Dithranol in the Treatment of Inflammatory Linear Verrucous Epidermal Nevus

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A case of inflammatory linear verrucous epidermal nevus (ILVEN) is reported. Short contact treatment with dithranol resulted in complete relief from itching and a remarkable clearing of all linear lesions except from a small verrucous band on the shin. In patients with ILVEN it is advisable to try dithranol therapy before carrying out surgical procedures such as excision, cryotherapy, electrocautery. The prompt response to dithranol is best explained by the assumption that most of the lesions in this case of ILVEN represented true linear psoriasis. *Key words: ILVEN; Psoriasis.* (Accepted June 22, 1988.)

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Inflammatory linear verrucous epidermal nevus (ILVEN) is a relatively rare but troublesome disorder. Based on the series of Altman & Mehregan (1) and a review of the more recent literature (2), the following criteria seem to be valid:

(i) early age of onset;
(ii) pruritic erythematous, slightly verrucous scaling of papules, which coalesce to linear plaques;
(iii) histopathological features resembling those of psoriasis;
(iv) persistent lesions showing marked refractoriness to treatment.

The criterion 'resistance to therapy', is a generally accepted feature of this nevus (3). Here we report on a patient whose lesions responded well to dithranol treatment.

CASE REPORT

A 44-year-old man presented with a severely itching linear skin disorder. At the age of 12 years a linear reddish scaling lesion appeared on the left shin following a minor trauma. During the following years similar lesions occurred on the dorsal aspect of the left lower leg, spreading over the dorsomedial aspect of the upper leg, genital region and the lower thoracic and lumbal areas of the left side of his body. From the age of 25 years onward the lesions had persisted without any response to treatment with UVB, PUVA and potent topical corticosteroid preparations.

Physical examination revealed erythematosquamous plaques arranged in a linear pattern following the lines of Blaschko (Fig. 1*a*, 2*a*) (4). On the left shin a slight verrucous aspect was seen. The erythematosquamous bands on the dorsomedial aspect of the left leg, genital region and lower half of the left side of the trunk had a more psoriasiform appearance. The finger nails showed multiple pits which appeared sharply punched out. On the scalp and on the extensor surfaces of the elbows sharply demarcated coin-sized erythematosquamous plaques were seen.

A 4 mm punch biopsy from the lesion on the left shin showed acanthosis with elongation of the rete ridges. There were areas with hyperkeratosis of the orthotype and areas with parakeratosis. The upper dermis showed a mild predominantly mononuclear infiltrate. Polymorphonuclear leukocytes were sparse and no exocytosis of these cells into the epidermis was seen.

The patient was treated with dithranol according to a short contact schedule. Treatment was started with dithranol 0.1% and 0.25% in a cream base (Psoricream, Essex, Amstelveen, The Netherlands). The lesions in the genital region markedly improved on dithranol 0.25%. For the treatment of the lesions on the left leg and trunk the concentration of dithranol was increased twice weekly according to the range 0.5%, 1%, 2%, 3%, 4%, 5%. Dithranol was applied in a stiffened paraffin base as described by Seville, containing 2% salicylic acid (5). After 8 weeks of treatment maximal improvement was reached (Fig.

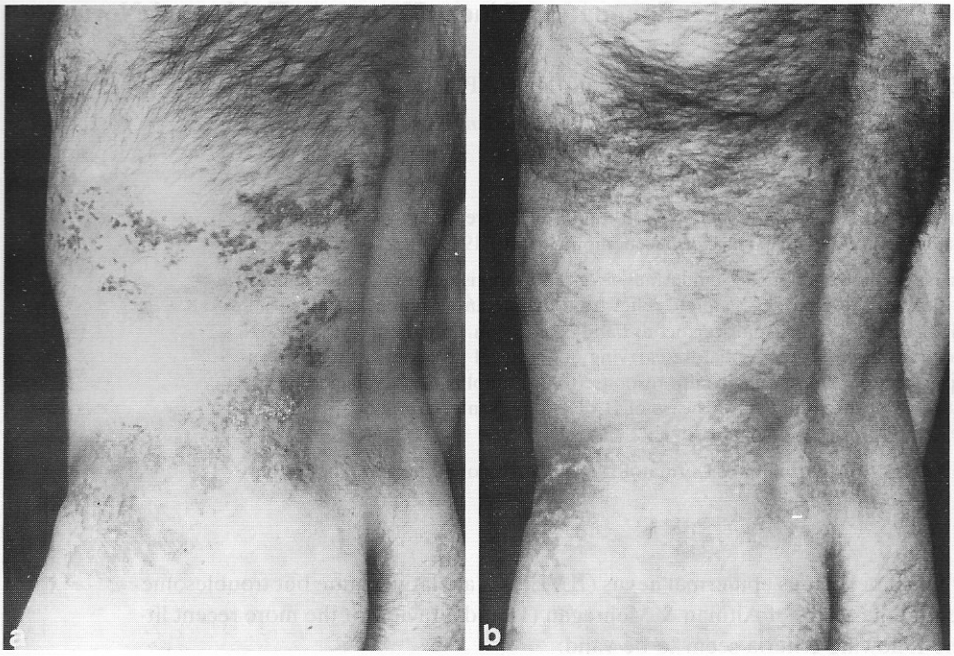

Fig. 1. Erythematosquamous plaques in a linear distribution on the trunk. (*a*) before treatment, (*b*) after treatment with dithranol.

