Atypical Morphology of Hyperkeratosis Lenticularis Perstans (Flegel’s Disease)

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Abstract

We report a forty-five-year old male who presented to dermatology clinic with mildly itchy whitish hyperkeratotic papules on the legs. Skin histopathology revealed typical features of Hyperkeratosis lenticularis perstans (HLP) or Flegel’s disease. A diagnosis of HLP was made, and to the best of our knowledge this is the first report of this atypical presentation of such a rare disease.

Introduction

Hyperkeratosis lenticularis perstans (HLP) is a rare chronic dermatosis, first described in 1958 by Flegel(1). This disorder of keratinization appears to be transmitted in autosomal dominant fashion(2). HLP is characterized clinically by asymptomatic multiple pink to reddish-brown hyperkeratotic papules occurring most commonly on dorsa of feet and lower parts of legs, commonly in middle to old age adults(1,3,4).

The typical histopathologic findings are hyperkeratosis with occasional areas of parakeratosis, atrophy of underlying epidermis and a well-circumscribed band-like lymphocytic infiltrate in the papillary dermis(1,4).

In the following case report we describe atypical morphologic presentation of Flegel’s disease.

Case Report

A 54-year-old male presents to the dermatology clinic with mild itchy skin lesions on both legs for 1 year. Past medical history, drug, allergy, and family history were not significant. Cutaneous examination was significant for multiple whitish hyperkeratotic papules with slight central depression varying in size from 5 mm to 8 mm; some of which has an erythematous rim. Lesions were mainly located over the extensor surface of both legs. (Figs. 1 and 2). The reminder of cutaneous and systemic examination was unremarkable.

The histopathologic of a skin biopsy revealed hyperkeratosis, epidermal atrophy overlying a band-like lymphocytic infiltrate of the papillary dermis (Fig. 3).

These findings were considered typical of HLP. Laboratory work-up including complete blood
count, fasting blood sugar, lipid profile, liver function test and renal profile were normal.

The patient was reassured about the benign nature of this presentation, and he was not willing to undergo any treatment.

**Fig. 1.** Multiple bilateral whitish keratotic papules on both shins

**Fig. 2.** Close-up view showing the erythematous rim and the keratotic atrophic white center.

**Discussion**

Hyperkeratosis lenticularis perstans (HLP) is a rare chronic dermatosis described in 1958 by Flegel. The disorder is characterized by 1 to 5 mm asymptomatic, reddish-brown keratotic papules located mainly on the lower parts of the legs and the dorsa of the feet. Involvement of pinnae, arms, palms, soles and even oral mucosa has been reported. Removal of the adherent horny scale above each papule causes pinpoint bleeding. Hyperkeratotic whitish papules have never been described before in HLP. So we present unreported morphology of this rare dermatosis.

**Fig. 3.** Dense lichenoid infiltrate associated with epidermal atrophy and compact Orthokeratosis (Hematoxylin and eosin stain x40)

Histologically HLP is characterized by foci of compact hyperkeratosis overlying a thinned malpighian layer and a well-circumscribed, band-like lymphoid infiltrate in the underlying papillary dermis. Several ultrastructural changes have been described in HLP. Several authors described quantitative, qualitative changes in membrane-coating granules, or Odland bodies, or both. However, these findings were not consistent and other work showed no abnormality. It remains unclear whether these altered or diminished granules are related to the pathogenesis of HLP.

The clinical and histologic picture of HLP is distinctive and helps one differentiate if from similar Keratotic diseases such as Kyrie’s disease, Stucco Keratosis, disseminated superficial actinic porokeratosis (DSAP) and porokeratosis of Mibelli.

Some authors have suggested that Kyrie’s disease and HLP are variants of the same disease. However, recently the concept that Kyrie disease...
and HLP are 2 separate entities has been accept-
ed(7). Kyrle’s disease differs from HLP by having clinically larger papules with conical hyperkeratotic plugs that may coalesce to form keratotic plaques. Histologically, Kyrle’s disease shows a parakeratotic plug filling an epidermal invagination that communicates with the dermis through epidermal disruption(13).

Stucco keratosis can be differentiated from HLP by having white to gray lesions on a nonerythema-
tous base and lacking the thinned stratum malpighii and band-like infiltrate of HLP(4).

Porokeratosis of Mibelli occurs commonly in children as single or few lesions, that are usually large (several centimeters in diameter), with a tendency to peripheral extension. Histologically there is horny invagination in the epidermis, with cornoid lamella(4).

DSAP usually appears in third to fourth decades as small plaques in sun-exposed skin. Small and raised peripheral border is commonly discerned. Histologically similar to porokeratosis of Mibelli, but the well-defined cornoid lamella is common(4).

In our patient, there is no conical hyperkeratotic papules rather there is some central atrophy, which is against Kyrle’s disease. Being in sun-protected areas, skin lesions in our patient are unlikely to be DSAP. A raised peripheral edge which is a characteristc feature in porokeratosis of Mibelli is lacking here. The histological features in this case are typical of HLP ‘and against the rest of the differential diagnoses.

Various treatment modalities for HLP have been described. Treatment with topical tretinoin was not helpful(14). Systemic retinoids gave variable success(14,15), and treatment results with topical steroids have not been consistent(16).

Topical 5% fluorouracil (5-Fu) cream seems to be the most effective(4,17). The mode of action of 5-Fu in HLP is unclear. DNA synthesis is blocked by 5-Fu and that is why it is used in treating actinic keratosis. However, in HLP, there is no evidence of neoplastic or preneoplastic process. A strong inflammatory response induced by 5-Fu, with subsequent “Shedding” of HLP lesions may explain the efficacy of this treatment in HLP(4).

Keeping in mind the rarity of HLP, and in the few reported cases, the presented case adds a new morphologic presentation which may aid in the clinical recognition of Flegel’s disease.

References
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27


