

Blaschkoid lichen planus: Throwing a “curve” in the nomenclature of linear lichen planus



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INTRODUCTION

Lichen planus (LP) is an inflammatory dermatosis that presents with pruritic papules, with LP lesions along the lines of Blaschko representing a rare variant.^{1,2} There are several terms for this entity, including *linear LP*, *Blaschkolinear LP*, and *Blaschkoid LP*. To minimize confusion and be more consistent with its distribution, we suggest this variant be termed what it truly is: *Blaschkoid lichen planus*. Although most cases of Blaschkoid LP have a unilateral distribution, we report a case of bilateral LP in a Blaschkoid distribution that subsequently resolved after treatment with oral glucocorticoids.

CASE REPORT

A 63-year-old African-American man presented for evaluation of a progressively worsening pruritic eruption of 3 months duration. The eruption initially involved the left side of his abdomen, for which he was treated with valacyclovir for presumed varicella zoster, without improvement. When evaluated in the dermatology clinic, the patient noted progression of the eruption to involve the left side of his trunk, back, and upper extremities. The patient noted pruritus but denied associated pain. There was no recent trauma to the area; however, he had a partial colectomy for stage I colon cancer 2 months before the development of the eruption. No personal or family history of atopy or psoriasis was elicited. The patient denied a history of varicella, recent immunizations, or travel. His medical history was notable for type 2 diabetes mellitus and hypertension, for which he was taking both metformin and lisinopril-hydrochlorothiazide, respectively, for the last

Abbreviation used:

LP: lichen planus

10 years. He denied any new medications or changes to his current medications.

Examination found multiple scaly, violaceous-to-erythematous, polygonal papules coalescing into thin linear plaques in an S-shape pattern on the left side of his abdomen (Fig 1), a V-shape pattern near the posterior midline, and in a linear distribution on his upper back with extension along the posterior left and right upper extremities (Fig 2). Involvement of his left first finger was appreciated with onychomadesis noted (Fig 3). There was no involvement of the genitals or oral mucosa.

A complete blood count and complete metabolic panel were unremarkable. Test results for hepatitis B virus, hepatitis C virus, human immunodeficiency virus, and rapid plasma reagin were negative. Shave biopsy from the left flank found compact hyperorthokeratosis, wedge-shaped hypergranulosis, irregular saw-tooth epidermal hyperplasia, vacuolar alteration of the basal layer, necrotic keratinocytes and a band-like predominantly lymphocytic infiltrate accompanied by scattered plasma cells, few eosinophils, and dermal melanophages (Fig 4). The clinicopathologic diagnosis was Blaschkoid lichen planus.

DISCUSSION

Although LP is a common mucocutaneous condition, LP in a Blaschkoid distribution is a rare

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Fig 1. Clinical photograph. Scaly, violaceous-to-erythematous, polygonal papules coalescing into thin linear plaques in an S-shape pattern on the left abdomen.

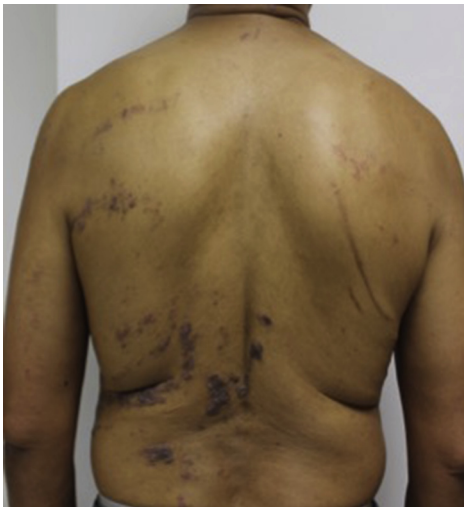


Fig 2. Clinical photograph. Scaly, violaceous-to-erythematous, polygonal papules and thin plaques in a V-shape pattern near the posterior midline, and in a linear distribution on the right and left upper back, with extension along the posterior upper extremities.

variant, estimated to affect 0.24% to 0.62% of all LP patients.¹⁻³ The term *linear LP* was first used in 1976 to describe lesions along the lines of Blaschko, demonstrating a characteristic S shape on the abdomen, V shape near the posterior midline, a linear pattern on the lower trunk and limbs, and whorls on the scalp and abdomen.² Since then, the term has been used to describe LP following various linear patterns. Given the existing inconsistencies with the term *linear LP*, we seek to clarify the current



Fig 3. Clinical photograph. Violaceous-to-erythematous, polygonal papules and thin plaques in a linear distribution with involvement of the proximal nail fold of the left first digit with associated onychomadesis.

nomenclature, suggesting that linear LP be referred to as *Blaschkoid LP* for this entity to be recognized for what it really is, lichen planus along the lines of Blaschko.

