

## Case Report

# A recalcitrant linear plaque since infancy: ILVEN or porokeratosis?

## 嬰兒期出現難治的線狀丘疹：炎症性線狀疣狀表皮痣或汗孔角化症？

SK Hui 許紹基 and KC Lee 李景中

---

A young lady presented with a recalcitrant linear scaly plaque on her right leg since infancy. The lesion showed partial response to cryotherapy. Histology reported inflammatory linear verrucous epidermal naevus (ILVEN) and porokeratosis at her age 8 and 19 respectively. The final diagnosis is ILVEN. The differential diagnosis of ILVEN, especially with porokeratosis will be discussed.

年輕女患者自嬰兒期出現右下肢線狀鱗狀丘疹。皮損經冷凍治療後有部分好轉。患者於 8 歲及 19 歲時皮膚活檢的組織學報告分別為炎症性線狀疣狀表皮痣及汗孔角化症。本例的最終診斷為炎症性線狀疣狀表皮痣。本文會討論此症的鑒別診斷，特別其與汗孔角化症的鑒別。

**Keywords:** Cornoid lamella, inflammatory linear verrucous epidermal naevus, porokeratosis

**關鍵詞：**角樣板層，炎症性線狀疣狀表皮痣，汗孔角化症

---

## Introduction

Inflammatory linear verrucous epidermal naevus (ILVEN) and linear porokeratosis are uncommon

---

**Social Hygiene Service, Department of Health, Hong Kong**

SK Hui, MRCP(UK), FHKAM(Medicine)

**Department of Pathology, Queen Elizabeth Hospital, Hong Kong**

KC Lee, FRCPA, FHKAM(Pathology)

Correspondence to: Dr. SK Hui

Sai Ying Pun Dermatology Clinic, 3/F, Sai Ying Pun Jockey Club Clinic, 134 Queen's Road West, Hong Kong

linear dermatoses. Clinically and histopathologically, both ILVEN and linear porokeratosis have characteristic features of their own. In the present case, pathological reports of ILVEN and porokeratosis were encountered from two different biopsies 11 years apart. These two specimens and relevant literatures were reviewed, the differentiation of the two conditions are discussed.

## Case report

A 19-year-old Chinese lady had a long history of an asymptomatic linear plaque on her right

lower limb. She first presented at aged 2 when the lesion was noted on her posterior right thigh. Subsequently, it extended to her buttock, right lower leg and foot till aged 5 (Figure 1). The patient enjoyed good general health and there was no family history of hereditary or congenital cutaneous disease. The clinical differential diagnoses were epidermal naevus, lichen striatus, linear porokeratosis and elastosis perforan serpiginosa. She was treated with cryotherapy but the lesion only partially improved. At age of 8, the first skin biopsy was performed and histopathology showed features compatible with ILVEN. Cryotherapy was continued, the lesions responded to treatment but then recurred later. The patient defaulted follow-up until she was 17. Examination showed a group of linear hyperkeratotic papules and plaques over her buttock, right posterior thigh and right foot. In some areas, the plaques are more psoriasiform (Figure 2). Residual linear hypo- and hyperpigmented marks were found on the course of the lesion, lying along the Blaschko's line from the right buttock to the right foot, which may represent the post-inflammatory pigmentation changes after cryotherapy. At age of 19, the patient was referred to plastic surgeon, offering her the possibility of surgical intervention, including skin graft. A second skin biopsy was performed with a pathological report of porokeratosis. In view of the considerations of porokeratosis, the patient was referred back to dermatologist for further assessment. Subsequently, the patient was reviewed clinically and the histology slides were reviewed.

Joined evaluation of the two skin biopsies concluded that the case was an ILVEN (Figure 3). In addition, cornoid lamellae were noted in these specimens (Figure 4). This porokeratosis-like changes contributed to the previous histologic confusion to a porokeratosis.

The patient is currently under the care of dermatologist. The scaly lesions and the



**Figure 1.** A linear brownish scaly plaque extended from the right buttock to the right foot.



**Figure 2.** Psoriasiform plaques over the right foot, persisted since childhood.

associated pruritus were effectively eradicated by cryotherapy, however, frequent recurrence was expected. Other treatment modalities, such as topical calcipotriol, topical retinoids and CO<sub>2</sub> laser will be considered. Though ILVEN was the

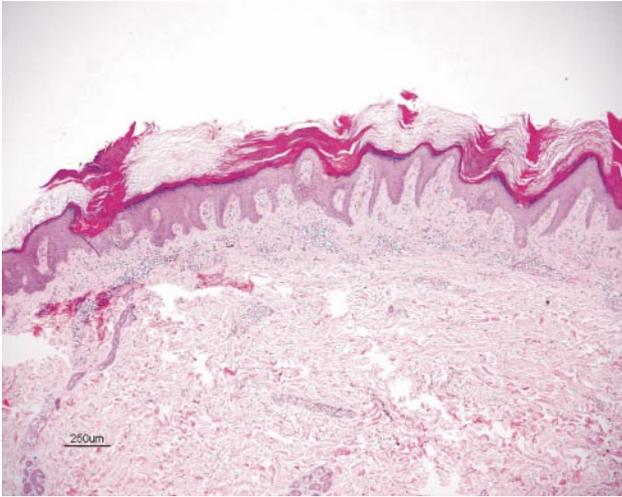
diagnosis, screening test for systematic illness and immune-compromised conditions were arranged for the patient, as that would be for a patient with linear porokeratosis (see below).

