DOI: http://dx.doi.org/10.3315/jdcr.2012.1111

Journal of Dermatological Case Reports

PHOTOLETTER TO THE EDITOR

Hyperkeratosis lenticularis perstans (Flegel's disease) with unusual clinical presentation. Response to isotretinoin therapy.

Ajay Krishnan, Sumit Kar

Department of Dermatology, Venereology & Leprosy, Mahatma Gandhi Institute of Medical Sciences (MGIMS), Sewagram, Wardha, Maharashtra, India — 442102.

Corresponding author:
Dr. Sumit Kar, Professor and Head, Department of Dermatology, Venereology & Leprosy, Mahatma Gandhi Institute of Medical Sciences (MGIMS), Sevagram, Wardha, Maharashtra, India — 442102. E-mail: *karmgims@gmail.com*

Abstract

Hyperkeratosis lenticularis perstans also known as Flegel's disease is a keratinisation disorder characterized by small keratotic papules with horny scales. Most cases have been reported in Europe with age of presentation between 35 and 60 years. We report a case of a 25-year-old man, who presented with 1-5 mm multiple asymptomatic hyperkeratotic papules of 15 years duration on both legs and hand along with lichenified plaques with Koebner phenomenon in the axillary folds, anticubital and popliteal fossae. Similar lesions were present in the eyelids also. There was no involvement elsewhere. Similar illness was found in the younger brother aged 13 years with the duration of illness for the past 5 years. Histopathology confirmed the clinical diagnosis. Patient was treated with oral isotretinoin in a daily dose of 20 mg per day to which he responded immediately with clearing of the lesions in two weeks. (*J Dermatol Case Rep*. 2012; 6(3): 93-95)

Key words:

Flegel's disease, hyperkeratosis lenticularis perstans, Koebner phenomenon, isotretinoin

Flegel originally described Hyperkeratosis Lenticularis Perstans (HLP) in 1958.[1] It was initially regarded as a variant of Kyrle's disease.[2] Most cases have been reported in Caucasian patients. Clinically asymptomatic small hyperkeratotic papules with adherent scales occur symmetrically on the extensors of extremities and the removal of the scale reveals a bright red base often with pinpoint bleeding. Involvement of pinna, palms, soles and oral mucosa have also been reported. We report a case of Flegel's disease of early onset with atypical distribution of the lesions in the flexural area and also periocular involvement.

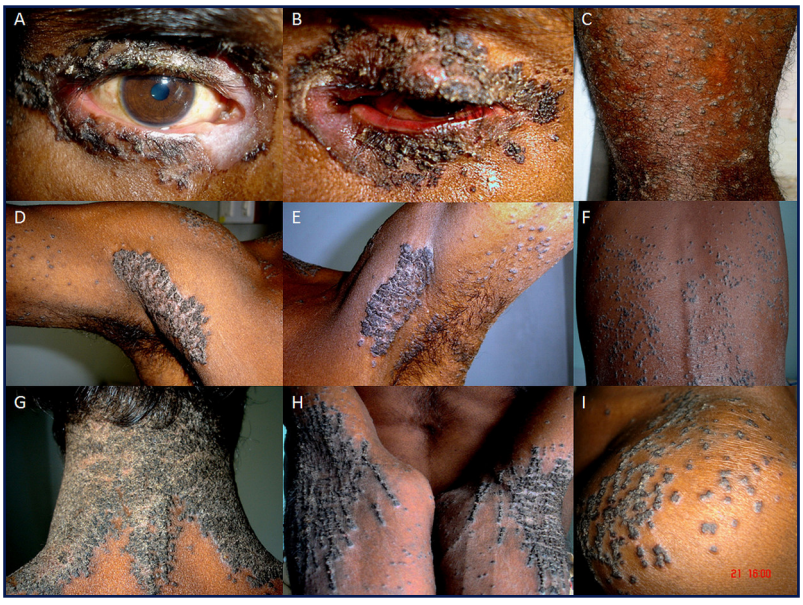

Figure 1

Flegel's disease showing involvement of the eyes (A,B), thigh (C), axillary folds (D,E), back (F), nape of neck (G), cubital fossa (H) and shoulder (I).

