

A Unique Porokeratotic variant of Inflammatory Linear Verrucous Epidermal Nevus

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Abstract

Inflammatory linear verrucous epidermal nevus is a rare variant of nevoid linear inflammatory dermatoses which appear most often in first six months of life and persists for many years or even whole life. Clinically it often shares many features with linear psoriasis, linear porokeratosis and other linear dermatoses which can usually be differentiated by an established set of clinical and histopathological criteria. Herein we are reporting an interesting and rare case fulfilling all the clinical and histopathological criteria of inflammatory linear verrucous epidermal nevus in a 13 year old boy which uniquely revealed cornoid lamella on histopathology so we termed it as “porokeratotic variant of inflammatory linear verrucous epidermal nevus”. To the best of our knowledge, only two such cases were reported previously in the literature and none from our country.

Keywords: Verrucous epidermal nevus; Linear dermatoses; Nevoid; Porokeratotic

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN) is clinically characterized by linear, erythematous and intensely pruritic scaly psoriasiform papules coalescing to form plaques following the Blaschko's lines usually involving the lower limb unilaterally [1]. ILVEN usually has early life onset and characteristically the lesions are persistent and resistant to treatment. Here we are reporting a unique case of ILVEN as a ‘porokeratotic variant of ILVEN’ in a 13 year old boy. To the best of our knowledge, only two such cases were reported in the previous literature and none from our country.

Case Report

A 13 year old boy came at the outpatient department presenting with multiple, extremely itchy papules, and plaques in a linear fashion on left lower limb since his infancy. During the infancy, lesions started to develop first on the dorsum of the left foot which subsequently extended to involve leg and posterior aspect of the thigh in a linear pattern over the next 3 years. No other body sites were affected with such lesions and there was no such family history.

On local cutaneous examination, multiple, intensely pruritic, psoriasiform and hyperkeratotic (at some places) coalescing papules and plaques on the lateral side of the left leg and posterior aspect of left thigh were noted on the hypopigmented and erythematous background which were arranged in a linear pattern along the blaschko's lines. On the dorsum of left feet, verrucous nodules were the predominant lesions (Figure 1).



Figure 1: A) verrucous nodules on dorsum of left foot, B) hyperkeratotic coalescing papules and plaques on the lateral side of left leg along the blaschko's lines.

The size of the lesions, erythema and scaling were variable. Examination of hair, nails, teeth, mucosa, palm and soles were normal. The child was normal on systemic examination and all laboratory parameters were within normal limits.

Clinically ILVEN, linear psoriasis, and linear porokeratosis were kept as differential diagnoses and to reach a final diagnosis, punch biopsy sample was taken and sent for the histopathological examination (HPE).

HPE revealed spongiosis, acanthosis, elongation of rete ridges and alternating areas of ortho- hyperkeratosis and hypergranulosis with parakeratosis and hypergranulosis. In two small foci, the epidermis showed an invagination having a column of poorly staining parakeratotic corneocytes (cornoid lamella). The granular layer was absent on the floor and the wall had hypergranulosis. A moderately dense superficial perivascular patchy lichenoid lymphocytic infiltrate was seen in the upper dermis (Figure 2).

