Hypothesis

CHILD + ILVEN = PEN or PENCIL

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Inflammatory Linear Verrucous Epidermal Naevus (ILVEN)¹ is a pruritic, red, scaly skin lesion which appears at birth or in early childhood, is orientated along Blaschko's lines, is usually unilateral with a sharp midline demarcation, and is resistant to therapy. From a review of over 80 case reports² it was concluded that there are no associated defects. However, Golitz and Weston³ reported one patient, and cited four others, with distal limb abnormalities, which in four specifically involved the limb underlying the naevus.

A year later these four cases were listed as examples of CHILD syndrome (Congenital Hemidysplasia with Ichthyosiform erythroderma and Limb Defects).⁴ Characteristically this syndrome comprises a red scaly eruption covering half the body, with sharp midline demarcation and severe ipsilateral limb hypoplasia. In some patients, spared areas of skin on the affected side show orientation along Blaschko's lines.

The similarities between these two disorders are remarkable. In both conditions the red scaly eruption is predominantly unilateral with sharp midline demarcation, follows Blaschko's lines, and usually spares the face. Histologically they are indistinguishable: there is both hyper- and parakeratosis with a granular layer correspondingly either thickened or absent, the epidermis shows spongiosis and psoriasiform elongation of the rete ridges, and there is a lymphohistiocytic infiltrate in the upper dermis.

Skeletal abnormality is pathognomonic of CHILD syndrome and exceedingly rare in ILVEN, but in both takes the form of distal bony deformity of the limb affected by the naevus. Both are commoner in females; ILVEN is four times more common in females, while CHILD syndrome has been reported in 15 females and one male (excluding the intermediate cases mentioned above). Both are usually sporadic disorders which have occasionally been found in related females. CHILD syndrome has been reported in two sisters and in the maternal aunt and great aunt of a proband, and ILVEN in a mother and daughter.

We conclude that CHILD syndrome and ILVEN are polar groups on a clinical spectrum, both reflecting an ectodermal defect variable in site and extent. If the defect involves the ectodermal proliferative zone overlying a limb bud, development of that limb is disrupted. Thus in CHILD syndrome the defect involves half the body and suppresses development of both ipsilateral limbs. In ILVEN the defect is very small relative to body surface area and the risk of it involving the limb bud proliferative zone is correspondingly lower, but if it does then the underlying limb will again be defective. The patchy distribution probably reflects mosaicism for a genetic disorder which is lethal if universal. The marked female excess suggests an X linked gene with mosaicism resulting from Lyonisation in females, or a somatic mutation in either sex. Familial cases could be the result of an unstable premutation or gonadal mosaicism.

In a recent study of a patient with CHILD syndrome, cultured fibroblasts from affected skin produced increased amounts of the inflammatory mediators prostaglandins E2 and I2 compared with cells from unaffected skin.⁵ It would be worth extending such studies to ILVEN, a pruritic, inflammatory disorder in which a defect of prostaglandin metabolism might be suspected. While both conditions have the advantage that unaffected skin provides a useful control for lesional skin, patients with ILVEN are much more readily available.

Combining these two disorders calls for a new

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name. The existing acronyms have been useful but not entirely appropriate. Inflammatory Linear Verrucous Epidermal Naevus is cumbersome and easily confused with Linear Verrucous Epidermal Naevus, which has neurological and ophthalmic associations. 'CHILD' applies to only the extreme (hemidysplastic) form of a variable condition. The simple descriptive term 'Psoriasiform Epidermal Naevus±Congenital Ipsilateral Limb defects' (PEN or PENCIL) is appropriate and memorable, at least to those who write on the subject.

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- 5 Goldyne ME, Williams ML. CHILD syndrome: phenotypic dichotomy in eicosanoid metabolism and proliferative rates among cultured dermal fibroblasts. *J Clin Invest* 1989;84: 357-60.