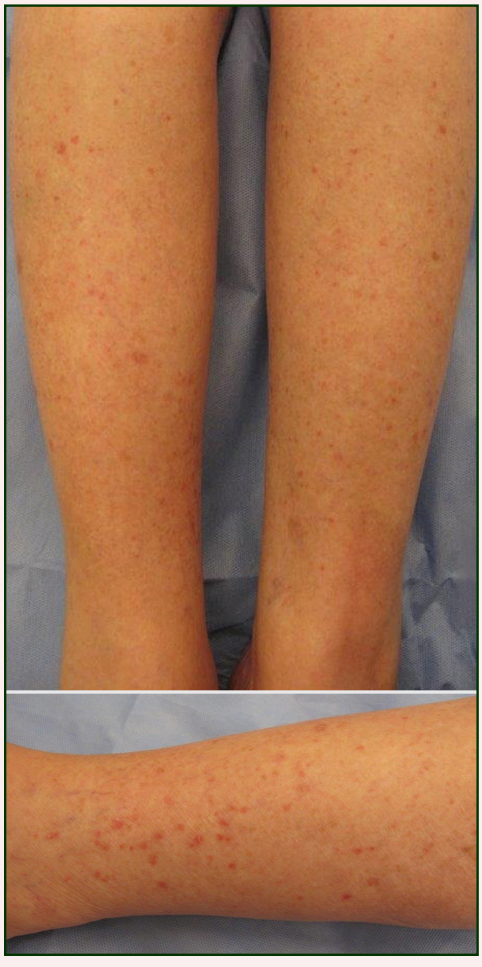
Erythematous Scaly Papules on the Shins and Calves



A 73-year-old white woman presented to the dermatology clinic with a mildly pruritic, slowly progressive rash on the shins and calves of 10 years' duration (top). She had been using triamcinolone ointment 0.1% prescribed by her primary care physician once daily for several weeks without improvement. On physical examination, multiple 1- to 4-mm erythematous, scaly papules were noted on the anterior and posterior aspects of the lower legs (bottom). No similar lesions were noted elsewhere on the body. Her medical history was remarkable for basal cell carcinoma on the face, diet-controlled diabetes mellitus, and hypothyroidism. Her family history was unremarkable.

What is your diagnosis?

a. disseminated superficial actinic porokeratosis
b. flat warts
c. hyperkeratosis lenticularis perstans
d. pityriasis lichenoides chronica
e. seborrheic keratoses

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The Diagnosis: Hyperkeratosis Lenticularis Perstans

A shave biopsy of a lesion on the right leg was performed. Histopathology revealed a discrete papule with overlying compact hyperkeratosis. There was parakeratosis with an absent granular layer and a lichenoid lymphocytic infiltrate within the papillary dermis (Figure). Given the clinical context, these changes were consistent with a diagnosis of hyperkeratosis lenticularis perstans (HLP), also known as Flegel disease.

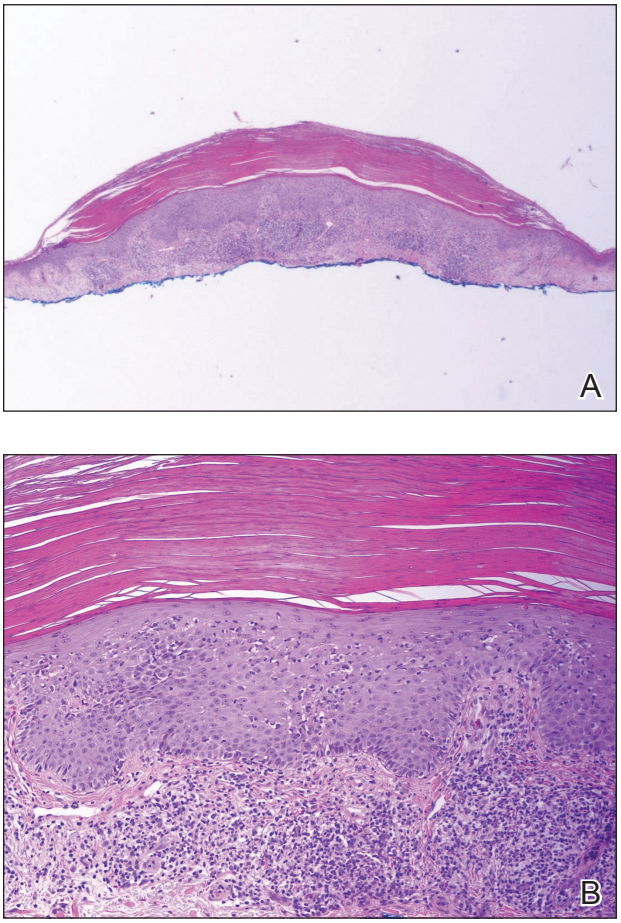
The patient was started on tretinoin cream 0.1% nightly for 3 months and triamcinolone ointment 0.1% as needed for pruritus but showed no clinical response. Given the benign nature of the condition and because the lesions were asymptomatic, additional treatment options were not pursued.

