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2. Inflammatory linear verrucous epidermal nevus

First described in 1971, inflammatory linear verrucous epidermal nevus (ILVEN) is a distinct, uncommon epidermal nevus variant that typically presents in early childhood, although adult onset has also been reported.¹⁻⁴ Clinically, it is characterized by persistent, intensely pruritic, unilateral, erythematous, verrucous, and scaly papules arranged in a Blaschko-linear distribution most commonly involving the lower extremities, pelvis, or buttocks.¹⁻⁴ Although familial cases have been reported, most ILVEN cases are sporadic.²⁻⁵ Histopathologically, ILVEN exhibits hyperkeratosis with parakeratotic foci, papillomatosis, psoriasiform epidermal hyperplasia with elongation and thickening of the rete ridges, occasional slight spongiosis with lymphocyte exocytosis, and a mild to moderate perivascular lymphohistiocytic infiltrate.²⁻⁴ A biopsy from our patient had similar characteristic features.

The etiopathogenesis of the condition is not clear. Most ILVEN cases typically occur as isolated findings, and although there have been infrequent reports of associated disorders such as autoimmune thyroiditis and arthritis, these most likely represent coincidental findings.^{2-4,6,7} While noninflammatory epidermal nevi are commonly associated with central nervous system, ocular, and skeletal abnormalities, ILVEN has been occasionally reported in association with ipsilateral skeletal and renal abnormalities. There has even been one report of ipsilateral undescended testes associated with ILVEN involving the left scrotal sac and penis.^{2,8,9} Our patient had no associated abnormality.

Differential diagnosis

Clinical differential diagnosis of ILVEN includes non-inflammatory epidermal nevi, linear psoriasis, and lichen

striatus.^{2,9,10} Noninflammatory epidermal nevi are typically asymptomatic, are usually associated with other abnormalities, and commonly do not have a predilection to involve the pelvis, buttocks, or lower extremities.^{2,9} Linear psoriasis is usually asymptomatic and, unlike ILVEN, typically responds completely to anti-psoriatic treatment.² Lichen striatus is usually asymptomatic, tends to resolve spontaneously, and has a different histopathologic picture.^{2,10}

Management

The natural history of ILVEN suggests that it has no tendency to remit or improve with time.^{2,3} Typically, it is resistant to therapy. Therapeutic options including topical and intralesional glucocorticoids, surgical excision, cryotherapy, and laser therapy have been tried with variable benefit.^{2,3}

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Competing interests

None declared

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