A 25-year-old male agricultural worker presented with multiple asymptomatic hyperkeratotic horny papules for the past 15 years. Lesions were noticed first in the extensor aspect of the lower limbs. Lesions also appeared on the axillary folds, flexural aspects of the upper limbs and nape of the neck. For the past seven years lesions started appearing around the eyes. He tried to remove the lesions by scratching which resulted in spreading of the lesions especially in the cubital fossa. Patient had no history of atopy or other significant medical history. Similar lesions were also present in the younger brother aged nine years who expired two years back in an accident.

Examination revealed multiple sharply defined hyperpigmented, hyperkeratotic papules of 1-5 mm size with central cone shaped adherent horny plugs on the extensor aspect of legs, arms, forearms and nape of the neck. Koebner phenomenon was observed over the lesions present in the axillae, cubital fossae and popliteal fossae where the lesions coalesced to form small linear plaques. Ocular lesions in the form involvement of the eyelids were also present (Fig. 1). Hair, nails and oral mucosa showed no involvement. Other physical and systemic examinations were nonspecific. Routine blood, urine and serological examinations were within normal limits.

Histopathological examination of the well formed lesion showed compact hyperkeratosis with parakeratosis along with flattening of the epidermis and lymphocytic infiltrate in the upper dermis. Broad orthokeratotic plugs were also present in the areas where elongation of epidermis is present. Abundant melanin was present within the plugs and that showed scanty parakeratosis. At the bottom of some of these plugs there was slight dyskeratosis (Fig. 2). Ultrastructural study of the skin cannot be performed due to lack of infrastructure.

Patient was given isotretinoin 20 mg orally daily along with emollients for topical application. The lesions showed marked improvement in 2 weeks. Ocular and skin lesions resolved almost completely leaving post inflammatory hypopigmentation (Fig. 3). Therapy was continued for another two months. Follow up of the patient showed no recurrence of the lesions in the six month duration.

Flegel's disease usually develops in the fourth or fifth decade.[3] The condition may be sporadic or inherited as an autosomal dominant condition. No instigating factor has been identified clearly however some investigators have implicated UV light. The main ultra structural finding is a morphological alteration of lamellar (Odland) bodies.[4] Ultra structural studies revealed

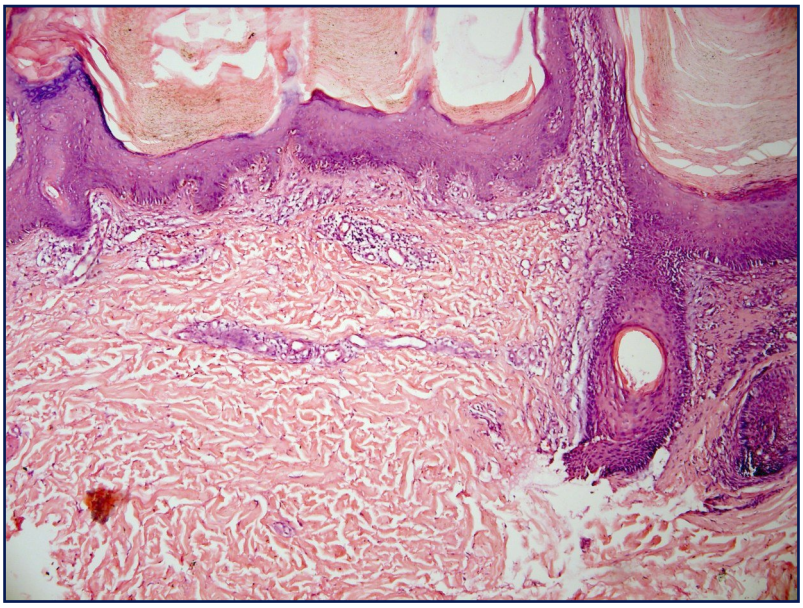

Figure 2

Flegel's Disease showing compact hyperkeratosis with parakeratosis along with flattening of the epidermis and lymphocytic infiltrate in the upper dermis. (H&E stain at 400X).

