

A Case of Lichen Striatus Following Blaschko Lines

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We present a case of lichen striatus (LS) in a 36-year-old woman with skin lesions on the right side of the chest distributed along Blaschko lines that showed a swirling s-shaped pattern. The differential diagnosis of LS includes a variety of nevoid and acquired conditions following Blaschko lines, particularly acquired relapsing self-healing Blaschko dermatitis. The extent to which these 2 conditions overlap remains contentious.

Lichen striatus (LS) is an uncommon dermatosis of unknown etiology that generally affects children. Clinically, it presents as a continuous or interrupted linear band of erythematous, slightly elevated, tiny papules on one of the extremities. However, some cases of LS show the bizarre pattern of a *v* on the spine, an *s* on the abdomen, or an inverted *u* from the breast to the upper arm, which might suggest a distribution along Blaschko lines.¹ Because a variety of conditions may occur along Blaschko lines,^{2,3} the differential diagnosis of LS can be difficult and should include other nevoid and acquired dermatoses following Blaschko lines. We describe a 36-year-old woman with unilateral, whorled, slightly pruritic lichenoid papules on the right side of the chest and discuss other conditions that should be considered in the differential diagnosis of LS.

Case Report

A 36-year-old woman presented with a 2-month history of unilateral, slightly pruritic eruptions following the appearance of Blaschko lines. Her medical and family history was unremarkable. On examination, erythematous to brown lichenoid papules and macules were present on the right side of her right breast,



FIGURE 1. Whorled, erythematous to brown lichenoid papules and macules on the right side of the right breast following Blaschko lines.

flank, and axilla (Figure 1). There were no other similar or mucous membrane lesions. Laboratory findings, including a complete blood count, urinalysis, liver function test, and chest x-ray, were normal or negative. A biopsy specimen showed hyperkeratosis, spongiosis, and some dyskeratotic cells in the epidermis. With a variable degree of exocytosis, focal, band-like, lymphocytic infiltrates occupied the papillary dermis and obscured the dermoepidermal junction (Figure 2). In the mid dermis, there were periadnexal lymphocytic infiltrates along the sweat glands. The interface dermatitis with both lichenoid and spongiotic features was consistent with LS. After 2 months of treatment with topical corticosteroids, the lesion flattened and improved, although postinflammatory hyperpigmentation remained.

Comment

Blaschko lines represent an observed pattern of cutaneous expression followed by multiple skin disorders.^{2,3} Nevoid conditions to be considered when they occur at birth or during infancy are incontinentia

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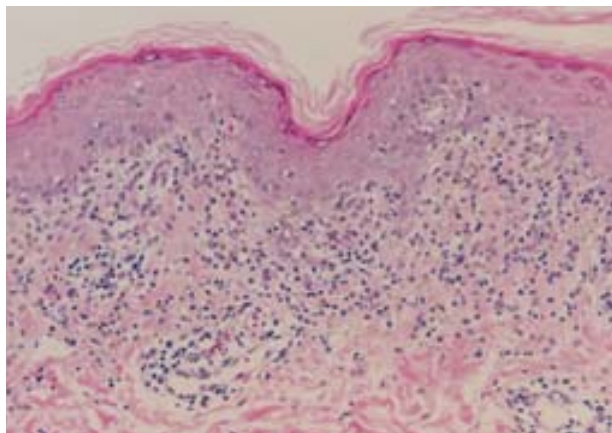


FIGURE 2. Bandlike inflammatory lymphocytic infiltrates obscure the dermoepidermal junction and extend into the lower portion of the epidermis (H&E, original magnification $\times 100$).

pigmenti, linear and whorled nevoid hypermelanosis, hypomelanosis of Ito, and epidermal nevi.³ Acquired conditions following Blaschko lines include psoriasis, porokeratosis, lupus erythematosus, pemphigus, scleroderma, fixed drug eruption, lichen planus, lichen nitidus, and LS.³ Blaschko lines are most commonly confused with dermatomes, the segments of skin defined by sensory innervation. In our patient, the differences between dermatomes and Blaschko lines were most apparent on the trunk where arcs on the upper chest, the s shape on the abdomen, and the v shape as the lesions approached the posterior middle line were visible.² The pattern had a streaky, whorled configuration following Blaschko lines and resembled that found in incontinentia pigmenti. However, erythematous to brown linear lichenoid papules, which are typical of LS, were also present. Incontinentia pigmenti differs from LS in the earlier onset in infancy, the presence of the preceding vesiculobullous stage, the association of systemic abnormalities, and the characteristic histologic findings of pigmentary incontinence.

