Inflammatory linear verrucous epidermal nevus and differential diagnosis with linear psoriasis: about a case

Maria Isabel Muniz Zemero[1], Maria Amélia Lopes dos Santos[1], Alena Margareth Darwich Mendes[1], Carla Andrea Avelar Pires[1],

ABSTRACT

Inflammatory Linear Verrucous Epidermal Nevus (ILVEN) is a rare clinical variety of verrucous epidermal nevus that manifests in early childhood as inflammatory lesions of keratosis surface, which coalesce and spread in band, following Blaschko lines. It makes a differential diagnosis with Linear Psoriasis; it is difficult to differentiate them given the clinical and histopathological aspects common to both, emphasizing the need to know the specific characteristics of each. The purpose of this report is to demonstrate a relatively rare affection, expressed in a 5-year-old girl, evolving from the first days of life with papulokeratosic plaques arranged linearly, followed by inflammatory signs and eroded areas, placed linearly, overtaking labia majora, perineum, the inner and upper face of the left thigh. Also, the patient showed keratotic papules and plaques in the posterior cervical region and external lateral border of the left foot plant, ascending along the posterior region of this limb. The clinical and histopathological criteria corroborate the diagnosis of ILVEN in differentiation with linear psoriasis, emphasizing the importance of establishing criteria/instruments to assist in distinguishing these two dermatoses in order to expedite the diagnosis, to optimize the treatment and minimize patients' discomfort. Long-term follow-up of patients with this disease is suggested due to the possibility, albeit minimal, of ILVEN malignancy.

Keywords: Skin diseases; Nevus; Psoriasis; Genitals; Diagnosis.

1. Universidade do Estado do Pará; Belém, (PA), Brasil.

Este é um artigo publicado em acesso aberto (Open Access) sob a licença Creative Commons Attribution, que permite uso, distribuição e reprodução em qualquer meio, sem restrições, desde que o trabalho original seja corretamente citado.

https://doi.org/10.11606/issn.2176-7262.rmrp.2021.176014

INTRODUCTION

Inflammatory Linear Verrucous Epidermal Nevus (ILVEN) is a rare clinical variety of verrucous epidermal nevus that manifests in early childhood[1]. It is an uncommon dermatosis, of unknown etiopathogenesis, probably due to somatic mutations that result in genetic mosaicism, possibly linked to an increase in the production of type 1 and type 6 interleukins and the tumor necrosis factor (TNF) Alpha[2].

The condition most frequently affects the female gender (proportion 4:1), with persistent lesions, which are expressed by elevated plaques with a keratotic surface, accompanied by inflammation and itching, which converge in a linear distribution, following the "Blaschko lines", in general, segmental, with a predilection for the lower limbs, genital involvement being uncommon[3].

Several treatments have been proposed, especially ablative lasers such as carbon dioxide (CO2) and erbium laser: yttrium-aluminum garnet (Er: YAG), but with little clinical resolution[4].

ILVEN is a dermatosis of unwieldy clinical management and difficult diagnostic confirmation, as it has several aspects similar to linear psoriasis. Beacuse of the medical literature and the epidemiological, clinical and histopathological findings, the most consistent diagnosis for the present case was ILVEN. Thus, this report aimed to demonstrate this relatively rare condition and the importance of its differentiation with linear psoriasis, speeding up the diagnosis, optimizing the treatment and minimizing the discomfort for these patients.

CASE DESCRIPTION

Girl, phototype II, 5 years old, presents papulokeratosic plaques from the first days of life, accompanied by inflammatory signs, taking up the labia majora, perineum and inner and upper face of the left thigh (Figure 01). Also keratotic papules and plaques in the posterior cervical region and external lateral border of the left foot's sole (Figures 02 and 03), ascending along the posterior region of this limb. Intense itching and burning referred by the mother, with worsening in the genital region during urination.

In family history, a deceased father with similar lesions.

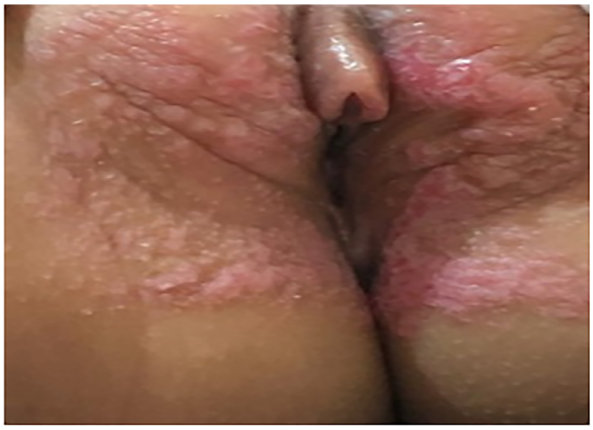

Figure 01: papulokeratosic plaques taking up the labia majora, perineum and internal and upper face of the left thigh.