## Discussion

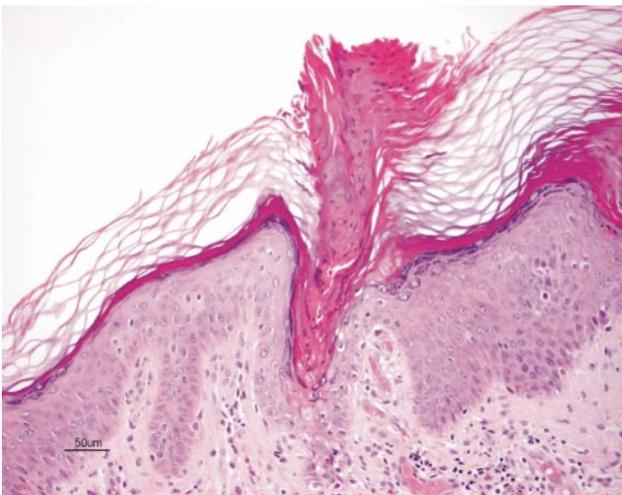
ILVEN is also known as dermatitic epidermal naevus. Altman & Mehregan first described this condition using the name inflammatory linear verrucous epidermal naevus in 1971.<sup>1</sup> ILVEN is a variant of epidermal naevus. It presents as linear psoriasiform papules and plaques on an extremity, they commonly persists for years despite treatment.<sup>2</sup> The definitive aetiology of ILVEN is not yet clear, but is considered to be related to the following aspects: 1) alteration of involucrin, which was increased in orthokeratotic area and decreased in parakeratotic area; 2) alteration of cytokines; 3) clonal deregulation of growth of keratinocytes and 4) somatic mosaicism.<sup>2,3</sup>

ILVEN usually appears in early childhood, but occasionally at birth or during adulthood. Seventy-five percent of patients have an onset before aged 5. It occurs equally in males and females, with no preference of sides of limbs. It presents as linear, erythematous and scaly papules coalescing to form a psoriasiform plaque, usually on a unilateral limb. The lesion typically follows the Blaschko's lines. Significant pruritus is common. ILVEN may show partial response to treatment, but usually recurs and persists, some may resolve spontaneously.<sup>4-6</sup>

Su reviewed 160 cases of epidermal naevi in 1982,<sup>7</sup> apart from the common type of epidermal naevus, nine variants were identified, including psoriasiform in cases of ILVEN and porokeratosis-like epidermal naevus. Two major histopathological changes were present in cases of ILVEN:<sup>7,8</sup> 1) psoriasiform changes, these included hyperkeratosis, parakeratosis,



**Figure 3.** Irregular psoriasiform change with superficial perivascular infiltrate. Note the scattered foci of parakeratosis alternating with orthokeratotic zones (H & E, Original magnification x 20).



**Figure 4.** Typical cornoid lamellae with a narrow zone of parakeratosis overlying region of keratinocyte degeneration in the epidermis, from the first biopsy specimen (H & E). Cornoid lamellation like area could be found in both biopsies specimens (H & E, Original magnification x 20).

acanthosis with thinned suprapapillary plates, tortuosity of the dermal capillaries and dermal mononuclear cell infiltrates; 2) sharply demarcated alternated areas of parakeratosis and orthokeratosis. The parakeratotic areas are slightly raised, with agranulosis, whereas the orthokeratotic areas are slightly depressed with hypergranulosis.

For the diagnoses of ILVEN, the following dermatoses should be considered for differentiation:<sup>5-7</sup> 1) Linear psoriasis. It is considered to be extremely rare. The early onset in ILVEN and the positive family history in psoriasis may help to differentiate the two. There may be scattered lesions elsewhere on the skin in psoriasis. Quantitative immunochemistry studies with T-cell markers and markers for keratinization showed different patterns in these two conditions.<sup>9</sup> 2) Linear lichen planus. The pathological pattern of linear lichen planus is that of lichenoid dermatitis, the post-inflammatory hyperpigmentation may be more prominent. 3) Lichen striatus. Lichen striatus is a self-limiting condition. It is not particularly itchy. The pathological pattern is lichenoid and/or spongiotic dermatitis. Eighty percent of these patients are atopic. 4) Ichthyosiform naevus in CHILD syndrome. In its less complete form, the skin lesion may follow Blaschko's line, favouring body folds. Histological finding may include foam cell in papillary dermis (verruciform xanthoma). 5) Linear porokeratosis. ILVEN and linear porokeratosis share many features in common, ILVEN is commonly itchy, and presents as scaly erythematous psoriasiform plaques; whereas in linear porokeratosis, the lesions are usually asymptomatic or only mildly itchy. They present as keratotic papules with raised borders, depressed and atrophic centres and longitudinal furrows. Histologically, a focus of parakeratosis with agranulosis (cornoid lamella) is typically found in porokeratosis; whereas in ILVEN, two major changes are present: i) psoriasiform dermatitis and ii) alternating areas of

parakeratosis with agranulosis and orthokeratosis with hyperkeratosis. Despite all these discriminating features, porokeratosis-like changes are present in certain case of epidermal naevi.<sup>8</sup> Further more, cornoid lamellation is by no means pathognomonic of porokeratosis, it can be found as an incidental phenomenon in a range of inflammatory, hyperplastic and neoplastic skin conditions.<sup>10</sup>

