Unilateral Psoriasis: A Case Individualized by Means of Involucrin

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Several authors have questioned the existence of unilateral (linear) psoriasis. These authors have suggested that the condition is actually an inflammatory linear verrucous epidermal nevus, or the result of an isomorphic effect on a pre-existing epidermal nevus. We report the case of a 25-year-old man, with no relevant personal or family history, who presented with a number of pruritic, punctiform erythematosquamous lesions that were linearly distributed over the left side of the body. Clinical examination and results of histopathologic and histochemical studies indicated unilateral psoriasis. Our findings confirm that involucrin immunohistochemistry can be a useful diagnostic tool in cases of this type. Treatment with keratolytics and topical calcipotriol led to a significant, but only temporary, improvement.

Unilateral (linear) psoriasis and inflammatory linear verrucous epidermal nevus (ILVEN) have very similar clinical and histopathologic characteristics,[1] making it often impossible to differentiate between the two. Indeed, a number of authors have suggested that unilateral psoriasis is merely an ILVEN. However, it has been reported that the involucrin expression in ILVEN differs from that in psoriasis.[2] We demonstrate how immunohistochemistry with involucrin can be used to differentiate between these two entities.

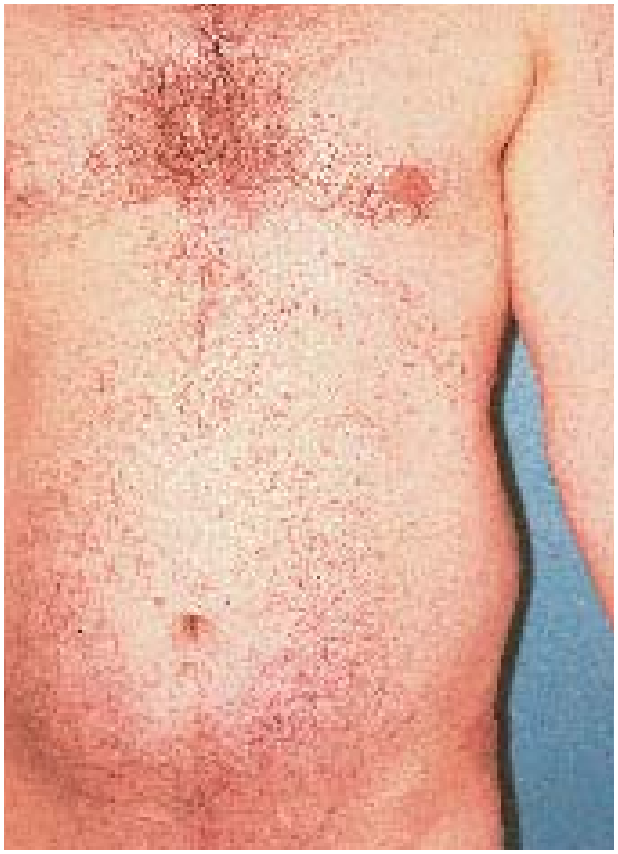

FIGURE 1. Erythematosquamous papules and plaques are distributed on the left half of the body and do not cross the midline.

Case Report

A 25-year-old man had erythematous lesions on the inner left arm first noted 3 years previously. The lesions had subsequently extended down the left torso and the left leg, and at no stage crossed the midline. The lesions were at times pruritic. The patient received topical treatment with emollients and steroids, and systemic treatment with anxiolytics and multivitamin preparations. In all instances, the treatments resulted in minimal improvement. When we saw him, the patient was not taking medications of any kind.

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FIGURE 2. Erythematosquamous papules and plaques are distributed on the left half of the body and do not cross the midline.

He reported no relevant history other than pulmonary tuberculosis, which had been treated appropriately. There was no family history of psoriasis or any other skin disorder.

Physical examination revealed numerous confluent papular erythematosquamous lesions, linearly distributed on the inner and outer left arm, down the left side of the thorax, on the anterointernal region of the thigh, and in the genital fold. At no point did the lesions cross to the right side of the body (Figures 1 and 2). No other skin lesions, or ungual, scalp, or mucosal lesions, were detected.

