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Case Report

Inflammatory linear verrucous epidermal nevus: A rare experience

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Abstract Inflammatory linear verrucous epidermal nevus (ILVEN) is caused by unknown postzygotic mutation rescued by genetic mosaicism. It presents as intensely pruritic, erythematous, psoriasiform and verrucous, papules which coalesce to form plaques along the lines of Blaschko, mostly involving the single leg. It has usually early life onset. However, since its first description, very few cases have been reported which were of adult onset. Herein, we present a case of ILVEN in a 22 year old male patient.

Key words
Linear verrucous, mosaicism, psoriasiform, epidermal nevus.

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN) is type of epidermal nevus, caused by unknown postzygotic dominant mutation rescued by genetic mosaicism. It presents as intensely itchy erythematous, psoriasiform and lichenified or verrucous, coalescing linear papules and plaques on the lines of Blaschko, mostly involving the single lower extremity.[1]

Most of the cases (75%) have less than 5 years age of onset and four times common in female patients. Herein, we report case of ILVEN in 22 year male.

Case report

A 22-year-old man presented with intensely pruritic, linear papules and plaques over left leg and thigh for 15 years. It started to appear initially on the left medial malleolus then slowly extended upwards up to the thigh. None of the family members were affected. On cutaneous examination, lesions comprised of erythematous and slightly hyper pigmented, scaly papules and plaques with some verrucosity. Such lesions were seen extending from left medial malleolus to mid thigh, arranged in a linear band following the Blaschko lines **(Figure 1).**

Except pruritus, no other symptoms were noted. Examination of other skin sites such as mucosa, nails and hair were normal. Systemic examination was unremarkable and routine laboratory parameters were also within normal limits. ILVEN, nevoid linear psoriasis, linear lichen planus and linear porokeratosis were kept as differential diagnosis.

Histopathological examination was done which revealed that the skin tissue showed hyperkeratosis and epidermal acanthosis.

The dermoepidermal junction showed focal lymphocytic infiltration along with pigment incontinence.

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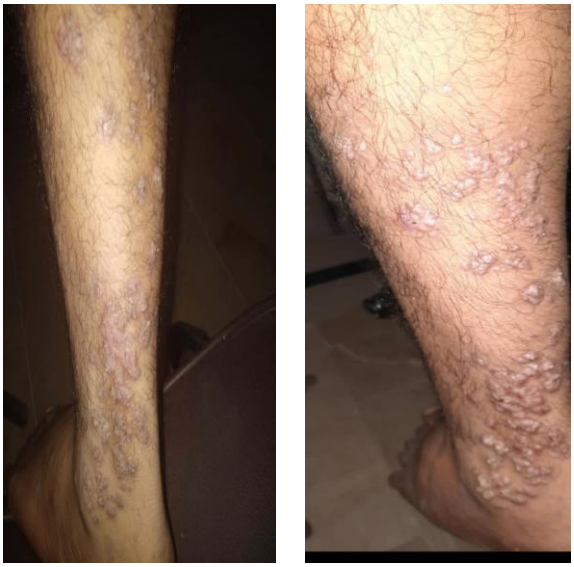

Figure 1 An erythematous and slightly hyper pigmented, scaly papules and plaques with some verrucosity.

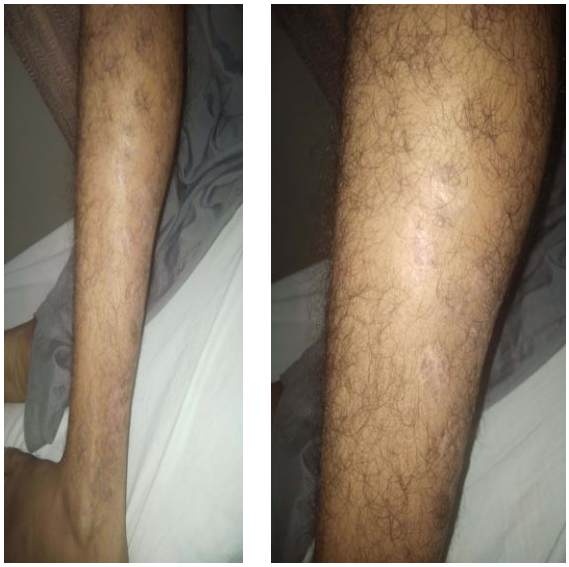

Figure 2

The clinical and histopathological features were consistent with the diagnosis of ILVEN. Therefore, it was diagnosed as a case of ILVEN in 22 year male.

