Bilateral Inflammatory Linear Verrucous Epidermal Nevus Associated with Psoriasis

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Inflammatory linear verrucous epidermal nevus is a benign hyperplasia of the epidermis. Bilateral distribution of inflammatory linear verrucous epidermal nevus is an extremely rare presentation. Although inflammatory linear verrucous epidermal nevus is generally accepted as a distinct entity, it has similar clinical and histopathologic features to psoriasis, or they occasionally may overlap. We report the case of an 8-year-old boy with bilateral inflammatory linear verrucous epidermal nevus who also had psoriasis.

Inflammatory linear verrucous epidermal nevus is a benign hyperplasia of the epidermis of unknown cause. It is a rare skin disorder that manifests as localized or systemic forms with hyperpigmented verrucous lesions arranged linearly along the lines of Blaschko. Most frequently, the lesions occur on the left lower extremity, but the right side can also be affected. Bilateral involvement is the least reported morphologic distribution. Inflammatory linear verrucous epidermal nevus may be present at birth; however, the majority of the cases appear during infancy and childhood. Although isolated reports of familial cases have been reported, the exact mode of transmission is not known.

The histologic and clinical similarities between inflammatory linear verrucous epidermal nevus and psoriasis have led some authors to speculate whether these two diseases are distinct entities. Moreover, in rare instances systemic epidermal nevus and psoriasis may be observed together. A common histopathogenic abnormality in the epidermal cell metabolism of inflammatory linear verrucous epidermal nevus and psoriasis has also been suggested.

We report a case of inflammatory linear verrucous epidermal nevus showing bilateral distribution associated with psoriasis.

Case Report
An 8-year-old boy presented with a multiple darkly pigmented papular eruption that had been present since age one month. The lesions spread in a linear fashion over the body and extremities and they were extremely pruritic. There was no consanguinity or family history of similar lesions. Dermatologic examination revealed multiple, linear, hyperpigmented verrucous papules and plaques, distributed bilaterally over the extremities and the trunk (systemic morphology), particularly along Blaschko's lines. The lesions were more prominent on the left side of the body. Erythematous, slightly scaly, round to oval psoriatic plaques were detected on the upper extremities, thighs, and buttocks (Figure 1). Mucous membranes were normal. The nails were dystrophic, particularly on the left hand. The remainder of the physical examination was unremarkable.

Histopathologic examination of a punch biopsy specimen of verrucous lesions showed hyperkeratosis, parakeratosis, areas of hypergranulosis, papillomatosis, acanthosis, elongation of the rete ridges, and a mild perivascular mononuclear cell infiltration in the upper dermis. Alternating parakeratosis without a granular layer and orthokeratosis with a thickened granular layer was demonstrated (Figures 2 and 3). These findings were compatible with inflammatory linear verrucous epidermal nevus. The pathologic examination of the psoriatic plaques demonstrated similar psoriasiform features to that of inflammatory linear verrucous epidermal nevus, however, it was diagnosed as psoriasis regarding the clinical appearance.

The patient was given occlusive treatment with betamethasone valerate. At the end of one month, the psoriasis plaques resolved completely, whereas the verrucous lesions showed little improvement (Figure 4).
**Comments**

Clinical criteria for the diagnosis of inflammatory linear verrucous epidermal nevus were defined as early age of onset, a predominance in female patients, frequent involvement of the left lower extremity, substantial pruritus, persistency of the lesions, and refractoriness to treatment. In the case presented, the verrucous lesions occurred at one month of age, there was intense pruritus, and the lesions showed minimal response to treatment. Although the lesions were more prominent on the left side, the right side was also involved showing a systematized morphologic distribution. Bilateral involvement of inflammatory linear verrucous epidermal nevus has been known to occur very rarely.

Histologic findings of inflammatory linear verrucous epidermal nevus have been reported by several authors. In general, a psoriasiform histologic pattern has been shown that is characteristic but not pathognomonic for inflammatory linear verrucous epidermal nevus. These consist of acanthosis, papillomatosis, spongiosis, and exocytosis leading to spotty areas of parakeratosis. This psoriasiform hyperplasia of the epidermis is commonly associated with alternating parakeratosis without a granular layer and orthokeratosis with a thickened granular layer. A mild perivascular lymphocytic infiltration is shown.

In our case, the histopathologic findings of the verrucous lesions confirmed the clinical diagnosis of inflammatory linear verrucous epidermal nevus.

Most investigators believe that inflammatory linear verrucous epidermal nevus is a specific entity. Nonetheless, the similarities between inflammatory linear verrucous epidermal nevus and psoriasis have prompted some others to dispute that these two diseases may overlap. At
the histologic level, parakeratosis with agranulosis is seen in both and psoriasis lesions can show linear arrangements clinically or psoriasis may overlap with the epidermal nevus. In the case presented, histopathologic findings of inflammatory linear verrucous epidermal nevus and psoriasis lesions were almost the same. However, the clinical picture of our patient showed two different lesions that had distinctive features. One of these had hyperpigmented, verrucous lesions showing linear distribution along Blaschko's lines, and the other showed erythematous, scaly, round to oval plaques scattered among the verrucous lesions. The widespread verrucous lesions had been present since one month of age, they were extremely pruritic, and showed little improvement with the treatment. On the other hand, the erythematous plaques had recent onset, were not pruritic, and resolved completely with the topical steroid therapy. Therefore, we believe that these lesions represent two distinct diseases: inflammatory linear verrucous epidermal nevus and psoriasis.

Ho et al. have suggested that although inflammatory linear verrucous epidermal nevus is a specific nevoid epidermal disorder, in histopathogenesis there may be some abnormalities in cell metabolism of the epidermal keratinocytes common to inflammatory linear verrucous epidermal nevus and psoriasis. Thus, inflammatory linear verrucous epidermal nevus may display histopathologic and immunohistochemical features similar to that in psoriasis. The association of inflammatory linear verrucous epidermal nevus and psoriasis in our case might support the explanation that they occur as the result of a common abnormality in the metabolism of epidermal keratinocytes.
REFERENCES


