

CASE REPORTS

INFLAMMATORY LINEAR VERRUCCOUS EPIDERMAL NEVUS (ILVEN)

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SUMMARY : *Inflammatory linear verrucous epidermal nevus is an uncommon dermatosis. The typical time of onset is childhood. The lesion is unilateral, pruritic and relatively resistant to therapy. There are some studies on topical calcipotriol for treatment. Here a case of inflammatory linear verrucous epidermal nevus in the inguinal region of a child is presented. The eruption showed a psoriasiform appearance histologically. Calcipotriol ointment (0.005%) has applied twice a day for 45 days. The case is discussed with its clinical, pathological findings and the differential diagnosis with a review of the literature.*

Key Words: *Childhood, Dermatitis, Inflammatory Linear Verrucous Epidermal Nevus.*

INTRODUCTION

Inflammatory linear verrucous epidermal nevus (ILVEN) is an uncommon entity first described by Unna in 1896 (1, 2). The typical time of onset is childhood (3). There is a 4:1 predominance in females (2). It may present at birth or more commonly, some time after (1). The lesion is unilateral, linear, intensely pruritic, persistent and resistant to therapy (1-3). It presents as erythematous, slightly verrucous, scaling papules arranged in one or several lines. The most common location is one of the lower extremities (3). The duration of the lesions in one large review ranged from three months to thirty years. The cause remains unknown. Welch et al suggested that ILVEN represents a clonal dysregulation in growth, probably secondary to an inflammatory stimulus (1). A few sporadic cases were reported associated with auto-immune

lymphocytic thyroiditis (4), lichen amyloidosis (5), HIV-1 infection (1) and arthritis (6). Isolated reports of atypical presentations of ILVEN include adult onset, generalized lesions, bilateral distribution and familial occurrence (2, 3). ILVEN has many similarities with psoriasis both clinically and histologically (1, 3, 7).

CASE REPORT

A 3-year-old boy presented with pruritic, erythematous, slightly verrucous, scaling papules with a linear fashion localized in the right inguinal region and inner thigh (Fig. 1). The lesion was present at birth. The following year it slowly increased in size and distribution. Otherwise his general physical and systemic examination were within normal limits. The family history is negative for skin disease including psoriasis. The clinical diagnosis of



Fig. 1: Erythematous, slightly verrucous, scaling plaque with a dermatomal distribution, localized in the right inguinal region and the inner thigh.

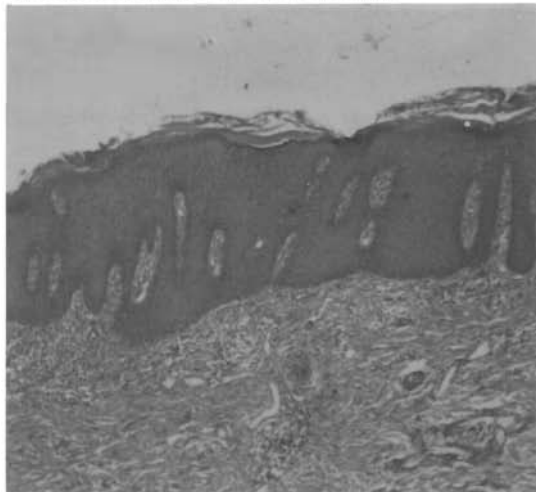


Fig. 2: Psoriasiform appearance, acanthosis, elongation and thickening of the rete ridges and mononuclear infiltration in the upper dermis (HEx40). lichen striatus, verrucous epidermal nevus, ILVEN, linear porokeratosis and psoriasis were considered. Auspitz sign was negative.

Previously, the patient was treated with topical steroids, retinoids, anti-fungal and anti-allergic agents, but there was little response. Calcipotriol ointment (0.005%) was applied twice daily. During the 16-week period of observation, the lesion remained unchanged, and no improvement was noticed.

Histopathologic examination of the skin biopsy revealed acanthosis, elongation and thickening of the rete ridges with a psoriasiform appearance (Fig. 2). Areas of hyperkeratosis with

a broadened granular layer were observed (Fig. 3), sharply demarcated from adjacent areas of parakeratosis with an absent granular layer (Fig. 4, 5). The upper dermis showed perivascular mononuclear infiltration (Fig. 2). The amyloid reaction was found to be negative with crystal violet stain.

DISCUSSION

ILVEN is considered to be a variant of epidermal nevi; however, it is described in the papulosquamous disorders due to its clinical and histologic similarity to psoriasis (3). Clinically, it manifests by the presence of pruritic verrucous inflammatory lesions that are refractory to treatment (8).

