A Case Report of Inflammatory Linear Verrucous Epidermal Nevus with Epidermolytic Hyperkeratosis.

Chuenkamol Uengpitakphan MD,

Phunamart Srivichien MD,

Pinnaree Kattipattanapong MD.

ABSTRACT :

UENGPITAKPHAN C, SRIVICHIEN P, KATTIPATTANAPONG P. A CASE REPORT OF INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS WITH EPIDERMOLYTIC HYPERKERATOSIS.

THAI J DERMAOL 2017; 33: 306-312.

INSTITUTE OF DERMATOLOGY, BANGKOK, THAILAND.

Inflammatory linear verrucous epidermal nevus is an uncommon subtype of epidermal nevus. It is characterized by linear hyperpigmented verrucous plaques along the lines of Blaschko. We report a 52 year-old Thai female presented with unilateral inflamed linear verrucous plaques distributed on left forearm along the lines of Blaschko since birth. Histologic findings revealed focal marked vacuolation with basophilic keratohyalin granules at granular cell layer coexisting with papillomatosis with alternated focal parakeratosis. The overall findings are compatible with ILVEN with unusual histopathologic pattern of epidermolytic hyperkeratosis.

Key words: Inflammatory linear verrucous epidermal nevus, Epidermolytic hyperkeratosis

บทคัดย่อ :

ชื่นกมล อึงพิทักษ์พันธุ์ พรรณมาตย์ ศรีวิเชียร ปิ่นนรี ขัตติพัฒนาพงษ์ รายงานผู้ป่วยโรค INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS ที่มีผลพยาธิวิทยา EPIDERMOLYTIC HYPERKERATOSIS

วารสารโรคผิวหนัง 2560; 33: 306-312.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

From: Institute of Dermatology–Ministry of Public Health, Bangkok, Thailand

Corresponding author: Chuenkamol Uengpitakphan MD., email: choco_maiii@yahoo.com

โรค Inflammatory linear verrucous epidermal nevus อยู่ในกลุ่มโรคที่เกิดจากการแบ่งตัวผิดปกติของชั้นหนังกำพร้ามาด้วยผื่นที่มีสีเข้ม ผิวขรุขระ เรียงตัวเป็นเส้นตาม blaschko's lines รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงไทยอายุ 52 ปีที่มาด้วยผื่นสีเข้ม ผิวขรุขระ เรียงตัวตาม blaschko's lines บนแขนซ้ายตั้งแต่กำเนิด ผลพยาธิวิทยาพบ focal marked vacuolationและ basophilic keratohyalin granules ที่ชั้น granular cell layer ร่วมกับมีpapillomatosis และ alternated focal parakeratosis จากลักษณะทั้งหมดเข้าได้กับ Inflammatory linear verrucous epidermal nevus ที่พบลักษณะทางพยาธิวิทยาเป็น epidermolytic hyperkeratosis ซึ่งพบได้ไม่บ่อยในโรคนี้

คำสำคัญ: โรค Inflammatory linear verrucous epidermal nevus, Epidermolytic hyperkeratosis

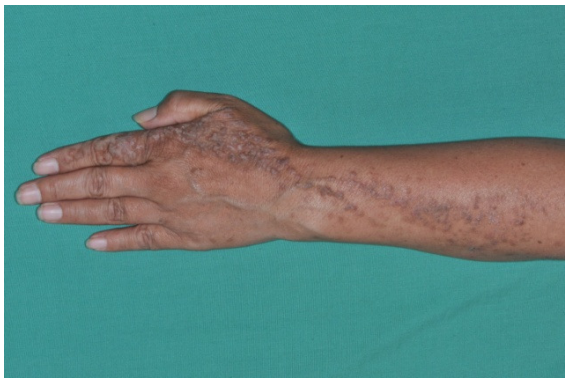

Figure 1 Unilateral linear brownish hyperkeratotic papules coalesced to verrucous plaque on left forearm along the lines of Blaschko.

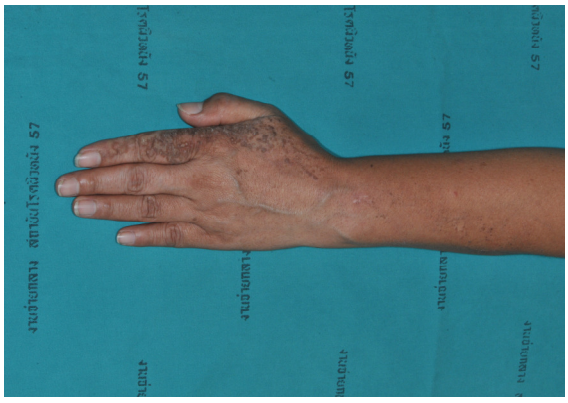

Figure 2 Post-treatment with topical 20% salicylic acid ointment and 0.05% betamethasone valerate for two months showed slightly improvement.

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare clinical subtype of epidermal nevus. The classical diagnostic criteria of ILVEN are composed of 1) early age of onset, 2) predominance in females, 3) frequent involvement of the left lower extremity, 4) pruritus, 5) marked refractoriness to therapy, and 6) distinctive inflammatory and psoriasiform histologic appearance.[1] Many specific and non-specific histopathologic features were reported. We reported a case of ILVEN with unusual histopathologic pattern of epidermolytic hyperkeratosis.

Case report

A 52 year-old Thai female presented with linear hyperkeratotic brownish papules distributed on her left index finger. The lesions first appeared since she was born, slowly progressed in size and thickness and extended to the left forearm during childhood. She also complained about the itching symptom on the affected area. The lesions had been stable for a

long period of time but over the past 3 months they were thickened and increased in numbers. She had neither underlying diseases nor family history of any systemic diseases. The dermatological examinations demonstrated multiple hyperpigmented hyperkeratotic papules coalesced into linear, verrucous, hyperpigmented plaques on the left index finger extending to the left forearm. The distribution followed the lines of Blaschko. (Figure 1). The remaining physical examination did not reveal any abnormalities.

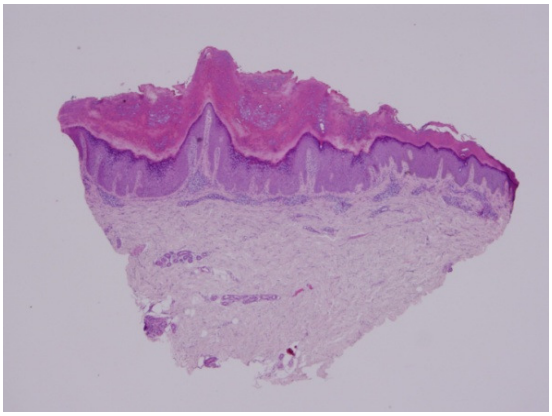

Figure 3 Histopathologic examination shows acanthotic epidermis with papillomatosis, hyperkeratosis and alternated focal parakeratosis. There is a marked vacuolation with basophilic keratohyalin granules at granular cell layer. (H&E, 4x)

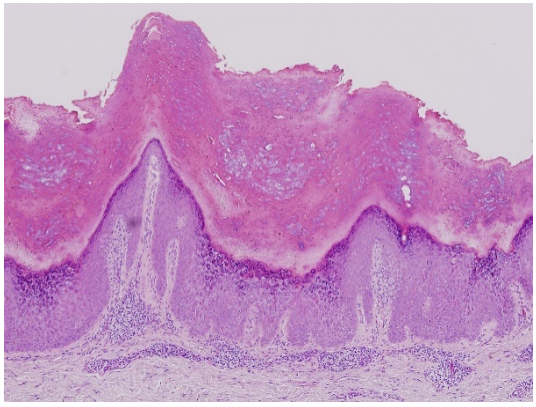

Figure 4 Histopathologic examination shows acanthotic epidermis with papillomatosis, hyperkeratosis and focal parakeratosis with alternated orthokeratosis. (H&E, 10x)

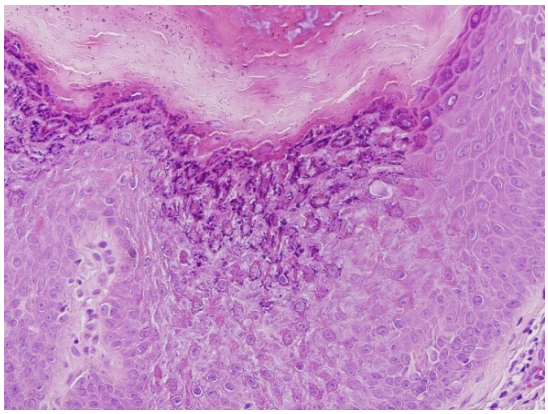

Figure 5 Histopathologic examination shows marked vacuolation with basophilic keratohyalin granules at granular cell layer. (H&E, 40x)

Histopathological examinations demonstrated acanthotic epidermis with papillomatosis, hyperkeratosis and focal parakeratosis with alternated orthokeratosis. There was a marked vacuolization with basophilic keratohyalin granules at granular cell layer. Neither interface

nor spongiotic change was seen. The dermis showed a subtle superficial perivascular lymphocytic infiltration. (Figure 3-5).

