Linear Violaceous Papules in a Child

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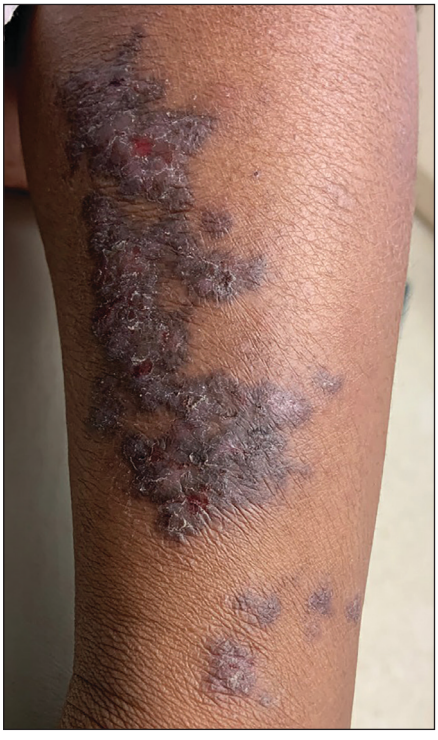

A 5-year-old Black girl presented to the dermatology clinic with a stable pruritic eruption on the right leg of 1 month's duration. Over-the-counter hydrocortisone cream was applied for 3 days with no response. Physical examination revealed grouped, flat-topped, violaceous papules coalescing into plaques with overlying lacy white striae along the right lower leg, wrapping around to the right dorsal foot in a blaschkoid distribution. The patient was otherwise healthy and up-to-date on immunizations and had an unremarkable birth history.

WHAT'S YOUR **DIAGNOSIS?**

a. incontinentia pigmenti
b. inflammatory linear verrucous epidermal nevus
c. lichen striatus
d. linear lichen planus
e. linear psoriasis

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The authors report no conflict of interest.

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THE **DIAGNOSIS:** Linear Lichen Planus

The patient was clinically diagnosed with linear lichen planus and was started on betamethasone dipropionate ointment 0.05% applied once daily with improvement in both the pruritus and appearance at 4-month follow-up. A biopsy was deferred based on the parents' wishes.

Lichen planus is an inflammatory disorder involving the skin and oral mucosa. Cutaneous lichen planus classically presents as flat-topped, violaceous, pruritic, polygonal papules with overlying fine white or grey lines known as Wickham striae.[1] Postinflammatory hyperpigmentation is common, especially in patients with darker skin tones. Expected histologic findings include orthokeratosis, apoptotic keratinocytes, and bandlike lymphocytic infiltration at the dermoepidermal junction.[1]

An estimated 5% of cases of cutaneous lichen planus occur in children.[2] A study of 316 children with lichen planus demonstrated that the classic morphology remained the most common presentation, while the linear variant was present in only 6.9% of pediatric cases.[3] Linear lichen planus appears to be more common among children than adults. A study of 36 pediatric cases showed a greater representation of lichen planus in Black children (67% affected vs 21% cohort).[2]

Cutaneous lichen planus often clears spontaneously in approximately 1 year.[4] Treatment in children primarily is focused on shortening the time to resolution and relieving pruritus, with topical corticosteroids as first-line therapy.[3,4] Oral corticosteroids have a faster clinical response; greater efficacy; and more effectively prevent residual hyperpigmentation, which is especially relevant in individuals with darker skin.[3] Nonetheless, oral corticosteroids are considered a second-line treatment due to their unfavorable side-effect profile. Additional treatment options include oral aromatic retinoids (acitretin) and phototherapy.[3]

Incontinentia pigmenti is characterized by a defect in the inhibitor of nuclear factor–κB kinase regulatory subunit gamma, *IKBKG*, gene on the X chromosome. Incontinentia pigmenti usually is lethal in males; in females, it leads to ectodermal dysplasia associated with skin findings in a blaschkoid distribution occurring in 4 stages.[5] The verrucous stage is preceded by the vesicular stage and expected to occur within the first few months of life, making it unlikely in our 5-year-old patient. Inflammatory linear verrucous epidermal nevus usually occurs in children younger than 5 years and is characterized by psoriasiform papules coalescing into a plaque with substantial scale instead of Wickham striae, as seen in our patient.[6] Lichen striatus consists of smaller, pink to flesh-colored papules that rarely are pruritic.[7] It is more common among atopic individuals and is associated with postinflammatory hypopigmentation.[8] Linear psoriasis presents similarly to inflammatory linear verrucous epidermal nevus, with greater erythema and scale compared to the fine lacy Wickham striae that were seen in our patient.[8]

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