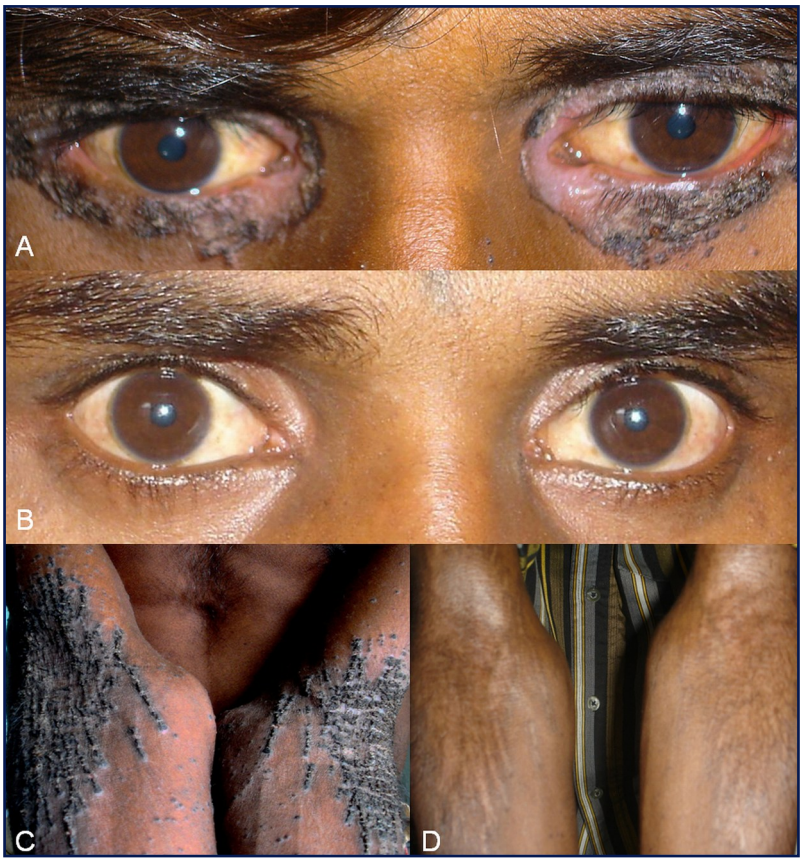

Figure 3

Flegel's Disease — before (A, C) and after (B, D) treatment with Isotretinoin.

the presence of many normal appearing Odland bodies in the keratinocytes of the old lesion whereas the organelles were not found in the keratinocytes of the early lesion.[5] No sex predilection is apparent. Except for the possibility that lesion may progress slowly and involve more proximal sites, prognosis for hyperkeratosis lenticularis perstans is excellent. No mortality has been reported.

Numerous reports exit in the literature that associate HLP with an endocrinopathy. Four reports were associated with adult onset diabetes and two with hypertension. A relationship between HLP and malignancies has been suggested. One report describes epithelial tumors (basal and squamous cell carcinoma) a possible association between cancers of the digestive system also has been implied.

Therapeutic options are limited to emollients, topical 5-FU, and retinoids, but none of these treatments is consistently helpful. In 1986, Gabrielson reported that HLP was effectively treated with etretinate. Our patient responded well to oral isotretinoin.

We are reporting this case due to the rarity of the condition, atypical distribution with involvement of the periocular region, flexural plaque lesions and presence of Koebner reaction. Our case also showed early onset of lesions with excellent response to oral isotretinoin.

References

1. Flegel H. Hyperkeratosis lenticularis perstans. *Hautarzt*. 1958; 9: 363-364. PMID: 13574669.

2. Price ML, Jones EW, MacDonald DM. A clinicopathological study of Flegel's disease (hyperkeratosis lenticularis perstans). *Br J Dermatol*. 1987; 116: 681-691. PMID: 2439113.

3. Cooper SM, George S. Flegel's disease treated with psoralen ultraviolet A. *Br J Dermatol*. 2000; 142: 340-342. PMID: 10730771.

4. Kanitakis J, Hermier C, Hokayem D, Schmitt D. Hyperkeratosis lenticularis perstans (Flegel's disease). A light and electron microscopic study of involved and uninvolved epidermis. *Dermatologica*. 1987; 174: 96-101. PMID: 3556701.

5. Ando K, Hattori H, Yamauchi Y. Histopathological differences between early and old lesions of hyperkeratosis Lenticularis Perstans (Flegel's disease). *Am J Dermatopathol*. 2006; 28: 122-126. PMID: 16625073.