The overall clinical manifestations and histopathologic features are compatible with inflammatory linear verrucous epidermal nevus with unusual histopathologic pattern of epidermolytic hyperkeratosis. The patient was treated with topical 20% salicylic acid ointment and topical 0.05% betamethasone valerate. The lesions had slightly improved after two months. (Figure 2)

Discussion

Inflammatory linear verrucous epidermal nevus (ILVEN) is an uncommon clinical subtype of epidermal nevus. It was first proposed by Altman and Mehregan in 1971. The classical clinical criteria for diagnosis of ILVEN are composed of 1) early age of onset, 2) predominance in females, 3) frequent involvement of the left lower extremity, 4) pruritus, 5) marked refractoriness to therapy, and 6) distinctive inflammatory and psoriasiform histologic appearance.[1] The definite pathogenesis is unknown. Some believe that ILVEN and psoriasis share the same pathogenesis. In ILVEN, the increased amount of involucrin in orthokeratosis and minimal involucrin in underlying area of parakeratosis are seen. While expression of involucrin in all layers is found in psoriasis.[2] Another cause of ILVEN is thought to be related to upregulation of interleukin 1, interleukin 6, tumor necrosis factor alpha and intercellular adhesion molecules.[3] Another rare subgroup is a mosaic disorder, resulting from post-zygotic mutations of the keratin 1 (K1) and/or keratin 10 (K10) genes.[4] Although ILVEN is usually sporadic, there were a few reports of familial cases. The exact mode of transmission was not clear.[5] It is typically presented at birth and in early childhood before 6 years of age. It is more common in females with female to male ratio of 4:1.[6] No racial predominance is reported.

The characteristic features of ILVEN are consisted of intensely pruritic linear erythematous verrucous plaques distributed along Blaschko's lines.[7] The unilateral involvement with left side predominance is more common. The most common affected parts are lower half of the body including legs, pelvis and buttock.[8] In addition, atypical areas such as genital and eyelid were reported.[9,10] Other associated abnormalities were rarely reported and they were likely to be coincidence.

The histopathological features of ILVEN are classified into two different types: nonspecific and specific. Specific findings present depressed hypergranulosis and overlying orthokeratosis alternating with areas of agranulosis and overlying parakeratosis. Anyway, these findings are not pathognomonic. Other microscopic findings including papillomatosis,

psoriasiform hyperplasia with elongation of the rete ridges, Munro's microabscesses as seen in psoriasis or lymphocytic dermal infiltration are nonspecific features. Moreover, there are uncommon histopathologic varieties including epidermolytic hyperkeratosis, acrokeratosis verruciformis-like, seborrheic keratosis-like, psoriasiform (ILVEN), verrucoid, focal acantholytic dyskeratosis and porokeratosis-like.[11,12] Epidermolytic hyperkeratosis (EH) may be found in only five to ten percent of the verrucous epidermal nevi.[3] Recently Meibodi et al. reported a case of ILVEN with epidermolytic hyperkeratosis. The clinical findings tend to be warty with skin fragility and sloughed off areas.[3]

Epidermolytic hyperkeratosis (EH) patterns are composed of hyperkeratosis, hypergranulosis, and epidermolysis. Within epidermolytic areas, there are characteristic epidermal cells with perinuclear vacuolization and indistinct peripheral boundaries in spinous and granular layers. The number of various shape and size of basophilic keratohyalin granules increase at any level of epidermis, predominantly in the stratum granulosum. There are two distinct histopathologic patterns in EH which are composed of 1) continuous involvement of the entire horizontal epidermis, and 2) focal involvement with skip areas of normal epidermis along the horizontal epidermis.[13] EH pattern is characteristic for generalized and mosaic epidermolytic hyperkeratosis.[13] But this same histopathologic feature can also be seen in various conditions including ILVEN, epidermolytic acanthoma, nevoid follicular epidermolytic hyperkeratosis, melanocytic nevus, solar keratosis, squamous cell carcinoma, basal cell carcinoma, pilar cyst, seborrheic keratosis, cutaneous horn, skin tag, leukoplakia, nevus comedonicus, solid hidradenoma. Moreover, it can be found incidentally in normal epithelium of oral mucosa and skin.[13,14]

Our patient presented with unilateral linear hyperpigmented verrucous plaques along Blaschko's line. The clinical findings should be differentiated from linear lichen planus, linear psoriasis, lichen striatus, and linear porokeratosis. Although non-specific histologic pattern of epidermolytic hyperkeratosis was found in our case, other typical findings, such as acanthotic papillomatosis with alternated focal parakeratosis helped to diagnose ILVEN. ILVEN with unusual histopathologic pattern of epidermolytic hyperkeratosis is confirmed as the definitive diagnosis.

