Case 15
A 60 year-old Thai woman from Ayutthaya

Chief complaint: Multiple pruritic papules on left alar nasi for 10 years

Present illness:
The patient developed multiple scaly erythematous papules with marked pruritus on her left alar nasi for 10 years. The lesions gradually extended to the left medial canthus and left upper eyelid. She was treated with topical mometasone cream with limited improvement. She was otherwise in good health

Past history: Anemia

Family history:
There was no family history of similar cutaneous lesions

Skin examination:
Multiple ill-defined scaly erythematous papules on left medial canthus, upper eyelid extending to left alar nasi

Physical examination:
Systemic examination other than skin lesions revealed no abnormality

Laboratory investigation:
KOH: negative

Histopathology: (S16-6817A, left eyelid)

- Alternating para/orthokeratosis and hypo/hypergranulosis in the stratum corneum and psoriasiform epidermal hyperplasia
- Superficial perivascular infiltrate of lymphocytes

Diagnosis: Inflammatory linear verrucous epidermal nevus

Treatment:
- Topical calcipotriol/betamethasone gel applied once daily
Cream base applied twice daily
- Cetirizine (10 mg) 1 tab oral twice daily
- Hydroxyzine (10 mg) 1 tab oral at bedtime
- Plan for CO₂ laser treatment

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**Discussion:**
Inflammatory linear verrucous epidermal nevus (ILVEN) is a relatively rare variant (accounted for 6%) of verrucous epidermal nevus which is a hamartomatous proliferation of the epithelium. It was originally described by Altman and Mehregan in 1971. It is a linear, psoriasiform, erythematous, scaly papule or plaque that follows Blaschko’s lines, unilaterally with more affected on the left side and is characteristically pruritic. It usually appears during childhood with 75% of nevi appearing before the age of 5 years, however late onset in adults has also been reported. It predominates in females in the ratio of 4:1. Most cases are sporadic, although familial cases have been described. The distribution was predominantly on the lower half of the body, especially on buttock area, and is relatively rare on genital and perigenital area. Only 16% of lesions were found on the upper half of the body, including the axillae, arm, and hand. There was one case report of ILVEN located on right upper eyelid. In our case, the lesions are located on the face including left upper eyelid, medial canthus and alar nasi which are unusual sites.

An estimated one-third of individuals with epidermal nevi have involvement of other organ systems especially ophthalmic, neurologic and skeletal system; called epidermal nevus syndrome, however it is seldom reported in ILVEN. Rarely, there are ipsilateral skeletal abnormalities, usually reduction deformities, suggesting that ILVEN is possibly a forme fruste of CHILD syndrome. ILVEN had also been reported in association with regional odontodysplasia, arthritis, autoimmune lymphocytic thyroiditis, hereditary mucinous nevus, undescended testis, and lichen amyloidosis. Moreover, there was one case report of malignant transformation of ILVEN into squamous cell carcinoma. In this case no other organ abnormalities or malignancy change was observed.

Inflammatory linear verrucous epidermal nevus is caused by somatic mutations that result in genetic mosaicism and although its physiopathology is still unclear, it is believed that it may be associated with an increase in the production of interleukins-1 and 6, tumor necrosis factor-alpha, and intercellular adhesion molecule.

The differential diagnosis of ILVEN includes other various dermatoses such as Darier’s disease, linear porokeratosis, linear lichen planus, linear psoriasis, and lichen striatus.

Skin biopsy is crucial in the diagnosis of ILVEN. There are two different histopathological features for ILVEN: nonspecific and specific. Areas of depressed hypergranulosis and overlying orthokeratosis alternating sharply with areas of agranulosis and overlying parakeratosis are specific features, but are not pathognomonic. The nonspecific findings includes an inflammatory dermal infiltrate, acanthosis, and papillomatosis of the psoriasis type with elongation of the epidermal ridges.

ILVEN is markedly refractory to therapy. Topical treatments such as topical corticosteroids with or without occlusion, intralesional steroid injections, and topical retinoids are rarely beneficial. Vitamin D analogs, as well as combination of potent topical steroid and topical calcineurin inhibitor, and medium to full-depth chemical peels were found to be helpful in some cases.

There are few reports of effective systemic treatments with acitretin and etanercept. Among the physical modalities; cryotherapy, photodynamic therapy, carbon dioxide laser therapy, and 585 nm flashlamp-pumped pulsed dye laser have...
been tried successfully.\textsuperscript{25,26,27} Although surgical excision tends to be followed by rapid recurrence and requires a generous depth of underlying dermis to be removed, there were some reported cases successfully treated with full-thickness surgical excision.\textsuperscript{18,28}

\textbf{References}