

Inflammatory Linear Verrucose Epidermal Nevus A Promising Treatment. " CASE REPORT "

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ABSTRACT

The description of Inflammatory Linear Verrucose Epidermal Nevus (ILVEN) was elaborated and clearly defined by Altman and Mehregan in their report of 25 cases in 1971. They considered (ILVEN) to be a distinct variety of linear epidermal nevus(1). The lesion of the(ILVEN)appears as an eczematous or psoriasiform band and is often mistakenly called linear psoriasis. The histological features of (ILVEN) are characteristic, and show moderate acanthosis , papillomatous thickening of the epidermis, elongation of the rete ridges, spongiosis with spotty areas of parakeratosis, and mild perivascular lymphocytic infiltrate in the upper dermis (2).

Altman and Mehregan in their report (1), summarized the features of their 25 cases as follows : Early age of onset , Predominance in females , Frequent in the left lower extremity, and Lesions showed marked resistance to treatment. We here present a case of Inflammatory Linear Verrucose Epidermal Nevus which first appeared at the age of 35 years, in a male, involving the right lower extremity and cleared completely after treatment by a combination of intralesional steroid and topical(steroid plus salicylic acid) without recurrence after one year follow up.

CASE REPORT

A 39 year- old , male patient was first seen at the out patient clinic, complaining of pruritic skin lesion of the right thigh and leg. The lesion first appeared at the age of 35, then spread in a linear fashion.

During the last 4 years the lesion covered the lower part of the right thigh to below the knee joint and mid part of the right leg . No other complaint. The patient general health was good and he had no previous personal or family history of similar disease. Examination of the lesion showed an erythematous, scaly, psoriasiform plaque in linear pattern affecting the lower two third of the right thigh and few centimeters below the knee joint.(fig. 1),and also affected the mid part of the right leg (fig. 2). No other site of the body was affected.



fig. 1. Inflammatory Linear Verrucose Epidermal Nevus
Linear psoriasiform plaque of the thigh .



fig. 2 . Verrucous lesion in a linear fashion at the mid of the leg.

The four m.m punch biopsy specimen taken from the lesion for histologic study showed hyperkeratosis, hypergranulosis, acanthosis, spongiosis, and elongation of rete ridges (fig. 3), raised table-like parakeratotic areas with absent granular layer alternating with depressed areas of orthokeratosis(fig. 4). The upper dermis showed a perivascular chronic inflammatory cell infiltrate consisting mainly of lymphocytes(fig. 5).

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The clinical appearance and the histological features confirmed the diagnosis of the Inflammatory Linear Verrucose Epidermal Nevus.

We tried a combination treatment of intralesional steroids and topical (steroid + salicylic acid) in this patient. Triamcinolone acetonide 40 mg/ml, diluted in distilled water in 1:4 concentration given intralesionally at the different parts of the lesion every month for three successive doses at the beginning of treatment, after four weeks and lastly on the eighth week. At the sametime, Clobetasol propionate .05% ointment plus 20% salicylic acid ointment in 1:1 ratio was applied to the lesion every night for three weeks. The lesion responded to the treatment within 3 weeks with an approximately 60% reduction of the pruritus, scales and erythema . After 5 weeks the lesion disappeared completely leaving a mild residual hypopigmentation evidently duo to steroid effect(fig.6)and(fig.7).

The patient was followed up for one year and he did not show any evidence of recurrence.