Blaschko lines represent migration pathways of epidermal cells during embryogenesis and highlight a pattern followed by several X-linked, congenital, and acquired skin conditions.² Blaschkoid LP is thought to be caused by a postzygotic mosaic alteration, in which a loss of heterozygosity occurs, leading to the formation of a keratinocyte clone that is more susceptible to the development of LP upon exposure to a trigger later in life.²⁻⁴ Although there are no definitive associations between specific

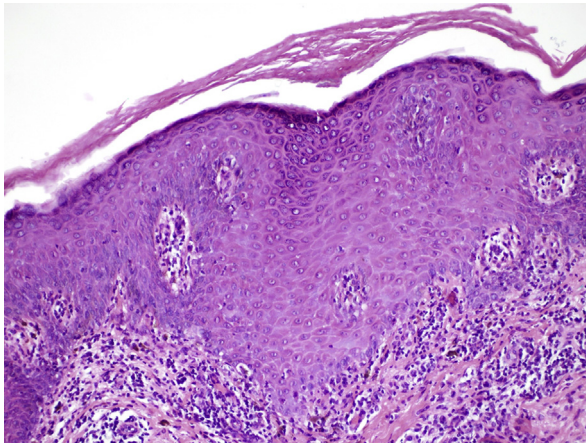


Fig 4. Shave biopsy shows compact hyperorthokeratosis, wedge-shaped hypergranulosis, irregular saw-tooth epidermal hyperplasia, vacuolar alteration of the basal layer, necrotic keratinocytes, and a band-like predominantly lymphocytic infiltrate accompanied by scattered plasma cells, few eosinophils, and dermal melanophages. (Hematoxylin-eosin stain; original magnification: $\times 20$.)

antigens and Blaschkoid LP, several cases are reported of Blaschkoid LP developing after vaccinations⁵ and in the setting of known neoplasms⁶ and recurring with successive pregnancies.⁷ Thus, it is speculated that the Blaschkoid distribution of LP may be predetermined during embryogenesis, allowing for development of LP lesions after exposure to antigenic trigger.³ Although no exact trigger was identified for our patient, one must consider his history of stage I colon cancer and as his surgical intervention.

Although a linear distribution of LP may occur secondary to the Koebner phenomenon or an isotopic (Wolf) response at the site of healed herpes zoster, the Blaschkoid variant of LP strictly describes lesions that follow the lines of Blaschko.² A recent immunohistochemical study found that varicella zoster virus antigens could be found in the eccrine epithelia of patients with LP distributed along dermatomes but not in the eccrine epithelia of patients with LP distributed along the lines of Blaschko.⁸ Distinguishing it from dermatomal LP or true linear LP will also help aid our future understanding of its pathogenesis.

In a review of 18 cases of Blaschkoid LP, the average age of onset was 33 years, with the lesions

developing over a period of 1 week to several months.² More than half of patients had associated pruritus, as in our patient's case. Additionally, most cases consisted of polygonal violaceous papules and plaques, although reports of annular, vesicular, and hypertrophic lesions have also been described.² Most cases of Blaschkoid LP tend to be unilateral in nature. Interestingly, in addition to our own case, only 2 other cases of bilateral multilinear LP have been reported.^{9,10}

The course of Blaschkoid LP is usually benign and self-limited, with the lesions often healing with pigmentary alteration.^{2,3} Topical glucocorticoids are considered first-line therapy, with additional therapies for refractory cases including oral glucocorticoids and phototherapy among many other alternatives.¹⁻³ Our patient was initially prescribed topical triamcinolone, but his condition progressed. He ultimately cleared after a course of oral prednisone at 60 mg per day, with a 20-mg taper every 5 days. His prognosis was excellent, without any recurrence and only residual hyperpigmentation on clinical examination.

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