

## Case Report

# A man with verrucous lesions over the right foot for 30 years: a case of ILVEN

## 一名男仕的右腳有著持續三十年的疣狀皮損：炎性線狀疣狀表皮痣之一病例

ACK Chung 鍾振堅 and WY Lam 林永賢

A 49-year-old man presented with itchy verrucous lesions over the right shin and the dorsum of the right foot for 30 years. There was no personal or family history of psoriasis or eczema. Skin biopsy showed findings consistent with inflammatory linear verrucous epidermal naevus. The patient had symptomatic relief with betamethasone valerate 0.1% cream. The clinical presentation, pathogenesis and management of this condition are discussed.

一名本身沒有銀屑病或濕疹病史或家族史的四十九歲男仕，因其右脛及右足背歷時三十年的疣狀皮損及瘙癢而求診。切皮檢查證實為炎性線狀疣狀表皮痣，患者病徵在使用倍他米松戊酸酯 0.1% 乳膏後有改善。本文將探討此病的臨床表現、致病機理及其治療。

**Keywords:** Inflammatory linear verrucous epidermal naevus

**關鍵詞：**炎性線狀疣狀表皮痣

### Case history

A 49-year-old man with a history of diabetes mellitus, hypertension, ischaemic heart disease

**Social Hygiene Service, Centre for Health Protection,  
Department of Health, Hong Kong**

ACK Chung, MBBS(HK), MRCP(UK)

**Department of Clinical Pathology, Tuen Mun  
Hospital, Hong Kong**

WY Lam, FRCPath, FHKAM(Pathology)

Correspondence to: Dr. ACK Chung

Cheung Sha Wan Dermatological Clinic, 3/F West  
Kowloon Health Centre, 303 Cheung Sha Wan Road,  
Kowloon

and hyperlipidaemia presented with itchy verrucous lesions over the right shin and the dorsum of the right foot which had been present for about 30 years. He had noticed that the lesions appeared after an injury by oyster shells and the lesions were very itchy all along.

Physical examination demonstrated that there was a linear verrucous lesion over the dorsum of the right foot which extended up to the shin with a hyperkeratotic surface (Figures 1a & 1b). The clinical differential diagnosis included infections like cutaneous tuberculosis infection, deep fungal infection and viral warts. Inflammatory conditions like linear prokeratosis, linear lichen planus, lichen striatus, linear psoriasis, linear Darier's disease and inflammatory linear verrucous

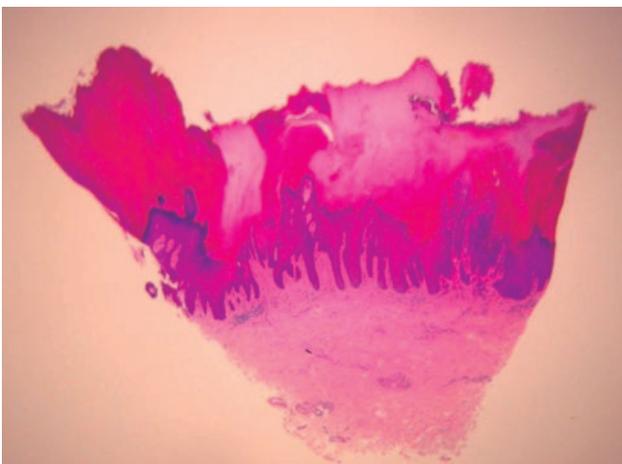
epidermal naevus (ILVEN) had also to be considered.

Incisional skin biopsy over the right shin showed discrete alternating zones of orthokeratotic and parakeratotic hyperkeratosis, digitated and

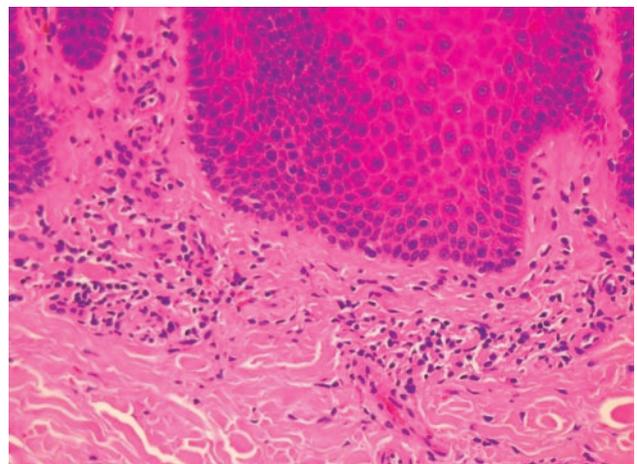
psoriasiform epidermal acanthosis, papillomatosis and a patchy superficial dermal perivascular lymphocytic infiltrate (Figures 2 & 3). There were no granulomata or fungi. The histological diagnosis was specific for inflammatory linear verrucous epidermal naevus.



**Figure 1.** (a) Linear verrucous lesion extending from the dorsal surface of the right foot to the right shin. (b) Close-up view showing brownish papules coalescing to form a linear plaque.



**Figure 2.** Low power field shows psoriasiform epidermis and overlying discrete pale zones of parakeratosis alternating with red zones of orthokeratosis, diagnostic for ILVEN .



**Figure 3.** High power field showing in the dermis beneath the acanthotic epidermis a perivascular inflammatory infiltrate of lymphocytes.