Results of a complete blood count, blood biochemistry tests, and urinalysis were normal. A serologic test for syphilis showed negative results. Neither a direct examination nor a culture of lesion samples revealed pathogenic fungi.

Histopathologic examination revealed the typical characteristics of psoriasis: acanthosis, papillomatosis, and parakeratosis in the epidermis and capillary dilation associated with chronic perivascular infiltration and exocytosis in the papillary dermis (Figure 3).

Results of immunohistochemical studies for involucrin revealed this protein throughout the epidermis, from the suprabasal keratinocyte layer upwards (Figure 4).

Comments

Unilateral psoriasis and ILVEN are very difficult to differentiate, and a number of authors have suggested that the former is simply an ILVEN. The typical (although by no means defining) characteristics of ILVEN were established by Altman and Mehregan[3] (Table I). These authors considered unilateral psoriasis to be an extremely rare disorder.

Diagnosis tends to be particularly difficult in cases that do not conform to the classic criteria for ILVEN, or in those with both nevoid and psoriatic lesions. Various histopathologic features (acanthosis, papillomatosis, the presence of ortho- and parakeratotic areas, and alterations of the papillary dermis) are typically observed in all difficult cases, and are clearly not useful for diagnosis.[1] In cases of this type, authors have generally chosen one of three diagnoses[4]: unilateral psoriasis,[5-8] ILVEN, or Koebner's phenomenon on a pre-existing epidermal nevus in a subject with psoriatic diathesis.[4,9-11] This latter possibility would explain cases in which inflammation arises over a linear epidermal nevus present since childhood, with or without psoriatic lesions in other locations. Such cases can be expected to respond well to classic antipsoriatic treatment.[11,12]

There has been one reported case of unilateral psoriasis associated with contralateral lichen striatus, possibly reflecting a common underlying cause.[8]

An objective test to differentiate between unilateral psoriasis and ILVEN is the pattern of involucrin expression in the epidermis. Involucrin, which is used as a marker of epidermal differentiation, is a protein precursor of the cornified envelope. In the normal epidermis, it is present in the upper part of the prickle-cell layer and in the granular layer, but in psoriasis it is detectable at deeper levels, from the suprabasal keratinocytes upwards.[13-15] Ito and colleagues[2] showed that in ILVEN, the involucrin expression is increased in orthokeratotic regions but is deficient in parakeratotic regions; by contrast, in parakeratotic areas of psoriasis, most suprabasal keratinocytes express involucrin.

Other authors have suggested that the behavior of other markers (elastin, antikeratin 10, and antiker-

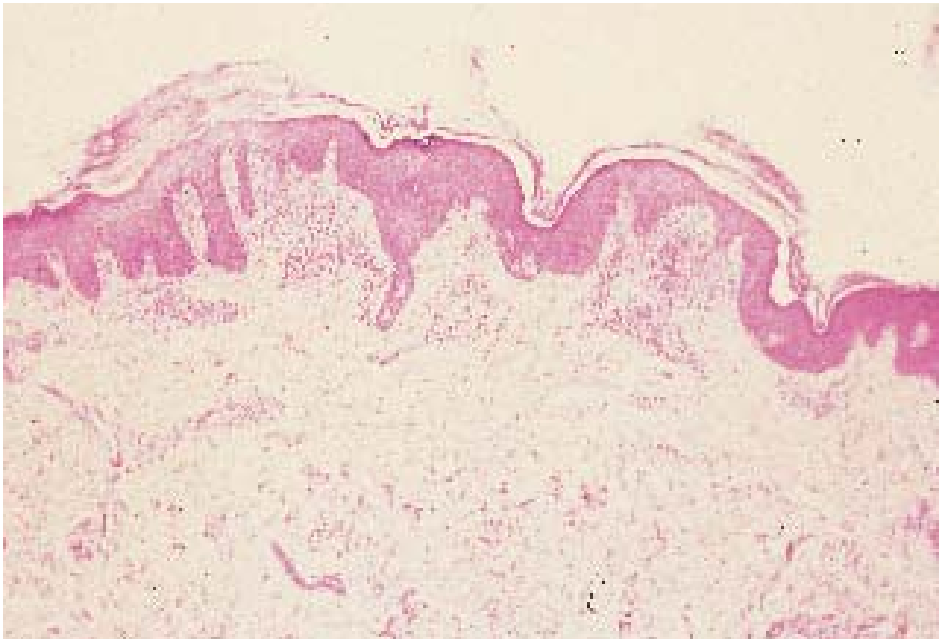

FIGURE 3. Histopathologic examination shows a psoriasiform image, with papillomatosis, acanthosis, ortho- and parakeratotic zones, and capillary dilation with a slight perivascular infiltrate in the papillary dermis (H&E; original magnification, X 25).

