Inflammatory Linear Verrucous Epidermal Nevus

Epidermal Protein Analysis in Four Patients

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• **Inflammatory linear verrucous epidermal nevus (ILVEN) may be difficult to differentiate clinically from psoriasis. We report four cases of ILVEN that confirm the observation that the analysis of epidermal proteins by sodium lauryl sulfate–polyacrylamide gel electrophoresis (SLS-PAGE) discloses a pattern different from that seen in psoriasis. We have also found, however, that the SLS-PAGE epidermal protein pattern is not identical in each case of ILVEN.**

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Inflammatory linear verrucous epidermal nevus (ILVEN) is a clinicohistopathologic entity characterized by an inflammatory psoriasiform lesion that is often intensely pruritic and refractory to treatment.[1] It is occasionally associated with congenital malformations of the skeletal or other systems.[2] Although diagnosis on clinical and histologic grounds is usually not difficult, differentiation from linear psoriasis, linear lichen simplex chronicus, linear Darier's disease, and lichen striatus may be challenging on occasion.

Adrian and Baden[2] showed that sodium lauryl sulfate–polyacrylamide gel electrophoretic (SLS-PAGE) analysis of epidermal fibrous proteins in ILVEN differed from the pattern observed in psoriasis in one case in which the clinical differentiation was difficult. The patient was a 22-month-old girl who also had tetralogy of Fallot. We have performed SLS-PAGE epidermal protein analysis on four additional patients with ILVEN in whom the diagnosis had been established or strongly considered to confirm the difference in pattern from psoriasis and to determine whether epidermal protein analysis in ILVEN would disclose any unique, consistent changes in all patients.

REPORT OF CASES

CASE 1.—A 7-year-old girl had had a two-year history of a pruritic, gradually extending linear rash on the right lower extremity. There had been no other clinical changes in the skin or its appendages. The family history was unknown as the patient had been adopted. Physical examination disclosed discrete and confluent psoriasiform, erythematous, scaling papules and plaques distributed in a linear array on the right lower extremity extending from about the mid thigh to the lower leg. The maximum width of the linear area of involvement was about 2 cm. There were no nail or other skin changes. Treatment of the patient with tar ointments, UV radiation, topical corticosteroids, and local preparations containing liquor carbonis detergens, salicylic acid, or 0.05% tretinoin each provided only minimal temporary benefit.

CASE 2.—A 27-year-old man had had an intensely pruritic rash on the right lower buttock and upper part of the thigh since birth. He had been healthy, and both physical and mental development had been normal. There were no other integumental abnormalities. The family history was noncontributory. Physical examination disclosed a linear erythematous scaling plaque involving the right lower part of the buttock and adjacent upper part of the thigh. Microscopic examination of a biopsy specimen of this lesion disclosed diffuse marked parakeratosis overlying psoriasiform epidermal hyperplasia, alternating with small foci of orthokeratosis. There were cup-shaped hypergranulotic epidermal depressions. A superficial perivascular lymphohistiocytic infiltrate was also observed. A diagnosis of ILVEN was made on clinical and histopathologic grounds. The lesion was refractory to treatment with numerous topical modalities.

CASE 3.—A 9-year-old girl in good health had had a minimally pruritic rash on the right lower extremity that had been present from birth and that had grown in proportion to her somatic growth. The family history was noncontributory. On physical examination, she had a linear array of discrete and confluent erythematous papules and plaques with overlying silvery scale on the right lateral thigh extending from the greater trochanter to just above the knee. The maximum width of the lesion was about 2 cm. There was some improvement of the patient's lesions in response to topical corticosteroid treatment.

CASE 4.—A 58-year-old man had had a 36-year history of a moderately pruritic lesion on the left lower extremity and a 25-year history of a similar lesion on the left side of the trunk and upper extremity. There were no other cutaneous abnormalities. Physical examination disclosed erythematous, hyperkeratotic plaques 1 to 2 cm wide in a linear distribution on the left arm, left thigh, and left leg, with extension along the left sole to the left great toe. The clinical diagnosis was ILVEN.

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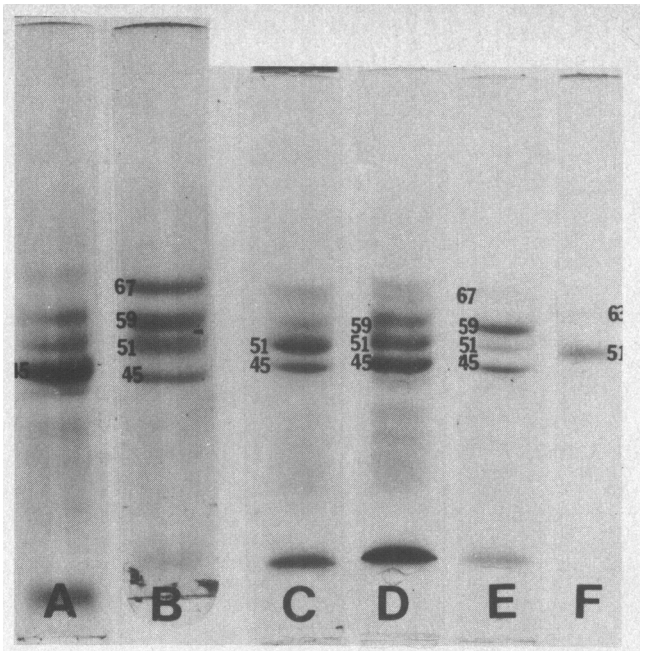