He was treated with betamethasone valerate 0.1% cream, with symptomatic relief. The treatment response was satisfactory and his pruritus improved with topical steroid.

## Discussion

Inflammatory linear verrucous epidermal naevus (ILVEN) was first described in the literature in 1971 by Altman and Mehregan. It is a distinct variety of epidermal naevus, which has an inflammatory and usually psoriasiform clinical appearance. Inflammatory linear verrucous epidermal naevus accounts for approximately 5% of patients with epidermal naevi.<sup>1,2</sup>

Patients usually present with persistent pruritic linear plaques along the Blaschko's lines on a limb in early childhood. The condition may be observed at birth, but it appears more frequently in infancy. The lesions are itchy, red and inflamed. The surface of the lesions may resemble eczema (dry, red, excoriated) or like psoriasis (red, scaly). Most cases are sporadic, although familial cases have been described. It is more common in females, with a female-to-male ratio of 4:1.

ILVEN belongs to a special subgroup of epidermal naevi. Epidermal naevi arise from the pluripotential germinative cells of the basal layer of the embryonic epidermis. Inflammatory linear verrucous epidermal naevus is distinct from psoriasis but both conditions may share some common pathogenic pathways.<sup>3</sup> These pathways are probably mediated by interleukin-1, interleukin-6, tumour necrosis factor-alpha, and intercellular adhesion molecule-1. Controversy exists as to whether linear psoriasis is a separate entity or subset of ILVEN.<sup>4-6</sup>

The histopathological appearance of ILVEN is characteristic, with discrete stubby zones of hypergranulosis and orthokeratotic hyperkeratosis alternating with similar well-defined zones of agranulosis and parakeratotic hyperkeratosis with

an associated lymphohistiocytic inflammatory infiltrate. This histology is specific for a distinctive type of epidermal nevus, and in particular the inflammatory verrucous epidermal nevus.<sup>7</sup>

It is rarely reported to be associated with autoimmune thyroiditis, arthritis and lichen amyloidosis. Association with syndromal disorders like congenital hemidysplasia with ichthyosiform erythroderma and limb defects syndrome (CHILD) were also reported. CHILD is extremely rare. It was first described in 1903 and is an X-linked dominant disorder. These patients present with lesions over half of the body which consist of birth defects with skin, gastrointestinal, musculoskeletal and central nervous system involvement, severe limb reduction, scoliosis and occasionally defects in internal organs. Limb defects are ipsilateral to the most prominent cutaneous lesions and range from digital hypoplasia to agenesis of the extremity. Some authorities even postulated that ILVEN might be a limited form of CHILD.<sup>8</sup>

Epidermal naevus syndrome occurs when epidermal naevi are associated with abnormalities in other systems, including ocular, neurological and skeletal defects. Thus, it is of paramount importance to evaluate for clinical evidence of involvement of other systems in children presenting with epidermal naevi.

Although uncommon, ILVEN is important to be identified due to the significant morbidity of intense pruritus. The lesions of ILVEN have no tendency to remit or improve with time.

Management of this condition is challenging as the lesions are usually resistant to topical steroids, intralesional steroids, topical retinoids and even cryotherapy. The aim of treatment is to relieve clinical symptoms and improve the patient's quality of life. As ILVEN is not a common disease, current literature review on therapy is mainly based on case reports or small case series. Using emollients and potent topical steroids under

occlusion can relieve itchiness. Vitamin D analogues, such as topical calcipotriol, can inhibit epidermal proliferation and promote keratinocyte differentiation and is useful in some cases.<sup>9</sup> Small lesions can be surgically excised. Destructive therapy by CO<sub>2</sub> laser, dermabrasion, and chemical peeling with 60% trichloroacetic acid (TCAA) can be offered to selected groups of patients. For patients with intractable pruritus refractory to the aforementioned therapy, full thickness surgical excision with primary closure, use of adjunctive tissue expansion, skin flaps and split-thickness skin graft have been described in the literature.

## References

1. Lee SH, Rogers M. Inflammatory linear verrucous epidermal naevi: a review of 23 cases. *Australas J Dermatol* 2001;42:252-6.
2. 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.
3. Miteva LG, Dourmishev AL, Schwartz RA. Inflammatory linear verrucous epidermal nevus. *Cutis* 2001;68:327-30.
4. Happle R. Linear Cowden nevus: a new distinct epidermal nevus. *Eur J Dermatol* 2007;17:133-6.
5. Happle R. Linear psoriasis and ILVEN: is lumping or splitting appropriate? *Dermatology* 2006;212:101-2.
6. Le K, Wong LC, Fischer G. Vulval and perianal inflammatory linear verrucous epidermal naevus. *Australas J Dermatol* 2009;50:115-7.
7. Ackerman AB, Guo Y, Vitale P. Clues to Diagnosis in Dermatopathology. Chicago:ASCP Press;1992. p.45-8.
8. Vidaurri-de la Cruz H, Tamayo-Sanchez L, Duran-McKinster C, de la Luz Orozco-Covarrubias M, Ruiz-Maldonado R. Epidermal nevus syndromes: clinical findings in 35 patients. *Pediatr Dermatol* 2004;21:432-9.
9. Zvulunov A, Grunwald MH, Halvy S. Topical calcipotriol for treatment of inflammatory linear verrucous epidermal naevus. *Arch Dermatol* 1997;133:567-8.