Linear Psoriasis of the Isolated Type

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Linear Psoriasis of the Isolated Type

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Case Presentation:

Patient: B.B. is an 18 year-old Caucasian female

History of Present Illness: This patient presented to our dermatology office complaining of a new onset pruritic linear rash on right arm, right chest and right leg present for 6 months. She was treated for an unrelated eczematous dermatitis in the past that resolved quickly and did not recur. Patient denied any triggering factors such as infections or medications and reported no associated symptoms, including arthralgia.

Medical History/Surgical History: Eczematous dermatitis

Family History: Atopic dermatitis, mother and brother; no history of psoriasis

Previous Treatments: Betamethasone dipropionate 0.05% ointment and tazarotene 0.1% cream

Current Treatments: Calcipotriene 0.005% ointment daily and Taclonex ointment for flares

Physical Examination: Linear, scaly, erythematous plaques along Blaschko lines on right arm, right chest (A) and right posterior leg (B) with a close-up of the right arm (C) on an 18 year-old female.

Figure 1: Distribution of linear, scaly, erythematous plaques along Blaschko lines on right arm, right chest (A) and right posterior leg (B) with a close-up of the right arm (C) on an 18 year-old female.

Figure 2: H&E (A-4x and B-10x) Punch biopsy from right upper arm, shows epidermal psoriasiform hyperplasia, superficial paller with diminished granular layer, incipient subcorneal spongiform pustule formation and staggered collections of neutrophils in a thickened parakeratotic horn.

Discussion:

Linear psoriasis (LPs) is an exceedingly rare variant of psoriasis. LPs may be associated with nonsegmental plaques of psoriasis vulgaris (superimposed linear psoriasis type) or it can occur as an isolated lesion (linear psoriasis of the isolated type). In the latter, the linear lesions tend to be more pronounced than those seen in the former type.

Debate exists whether LPs is a distinct clinical entity or merely an inflammatory linear verrucous epidermal nevus (ILVEN). This has been disputed in the published work over recent decades. Differentiating between the two conditions may be difficult or even impossible on clinical and histological criteria alone. Only a detailed history and the time course of the skin lesions may help establish an accurate diagnosis.

ILVEN tends to develop during the first months of life, progresses slowly, can be very pruritic and is relatively unresponsive to treatment. LPs, however, tends to develop later in life, has a positive family history, may progress rapidly, can be occasionally pruritic and responds well to antipsoriatic treatment.

Histopathologically, ILVEN differs from LPs by areas of hypergranulosis with orthohyperkeratosis alternating with areas of agranulosis with parakeratosis. In uncertain cases, immunohistochemical techniques may be helpful. ILVEN, in contrast to LPs, has a parakeratotic epidermis lacking involucrin expression, tends to have lower expression of the proliferation marker Ki-67 but shows an increase in keratin-10.

The pathogenesis of LPs is not clear, but could be explained by the well-established concept of genetic mosaicism. Individuals affected with LPs are similarly heterozygous for many gene loci involved in psoriasis. During early embryogenesis, crossing over may occur in a somatic cell. This results in loss of heterozygosity involving one of the predisposing genes, giving rise to daughter cells that will distribute along Blaschko’s lines. For the ultimate manifestation of LPs, the presence of other predisposing genes as well as environmental factors would be necessary. This explains why linear psoriasis is mostly absent at birth but develops later in life.

References: