

BRIEF ARTICLES

Linear Psoriasis: A Case Report of a 71-Year-Old Female

Chun-Hui Yi MD PhD^a, Chen Chen MD^b, Allen N Sapadin MD JD^c, Robert G Phelps MD^b

^aDepartment of Pathology, Mount Sinai Health System, St. Luke's-Roosevelt Hospital and Beth Israel Medical Center, New York, NY

^bDepartment of Pathology, Mount Sinai School of Medicine, New York, NY

^cAllen Sapadin Dermatology and Dermatologic Surgery, Hackensack, NJ

ABSTRACT

A 71-year-old woman presented with a one-month history of pruritus, burning, flaky, and erythematous plaques. The lesions had a very narrow lineal distribution, extending from the lateral left sole to the left popliteal region. Microscopically, the skin biopsy sample revealed orthokeratosis, parakeratosis and traumatized Koebnerization. Elongation of rete ridges was not prominent. The patient had a good clinical response to oral prednisone. Linear psoriasis is a rare variant of psoriasis of unknown etiology. Approximately 23 reports have been published in the English literature. The main differential diagnosis is Inflammatory linear verrucous epidermal nevus (ILVEN). Thorough clinicopathologic findings and response to treatment are all required in the diagnosis and management of this rare form of psoriasis.

CASE REPORT

A 71-year-old woman presented to the clinic with a one-month history of pruritic, burning, flaky pink to dark red erythematous plaques. The lesions had a very narrow, linear, Blaschkoid distribution which extended from the left lateral sole to the popliteal region (Figure 1). The right leg also had a similar erythematous plaque located at the pretibial region. The patient had no other lesions or previous history of psoriasis. Punch biopsies were performed which revealed orthokeratosis and mounds of parakeratosis, and traumatized Koebnerization. Elongation of rete ridges was not prominent, however, mild psoriasiform hyperplasia and suprapapillary thinning of the epidermis with

mildly increased vessels in the dermal papillae and a lymphocytic infiltrate in the upper dermis were observed. (Figure 2A-D). The findings were consistent with features of an early psoriasis lesion.

PAS and the direct immunofluorescence (DIF) stains were negative, but Ki-67 showed increased expression (Figure 2E). Together with the clinical presentation, a diagnosis of linear psoriasis was favored.

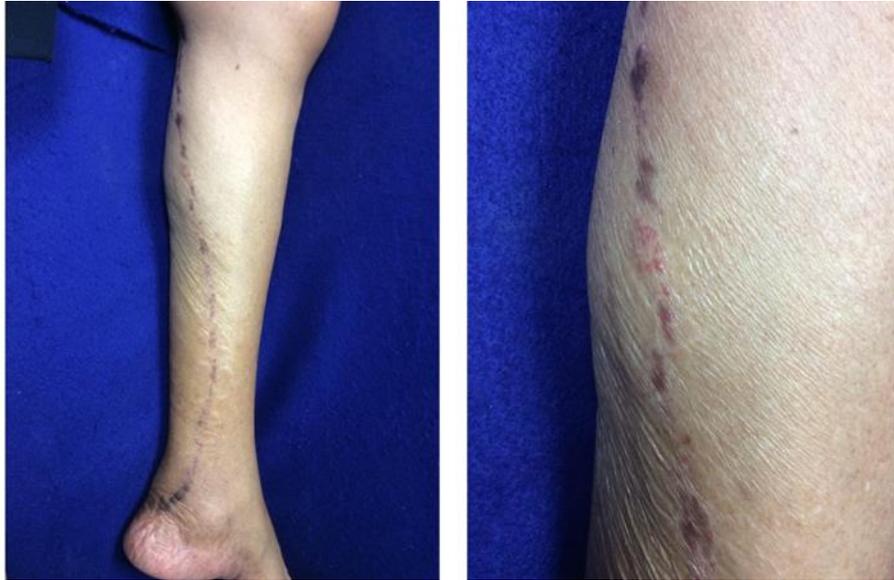


Figure 1. The patient presented with a one-month history of pruritic, burning, flaky pink to dark red erythematous plaques. The lesions had a very narrow, linear, Blaschkoid distribution extending from the left lateral sole to the popliteal region.

