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Inflammatory Linear Verrucous Epidermal Nevus (ILVEN)

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Abstract. A 12-year-old boy with inflammatory linear verrucous nevus is described. The lesions were pruritic and consisted of small, erythematous, slightly scaling papules coalescing to form linear, lichenified and excoriated plaques. The histological picture was psoriasiform but features of eczema were also seen.

Key words: Linear epidermal nevus; Inflammatory psoriasiform features

In 1971 Altman & Mehregan, basing their investigation on 25 cases, described inflammatory linear verrucous epidermal nevus (ILVEN) as a distinct clinical and histological variety of linear verrucous nevus or nevus unius lateris (1). Sporadic cases had earlier been reported as "cases for diagnosis" (4), atypical cases of linear psoriasis (2, 6), or as psoriasiform linear nevus (7). Additional cases have been presented in a few reports (3, 5, 8) following establishment of the condition as a distinct disease entity.

The following features are characteristic of ILVEN:

1. Unilateral, usually pruritic lesions, consisting of small, discrete, erythematous, slightly verrucous, scaling papules which tend to coalesce to form linear plaques.
2. Clinical and histological resemblance to either psoriasis or eczema.
3. Absence of typical psoriasis.
4. Female predominance of 4:1.
5. Early onset of lesions, 50% before the age of 6 months, 75% prior to the age of 5 years.
6. Minimal response to treatment.

CASE REPORT

A 12-year-old boy was admitted to the Department of Dermatology, Odense University Hospital, with an extremely pruritic, linear lesion on his right leg extending from the ankle to the loin (Fig. 1). There was no family history of psoriasis or atopic disease. The lesion had developed at the age of 6 years, starting in the popliteal area

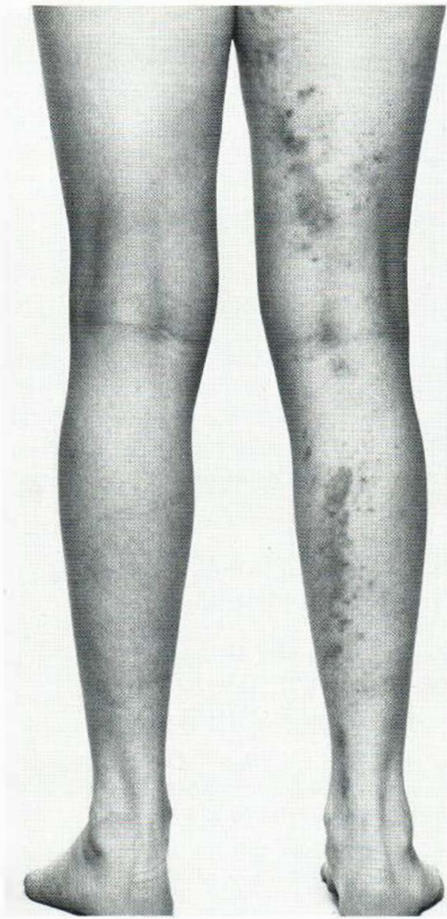


Fig. 1. Linear cluster of excoriated, slightly scaling erythematous papules on right buttock, posterior part of and lower leg.

and extending both centrifugally and centripetally, reaching the present localization in one year. A similar zosteriform lesion of one year's duration had been present on the right part of the abdomen but had vanished recently leaving slight hyperpigmentation. The eruption consisted of small excoriated or slightly scaling papules forming linear, lichenified, erythematous plaques. Treatment with potent steroid ointments was ineffective except for some amelioration of the itching. During hospitalization the patient was given Bctamethasone dipropionate 0.05 % cream (Diproderm, Essex) under plastic occlusion at night and crude coal tar application in the morning. The lesions responded within one week with an approximately 50% reduction in pruritus, infiltration and erythema. Only a slight further improvement was achieved.

Histopathological examination revealed features consistent with psoriasiform sub-acute dermatitis (Fig. 2). The scaling was mainly orthokeratotic but areas of parakeratosis associated with absence of the granular cell layer were also seen. Epidermis showed acanthosis, papillomatosis and elongation of rete ridges. Interdermal edema, epidermal infiltration with small amounts of lymphocytes and granulocytes and subcorneal vesiculation were present. No Munro's microabscesses could be found. A perivascular infiltration with lymphocytes and histiocytes was present in the upper dermis.

DISCUSSION

The case presented showed typical clinical and histological features of ILVEN. Linear neurodermatitis and linear psoriasis are unlikely to start in early childhood and to remain in the same localization for 6 years independent of treatment or seasonal variation. Lichen striatus appears primarily on the extremities of children, but itching is not a prominent feature and even prolonged courses do



Fig. 2. Biopsy of inflammatory linear verrucous nevus with papillomatous acanthosis, subcorneal vesiculation, spongiotic edema and polymorphonuclear leukocytes at tip of papilla.

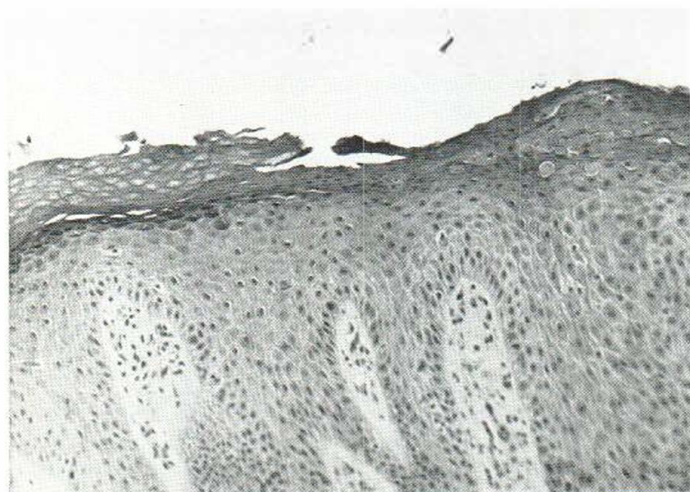


Fig. 3. The scaling is partly parakeratotic, with loss of granular layer.

not exceed one year. Finally, lichen planus and Darier's disease can be excluded on histological grounds.

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Oral Methoxsalen Photochemotherapy of Uncommon Photodermatoses

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Abstract. Three uncommon idiopathic photodermatoses, refractory to other treatment, responded to oral psoralen photochemotherapy. Two females with photosensitivity in association with atopy, one female with persistent light reaction following a systemic drug-induced photosensitivity and an elderly male with actinic reticuloid were treated.

Key words: PUVA; Photosensitivity; Actinic reticuloid

Patients with severe idiopathic photodermatoses are greatly disabled by their condition leading to an almost hermit-like existence. These patients usually receive little or no benefit from topical sunscreens and, therefore, new approaches to treatment are constantly being sought. Antimalarials have been used for many years and help some patients but are not without risk. Recently β -carotene has been found to produce some alleviation of symptoms in a proportion of patients (4, 9, 10). Recent findings that oral psoralen photochemotherapy (PUVA) is beneficial in atopic eczema (5), mycosis fungoides (2), and polymorphic light eruption (3, 7) prompted us to try this therapeutic modality in patients with