The natural course of ILVEN is chronic without tendency to remit or improve with time. It is markedly refractory to therapy and the results are quite variable. Intralesional corticosteroids or potent topical corticosteroids under occlusion are the first line and the most widely used treatments. The relief of symptoms

is temporary. Topical calcipotriol was reported to have benefit in pruritus, koebnerization or ILVEN with superimposed psoriasis. Chu, G. Y. et al reported sustained remission with acitretin 25 mg twice daily in relapse cases.[15] Other non-invasive therapeutic choices include topical tretinoin combined with 5-fluorouracil, destructive therapies, such as liquid nitrogen, electrodesiccation, ablative laser, dermabrasion, pulsed dye laser. Full thickness surgical excision should be considered in patients who failed non-operative treatment. It can reduce risk of recurrence but may result in scarring.[16] Our patient was treated with topical 20% salicylic acid ointment in combination with topical 0.05% betamethasone valerate. After two months the lesions showed slightly improvement.

In conclusion, ILVEN is a rare type of epidermal nevus which can be presented with varieties of histopathologic features. EH is also one of the uncommon histologic findings in ILVEN. The correlation of clinical manifestation and histopathology is very important for diagnosis.

References

1. Altman J, Mehregan AH. Inflammatory linear verrucose epidermal nevus. Arch Dermatol 1971; 104: 385-9.
2. Requena L, Requena C, Cockerell CJ. Benign epidermal tumors and proliferations. In: Bolognia JL, Jorizzo JL, Schaffer JV, editors. Dermatology. Vol One. 3rd ed. Philadelphia, London: Elsevier Saunders, 2012: 1810-11.
3. Meibodi NT, Nahidi Y, Javidi Z. Epidermolytic hyperkeratosis in inflammatory linear verrucous epidermal nevus. Indian J Dermatol 2011; 56: 309-12.
4. Guite PD, Gunto H, Das K. Epidermolytic hyperkeratosis in verrucous epidermal nevus. J Med Soc 2014:47-8.
5. Alsaleh QA, Nanda A, Hassab-el-Naby HM, Sakr MF. Familial inflammatory linear verrucous epidermal nevus (ILVEN). Int J Dermatol 1994; 33: 52-4.
6. Sugarman JL. Epidermal nevus syndromes. Semin Cutan Med Surg 2007; 26: 221-30.
7. Simonart T, Heenen M. Inflammatory verrucous epidermal naevus presenting as a psoriasiform plaque. Dermatology 2007;215:167-8.
8. Lee SH, Rogers M. Inflammatory linear verrucous epidermal naevi: a review of 23 cases. Australas J Dermatol 2001; 42: 252-6.
9. Bandyopadhyay D, Saha A. Genital/Perigenital Inflammatory Linear Verrucous Epidermal Nevus: A Case Series. Indian J Dermatol 2015; 60: 592-5.
10. Nakai N, Ohshita A, Katoh N. A Case of Inflammatory Linear Verrucous Epidermal Nevus on the Upper Eyelid. Indian J Dermatol 2015; 60: 323.
11. Podder I, Das A, Ghosh A, Shome K. Epidermolytic blaschkoid verrucous epidermal nevus: Report of two cases. Indian J Dermatopathol Diagn Dermatol 2015;2:46-8.

12. Su WP. Histopathologic varieties of epidermal nevus. A study of 160 cases. Am J Dermatopathol 1982; 4: 161-70.
13. Ross R, DiGiovanna JJ, Capaldi L, Argenyi Z, Fleckman P, Robinson-Bostom L. Histopathologic characterization of epidermolytic hyperkeratosis: a systematic review of histology from the National Registry for Ichthyosis and Related Skin Disorders. J Am Acad Dermatol 2008; 59: 86-90.
14. Hayashida MT, Mitsui GL, Reis NI, Fantinato G, Jordão Neto D, Mercante AM. Epidermolytic Hyperkeratosis-case report. An Bras Dermatol 2015; 90: 888-91.
15. Chu GY, Hu SC, Lan CC. Unusual presentation of inflammatory linear verrucous epidermal naevus mimicking linear psoriasis successfully treated with oral retinoid. J Eur Acad Dermatol Venereol 2015; 29: 2255-7.
16. 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.