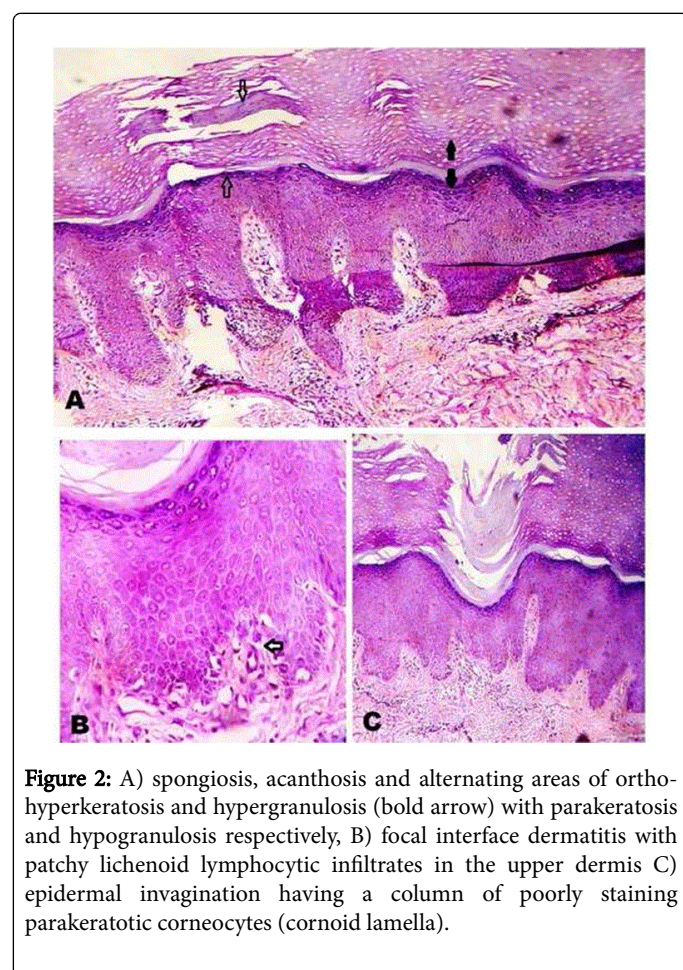


Figure 2: A) spongiosis, acanthosis and alternating areas of ortho-hyperkeratosis and hypergranulosis (bold arrow) with parakeratosis and hypogranulosis respectively, B) focal interface dermatitis with patchy lichenoid lymphocytic infiltrates in the upper dermis C) epidermal invagination having a column of poorly staining parakeratotic corneocytes (cornoid lamella).

In past few years, the child was treated with topical emollients, potent topical corticosteroid, and topical tacalcitol, but none of them appeared to be effective.

The detailed history, clinical features, histopathological findings and marked failure of response to antipsoriatic medications favored the diagnosis of ILVEN to be made and this case was considered as a 'porokeratotic variant of ILVEN'. The patient was started on oral isotretinoin therapy with topical keratolytics but he did not turn up thereafter.

Discussion

In 1896, the term 'inflammatory linear verrucous epidermal nevus' was proposed by Unna but this rare linear dermatosis was first described separately by Altman and Mehregan in 1971 [1]. Usually, it presents as markedly pruritic, variably erythematous, scaly, coalescing papules and plaques along the Blaschko's lines typically involving the single lower limb and persists for many years of life [1,2].

The aetiopathogenesis of ILVEN is still unclear but owing to its sporadic occurrence, extremely rare familial cases and resemblance to other nevoid inflammatory linear dermatoses, it was thought to be the result of an early, potentially lethal and unidentified post-zygotic dominant mutation rescued by genetic mosaicism [3]. Hypothetically in 2002, Happle has suggested that ILVEN might be a reflection of the action of a 'retrotransposon' that is partly expressed and partly silenced at an early developmental stage [4].

Based on genetic and molecular markers, there are some suggested cascade of underlying pathological changes which eventually results in the clinical manifestation and they are: 1) elevated ICAM-1, ECAM-1, HLA-DR and keratin 10 expressions; 2) increased IL-1, IL-6 and TNF- α ; 3) upregulation of involucrin expression in orthokeratotic area and decreased expression in parakeratotic area; 4) clonal dysregulation of growth of keratinocytes [5].

To differentiate from other pathogenetically and clinically related nevoid linear inflammatory dermatoses, following distinct diagnostic clinical criteria have been given by Altman and Mehregan in 1971 [1] which were later modified in 1985. Further histopathological criteria were also proposed by Dupre and Christol in 1977 to increase the diagnostic accuracy [6].