The treatment of ILVEN is difficult. Though surgical excision is curative, it is commonly associated with scarring. Treatment with dermabrasion or cryotherapy is commonly associated with recurrence by superficial procedures and scarring by deeper procedures.<sup>11</sup> Topical agents like calcipotriol, 5-fluorouracil and retinoids were reported to be effective in some patients.<sup>12-14</sup> Laser therapy with CO<sub>2</sub> laser, Erbium-YAG ablation laser and 585 nm flashlamp-pumped pulsed dye laser also showed curative results in individual cases.<sup>15</sup>

The different management of linear porokeratosis makes the differential diagnosis from ILVEN significant. For cases of linear porokeratosis, there is: 1) A necessity of assessment of the immunity functions and systemic illness, as porokeratosis is associated with HIV infection, end-stage hepatic or renal diseases occasionally; and 2) A necessity of monitoring of malignant changes, since up to 7.5-11% of porokeratosis may have malignant changes, especially in extensive and long standing lesions of linear porokeratosis.

In summary, we reported a young Chinese lady who had a recalcitrant linear plaque over her right buttock and lower limb since infancy. The presence of cornoid lamellae caused an initial diagnostic confusion between ILVEN and linear porokeratosis. Clarification of these two conditions is of practical importance for these two conditions are different in aetio-pathogenesis, management and outcome.

## References

1. Altman J, Mehregan AH. Inflammatory linear verrucose epidermal nevus. *Arch Dermatol* 1971;104:385-9.
2. Welch ML, Smith KJ, Skelton HG, Frisman DM, Yeager J, Angritt P, et al. Immunohistochemical features in inflammatory linear verrucous epidermal nevi suggest a distinctive pattern of clonal dysregulation of growth. Military Medical Consortium for the Advancement of Retroviral Research. *J Am Acad Dermatol* 1993;29 (2 Pt 1):242-8.
3. Ito M, Okuda C, Shimizu N, Tazawa T, Sato Y. Histopathogenesis of inflammatory linear verrucose epidermal naevus: histochemistry, immunohistochemistry and ultrastructure. *Arch Dermatol Res* 1991;283:491-9.
4. Hodge SJ, Barr JM, Owen LG. Inflammatory linear verrucose epidermal nevus. *Arch Dermatol* 1978;114:436-8.
5. Rogers M, McCrossin I, Commens C. Epidermal nevi and the epidermal nevus syndrome. A review of 131 cases. *J Am Acad Dermatol* 1989;20:476-88.
6. Lee SH, Rogers M. Inflammatory linear verrucous epidermal naevi: a review of 23 cases. *Australas J Dermatol* 2001;42:252-6.
7. Su WP. Histopathologic varieties of epidermal nevus. A study of 160 cases. *Am J Dermatopathol* 1982;4:161-70.
8. Dupre A, Christol B. Inflammatory linear verrucose epidermal nevus. A pathologic study. *Arch Dermatol* 1977;113:767-9.
9. Vissers WH, Muys L, Erp PE, de Jong EM, van de Kerkhof PC. Immunohistochemical differentiation between inflammatory linear verrucous epidermal nevus (ILVEN) and psoriasis. *Eur J Dermatol* 2004;14:216-20.
10. Weedon D. Disorders of epidermal maturation and keratinization. In: *Skin pathology*. 2nd ed. London: Churchill Livingstone, 2002;9:292-3.
11. Lee BJ, Mancini AJ, Renucci J, Paller AS, Bauer BS. Full-thickness surgical excision for the treatment of inflammatory linear verrucous epidermal nevus. *Ann Plast Surg* 2001;47:285-92.
12. Gatti S, Carrozzo AM, Orlandi A, Nini G. Treatment of inflammatory linear verrucous epidermal naevus with calcipotriol. *Br J Dermatol* 1995;132:837-9.
13. Mitsushashi Y, Katagiri Y, Kondo S. Treatment of inflammatory linear verrucous epidermal naevus with topical vitamin D3. *Br J Dermatol* 1997;136:134-5.
14. Kim JJ, Chang MW, Shwayder T. Topical tretinoin and 5-fluorouracil in the treatment of linear verrucous epidermal nevus. *J Am Acad Dermatol* 2000;43(1 Pt 1):129-32.
15. Alster TS. Inflammatory linear verrucous epidermal nevus: successful treatment with the 585 nm flashlamp-pumped pulsed dye laser. *J Am Acad Dermatol* 1994;31(3 Pt 1):513-4.