The patient successfully improved on topical keratolytic agent and Intralesional steroids at a dose of 10mg per lesion at two weeks intervals for 12 weeks **(Figure 2).**

Discussion

The term "Inflammatory linear verrucous epidermal nevus" (ILVEN) was coined by Unna in 1896 but it took many decades to have its first detailed description by Altman and Mehregan in 1971 who reviewed 25 such cases and suggested 6 characteristic clinical features of ILVEN.[2]

These are: 1) Age of onset early; 2) Female involvement (4:1 F:M ratio); 3) Involvement of the lower limbs; 4) Marked itching; 5) Does not respond to therapy; and 6) Distinctive psoriasiform and inflammatory histopathology.

Thus, ILVEN was accepted as a distinct clinical and histopathologic type of epidermal nevus. ILVEN presents as intensely itchy erythematous, psoriasiform, scaly papules which coalescing to plaques on the Blaschko's lines, usually involving a single lower extremity.[2] The lesions tend to persist for many years despite treatment and may become verrucous and lichenified.

Since postzygotic mutation followed by somatic mosaicism is the underlying molecular basis of its origin, family history is usually absent.[3]

Some closely mimicking blaschko linear dermatoses must be considered as differential diagnoses of ILVEN. These are nevoid linear psoriasis, blaschko linear lichen planus, linear porokeratosis, porokeratotic eccrine ostial dermal duct nevus (PEODDN), lichen striatus and linear lichen simplex chronicus.[5]

The important salient features of such dermatoses have been summarized in a table **(Table 1)**.

Table 1 Differentiating diagnosis of ILVEN

Features	*ILVEN*	*Linear nevoid psoriasis*	*Blaschkolinear lichen planus*	*Adult Blaschkitis*	*Lichen striatus*	*PEODDN*	*Linear porokeratosis*
Suggested etiology and inheritance	Mosaicism and Sporadic	Mosaicism and Sporadic	Mosaicism and Sporadic	Mosaicism and Sporadic	Mosaicism and Sporadic	Mosaicism and Sporadic	Mosaicism and Autosomal dominant > sporadic
Age at onset	Birth or infancy	Young adults or childhood	30-40 years	Adult (mean age is 40 years)	5-15 years	Birth or early childhood	Infancy or childhood
Gender	F>M (4:1)	Not clear	M>F	M>F	M=F	M=F	M>F (2:1)
Common location	Extremity	Extremity	Extremity	Trunk	Extremity	Palm and sole	Extremity
Clinical presentation	Erythematous, psoriasiform and verrucous, coalescing papules and plaques following the lines of Blaschko	Linear band of erythematous, scaly confluent and discrete papules and plaques along the lines of Blaschko	Violaceous confluent papules and plaques arranged in a band like fashion along the line of Blaschko	Relapsing inflammatory linear eruption presenting as multiple lines of itching papules and vesicles following the Blaschko lines	Continuous or interrupted, single or multiple linear bands consisting of small, 1–2 mm flat-topped, scaly, erythematous papules along Blaschko's line	Linear punctate pits or pitted papules with comedo like plugs	Sharply demarcated hyperkeratotic grouped annular plaques with distinct keratotic edge and atrophic centre, arranged in linear fashion
Pruritus	Intense	Absent or mild	Variable	Absent	Absent	Absent or mild	Absent
Histopathology	Psoriasis like changes, and sharply demarcated alternate areas of parakeratosis with attenuated granular layer, and orthokeratosis with hypergranulosis	Classical psoriatic histopathology	Classical of lichen planus	Predominantly spongiotic dermatitis	Lichenoid interface dermatitis	Epidermal invaginations containing cornoid lamella typically involving the eccrine ducts	Classical of porokeratosis
Clinical course	Persistent with slow progression	Persistent lesions	Persistent lesions	Slow self resolution	Rapid self resolution	Persistent, may progress slowly	Persistent, may progress slowly
Malignant transformation	No	No	No	No	No	No	Yes
Therapeutic response	Poor	Variable	Variable	Good	Good	Poor	Variable

In spite of the classical description, atypical cases of ILVEN have also been reported such as having extensive/bilateral involvement, positive family history, onset in adult age and association with other cutaneous or systemic conditions. Therefore, it encouraged us to report our case here.

Being poorly responsive to antipsoriatic medications such as coal-tar, topical corticosteroids and calcipotriol, various other treatments have been tried with variable results. They are oral acitretin and etanercept, laser ablation, cryotherapy, and full-thickness surgical excision.

Our patient dramatically responded to topical keratolytic agent and Intralesional steroids at a dose of 10mg per lesion at two weeks intervals for 12 weeks **(**compare **Figure 1** and **Figure 2).**

Conclusion

ILVEN is a type of congenital epidermal nevus, present with a itchy verrucous or psoriasiform papules and coalescing to form plaques in a linear pattern mostly over lower limbs.

Proper history, clinical examination and the histopathological reports are very important to diagnose the disease.

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