Altman and Mehregan's clinical criteria [1]

1. Early age of onset (75% of cases with onset before 5 years of age, and 50% before 6 months)
2. Predominance in women in a 4:1 proportion (omitted in the modified criteria)
3. Left side more commonly affected
4. Pruritus
5. Psoriasis-like morphology
6. Persistence and resistance to treatment

Dupre and Christol's histopathological criteria [6]

1. Psoriasiform changes - parakeratosis, acanthosis, elongation of rete ridges, thinned suprapapillary plates, tortuosity of the dermal capillaries and upper dermal lymphocytic infiltrates.
2. Sharply demarcated alternate areas of parakeratosis with agranulosis or hypogranulosis and orthokeratosis with hypergranulosis.

Following linear dermatoses should be kept as close clinical differential diagnoses: linear psoriasis, linear lichen planus, linear porokeratosis, lichen striatus and ichthyosiform nevus in CHILD syndrome [7].

All these dermatoses can be ruled out by the above-mentioned criteria. Although linear psoriasis is the closest differential but with the help of clinical criteria of ILVEN such as the very early age of onset, usually negative family history, intense pruritus and resistance to antipsoriatic medications (as noted in our case), it can be ruled out.

Furthermore, wherein the facility is available, the genetic and quantitative immunohistochemical markers can also be used such as increased keratin-10 expression and reduced Ki-67, CD-4, CD-8, CD-45RO, CD-2, CD-25, CD-94 and CD-161 positive cells in ILVEN compared to psoriasis [8]. But quantification of these markers could not be done in our case due to financial constraints and poor facility.

In our case, all the clinical and histopathological features were much more in favor of ILVEN than the rest of the differentials. The mere presence of coronoid lamella was not enough to allow reckless ignorance of all those features which were all diagnostic of ILVEN.

Similar to our case, only two such examples exist in the medical literature, one in an 18 month old girl child having linear verrucous lesions on the right half of chest along the lines of Blaschko, and another case in a 19 year old young lady with recalcitrant linear scaly plaque since her infancy. Both cases revealed coronoid lamella on HPE but considered as 'linear porokeratotic epidermal nevus' in the first case and ILVEN in the second [7,9]. Further in 1982, Su identified the porokeratotic changes in epidermal nevus and gave the term 'porokeratotic epidermal nevus' [10]. Therefore, coronoid lamellation is by no means pathognomonic of porokeratosis as it can be found in a range of inflammatory, hyperplastic and neoplastic skin conditions as described by Wade and Ackerman in 1980 [11]. Hence, the presence of coronoid lamella does not necessarily mean that the condition is primarily porokeratosis.

As our case fulfilled all the required established criteria of ILVEN and also demonstrated the presence of coronoid lamella, it was justified to consider this case as 'porokeratosis variant of ILVEN' rather than making a hasty diagnosis of linear porokeratosis.

For therapeutic point of view, ILVEN is notoriously resistant to treatment, occasionally having a fortunate course of spontaneous resolution. Potent topical corticosteroids under occlusion and intralesional injections of triamcinolone acetonide may provide symptomatic relief. Topical retinoids and vitamin D analogs (calcipotriol, tacalcitol) appear to provide either no benefit or merely reduce the itching and redness.

Systemic treatments in the form of oral administration of acitretin and etanercept have been proved effective [8,10]. Treatment with dermabrasion or cryotherapy is commonly associated with recurrence after superficial procedures and scarring following deeper procedures.

Surgical excision may be curative if done up to the appropriate depth but postoperative scarring is the obvious sequelae. Successful treatment has also been reported with the Erbium-YAG ablation laser,

585 nm flash lamp-pumped pulsed tunable dye laser in some cases [12].

Conclusion

Considering the major differences in the therapeutic approach for linear porokeratosis and ILVEN, it is of utmost importance to differentially diagnose these two conditions on the basis of detailed clinical and histopathological evaluation. Hence we have reported this extremely rare entity diagnosing as porokeratotic variant of ILVEN.

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