CHILD naevus is not ILVEN

In a recent article, Moss and Burn1 advanced the hypothesis that CHILD syndrome and ILVEN are “polgar groups on a clinical spectrum, both reflecting an ectodermal defect variable in site and extent”. They proposed the new descriptive term ‘psoriasis-form epidermal naevus (PEN)’, sometimes associated with ‘congenital ipsilateral limb defects (PENCIL)’.

For the following reasons, however, the equation CHILD naevus + ILVEN = PEN or PENCIL” is mistaken. The epidermal naevus associated with the CHILD syndrome is definitely not ILVEN but a distinct cutaneous entity that should be called “CHILD naevus”.2

(1) CHILD naevus can be distinguished from ILVEN by the presence of yellow, wax-like scales, resulting in a distinctive ‘ichthyosiform’ appearance. (2) This naevus shows a tendency to a non-linear arrangement, often involving one half of the trunk in a diffuse manner. In contrast, ILVEN is always linear. (3) This naevus displays a pronounced affinity for the body folds, or psychotropism;2 by contrast, ILVEN is not psychotropism. (4) CHILD naevus causes no, or only minimal, pruritus whereas in patients affected with ILVEN itching often constitutes a serious problem. (5) CHILD naevus may show the histopathological features of ‘verruciform xanthoma’, a phenomenon characterised by abundant foamy histiocytes occupying the dermal papillae. Such xanthomatous transformation has so far not been observed in ILVEN. (6) CHILD naevus is a well defined genetic entity inherited as an X linked dominant trait, constituting the cutaneous hallmark of the CHILD syndrome. It occurs almost exclusively in females because the underlying X linked mutation is lethal in male embryos. By contrast, the genetic basis of ILVEN is unclear and possibly heterogeneous.

Furthermore, I disagree with Drs Moss and Burn that CHILD should apply only to the extreme ‘hemi-dysplastic’ form of a variable condition. It is true that the term was originally suggested as an acronym for ‘congenital hemiedysplasia with ichthyosiform erythroderma and limb defects’,3 but it has now become evident that the associated skin disease should be classified more appropriately as a naevus, and therefore the following modified interpretation of the acronym has been proposed: ‘congenital hemiedysplasia with ichthyosiform naevus and limb defects’.

In conclusion, there are different epidermal naevi giving the impression of a psoriasisform skin lesion, and it seems unjustifiable to lump them together under the term ‘psoriasisform epidermal naevus’.

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Letters to the Editor