

Inflammatory linear verrucous epidermal nevus syndrome

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Sir,

A 26-year-old male patient, who was a born of a nonconsanguineous marriage came to our outpatient clinic with a lifelong history of pruritic red, scaly lesions over left side of his body, left upper and lower limbs. His past medical and family history were unremarkable except the fact that he had been prescribed for skin lesions for several times without improvement. Upon dermatological examination, we observed multiple erythematous scaly papules coalescing into linear plaques following the lines of Blaschko on the left side of his trunk, medial aspect of his left arm and elbow, also medial aspect of his left leg down to the plantar region (Figs 1-3). The scale was obviously adherent and silvery at some points while semi-adherent in other regions (Fig. 3). It was also noticed that on the plantar region the lesion was more likely a verrucous plaque with an irregular serpiginous outline and an erythematous border. Clinical assessment of the patient implied that he had neuropsychological problems particularly mental retardation. However, the patient did not accept neither any laboratory and medical imaging procedures, nor the neurological consultation. Histopathological examination of the lesional skin biopsy revealed hyperkeratosis, acanthosis and elongation of the rete ridges with mild spongiosis in the epidermis (Fig. 4). Although we could not perform further evaluation, based on clinical and histopathological findings we made a diagnosis of ILVEN syndrome and prescribed topical keratolytics and topical corticosteroids.

Prior to the study, patient gave written consent to the

examination and biopsy after having been informed about the procedure.

Inflammatory linear verrucous epidermal nevus (ILVEN), is a type of epidermal nevus which is characterized by psoriasiform papules coalescing to form linear plaques following the lines of Blaschko. It is fourfold more likely to occur in females than males and although occasional isolated adult cases have been described, ILVEN most commonly appears during the first five years of life [1-3]. The etiology of ILVEN is obscure. Many hypotheses brought forward including that ILVEN is nothing but a mosaic form of psoriasis [4], and that ILVEN reflects the action of a retrotransposon which is actually a transposable DNA element that is partly expressed and partly silenced at an early developmental stage [5]. The histopathological resemblance of ILVEN to psoriasis is striking. Indeed, ILVEN appears as psoriasiform papules and plaques in a Blaschko-linear distribution. Lesions are generally on a limb, most frequently left lower extremity, although rare bilateral and widespread involvements have been described. Moreover, since CHILD syndrome (congenital hemidysplasia with ichthyosiform erythroderma and limb defects) is characterized by unilateral erythematous verrucous lesions, infrequent reports of limb reduction in association with ILVEN have prompted the idea that ILVEN represents a forme fruste of CHILD syndrome [2,3].

On the other hand, ILVEN has been considered as a specific group within the epidermal nevi and since in about one third of cases with epidermal

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Figure 1: Erythematous papules coalescing into linear plaques following the lines of Blaschko



Figure 3: Blaschkoid papules in a closer view



Figure 2: Psoriasiform papules and plaques in a Blaschko-linear distribution

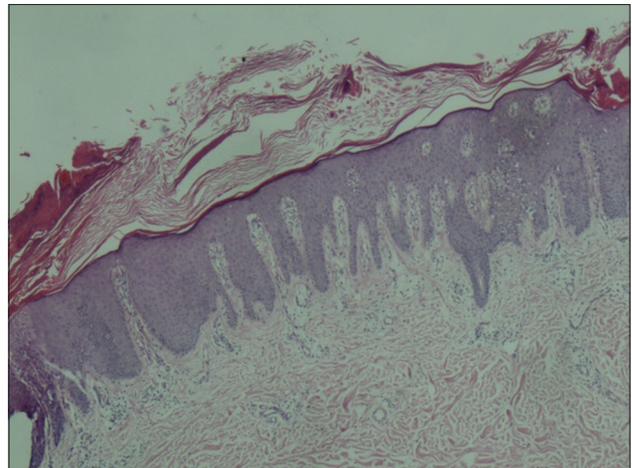


Figure 4: Histopathological findings, (H&Ex40)

nevi have extracutaneous manifestations, ILVEN may display other organ defects and present as a component of an epidermal nevus syndrome [6-8]. Cerebrovascular malformations, skeletal abnormalities, visceral anomalies of eye, heart and kidney have been reported in patients with ILVEN syndrome. However, CNS complications are the most common extracutaneous manifestation and among the other neurological abnormalities like hydrocephalus, cortical atrophy hemiplegia, cranial nerve palsies, intracerebral calcification and hemimegalencephaly, mental retardation is the most common one as in our case [1,6,8,9].

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

REFERENCES

1. Ruggieri M, Sugarman JL. Inflammatory Linear Verrucous Epidermal Nevus (Ilven). In: Ruggieri M, Pascual-Castroviejo I, Rocco CD, eds. Neurocutaneous Disorders Phakomatoses and Hamartoneoplastic Syndromes. 1st ed. Germany: Springer-Verlag/Wien; 2008. p. 575-580.
2. Moss C, Shahidullah H. Naevi and other Developmental Defects. In: Burns T, Breathnach S, Cox N, Grittiths C, eds. Rook's Textbook of Dermatology. 8th ed. Oxford: Wiley—Blackwell; 2010. p.18.19-21.
3. Khachemoune A, Janjua SA, Guldbakke KK. Inflammatory linear verrucous epidermal nevus: a case report and short review of the literature. *Cutis*. 2006;78:261-7.
4. Sengupta S, Das JK, Gangopadhyay A. Naevoid Psoriasis and ILVEN: Same Coin, Two Faces? *Indian J Dermatol*. 2012;57:489-91.
5. Happle R. Transposable elements and the lines of Blaschko: a new perspective. *Dermatology*. 2002;204:4-7.
6. Kumar CA, Yeluri G, Raghav N. Inflammatory linear verrucous epidermal nevus syndrome with its polymorphic presentation – A rare case report. *Contemp Clin Dent*. 2012;3:119-22.
7. Rogers M, McCrossin I, Commens C. Epidermal nevi and the epidermal nevus syndrome. A review of 131 cases. *J Am Acad Dermatol*. 1989;20:476-88.
8. Golitz LE, Weston WL. Inflammatory linear verrucous epidermal

- nevus. Association with epidermal nevus syndrome. Arch Dermatol. 1979;115:1208-9.
9. Surve TY, Muranjan MN, Deshmukh CT, Warke CS, Bharucha BA. Inflammatory linear verrucous epidermal nevus syndrome with bilateral vertebral artery occlusion. Indian Pediatr. 1999;36:820-3.

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