvement of hyperkeratotic papules.

hyperkeratotic papules 1 to 5 mm in diameter, with an initial erythematous coloration that with time acquires brownish coloration. When the papules are dislodged, they give rise to a depression with punctate bleeding. It runs a chronic course, and lesions persist indefinitely.

Histologically, there is compact hyperkeratosis with focal parakeratosis on an atrophic epidermis, with a thinned or absent stratum granulosum. In the papillary dermis, there is a band of lymphocytic infiltrate with dilatation and proliferation of superficial vessels.[6] Given the rarity of cases, no standard modality exists for treatment. Reported treatments include topical tretinoin, fluorouracil cream, and vitamin D derivates.[7,8] Systemic retinoids have been used with good results.[9,10]

Differential diagnosis of HLP includes causes of acral keratosis, particularly stucco keratoses, disseminated superficial actinic porokeratosis, and Kyrle disease. Stucco keratoses are usually located on the distal portion of the extremities and clinically manifest as small, scaly white or greyish hyperkeratotic papules, but they do not bleed when scraped off, and histopathologic evaluation reveals hyperkeratosis with epidermal hyperplasia instead of atrophy of the epidermis. Disseminated superficial actinic porokeratosis is characterized by brownish hyperkeratotic papules predominantly found on the arms and legs, and symptoms appear later in adult life. The papules have a hyperkeratotic ridge at the periphery, and histologic results show the presence of cornoid lamellae. Acquired reactive perforating dermatosis (Kyrle disease) presents with keratotic papules up to 1 cm in diameter that are seen primarily on the legs and arms. It is most often seen in conjunction with diabetes or renal failure. Kyrle disease has more prominent keratotic plugs, and there is transepidermal elimination of connective tissue.

This patient had an uncommon presentation of HLP given the rarity of unilateral presentation. It is important for clinicians to recognize HLP because it may mimic other more common dermatoses such as porokeratosis or stucco keratoses.

ARTICLE INFORMATION

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