

Dermoscopy of an Inflammatory Linear Verrucous Epidermal Nevus (ILVEN)

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Image Article

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Abstract

Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare form of epidermal nevus characterized by intensely pruritic inflammatory papules coalescing into verrucous plaques with Blaschko linear distribution. Various therapeutic modalities have been described, but no one therapy has been successful consistently. We report a case of a 1 and half year-old infant with ILVEN with special concern to its dermoscopic features.

Keywords: ILVEN; Inflammatory Linear Verrucous Epidermal Nevus; Verrucous Nevus; Dermoscopy

Case Report

A 1 and half year-old Moroccan infant presented with a large painless erythematous and squamous papillary lesion that was present since the age of 2 months. Detailed medical history revealed no consanguineous marriage of the parents nor similar cases in the family or any pathological antecedent. The lesion was first small, slightly squamous and itchy, with heat-aggravated pruritus, sitting in the right forearm, then gradually increased in size extending to the ipsilateral arm and shoulder. Dermatological examination showed two erythematous large and well demarcated patches, of about 3.5 cm each, with keratotic and verrucous surface and areas of hemorrhagic crusts, resulting of the confluence of multiples plaques and papules [Figure 1(A)].

These lesions were associated to linear and yellowish less infiltrated and painless macules, all having a blaschkoid and linear distribution, at the posterior face of the right upper limb arriving to the right shoulder [Figure 1(B)]. Dermoscopic examination revealed filiform areas

with irregular linear vessels, more accentuated at the periphery of the lesions, on an erythematous background, along with some large brown circles at the center [Figure 2]. In association to yellowish homogenous areas [Figure 3]. Due to the presence of multiple unilateral wart-like patchy plates, the diagnosis of ILVEN syndrome was made. Histological examination was delayed according to the parents' wishes, because of his young age. The patient was then evaluated twice a year to assess the course of the disease, which revealed a stationary evolution.

Discussion

Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare form of epidermal nevus [1]. It usually appears at birth or within the first five years of life, although an adult onset has been described too [2]. It's usually a sporadic disease, even if familial cases have been reported [3]. Clinically, it is characterized by intensely pruritic, erythematous, inflammatory papules coalescing into well-demarcated verrucous plaques [1]. The latter's usually occur on a limb following Blaschko lines, more

rarely on the trunk, in a curvilinear pattern [2]. Various therapeutic modalities have been described, but no one therapy has been successful consistently [3]. Surgical excision of lesions is not preferable due to extensive scarring and relapse of disease [1].

This type of epidermal nevus is rare and sometimes misdiagnosed. Moreover, there are few publications concerning its dermoscopic aspects. Hence the importance of reporting all cases and describing the different dermoscopic aspects encountered.



Figure 1: Clinical image: (A) : two erythematous large well demarcated patches, made of the confluence of multiple keratotic papules, of about 3.5 cm each, with keratotic and verrucous surface and areas of hemorrhagic crusts. (B): linear and yellowish macules of blaszkoid distribution, on right upper limb arriving to the right shoulder.

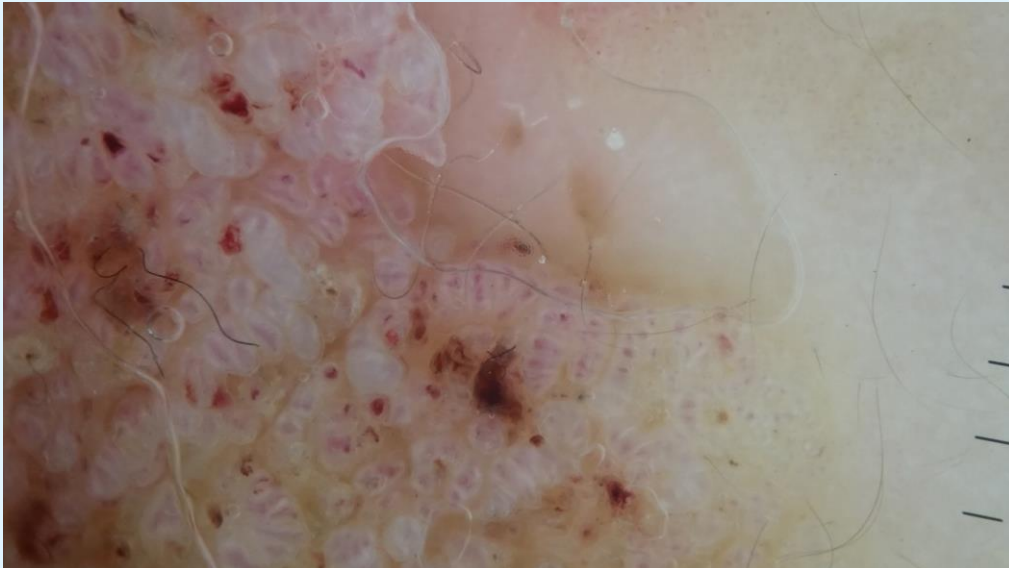


Figure 2: Dermoscopic image showing filiform areas with irregular linear vessels, on an erythematous background, in association to some large brown circles at the center.

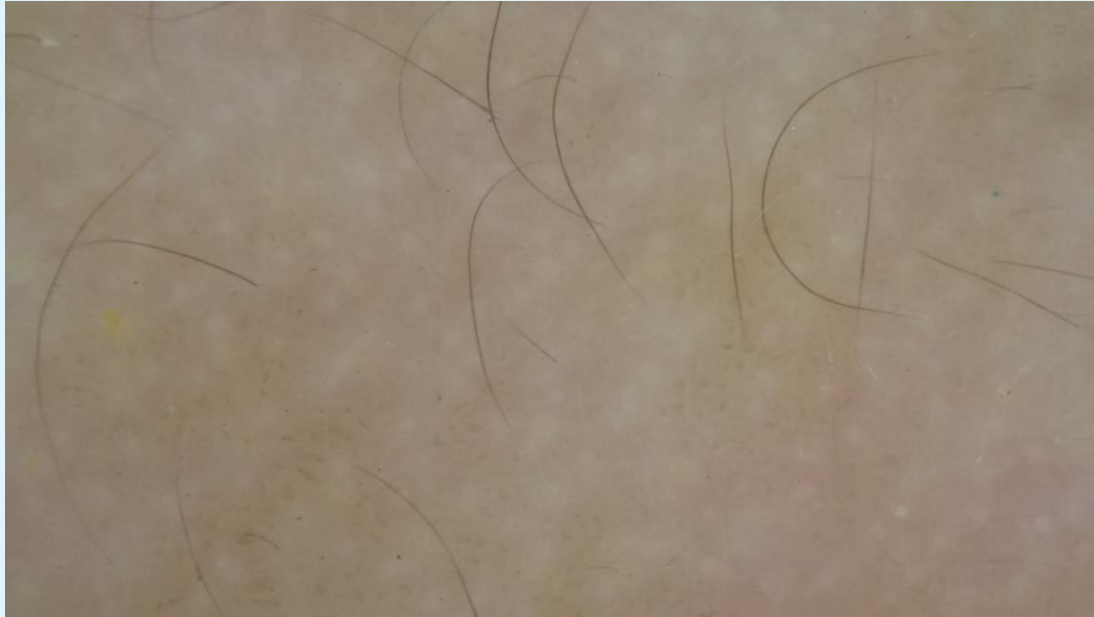


Figure 3: Dermoscopic image showing yellowish homogenous well demarcated rounded areas.

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