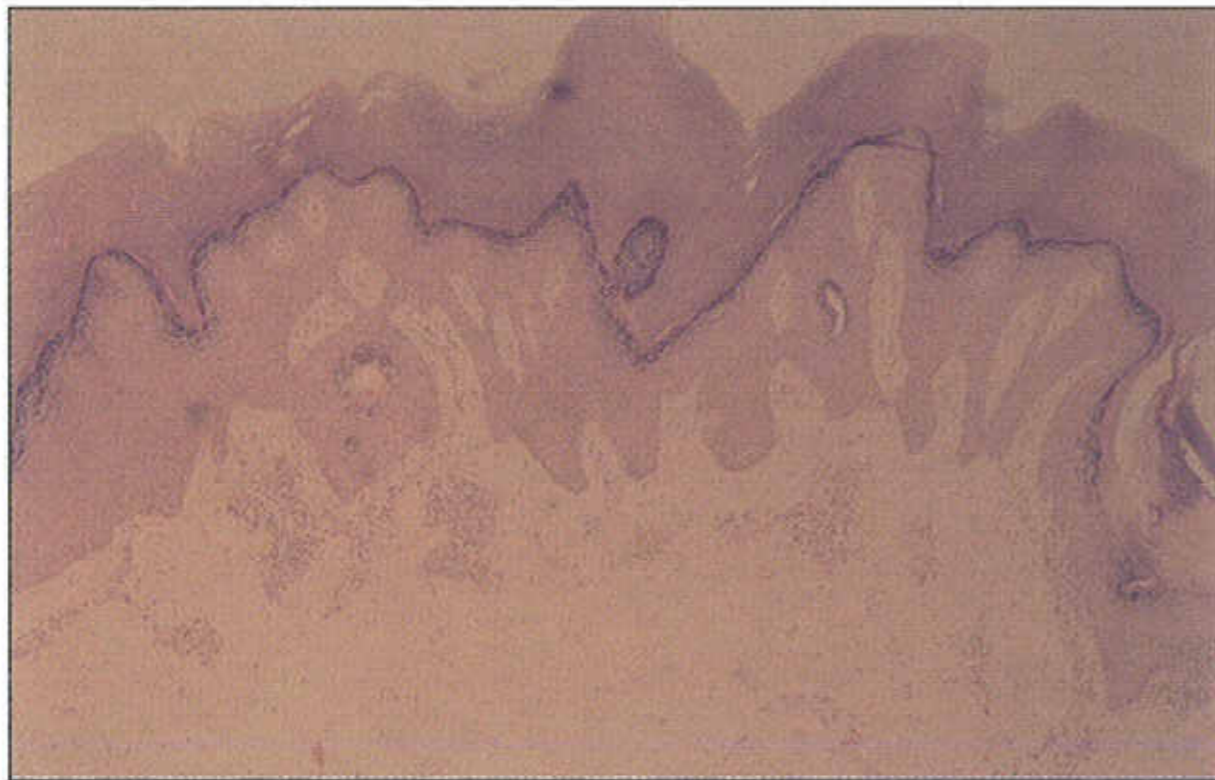


fig. 3 . Hyperkeratosis, hypergranulosis, acanthosis, spongiosis, nd elongation of rete ridges