1*b*, 2*b*). The troublesome pruritus had disappeared totally and a remarkable improvement up to virtually total clearing was reached in all areas with the exception of a verrucous zone on the shin that persisted virtually unchanged.

DISCUSSION

The skin lesions in the present case fulfill the criteria for the diagnosis ILVEN: (i) onset before puberty; (ii) pruritic sensation and typical clinical appearance; (iii) psoriasiform histological features; (iv) resistance to topical corticosteroids, PUVA and UVB treatment. However, treatment with dithranol—a therapy which is effective specifically in psoriasis and seborrhoeic dermatitis—proved to be effective in all linear lesions except for a limited area on the shin. From a clinical point of view a diagnosis of linear psoriasis would be acceptable for most of the linear lesions in the present case. The presence of signs of psoriasis in other areas (nails, elbows, scalp) further supports this diagnosis.

In the past, many authors have discussed the nosological significance of the ILVEN. It has been argued that at least some cases of ILVEN represent *true linear psoriasis* (6–8). In two of these cases (6, 7) classical manifestations of psoriasis were seen on other skin sites and in all 3 cases antipsoriatic therapies such as chrysarobin, tar and ultraviolet radiation resulted in complete clearing of the lesions. Alternatively, an *epidermal nevus of the common non inflammatory type* may constitute a *site of predilection for psoriasis* as described by Sugai et al. and Bennett et al. (9, 10). Theoretically, the clinical and histological appearance of such a case may be indistinguishable from classical ILVEN. In the case reported by Bennett et al. (10) the systematized nevus adopted a psoriatic appearance and typical psoriatic lesions were seen on other sites. Treatment with dithranol resulted in resolution of the typical psoriatic lesions, whereas the linear lesions remained unaffected. Ogino and Higuchi (11) described a

Fig. 2. The dorsal aspect of the left lower leg, showing erythematosquamous lesions in a linear distribution. (*a*) before treatment, (*b*) after treatment with dithranol.

patient with generalized pustular psoriasis associated with ILVEN. With methotrexate an almost complete clearance of pustular psoriasis was achieved. However, pustule formation remained within the nevus, indicating that ILVEN might represent a 'locus minoris resistentiae' to psoriasis. A third subclass of ILVEN may be *'ILVEN SUI GENERIS'* (1, 2, 12, 13). This subclass has been claimed to be remarkably resistant to topical treatment.

The present case apparently represents a manifestation of true linear psoriasis, extending far beyond the clinical boundaries of 'ILVEN SUI GENERIS' (localized on the left shin). This case demonstrates that the ILVEN is not only a site of predilection for superimposing psoriasis (9, 10, 11), but that even new psoriatic lesions may develop in linear zones following the lines of Blaschko. A similar distribution of linear psoriasis outside the limits of an epidermal nevus has been described by Bondi (14). This distribution is not at random but is arranged according to the lines of Blaschko that visualize the migration of mutant cells derived from a small group of stem cells during embryogenesis (15).

The present observation indicates that therapeutical defeatism in the management of patients with ILVEN is not always justified. As some cases may represent true linear psoriasis, an attempt to treat the lesions with dithranol seems worthwhile.

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Psoriasis of the Nails Treated with Grenz Rays: A Double-blind Bilateral Trial

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Lindelöf B. Psoriasis of the nails treated with grenz rays: A double-blind bilateral trial. Acta Derm Venereol (Stockh) 1989; 69: 80–82.

The effect of grenz ray therapy in the treatment of psoriatic nails was assessed in 22 patients by randomly allocating active treatment to the psoriatic nails of one hand while the other one, which received simulated therapy, served as a control. Five Gy of grenz rays were applied on 10 occasions at intervals of 1 week. There was a significantly better response to active treatment compared with the untreated control. However, the therapeutic response was moderate. It is concluded that grenz ray therapy could be useful only when the psoriatic nails are of normal thickness. (Received June 1, 1988.)

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Various therapeutic principles are used in the treatment of psoriatic nails, but the results are usually unpredictable and not always successful. For a recent review see Scher (1). X-rays (1, 2) as well as PUVA (3) have been used to irradiate psoriatic nails with good to moderate response. Grenz rays (ultrasoft X-rays) have also been widely used either alone, or in combination with topical medicaments. However, little has been written about the effect of grenz ray on the nail changes of psoriasis. The present double-blind placebo controlled trial was, therefore, designed to assess the efficiency of grenz rays on psoriatic nails. The exposure dose, kV and fractionation of the grenz ray therapy is the standard regime which has been used at the Department of Dermatology, Karolinska Hospital, Stockholm, for several years.

PATIENTS AND METHODS

Patients

Twenty-four patients with psoriasis of the nails of both hands took part in the study. Age range was 29–75 yr. Duration of disease was 1–15 yr. The patient had been untreated for at least 6 months before the start