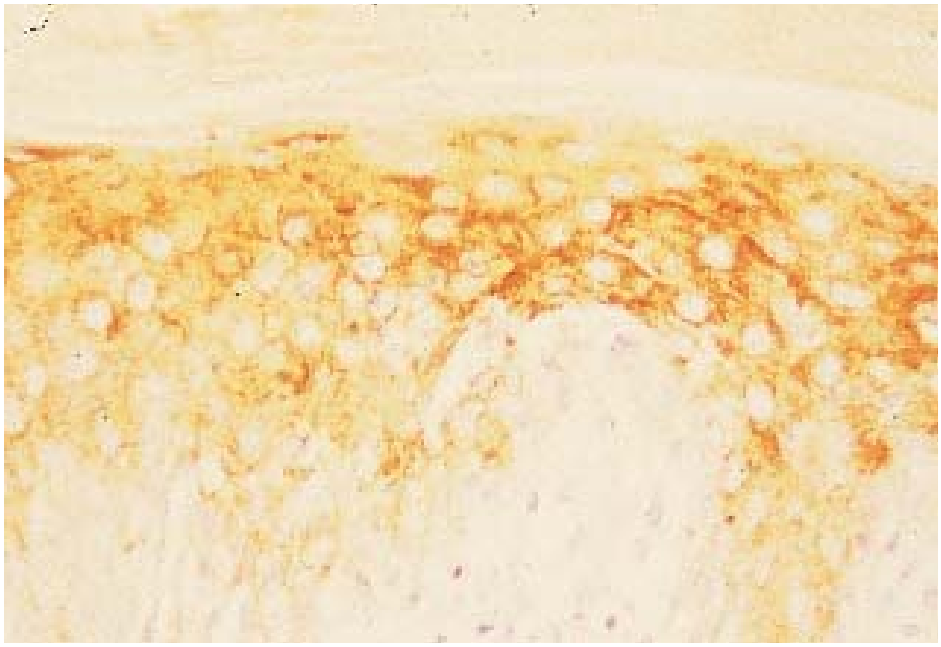

FIGURE 4. Involucrin is present from the suprabasal keratinocytes up to the granular layer, both in orthokeratotic and parakeratotic areas. This pattern is typical of psoriasis and different from that of ILVEN (original magnification, X 400).

atin 16) may prove useful for differentiating between unilateral psoriasis and ILVEN.[1]

The case described here clearly displays the characteristics of unilateral psoriasis. The patient's late onset of symptoms and his temporary response to treatment with topical calcipotriol argue against the Altman and Mehregan criteria for ILVEN. The subsequent appearance of lesions of inverse psoriasis also argues against a diagnosis of ILVEN. We did not detect any evidence of a linear epidermal nevus prior to the onset of symptoms, which argues against the possibility that the observed lesions arose as an isomorphic effect. The results of involucrin immunohistochemistry testing provide evidence that the patient had unilateral psoriasis.

Differentiation between ILVEN and unilateral psoriasis is clearly important, since the two disorders respond to treatment in different ways. Unilateral psoriasis usually shows some response, whereas ILVEN is characterized by its persistence despite numerous

Table I

Altman and Mehregan's Criteria for the Diagnosis of ILVEN[3]

1. Early onset (before 5 years of age in 75% of cases)
2. Four times more common among women than men
3. Left leg frequently affected
4. Pruritus
5. Psoriasiform appearance
6. Persistent and resistant to treatment

attempts at treatment. This differentiation can especially avoid expensive or aggressive treatments in patients with the latter disorder.

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