Sir,

A 22-year-old female patient presented with complains of gradually increasing asymptomatic skin lesions all over body for last 2 years. On examination, multiple, tiny, discrete, erythematous, scaly, folliculo-papular, bilaterally symmetrical lesions were present all over her body, except face and scalp [Figures 1-4]. Lesions were dense on distal extremities as compared to proximal extremities and trunk. Few discrete lesions were present on palms and soles also. Skin lesions were asymptomatic, only occasional pruritus was reported by patient. Nail and mucosa were unaffected. Lesions started on distal extremities and gradually spread proximally. Patient applied various topical medicines including topical steroids, moisturizers etc., without significant improvement.

Patient was unmarried and healthy, had no history of any drug taken. Her menstrual periods were normal. She had history of regular hair removal only from extremities by waxing since the age of 14 years. She had no history of tuberculosis or any other major illness in past. No other family member had history of similar lesions.

Her complete blood count, blood sugar level, thyroid function tests, and X-ray chest were normal.

Skin biopsy showed dense, focal hyperkeratosis and parakeratosis, absent granular layer, acanthosis, and upper dermal lymphocytic infiltrate in a band-like manner. Changes showed sharp demarcation from normal skin, which showed basket weave hyperkeratosis [Figure 5]. Clinical and histo-pathological correlation confirmed the diagnosis of hyperkeratosis lenticularis perstans (HLP) or Flegel’s disease.

Patient was prescribed oral vitamin A (5000 IU daily for 15 days), topical tretinoin cream 0.05% once-a-day application, and emollients with mild improvement in appearance noted after 3 weeks [Figure 6].

HLP was first described by Flegel in 1958[1] and hence also called as Flegel’s disease.

Pathophysiology of HLP is unknown, but role of ultraviolet light in induction of lesions, role of cell-mediated cytotoxicity against epidermal cells are some of the mechanisms suggested. Ultrastructurally, loss of or decreased number of membrane-coating granules or Odland bodies have been reported.[2] Odland bodies are involved in normal process of desquamation. There absence may be associated with abnormal keratinisation.
Incidence and prevalence of HLP are not known. Both familial and non-familial cases have been reported. In familial cases, autosomal dominant transmission is shown. Almost all reported cases are from white population. No sex predilection is seen. HLP is most common during mid to old age but has been reported in much younger patients also.

HLP is clinically characterized by small, red-brown, hyperkeratotic, 1-5 mm papules on the lower extremities. Involvement of the ear pinnae, arms, palms, soles, and the oral mucosa has been reported, although these reports are rare. Involvement of the trunk has been reported but remains an unusual variant. Removal of the scale reveals a bright red base, often with pinpoint bleeding. A localized unilateral variant has been reported. The trunk tends to be spared; absence of axial lesions is characteristic. Keratosis pilaris, Darier disease, follicular lichen planus, skin lesions of scurvy, and other follicular keratotic lesions should be differentiated from HLP. Histopathology from a well-developed lesion shows greatly thickened, compact, strongly eosinophilic horny layer standing out in sharp contrast to the less heavily stained basket-weave keratin of the uninvolved epidermis. The underlying stratum malpighii is flattened with thin or absent granular layer. A lymphoid infiltrate with occasional histiocytes in a bandlike pattern in the papillary dermis typically is seen close to the epidermis. The lateral edge of the lesion demonstrates abrupt hyperkeratosis and a combination of epidermal atrophy and acanthosis.

Various treatment options including topical 5-fluorouracil, topical and oral retinoids, oral vitamin A, topical calcipotriol, topical steroids, emollients, PUVA therapy, and dermabrasion have shown variable success rate. To our knowledge, this is the first case reported in Indian literature. Apart from rarity of this condition in Indian population, our patient also had extensive involvement with involvement of trunk, which is a rare finding. Rarity and chance of misdiagnosis, if biopsy is not done, prompted us to present this case.

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