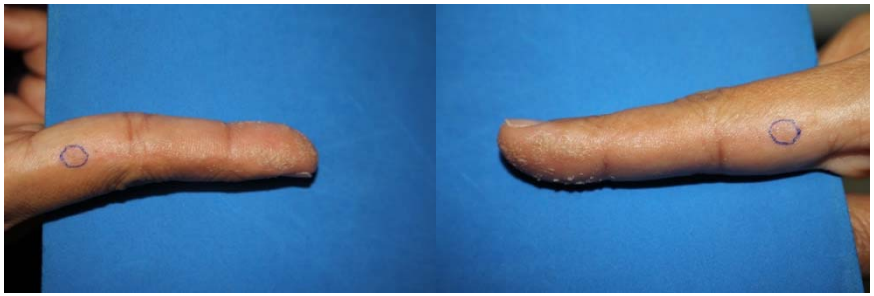
**Case 14**
A 38-year-old Thai woman from Bangkok
**Chief complaint:** Thick, hard, pruritic rash on right index finger for many years


(Fig. 14.1)

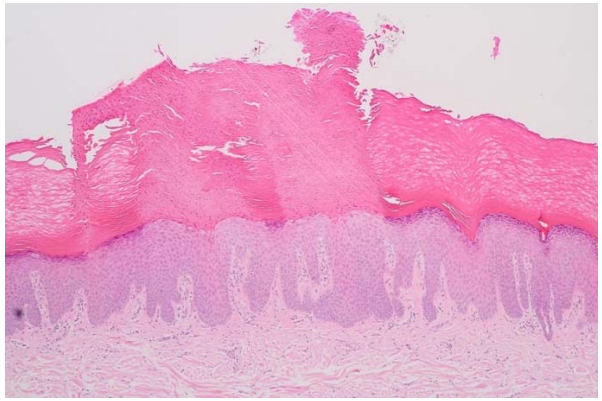
**Present illness:** At the age of 10, she noticed mild skin thickness on the lateral side of right index finger, without any other symptoms**.** At age 33, the lesion developed into a hyperkeratotic plaque with scales in linear distribution**.** She has been treated with topical keratolytics and cryotherapy, with minimal improvement**.**

**Past history:** No underlying diseases, no drug allergies

**Physical examination:**
HEENT: Mildly pale conjunctivae, anicteric sclerae
Lymph node: Not palpable
Extremities: No swelling or deformities of extremities

**Dermatological examination:** (Fig 14.1)
Hyperkeratotic, scaly linear plaque along the lateral side of right index finger

**Histopathology:** (S17 - 08243, Right index finger) (Fig. 14.2)


(Fig. 14.2)

- Alternate orthokeratosis overlying hypergranulosis, and parakeratosis overlying hypogranulosis of the psoriasiform epidermis

**Diagnosis:** Inflammatory Linear Verrucous Epidermal Nevus

**Treatment:**
- 20% urea cream apply on the lesion twice daily
- 0.064% betamethasone/3% salicylic ointment apply on the lesion once daily
- 20% salicylic acid cream apply on the lesion once daily

**Presenter:** Eesha Patel, MBBS

**Consultant:** Ruedee Phasukthaworn, MD

**Discussion:**

Inflammatory Linear Verrucous Epidermal Naevi (ILVEN) were first described in the literature in 1971.[1] They are a distinct variety of keratinocytic epidermal nevus that is clinically inflammatory and is usually psoriasiform, with regular acanthosis on histopathology. Unlike non-inflammatory epidermal nevi, ILVEN are far less common, usually erythematous and intractably pruritic.[2]

Clinically, it presents with erythematous and verrucous papules with intense pruritus and linear distribution following Blaschko's lines.[3] Although cases of bilateral involvement have been described, the disease is often unilateral and localized in one extremity.

Children are more commonly affected and it predominates in females in the ratio of 4:1. Most cases are sporadic, although familial cases have been reported.[4] A majority of the lesions resolve spontaneously by adulthood.[5] Possible association of ILVEN with arthritis has been described.[6]

ILVEN is caused by somatic mutations that result in genetic mosaicism and, although its pathophysiology is still unclear, it is believed that it may be associated with an increase in the production of interleukins 1 and 6, tumoral necrosis factor-alpha and intercellular adhesion molecule 1.[7]

ILVEN is diagnosed by clinical and histological examination. The classical clinical criteria for diagnosis, suggested by Altman and Mehregan in 1971 and later modified by Morag and Metzker in 1985, are: early age of onset, predominance in females, frequent involvement of the left leg, pruritus, distinctive psoriasiform appearance and marked refractoriness to therapy.[8,9]

Histological alterations of ILVEN were described by Dupre & Christol.[9] Specific signs include alternation between orthokeratosis and parakeratosis and the presence or absence of the granular layer, although the latter are not pathognomonic. Other microscopic findings include: papillomatosis, acanthosis, lymphocytic dermal infiltration or even Munro's microabscesses, but these are unspecific markers.

The differential diagnosis of ILVEN must be done with various dermatoses, such as other epidermal nevi, linear psoriasis, and lichen striatus.[10]

ILVEN is markedly refractory to therapy. There are reports of the use of many alternatives in its control: topical glucocorticoids applied under occlusion, intralesional corticosteroid, combination of tretinoin 0.1% and fluorouracil 5%; anthralin, tar, vitamin D3 analogues, surgical excision, cryotherapy with liquid nitrogen, and laser therapy with carbon dioxide.[7] Pulsed dye laser has been used successfully in some cases. Its mechanism of action is thought to involve destruction of small blood vessels in the papillary dermis that supply the overlying epidermis.[11] Effective topical treatment of ILVEN with the synthetic vitamin D analogue calcipotriol has been reported.[12] Pierini et al.[13] reported a case of inflammatory verrucous epidermal nevus which had good therapeutic results with the use of TCA peels. There are anecdotal reports of improvement with etanercept.[14]

Arthritis in association with ILVEN, although rare, can lead to significant morbidity. Early recognition is critical. Treatment is the same as for psoriatic arthritis.[13]

**References:**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