Linear and whorled nevoid hypermelanosis is another pigmentary anomaly that should be considered in the differential diagnosis of an infant with hyperpigmented streaks following Blaschko lines. A review of the literature indicates that several cases of similarly patterned hypermelanosis have been reported as reticulate hyperpigmentation distributed in a zosteriform fashion, zebra-like hyperpigmentation, and zosteriform lentiginous nevus.⁴ The skin lesions of these conditions are not lichenoid papules or plaques, as in our case, but streaky and whorled hyperpigmented macules. Clinically, these macules closely resemble that of hypomelanosis of Ito but are inversely pigmented.

Histologically, there is only an increased pigmentation of the basal layer without incontinence of pigment.

An epidermal nevus is a linear, persistent, pruritic dermatosis consisting of warty or scaling papules that usually present at birth or during infancy. At this stage, the condition may be barely palpable, but with time it becomes papillomatous and hyperkeratotic. Linear epidermal nevi can be distinguished histologically from LS on the basis of hyperkeratosis, papillomatosis, acanthosis with elongated rete ridge, and, occasionally, epidermolytic hyperkeratosis.

In our patient, histologic examination revealed a bandlike infiltrate of lymphocytes and histiocytes in the papillary dermis, which is consistent with LS but is also typically exemplified in lichen planus.⁵ Clinically, lichen planus usually consists of a single discrete or a few aggregated lesions, but a linear pattern has been reported.^{6,7} Some cases of linear lichen planus show a swirling or s-shaped pattern on the trunk, which might suggest a distribution along Blaschko lines.^{3,8} Linear lichen planus also may occur as an isomorphic response to injury (Köbner's phenomenon), but it differs from idiopathic linear lichen planus in that the streaks and bands may be narrower or shorter.^{8,9} If the extent and linearity of the lichen planus are unusually prominent and greatly exceed that which is normally associated with it, the differential diagnosis of LS can be difficult. Lichen planus and LS share many common histologic characteristics. However, there are several clues that favor a diagnosis of LS: intraepidermal spongiosis, dyskeratotic cells in epidermis, focal bandlike lymphocytic infiltrates with variable exocytosis, and the alignment of the infiltrate with hair follicles and eccrine ducts.^{5,10} Furthermore, the clinical feature of LS is not consistent with a pruritic, keratotic papular disease, but rather is a mixed erythematous and eczematoid eruption of transient duration with an occasional papular component.^{1,11}

In 1990, Grosshans and Marot¹² reported relapsing acquired unilateral skin lesions in adults that follow Blaschko lines. In 1994, Megahed et al¹³ proposed the term *acquired relapsing self-healing Blaschko dermatitis* to differentiate this condition from LS. They emphasized that, unlike LS, there is an adult onset, relapsing history, vesicular lesions, and a greater tendency toward a spongiosis rather than a lichenoid pattern. However, according to Gianotti et al,¹⁰ the histopathology of LS often shows a polymorphic epidermal reaction process with variable lichenoid and spongiosis changes. This process has no specific histopathological criteria because microscopic findings change with the age of the lesion. Furthermore,

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LS often presents quite a different clinical picture in adults than in children. In an adult, the condition is more vesicular, extensive, and pruritic.¹¹

Conclusion

It is not clear whether the distinction between these 2 diseases is justified or whether they merely represent either end of a spectrum, but adult onset and relapsing history may not be sufficient to propose the new term *acquired relapsing self-healing Blaschko dermatitis*. Instead of LS, the acronym BLAISE (Blaschko Linear Acquired Inflammatory Skin Eruption) was suggested based on the correlation between the predominant erythema and eczematous features and the comparative insignificance of the lichenoid component.¹

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