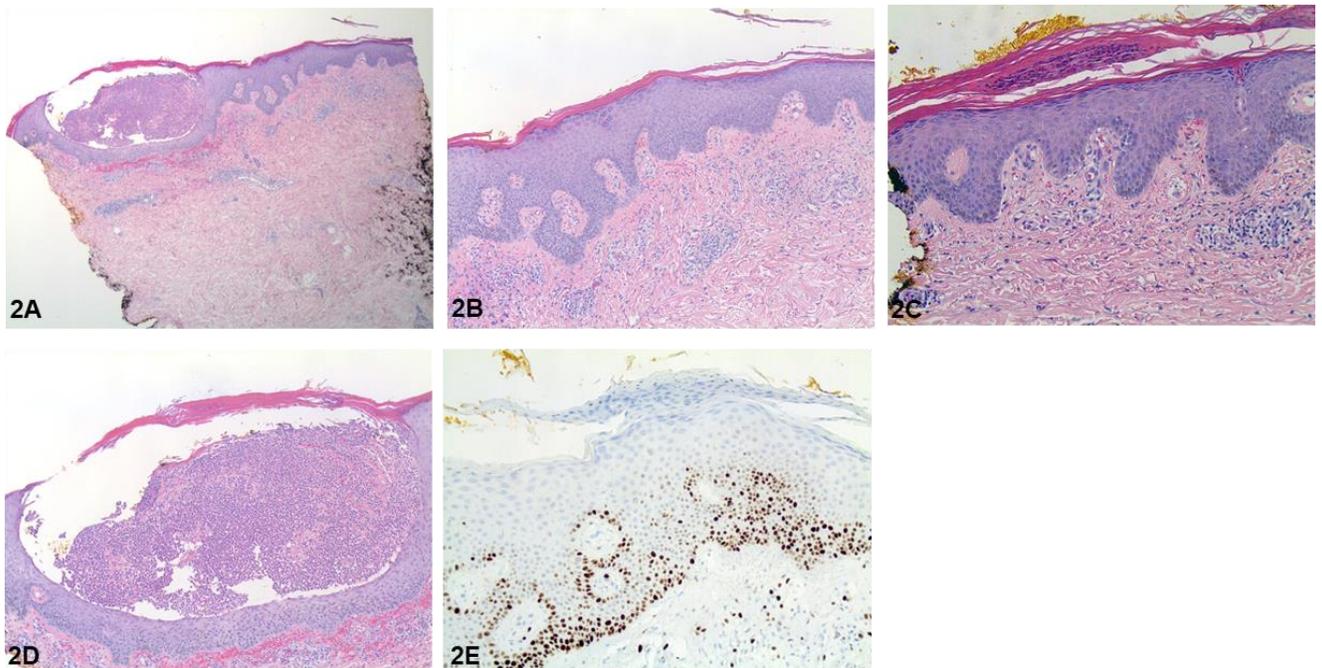


Figure 2. Histologic features of the pruritic erythematous plaque. Biopsy of the plaque showed mounds of parakeratosis in the stratum corneum, retention of the granular layer, mildly increased vessels in the dermal papillae, and lymphocytic infiltrates in the upper dermis (A-C). Focal Munro pustules were also present (D). Ki-67 was found to be increased (E).

DISCUSSION

Linear psoriasis is a rare variant of psoriasis. Approximately 23 reports have been published in the English literature. It is characterized by a linear distribution of the psoriatic lesions along Blaschko's lines.¹ It typically occurs in the lower extremities of older individuals, although rare occurrences in children also have been reported. The etiology of linear psoriasis is not known. Happle suggested that it may be the result of somatic recombination of genes that predisposes the patients to segmental mosaicism.² There may also be an association between human leukocyte antigen class I alleles and development of the disease.³ Linear psoriasis typically responds well to local treatment.

The main differential diagnosis of this entity is inflammatory linear verrucous epidermal nevus (ILVEN), psoriasis superimposed upon an epidermal nevus in a linear growth pattern or other linear dermatosis.⁴ ILVEN may have a similar clinical presentation to linear psoriasis as highly pruritic psoriasiform skin lesions that tend to occur in the left lower extremity. However, ILVEN usually occurs in early childhood with a female predominance whereas linear psoriasis frequently occurs in the elderly. The rash of linear psoriasis usually has mild pruritus or no symptoms.^{5,6} It is interesting to note that several reported cases of linear psoriasis in the literature also revealed a tendency to involve the left side of the body.^{3,7} Previously reported cases of linear psoriasis showed wide, band-like linear lesions, however, the rash on this patient showed a very narrow linear distribution similar to ILVEN or epidermal nevus.

