

**Case 15**

A 51 year-old Thai female from Prae

**Chief complaint:** Multiple pruritic violaceous rash on left leg for 1 year



(Fig. 15.1)

**Present illness:** A linear violaceous rash gradually extending from left thigh to the lower leg and foot with intense itching for a year.

She denied history of contact with any specific materials.

**Past history:** She had no underlying medical conditions.

**Physical examination:**

HEENT: No pale conjunctivae, anicteric sclerae

Lymph node: Not palpable

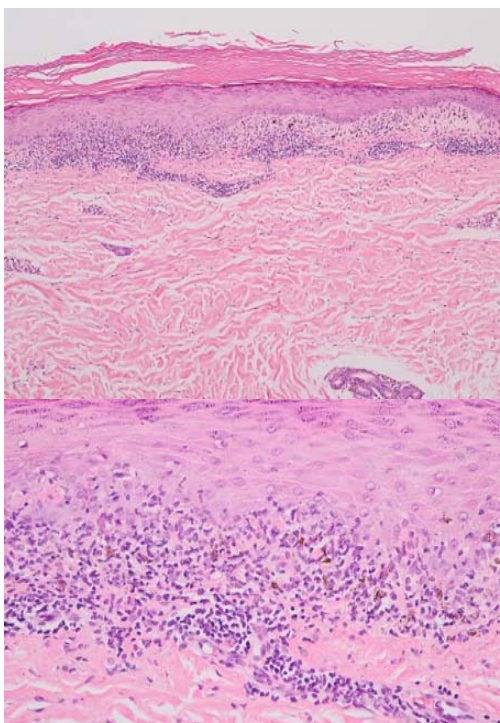
Heart&lung: Normal

Abdomen: Soft, not tender, no hepatosplenomegaly

**Dermatologic examination:** (Fig. 15.1)

- Linear purplish macules and papules on left buttock, leg and foot following Blaschko line
- No lesions seen on scalp, oral mucosa, or nails

**Histopathology:** (S17-04664, Left leg) (Fig. 15.2)



(Fig. 15.2)

- Irregular epidermal acanthosis, wedge-shaped hypergranulosis, and compact orthokeratosis
- Lichenoid cell infiltrate predominantly of lymphocytes with numerous melanophages in the papillary dermis with obscuring the dermoepidermal junction

**Diagnosis:** Linear lichen planus

**Investigation:** Anti-HCV: Negative

**Treatment:**

- 0.05% clobetasol propionate ointment apply on the lesion twice daily
- Oral prednisolone 20 mg/day for 7 days
- Oral cetirizine 10 mg/day

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**Discussion:**

Lichen planus (LP) is a common inflammatory disease of the skin and mucous membrane. The disease usually occurs in adults with the prevalence of 0.2 – 1%.<sup>1</sup>

Classic LP typically presents with the four P's: purple, pruritus, polygonal, and papules/plaques. A thin, transparent, branny scale may be seen on top of the lesion and also "Wickham's striae", a network of fine white lines. Many morphological variants have been described including annular, atrophic, actinic, bullous, eruptive, hypertrophic, inverse, linear, ulcerative, LP pigmentosus, lichen planus-lupus erythematosus overlap syndrome, lichen planus pemphigoides, vulvovaginal-gingival, drug-induced and lichen planopilaris. Clinical presentation of rare variant lesions may be atypical as those of classic LP, and therefore difficult to diagnose based solely on clinical examination.<sup>2</sup>

In classic LP, the lesions are typically symmetric distribution. The predilection sites include the upper and lower extremities, particularly the extensor surfaces of lower legs and the volar aspect of the wrists and forearms as well as the trunk and the lumbar region. The face does not usually affect.<sup>3</sup>

Linear lichen planus is a rare variant of LP, accounts for less than 0.2% of all LP cases. Unlike classic LP, this variant is often seen

in children and adolescent.<sup>4</sup> However, adult onset is possible.<sup>5</sup> Individual lesions may be typical flat-topped papules but vesicular, hyperkeratotic and annular morphologies may be observed. Clinical features of linear lichen planus are characterized by streak-like manifestation. The linear distribution can be limited to the dermatomes or follow the lines of Blaschko.<sup>6,7</sup> Cases associated with Blaschko-like distribution patterns have been proposed to be caused by a loss of heterozygosity in skin with a predisposition to the disease.<sup>8</sup>

The precise cause of linear lichen planus is unknown but associated factors and disease conditions seen include drugs such as ibuprofen and olanzapine, dental metal allergy, bone marrow transplant, multiple deliveries and they may occur as an isotopic response in areas of prior zoster, and that may in turn be related to HCV infection<sup>9-17</sup>.

For the pathogenesis, it is evident that LP represents T-cell mediated autoimmune, with preferential accumulation of CD8+ cells, damage to basal keratinocytes that express altered self-antigens on their surface.<sup>18</sup> Due to the rarity of linear lichen planus, the definite diagnosis depends on pathological examination.

The pathological findings of lichen planus include wedge-shaped hypergranulosis with no parakeratosis except when rubbed or oral, irregular acanthosis with saw-toothed rete ridges, liquefaction degeneration of the basal layer, lichenoid lymphocytic infiltrate, Colloid bodies and melanin incontinence often seen. Direct immunofluorescence (DIF) often reveals characteristic findings of immunoglobulin – mainly IgM, complement, and fibrin staining of colloid bodies in the deep epidermis and superficial dermis. Note that DIF of LP is not usually necessary.<sup>6, 19</sup>

In this case, topical and oral corticosteroids as well as oral antihistamines are prescribed. These medications are mostly used in

the treatment of lichen planus following the lines of Blaschko with good response; however, persistent pigmentation might remain.<sup>7,20</sup> Additionally, tacrolimus ointment has been reported to be an effective treatment for linear lichen planus.<sup>21</sup>

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