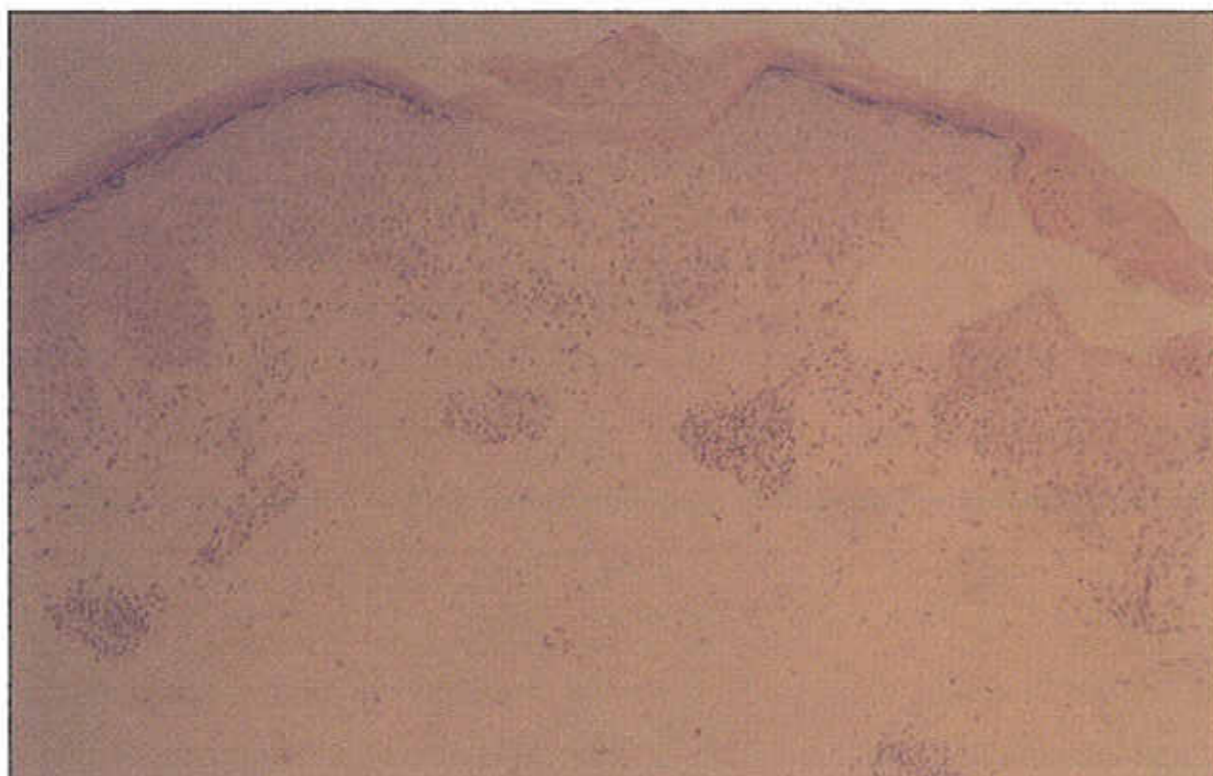


fig. 4 . Raised parakeratosis alternating with depressed areas of orthokeratosis.

