First case of Flegel's disease in Sri Lanka

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Abstract

A case of Flegel's disease occurring in a 26 year old female from North-Eastern part of Sri Lanka is reported. Patient presented with chronic asymptomatic papular eruption affecting the upper arms and ear lobes. Clinical diagnosis of Flegel's disease was confirmed by histopathological findings. Topical treatment with calcipotriol plus betamethasone dipropionate resulted in rapid resolution. This is the first published report of this rare inherited disease from Sri Lanka.

Case report

A 26 years old female presented to the Skin Clinic, Base Hospital, Kantale, Sri Lanka with 10 years history of asymptomatic skin eruption on arms and ear lobes (Figure 1). Skin lesions have progressed at a very slow rate. She has no diabetes, chronic renal disease or other significant medical illnesses. There was no family history of similar skin lesions. She was previously treated with various topical steroids and emollients with no satisfactory improvement.

On examination, there were numerous red brown hyperkeratotic 2-3mm papules with discrete irregular margins on extensor aspects of both upper arms and ear lobes. When keratotic scales were removed from a lesion, a non exudative red base was revealed (i.e. ''cornflake sign'') while slightly depressed bleeding surfaces could be seen in others. Clinical appearance of the skin eruption was typical of Flegel's disease.

Basic investigations including complete blood count, blood sugar, ESR, UFR, renal and liver function tests were all normal. Histopathological examination of lesional skin biopsy showed hyperkeratosis of the epidermis with focal confluent parakeratosis and mild acanthosis with diminished granular layer at the parakeratotic plaque (Figure 3). Suprapapillary thinning of epidermis at the parakeratotic plaques was not seen. The underlying epidermis showed mild perivascular lymphocytic cell infiltrate and oedema. Histological features are compatible with the clinical diagnosis of Flegel's disease.

Patient was treated with combined calcipotriol and betamethasone dipropionate ointment once daily for two weeks and near complete resolution was observed at the end. Same treatment was continued together with emollients for another 6 months with no relapse.

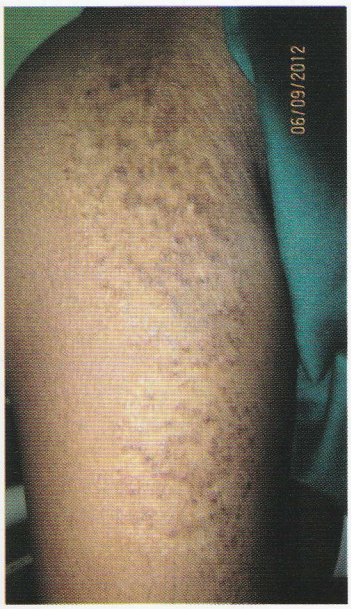

Figure 1. *Upper arm on presentation.*

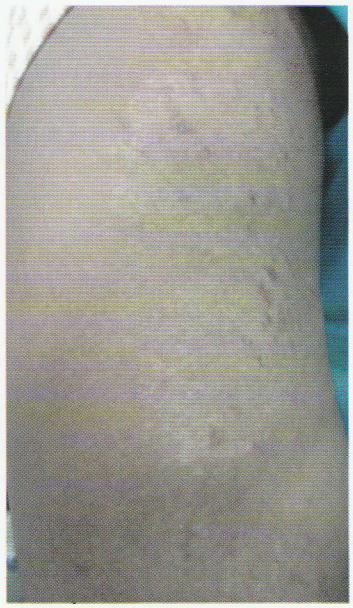

Figure 2. *Upper arm after two weeks of treatment.*

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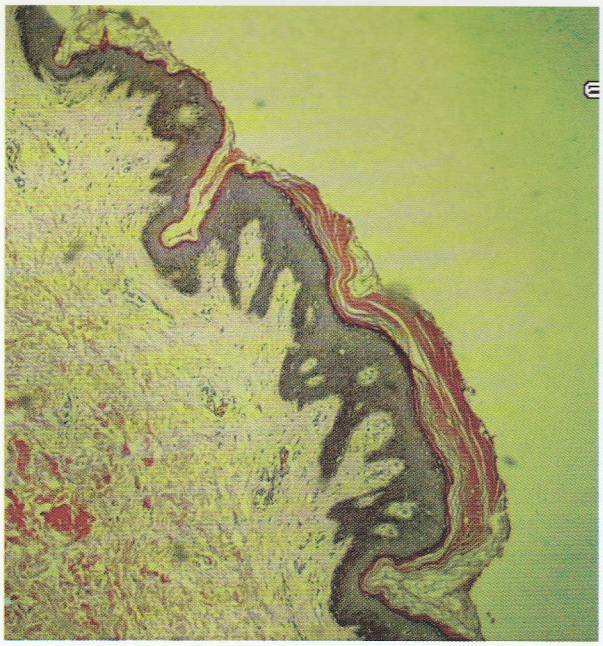

Figure 3. *Histology of skin lesion.*

Discussion

Flegel's disease (Hyperkeratosis Lenticularis Perstans —HPL) is inherited as an autosomal dominant condition although many cases, (including this case) were sporadic[1]. It is characterized by a chronic eruption of numerous symmetric hyperkeratotic 1-5 mm papules, located primarily on the dorsal feet and lower legs, with a decreasing likelihood of manifestation proximally.

Although lower extremities are commonly affected in this condition, involvement of the ear pinnae, arms, palms, soles, and the oral mucosa has been reported in several cases. Our patient also had typical skin eruption involving the earlobes and upper arms. HLP is a disorder of keratinisation and its differential diagnosis include arsenical keratosis, Kyrle's disease and porokeratosis.

Most characteristic feature in HPL is the absence or reduction of membrane coating granules (Odland bodies), which affect the normal process of desquamation resulting in retention hyperkeratosis[2]. Cell mediated cytotoxicity against epithelial cells may also involved in the pathogenesis of HPL.

Management of this disease has been difficult[2]; many treatments including emollients, topical steroids, cryotherapy, PUVA, topical retinoids, calcipotriol 5-fluorouracil and topical 5-aminolevulinic acid-based photodynamic therapy and oral isotretinoin, have been tried with variable success[4].

However the clinical response to topical therapy with combined calcipotriol plus betamethasone dipropionate in our patient was very impressive. This is the first case of Flegel's disease reported in Sri Lanka.

References

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