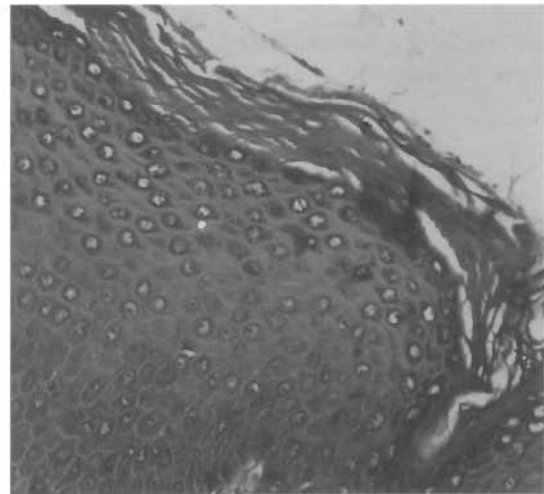


Fig. 3: Areas of hyperkeratosis with a broadened granular layer (HEx400).

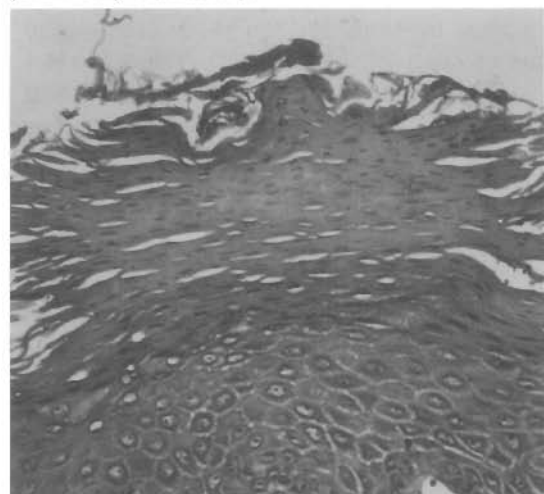


Fig. 4: Areas of parakeratosis with an absent granular layer (HEx400).

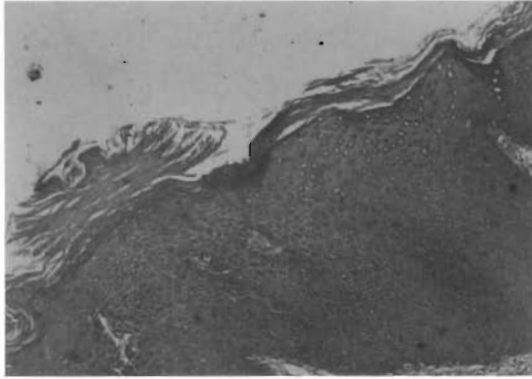


Fig. 5: The sharp demarcation between the areas hyperkeratosis and parakeratosis (HEX200).

The histology of ILVEN has been described in detail by Dupre and Christol. The alternation of relatively short, broad areas of parakeratosis overlying agranulosis, sharply demarcated from depressed areas of hypergranulosis with overlying hyperkeratosis is characteristic (9).

Neither clinical nor pathological examinations alone are sufficient to make a definitive diagnosis of ILVEN. Entities in the differential diagnosis include; psoriasis, lichen striatus, lichen planus, ichthyosis hystrix, Darier's disease and lichen simplex chronicus (2, 3). Although the differential diagnosis of psoriasis and ILVEN is sometimes difficult, the dermatomal distribution, lack of other lesions over a prolonged period, or response to therapy, as well as lack of a personal or family history of psoriasis favors a diagnosis of ILVEN (1). However lichen striatus is nonpruritic, histologically demonstrates a lichenoid infiltrate and lack epidermal acanthosis, the clinical appearance of lichen striatus and ILVEN may be indistinguishable (3). Furthermore the characteristic lesion, alternate areas of parakeratosis without a granular layer were demonstrated in our case. Lichen planus was excluded by the absence of a band-like infiltrate, destruction of the basal layer and beaded hypergranulosis. Ichthyosis hystrix, verrucous bands of epidermal nevi, lack the erythematous pruritic component of ILVEN and on biopsy alternating areas of hyperkeratosis and parakeratosis are not evident (2).

Darrier's disease was excluded by the absence of acantholysis and dyskeratosis. The last lichen simplex chronicus, was not considered because of the absence of lichenification and changes of chronic dermatitis.

Although partial response to topical calcipotriol treatment was reported (10,11,12), there was no improvement in our patients' eruptions.

Our patients' lesion confirmed to the diagnosis of ILVEN clinically and histologically.

A case of ILVEN is reported because of its rarity with its clinical, pathological findings and differential diagnosis with a review of the literature.

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