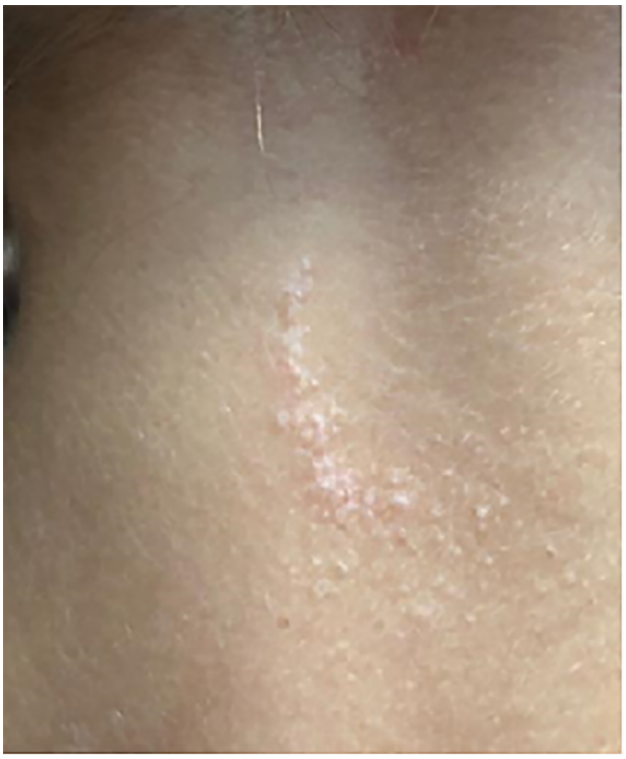

Figure 02: papulokeratosic plaques on the nape of the neck

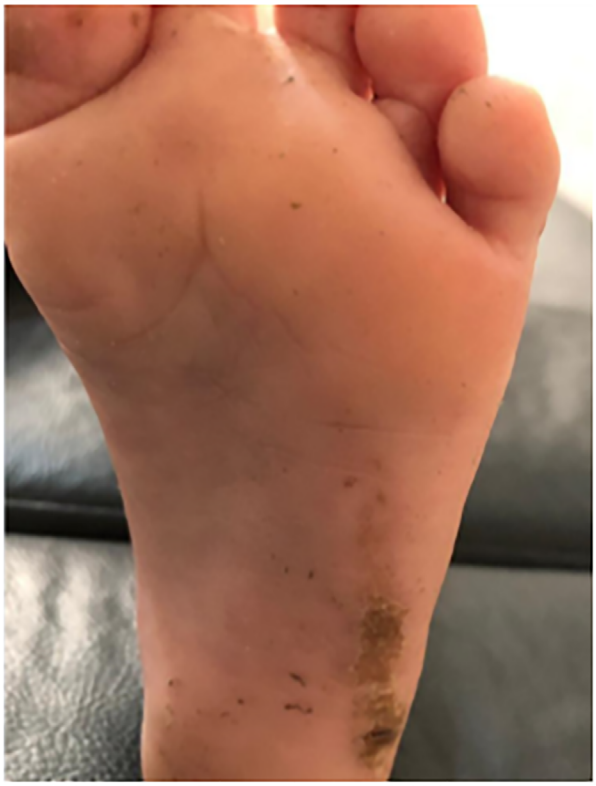

Figure 03: irregular papulokeratosic plaques on the outer lateral edge of the left foot

An incisional skin biopsy was performed on the external genitalia whose histopathological examination (Figure 04 and Figure 05) showed epidermis with hyperkeratosis, hypergranulose, irregular acanthosis, elongation of epithelial ridges, and a dermal inflammatory infiltrate, compatible with ILVEN.

It was prescribed topical treatment with hydrocortisone, desonide and clobetasol propionate associated with 3% salicylic acid, 5% dexpanthenol, which resulted in the reduction of keratosis and inflammatory signs and in the symptom control.

