INTRODUCTION

Inflammatory linear verrucous epidermal nevus (ILVEN) is a unilateral, benign, pruriginous, cutaneous hamartoma first described in literature in 1971.[1] Although the etiopathogenesis is not known exactly, it is thought to be associated with the upregulation of interleukin 1 and interleukin 6, tumor necrosis factor α, and intercellular adhesion molecule 1.[2]

Typically, ILVEN is resistant to treatment. Therapeutic options include topical and intralesional glucocorticoids, dithranol, calcipotriol, excision, cryotherapy, laser therapy,[2,3] and systemic retinoids.

Systemic retinoids have been the treatment of choice for many disorders of keratinization. They possess anti-keratinizing, antiproliferative, and anti-inflammatory properties. We present our experience of acitretin therapy in 2 cases of ILVEN.

CASE REPORTS

Case 1

An 18-year-old unmarried girl presented for evaluation of a pruritic, linear, plaque over her left-hand dorsa that appeared at 8 years of age with intermittent flare-ups and incomplete remission. The eruption and the associated pruritus did not respond to previous therapy with topical salicylic acid preparations and topical retinoids. The family history was not significant.

On dermatological examination, she had a linear crusted erythematous plaque over her dorsal left index finger extending over the adjacent web space and left-hand dorsa [Figure 1]. The rest of the physical examination was unremarkable, and no associated physical abnormality was found.

Results from the skin biopsy specimen from the lesion revealed an acanthotic epidermis with zones of parakeratosis devoid of granular cell layer alternating with zones of orthohyperkeratosis confirming the clinical diagnosis of ILVEN.

The patient was started on acitretin therapy after appropriate laboratory workup including fasting lipid profile and liver function tests (LFTs). A careful menstrual history and counseling regarding the importance of not becoming pregnant 1 month before, during therapy, and 3 years after therapy was impressed on the patient.

The patient was started on acitretin 25 mg PO daily (0.5 mg/kg) for 6 weeks and then tapered to 25 mg PO every other day for 6 weeks and then tapered to 25 mg PO every other day...
for 12 weeks and then acitretin was stopped. Monthly monitoring of lipid profile and LFT was undertaken. There was a significant reduction in the hyperkeratosis and erythema at the end of the 4 weeks with almost complete clearance of the lesions at the end of therapy [Figure 2]. The patient remained lesion free for more than a year post discontinuation of acitretin. Subsequently, she lost to follow-up.

**Case 2**

A 12-year-old boy presented with a pruritic, linear, hyperkeratotic, scaly plaque over the right forefoot dorsa since 4 years of age [Figure 3]. There was no family history of similar complaints.

Cutaneous and systemic examination did not reveal any other abnormality. Histopathological examination confirmed the diagnosis of ILVEN.

His baseline investigations being normal, the patient received 25 mg PO of acitretin for 6 weeks with tapered dose of 25 mg every other day for another 12 weeks.

The lesion distinctly improved with a marked reduction in hyperkeratosis by 4 weeks into the treatment [Figure 4]. After discontinuation of therapy, the patient remained almost lesion free up to 1 year till he was lost to follow-up. The biochemical tests repeated at the end of treatment were normal.

Both patients did not have any significant adverse effect secondary to acitretin except for minimal skin dryness.

**DISCUSSION**

The introduction of retinoids in the armamentarium of dermatologic therapy has allowed many difficult-to-treat dermatoses to be now in a position of controllable diseases. ILVEN is a rare chronic skin disorder believed to be resistant to therapy which may trouble the patient considerably. It has been shown to respond to a variety of antipsoriatic therapies, leading to some authors to believe that it is a nevoid form of psoriasis.

Acitretin is a second-generation monoaromatic retinoid used successfully in keratinization disorders in children.[4-7] Anecdotal case reports have documented the efficacy of systemic retinoids for the treatment of ILVEN. Renner et al. have documented the disappearance of ILVEN lesions...
after 8 weeks of incremental acitretin (up to 30 mg) in a 36-year-old woman.\cite{8}

In our case, long-standing ILVEN lesions in children resistant to topical first-line therapies responded very well to second-line oral therapy with acitretin with no major side effects and remained documented lesion free for up to a year after cessation of therapy. There are few reported case reports of acitretin use in ILVEN.

It is not clear whether ILVEN represents true form of inflammatory epidermal nevus. Review of literature suggests that some ILVEN cases respond to anti-psoriatic and anti-inflammatory treatment with only a slight reduction in itching and the inflammation.\cite{9} These cases are due to an underlying ( verrucous) epidermal naevus. In contrast, cases treated successfully with anti-inflammatory and anti-psoriatic therapy implicate that they have no underlying naevus. According to Hofer, these should rather be called inflammatory linear verrucous eruption (ILVEN).\cite{10}

We present these two case reports as an anecdotal evidence of successful judicious use of acitretin for the treatment of ILVEN.

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Conflicts of interest
There are no conflicts of interest.

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