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Case Report

Adult onset of inflammatory linear verrucous epidermal nevus: truly a rare experience

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Abstract Inflammatory linear verrucous epidermal nevus is an uncommon variant of epidermal nevus, thought to be caused by unknown postzygotic dominant mutation rescued by genetic mosaicism. Clinically, it presents as intensely pruritic, erythematous, psoriasiform and verrucous, papules and plaques following 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. We have been able to find less than 10 such cases globally. Herein, we report an adult case of ILVEN in a 24‑year‑old male.

Key words
Linear verrucous, mosaicism, psoriasiform, nevus, adult onset.

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN) is an uncommon variant of epidermal nevus, thought to be caused by unknown postzygotic dominant mutation rescued by genetic mosaicism. Clinically, it presents as intensely pruritic, erythematous, psoriasiform and lichenified and/ or verrucous, coalescing papules and plaques following 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 nearly all the cases manifest before reaching adulthood.[1] However, since its first description, very few cases have been reported which were of adult onset. We have been able to find only one case of ILVEN of adult onset reported in Indian literature and less than 10 cases from other parts of the world. Herein, we report another case of ILVEN of adult onset in a 24-year-old male.

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Case report

A 24-year-old man presented with intensely pruritic, linearly arranged papules and plaques over left leg and thigh for past one and half years. It started to appear initially on the medial malleolus then slowly extended upwards up to the thigh. None of the family members were affected.

On cutaneous examination, lesions were comprised of erythematous and slightly hyperpigmented, confluent papules and scaly plaques with some lichenification and verrucosity. Such lesions were seen extending from medial malleolus to mid thigh, arranged in interrupted linear band-like fashion with maximum width 2 cm, following the Blaschko lines **(Figure 1a & 1b)**. Except pruritus, no other symptoms were noted. Examination of other skin sites, mucosa, nails and hair were normal. Systemic examination was unremarkable and routine laboratory parameters were also within normal limits. ILVEN, nevoid linear psoriasis, blaschkolinear lichen planus, linear porokeratosis and adult blaschkitis were kept as differential diagnosis.

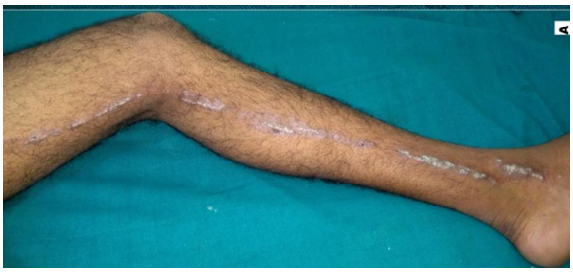

Figure 1a Erythematous and hyperpigmented scaly plaques arranged in interrupted linear band-like fashion following the lines of Blaschko

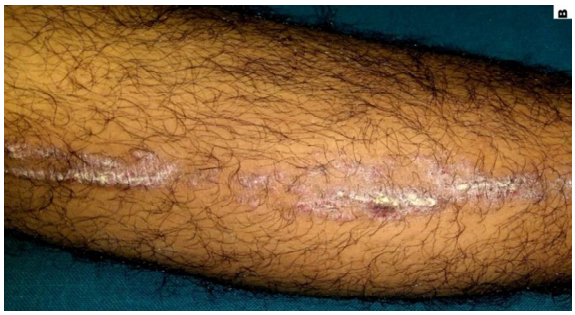

Figure 1b Psoriasiform erythematous plaques with apparent lichenification and mild verrucosity

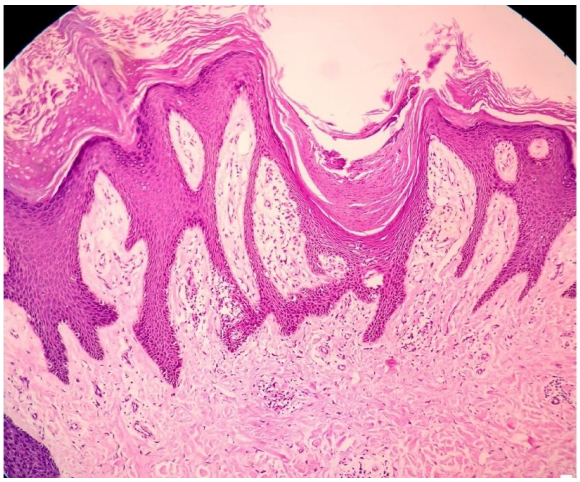

Figure 1c Psoriasiform changes along with characteristic alternating areas of 'parakeratosis with agranulosis/hypogranulosis' and 'orthokeratosis with hypergranulosis' (H& E, × 200)