Originally described by Flegel[1] in 1958, HLP is a rare skin disorder commonly seen in white individuals with onset in the fourth or fifth decades of life.[1,2] While most cases are sporadic,[3-6] HLP also has been associated with autosomal dominant inheritance.[7-10]

Patients with HLP typically present with multiple 1- to 5-mm reddish-brown, hyperkeratotic, scaly papules that reveal a moist, erythematous base with pinpoint bleeding upon removal of the scale. Lesions usually are distributed symmetrically and most commonly present on the extensor surfaces of the lower legs and dorsal feet.[1,2,7] Lesions also may appear on the extensor surfaces of the arms, pinnae, periocular region, antecubital and popliteal fossae, and oral mucosa and also may present as pits on the palms and soles.[2,4,7,8] Furthermore, unilateral and localized variants of HLP have been described.[11,12] Hyperkeratosis lenticularis perstans usually is asymptomatic but can present with mild pruritus or burning.[3,5,13]

The etiology and pathogenesis of HLP are unknown. Exposure to UV light has been implicated as an inciting factor[14]; however, reports of spontaneous resolution in the summer[13] and upon treatment with psoralen plus UVA therapy[15] make the role of UV light unclear. Furthermore, investigators disagree as to whether the primary pathogenic event in HLP is an inflammatory process or one of abnormal keratinization.[1,3,7,10] Fernandez-Flores and Manjon[16] suggested HLP is an inflammatory process with periods of exacerbations and remissions after finding mounds of parakeratosis with neutrophils arranged in different strata in the stratum corneum.

Histologically, compact hyperkeratosis usually is noted, often with associated parakeratosis, epidermal atrophy with thinning or absence of the granular layer, and a bandlike lymphohistiocytic infiltrate in the papillary dermis.[1-3] Histopathologic differences between recent-onset versus longstanding lesions have been found, with old lesions lacking an inflammatory infiltrate.[3] Furthermore, new lesions often show abnormalities in quantity and/or morphology of membrane-coating granules, also known as Odland bodies, in keratinocytes on electron microscopy,[3,10,17] while old lesions do not.[3] Odland bodies are involved in normal desquamation, leading to speculation on their role in HLP.[10]



Discrete papule with overlying compact hyperkeratosis and an accompanying lichenoid inflammatory infiltrate (A) (H&E, original magnification ×2). Higher power view highlighting the lichenoid inflammation and loss of the granular layer with overlying parakeratosis (B)(H&E, original magnification ×10).

Currently, it is unclear whether abnormalities in these organelles cause the retention hyperkeratosis seen in HLP or if such abnormalities are a secondary phenomenon.[3,17]

There are questionable associations between HLP and diabetes mellitus type 2, hyperthyroidism, basal and squamous cell carcinomas of the skin, and gastrointestinal malignancy.[4,9,18] Our patient had a history of basal cell carcinoma on the face, diet-controlled diabetes mellitus, and hypothyroidism. Given the high prevalence of these diseases in the general population, however, it is difficult to ascertain whether a true association with HLP exists.

While HLP can slowly progress to involve additional body sites, it is overall a benign condition that does not require treatment. Therapeutic options are based on case reports, with no single treatment showing a consistent response. Therapies that have been most effective include dermabrasion, excision,[19] topical 5-fluorouracil,[2,17,20] and oral retinoids.[8] Hyperkeratosis lenticularis perstans generally is resistant to topical steroids, retinoids, and vitamin D_3 analogs, although success with betamethasone dipropionate,[5] isotretinoin gel 0.05%,[11] and calcipotriol have been reported.[6] A case of HLP with clinical response to psoralen plus UVA therapy also has been described.[15]

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