CHILD (Congenital hemidysplasia with ichthyosiform erythroderma and limb defects) syndrome also presents with unilateral linear skin rashes, characterized by erythroderma and ichthyosis with an equal left- or right-sided predominance. It is an X-linked dominant hereditary disease that only occurs in girls, since the mutation is lethal to male embryos, and it is treated with topical cholesterol and simvastatin.^{8,9} Patients afflicted with this disorder also have hemidysplasia and developmental defects in other organ systems. Unlike CHILD syndrome, linear psoriasis and ILVEN are not hereditary and present later in life.

Distinguishing between ILVEN and linear psoriasis is difficult in some cases since the conditions have overlapping clinical and histological features. Recently, a few immunohistochemistry markers have been shown to be helpful in this diagnosis, such as Ki-67, keratin 10, and involucrin. Ki-67 index is usually high in linear psoriasis but remains low in ILVEN.¹⁰ Keratin 10 is rarely expressed in psoriasis, but the level is normal in ILVEN.¹ Involucrin is a transglutaminase substrate protein present in keratinocytes of epidermis. It is cross-linked to other membrane proteins and contributes to the formation of the cornified cell envelope.⁵ While it is present in the upper portion of the spinous and granular layer of the normal epidermis, it is expressed in the subbasal layer in psoriasis. In ILVEN, transglutaminase expression is increased in areas of the orthokeratosis but decreased in areas of parakeratosis.⁵ Furthermore, ILVEN is refractory to antipsoriatic treatment, whereas linear psoriasis responds well to topical

antipsoriatic agents or narrow-band UVB phototherapy.¹¹

In summary, we report a rare case of unilateral linear psoriasis involving the lower leg of a 71-year-old female, and discuss the clinical, histological features of the lesion, and the most common differential diagnosis. Unilateral linear psoriasis is rarely reported and often misdiagnosed as ILVEN.

Conflict of Interest Disclosures: none.

Funding: none.

Corresponding Author:

Chun-Hui Yi, MD, PhD
 Department of Pathology
 Mount Sinai Health System
 St. Luke's-Roosevelt Hospital and Beth Israel
 Medical Centers
 1000 Tenth Avenue
 New York, NY 10019
 212-523-8631 (Office)
 212-523-7232 (Fax)
 Chunhui.yi@mountsinai.org

References:

1. Figueiras Dde A, Cauas RC, Takano DM, Ramos TB, Marinho AK, Bezerra MS. Linear psoriasis: case report on three year old child. *An Bras Dermatol*. 2015;90(3 Suppl 1):194-196.
2. Happle R. Somatic recombination may explain linear psoriasis. *J Med Genet*. 1991;28(5):337.
3. Li W, Man XY. Linear psoriasis. *CMAJ*. 2012;184(7):789.
4. Happle R. Superimposed linear psoriasis: a historical case revisited. *J Dtsch Dermatol Ges*. 2011;9(12):1027-1028.
5. Ferreira FR, Di Chiacchio NG, Alvarenga ML, Mandelbaum SH. Involucrin in the differential diagnosis between linear psoriasis and inflammatory linear verrucous epidermal nevus: a report of one case. *An Bras Dermatol*. 2013;88(4):604-607.
6. Akelma AZ, Cizmeci MN, Kanburoglu MK, Mete E. A diagnostic dilemma: inflammatory linear verrucous epidermal nevus versus linear psoriasis. *J Pediatr*. 2013;162(4):879-879.
7. Singh N, Noorunnisa N. Linear psoriasis: A rare presentation. *Indian Dermatol Online J*. 2012;3(1):71-73.
8. Mi XB, Luo MX, Guo LL, Zhang TD, Qiu XW. CHILD Syndrome: Case Report of a Chinese Patient and Literature Review of the NAD[P]H Steroid Dehydrogenase-Like Protein Gene Mutation. *Pediatr Dermatol*. 2015;32(6):e277-282.
9. Noguera-Morel L, Hernandez-Ostiz S, Casas-Fernandez L, et al. CHILD syndrome with minimal limb abnormalities. *J Eur Acad Dermatol Venereol*. 2015.
10. Yin B Fau - Ran Y-p, Ran Yp Fau - Wang P, Wang P Fau - Lama J, Lama J. Is it inflammatory linear verrucous epidermal nevus or linear psoriasis? *Chinese medical journal*. 2013;126(0366-6999 (Print)):1794-1795.

11. Brinca A, Santiago F, Serra D, Andrade P, Vieira R, Figueiredo A. Linear psoriasis - a case report. *Case Rep Dermatol.* 2011;3(1):8-12.