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Linear psoriasis or ILVEN? A pediatric case unraveling the mosaic puzzle

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Abstract

Linear psoriasiform eruptions in children are diagnostically challenging, particularly when they follow Blaschko's lines. The main differential diagnosis includes inflammatory linear verrucous epidermal nevus (ILVEN) and linear psoriasis (LP), a rare mosaic variant. We describe the case of a 5-year-old boy with extensive unilateral lesions clinically suggestive of both entities. Histology revealed Munro microabscesses and psoriasiform hyperplasia. The patient showed rapid improvement with topical calcipotriol/betamethasone, followed by long-term remission. Notably, after 8 years of follow-up, the lesions remained strictly confined to the original Blaschkoid distribution, with no extracutaneous involvement and only mild recurrences. This clinical course strongly supports the diagnosis of isolated linear psoriasis (type 1 mosaicism). Few pediatric cases in the literature have documented such a long and stable evolution, making this report a valuable contribution to the distinction between LP and ILVEN. The case highlights the importance of integrating clinical, histological, and longitudinal data when evaluating linear dermatoses in children.

Introduction

Linear psoriasiform eruptions in childhood represent a rare and often challenging diagnostic entity, particularly when they follow the lines of Blaschko. The two main considerations are inflammatory linear verrucous epidermal nevus (ILVEN) and linear psoriasis (LP), the latter being a mosaic form of psoriasis first clearly distinguished by Happle's classification.¹⁻³ Differentiating these conditions is critical, especially in pediatric patients, due to their differing natural histories and therapeutic responses.^{4,5}

Case Report

A 5-year-old boy presented with erythematous scaly plaques in a Blaschkoid distribution involving the left hemibody (Figure 1 a-c). The lesions had emerged several months earlier, following a streptococcal infection, and had gradually worsened. Pruritus was mild. Examination revealed widespread psoriasiform plaques with silvery scale on the left trunk, limbs, and retroauricular area. Fingernails and toenails were unaffected. Mild scalp flaking was observed. Family history for psoriasis was negative. A 4-mm punch biopsy from the thigh showed marked acanthosis, hypogranulosis, and parakeratosis with neutrophils forming Munro microabscesses (Figure 1d).² Based on the histopathology and clinical appearance, both ILVEN and LP were considered.^{5,6} The patient responded rapidly to a 4-week course of topical calcipotriol/betamethasone. Maintenance therapy with emollients and intermittent corticosteroids followed. At a recent telephone follow-up,

the patient's family reported that, after 8 years, the lesions had remained strictly confined to the original Blaschkoid distribution, with only mild recurrences managed with topical therapy.

Discussion

According to the classification proposed by Happle, LP can manifest as type 1 (isolated), with lesions limited to the mosaic pattern, or type 2 (superimposed), where segmental lesions precede or coexist with generalized psoriasis.¹⁻³ In our patient, the absence of extracutaneous involvement and the persistence of Blaschkoid lesions over 8 years support a diagnosis of type 1 isolated mosaicism. Differentiating LP from ILVEN remains difficult in the early stages. ILVEN tends to begin in infancy, presents with intense pruritus, shows a chronic course, and is typically refractory to antipsoriatic treatments.⁴⁻⁶ Histology may be similar, though immunohistochemistry (involucrin positivity in LP, Ki-67/K10 patterns) may help when available.⁶ In this case, the therapeutic response, lesion distribution, and long-term evolution all favored LP over ILVEN, with clinical features ultimately consistent with type 1 mosaicism.

Conclusions

This case emphasizes the need for integrating clinical, histological, and therapeutic data when evaluating linear psoriasiform eruptions in children. Long-term follow-up was instrumental in confirming the diagnosis of isolated linear psoriasis (type 1 mosaicism).

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Figure 1. Clinical and histological photographs of the patient. (a-c) Cutaneous examination showed scaly plaques arranged in linear bands along the lines of Blaschko extending from the scalp to the feet, with a marked midline cutoff. (d) Skin biopsy from a scaly plaque of the thorax showed parakeratosis, areas of agranulosis, papillomatosis with dilated capillaries in the tips of papillae and Munro's microabscesses (H&E, 10x magnification).