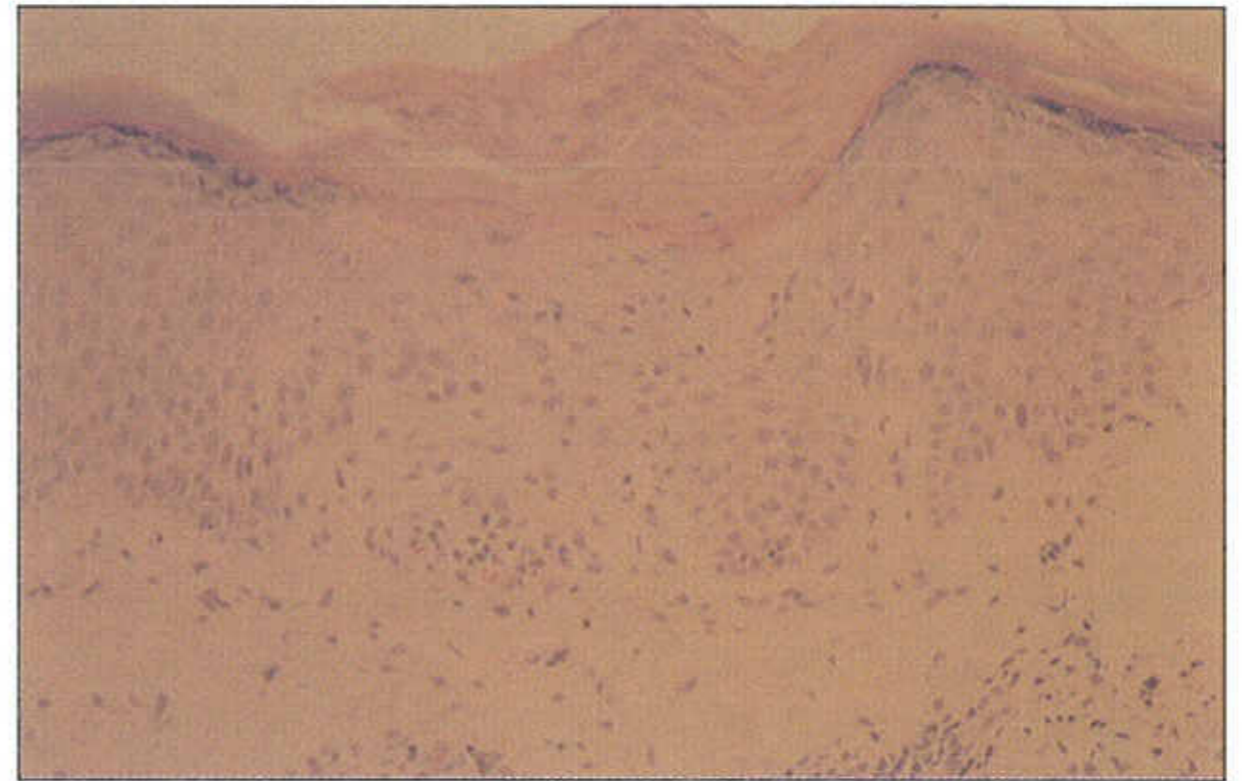


fig. 5 . Magnification of (fig. 4), showing also a perivascular inflammatory cell infiltrate.



fig. 6 Same lesion as in (fig. 1) after treatment.



fig.7 . Same lesion as in (fig. 2) after treatment.

DISCUSSION

ILVEN is considered a variant of epidermal nevus(3). The epidermal nevus consists of single or multiple wart-like overgrowth of the epidermis in a linear unilateral or bilateral and sometimes show extensive distribution. The linearity pattern is ex-

plained by boundaries of the areas innervated by the main cutaneous nerves or lines of cleavage or Blaschko's lines and embryonic suture lines (3). Epidermal nevi include many clinical variants such as nevus unius lateris, ichthyosis hystrix and bilateral epidermal nevi. The epidermal nevi form part of a syndrome (The epidermal nevus syndrome) which includes skeletal and CNS abnormalities(4).

The presence of extensive epidermal nevi justifies careful examination of the patient to find any possible associated anomaly(4).

Epidermal nevus may occasionally develop basal cell carcinoma, squamous cell carcinoma(5), keratoacanthoma(6), oral mucosal involvement(7), haemangiomas and lymphangiomas(3). The nevus sebaceous of Jadassohn may start as an epidermal nevus and only at puberty do the sebaceous and apocrine glands proliferate(3).

ILVEN was described by Altman and Mehregan(1). Kaidbey and Kurban (8) described five cases whose histological features suggested that the concept of "epidermal nevus" needs to be widened to include nevi characterized by a dermatitic component.

ILVEN is formed of erythematous slightly verrucous and scaly papules which coalesce to form linear plaques which most commonly affect legs and thighs .

The next common sites affected are the buttocks, genitalia, groins and more than one site may be affected(1). The majority of lesions are itching but are sometimes asymptomatic(1). 75% of cases occur before age of 5 years and 50% occur before the age of 6 months(1).

ILVEN has to be differentiated from lichen striatus, lichen simplex chronicus, linear psoriasis, linear lichen planus, linear Darier's disease(9) and unilateral keratosis follicularis(10).

Although differentiation could be reached on clinical bases/ yet the clinical entity of ILVEN has a specific histologic appearance which is characterized by raised table-like parakeratotic areas with absent granular cell layer alternating at regular spatial intervals with depressed cup-like areas of orthokeratosis with increased granular cell layer (4-8 layers).

The dermal pericapillary lymphohistiocytic inflammatory infiltrate seen in ILVEN is the result rather than the cause of the orthokeratosis or

parakeratosis(2). It has been recommended that all linear epidermal nevi have to be biopsied for diagnosis. Inflammatory linear verrucose epidermal nevus was reported as a component of epidermal nevus syndrome in a newborn female who also had congenital bony abnormalities (11).

The lesions of ILVEN proved to be very resistant to many forms of treatment including topically applied steroids, with or without occlusion, intralesional steroids and topically applied tar preparations(1). Desiccation and curettage is likely to be unsuccessful or disfiguring. Dermabrasion offers another modality of treatment also often followed by recurrence (3).

In this report we presented a case of ILVEN affecting the right thigh and leg in a male at the age of 35 years. ILVEN lesions were reported to appear for the first time at age of 49 years (1).

The diagnosis of linear psoriasis, linear lichen planus, and linear lichen simplex chronicus were excluded clinically and histologically. The lesions resisted conventional treatment for four years and had a dramatic response without recurrence on the modality of treatment that we followed. This modality of treatment could be tried in more cases of ILVEN and may be in the other types of epidermal nevi as well for assessment of its effectiveness.

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