To confirm, histopathological examination was done which revealed regular psoriasiform hyperplasia and mounds of parakeratosis containing small collections of neutrophils. The papillary dermis was hyperplastic with papillomatosis containing tortuous dilated capillaries. There were moderately dense superficial perivascular mixed infiltrate of lymphocytes and neutrophils with papillary dermal edema and extravasation of RBCs. Of note, agranulosis and parakeratosis alternating with hypergranulosis and orthokeratosis were present **(Figure 1c)**.

Although age of onset was atypical, clinical and histopathological features were consistent with the diagnosis of ILVEN. Therefore, it was diagnosed as a case of ILVEN of adult onset. Since there was no significant improvement with topical keratolytics, oral zinc and acitretin (25mg daily) was added and patient was advised to come for regular follow up.

Discussion

The term "Inflammatory linear verrucous epidermal nevus" 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: early age of onset; female preponderance (4:1 female-male ratio); frequent involvement of the left leg; marked pruritus; refractoriness to therapy; and a distinctive psoriasiform and inflammatory histopathology. Besides psoriasiform histology, distinctive histopathologic findings were consistently present in all the subsequently reported cases consisting of 'sharply demarcated alternate areas of parakeratosis with attenuated granular layer, and orthokeratosis with hypergranulosis'. Thus, ILVEN was accepted as a distinct clinical and histopathologic variant of epidermal nevus.

ILVEN classically presents as intensely pruritic, variably erythematous, psoriasiform, scaly, coalescing papules and plaques along the Blaschko's lines, usually involving the single lower extremity.[2] The lesions tend to persist for many years despite of 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 blaschkolinear dermatoses must be considered as differential diagnoses of ILVEN. These are nevoid linear psoriasis, blaschkolinear lichen planus, adult blaschkitis, linear porokeratosis, porokeratotic eccrine ostial dermal duct nevus (PEODDN), lichen striatus and linear lichen simplex chronicus.[5] All except linear psoriasis can be easily ruled out by characteristic clinical and histopathologic features of ILVEN. The important salient features of such dermatoses have been summarized in a table **(Table 1)**. Considering linear nevoid psoriasis, although its true existence is very disputed, it does have clinical and histological resemblance. ILVEN differs from linear nevoid psoriasis by very early age of onset, presence of intense pruritus, and very poor response to anti-psoriatic treatments. Moreover, in two-third cases of linear psoriasis, other sites are also involved. Quantitative molecular and immunohistochemical markers can also be used to differentiate these two entities.[2]

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.[4] As our case was of adult onset, we searched literature. It was interesting to find only 9 cases of ILVEN with onset in adult age (more than 18 years) among which only one case was reported in Indian literature. Therefore, it encouraged us to report our case here. All such previously reported cases are summarized in a table **(Table 2)**.[1,5-12] 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.[4]

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

Table 2 Clinical profile of previous cases of ILVEN of Adult onset

Serial number	*Author*	*Year*	*Age of Onset (years)*	*Age at presentation (years)*	*Gender*	*Family History*	*Site*
01	Altman and Mehregan[1]	1971	49	Not mentioned	Not mentioned	Absent	Not mentioned
02	Toribio and Quinones[6]	1975	20	35	Female	Absent	Left lower leg
03	Hodge et al[7]	1978	28	29	Female	Absent	Left groin
04	de Jong et al[8]	1991	24	37	Male	Absent	Leg
05 &	Goldman and Don[9]	1994	56	56	Female	Mother	Neck and chest
06			33	33	Female	Daughter	Face, trunk and both limbs
07	Kawaguchi et al[10]	1999	43	44	Male	Absent	Left whole leg
08	Kosann[11]	2003	19	23	Male	Absent	Right arm
09	Vukicevic et al[12]	2011	48	68	Female	Absent	Right side of face, neck, trunk, upper and lower limb

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