

# WHAT IS YOUR DIAGNOSIS?

## Case for diagnosis

### Caso para diagnóstico

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#### HISTORY OF THE DISEASE

Female patient, 15 months old, white, has presented from birth erythematous lesions, of linear or grouped distribution, involving trunk and lower extremities bilaterally. Lesions were extremely pruriginous and aggravated with heat. There were no pathological antecedents or previous family history of the disease. Dermatologic examination revealed papular lesions and erythematous plaques with areas of scal-

ing and crusts, linearly distributed in the lower limbs, also affecting the inguinal region, buttocks, abdomen, and both sides of the thorax (Figures 1-3). Biopsy of the lesion showed irregular acanthosis, hyperparakeratosis, hyperpigmentation of the basement layer with discreet perivascular lymphocytic infiltrate in the papillary dermis, absence of vesicles in the epidermis and of melanosis in the dermis (Figure 4).



FIGURE 1: Erythematous lesions of linear distribution in the hips and legs



FIGURE 2: Linear lesions in the left leg (the most affected site)



FIGURE 3: Details of lesions close to the knee

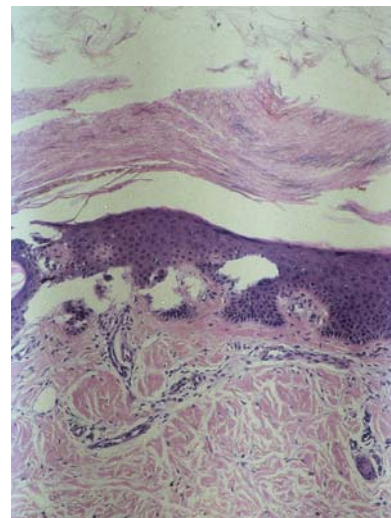


FIGURE 4: Skin with acanthosis, hyperparakeratosis and discreet perivascular lymphocytic infiltrate in the papillary dermis (HE, 10x)

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## COMMENTARIES

Epidermal nevi are congenital hamartomas of embryonal ectodermal origin classified on the basis of their main component, which can be sebaceous, apocrine, eccrine, follicular or keratinocytic. About 5% of epidermal nevi are represented by inflammatory linear verrucous epidermal nevus (ILVEN).

ILVEN is a variant of verrucous epidermal nevus and is characterized by recurrent inflammatory phenomena with chronic eczematous or psoriasiform aspect. Clinically, it presents with erythematous and verrucous papules with intense pruritus and linear distribution following Blaschko's lines.<sup>1</sup> Although cases of bilateral involvement have been described, the disease is often unilateral and localized in one extremity. Oddly, the left leg is more affected. Children are more commonly affected and it predominates in females in the ratio of 4:1. Most cases are sporadic, although familial cases have been reported.<sup>2</sup>

ILVEN is caused by somatic mutations that result in genetic mosaicism and, although its pathophysiology is still unclear, it is believed that it may be associated with an increase in the production of interleukins 1 and 6, tumoral necrosis factor- $\alpha$  and intercellular adhesion molecule 1.<sup>3</sup>

ILVEN is diagnosed by clinical and histological examination. The classical clinical criteria for diagno-

sis, suggested by Altman and Mehregan in 1971 and later modified by Morag and Metzker in 1985, are: early age of onset, predominance in females, frequent involvement of the left leg, pruritus, distinctive psoriasiform appearance and marked refractoriness to therapy.<sup>4,5</sup> Histological alterations of ILVEN were described by Dupre & Christol.<sup>5</sup> Specific signs include alternation between orthokeratosis and parakeratosis and the presence or absence of the granular layer, although the latter are not pathognomonic. Other microscopic findings include: papillomatosis, acanthosis, lymphocytic dermal infiltration or even Munro's microabscesses, but these are unspecific markers.

The differential diagnosis of ILVEN must be done with various dermatoses, such as other epidermal nevi, linear psoriasis,<sup>6</sup> and lichen striatus.

ILVEN is markedly refractory to therapy. There are reports of the use of many alternatives in its control: topical glucocorticoids applied under occlusion, intralesional corticosteroid, combination of tretinoin 0.1% and fluorouracil 5%; anthralin, tar, vitamin D<sub>3</sub> analogues, surgical excision, cryotherapy with liquid nitrogen, and laser therapy with carbon dioxide.<sup>3</sup> However, no research has shown consistent results about the superiority of any one of these therapies. □

**Resumo:** Nevo epidérmico verrucoso inflamatório linear (Nevil) é uma variante do nevo epidérmico verrucoso que acomete mais comumente o sexo feminino. Clinicamente, é caracterizado por fenômenos inflamatórios recorrentes, conferindo aspecto de dermatite eczematosa crônica ou psoriasiforme, frequentemente unilateral, com prurido intenso, de aparecimento desde o nascimento e de difícil tratamento.

Palavras-chave: Diagnóstico; Nevo; Psoríase

**Abstract:** Inflammatory linear verrucous epidermal nevus (Ilven) is a rare variant of epidermal verrucous nevus that commonly affects females. Clinically is characterized by the appearance, since birth, of recurrent inflammatory phenomena with chronic eczematous or psoriasiform aspects, usually unilateral, with severe pruritus, and refractory to therapy.

Keywords: Diagnosis; Nevus; Psoriasis

## REFERENCES

1. Simonart T, Heenen M. Inflammatory verrucous epidermal naevus presenting as a psoriasiform plaque. *Dermatology*. 2007;215:167-8.
2. Pierson D, Bandel C, Ehrig T, Cockerell C J. Benign etidermal tumor and proliferations. In: Bologna JL, Jorizzo JL, Rapini RP. *Dermatology*. London: Mosby; 2003. p. 1710-11.
3. Ulkur E, Celikoz B, Yuksel F, Karagoz H. Carbon dioxide laser therapy for an inflammatory linear verrucous epidermal nevus: a case report. *Aesthetic Plast Surg*. 2004;28:428-30.
4. Schwartz RA, Jozwiak S. Epidermal nevus syndrome. [cited 2009 Jul 2]. Available from: <http://emedicine.medscape.com/article/1117506-overview>.
5. Renner R, Rytter M, Sticherling M. Acitretin treatment of a systematized inflammatory linear verrucous epidermal naevus. *Acta Derm Venereol*. 2005;85:348-50.
6. Romiti R, Maragno L, Arnone M, Takahashi MDF. Psoríase na infância e na adolescência. *An Bras Dermatol*. 2009;84:9-22.

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