Sodium lauryl sulfate-polyacrylamide gel electrophoresis patterns of proteins extracted from scales. A, B, C, and D are from patients with inflammatory linear verrucous epidermal nevus (ILVEN) (cases 1, 2, 3, and 4, respectively). E is from a psoriatic plaque and F is from normal skin. Specimens A, B, and F were analyzed separately, and C, D, and E were analyzed together. Standards were included in all runs to determine molecular weights; 45 indicates 45,000; 51, 51,000; 59, 59,000; 63, 63,000; and 67, 67,000. Note that normal pattern (F) includes prominent bands only at molecular weight of 51,000 and 63,000. In contrast, psoriatic pattern (E) includes prominent bands at molecular weight of 45,000 and 59,000 that are not present in normal skin. Cases of ILVEN (A through D) differ from both of these and from each other in relative proportions of bands.

Microscopic examination of a biopsy specimen from the lesion on the left side of the chest wall showed parakeratosis, elongation and widening of the rete ridges, and a chronic perivascular inflammatory cell infiltrate. Small collections of polymorphonuclear cells were found in areas of spongiosis immediately below the granular layer. The findings were interpreted as "compatible with psoriasiform dermatitis." A microscopic examination of a biopsy specimen from the left leg showed hyperkeratosis with parakeratosis and rather regular downward proliferation of the prickle cell layer. Treatment with numerous modalities, including topical corticosteroids, emollients, 0.1% tretinoin cream, and a modified Goeckerman regimen, failed to have any notable effect on the patient's skin lesions.

Epidermal Protein Analyses

Specimens of scale from involved areas were obtained from each of the aforementioned patients whose conditions were diagnosed as ILVEN. These specimens were processed for SLS-PAGE according to the procedure outlined by Baden et al.[3] Briefly, the tissue was extracted with 0.05M TRIS buffer, pH 9.5, containing 6M urea and 0.1M mercaptoethanol and then dialyzed against 0.05M TRIS buffer at pH 7.5 to obtain the fibrous proteins that were precipitated out of solution. When SLS-PAGE, which separates polypeptides on the basis of size, was performed on the protein isolated from stratum corneum scraped from the epidermal surface of normal individuals and from uninvolved skin of patients with psoriasis, two bands were seen at a molecular weight of 51,000 and 63,000 (Figure, F). In contrast, the pattern we have observed in affected areas in more than one hundred individuals with untreated psoriasis showed two strong bands at a molecular weight of 45,000 and 59,000 and very weak bands at a molecular weight of 51,000 and 67,000 (Figure, E).

The patterns obtained from the four patients with ILVEN are shown in the Figure (A through D) and differ from those seen in psoriasis and normal skin in the relative proportion of the bands. In patient 1 (Figure, A), the band with a molecular weight of 45,000 was very strong. In patient 2 (Figure, B), the bands with a molecular weight of 45,000, 51,000, 59,000, and 67,000 were of equal intensity. In patient 3 (Figure, C), the bands with a molecular weight of 45,000 and 51,000 were very strong. In patient 4 (Figure, D), the bands with a molecular weight of 45,000, 51,000, and 59,000 were strong and of almost equal intensity.

COMMENT

Although the patterns of epidermal fibrous polypeptides in these four cases of ILVEN were not the same, they differed from the patterns described for psoriasis and normal superficial stratum corneum. Case 3, however, did have the same pattern described in a previous case of ILVEN.[2]

Features said to be characteristic of ILVEN include the following: early age at onset; 4:1 female-male ratio; frequent involvement of the left side of the body; notable pruritus; persistent lesions refractory to treatment; and distinct inflammatory, psoriasiform histopathologic features.[1,2] However, none of these is entirely conclusive in a single patient. For example, age at onset may vary considerably. In two of our patients, ILVEN was present at birth, but, in the other two, disease onset occurred at the ages of 5 and 22 years, respectively. Of the 25 patients with ILVEN whose cases were reported by Altman and Mehregan,[1] lesions were present at birth in only four patients. Age at onset in the remainder of the cases varied from less than 6 months for approximately 50% of the patients to as late as age 49 years.

Our results indicate that scale analysis provides additional data that are useful in distinguishing ILVEN from psoriasis. They further demonstrate, for the first time, that the SLS-PAGE pattern of epidermal fibrous proteins from ILVEN may differ from case to case and, thus, that a single unique pattern does not exist for that disease.

References

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