DISCUSSION

ILVEN is a rare condition, consisting of hyperplasia of normal epidermal components. It mainly affects the female sex at the early life stages, being rare to appear in adulthood. Clinically characterized by persistent lesions, erythematous

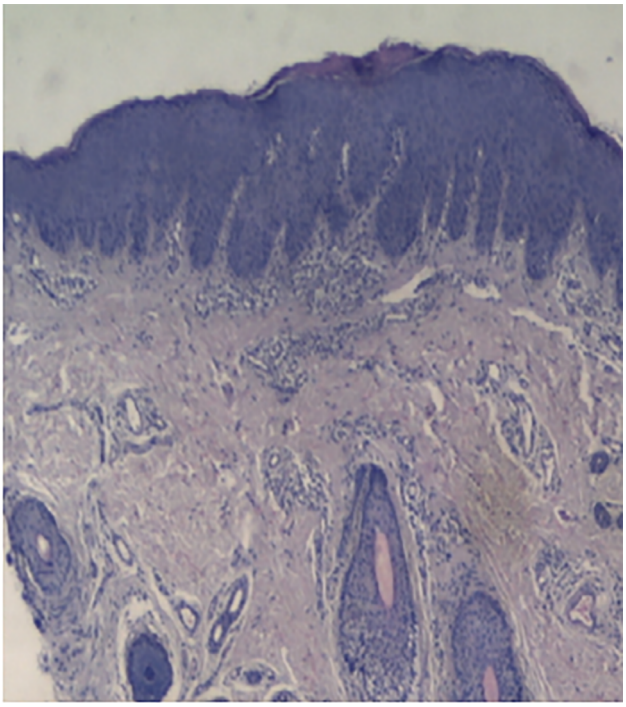

Figure 04: epidermis with hyperkeratosis, hypergranulose, irregular acanthosis, elongation of epithelial ridges and dermal inflammatory infiltrate (HE, 4x)

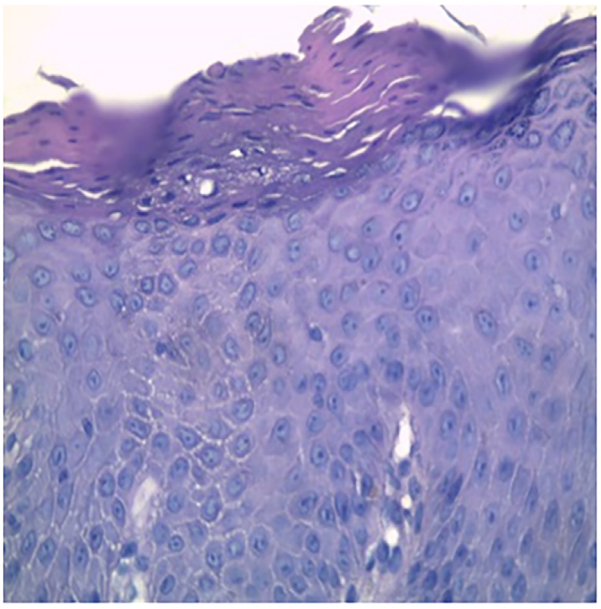

Figure 05: hyperkeratosis, hypergranulose, irregular acanthosis and elongation of epithelial ridges in greater (HE, 40x)

or grayish-brown papules with a keratotic and/or scaly surface, accompanied by inflammatory signs and pruritus, which converge and are distributed in a band, usually segmental, with a predilection for the lower limbs, rarely affecting the genital region[5].

Linear psoriasis (LP) is the main differential diagnosis of ILVEN. Differentiation between the two is difficult, due to both clinical and histopathological similarities, which highlights the need for further studies on the subject, in order to identify specific aspects of each one, which is fundamental for a better clinical follow-up and an adequate therapeutic approach, since for ILVEN there is the possibility of surgical treatment [6,7].

Some authors list criteria that favor the diagnosis of ILVEN in relation to PL; they are: inflammatory keratotic papule lesions arranged in a band; intense itching; appearance in early childhood; therapeutic refractoriness, and histopathology with psoriasiform aspect[8]. The studied case fulfills the above criteria: compatible clinical, linearly distributed in the lower left segment, appearing in the first months of life, resistant to treatment; in addition to the fact that the clinical feature, over the child's 5 years lifetime, is limited to the areas of initial involvement, which is uncommon in psoriasis, whose lesions tend to change in pattern, and generalization and evolution in remission burst and clinical worsening.

Proposed clinical and pathological criteria currently fail when there is a significant degree of overlap between ILVEN and linear psoriasis. ILVEN findings can be observed in psoriasis such as: papillomatosis, acanthosis, and chronic perivascular and dermal lymphomonocytic infiltrate, on the other hand, Munro's microabscesses, considered as characteristic criteria of psoriasis, can be seen in ILVEN and other pathologies[9].

There is a feature that has been reported in the medical literature to distinguish both, Involucrine Studies mentions that the pattern of involucrin expression in the epidermis is a very well-established tool as an adjunct in the differential diagnosis of ILVEN and linear psoriasis, despite the limited access[10]. According to Ginarte et al, as it represents a marker of epidermal differentiation, it acts as one of the first corneified envelope protein precursors of different manifestations in the two entities. In ILVEN, involucrin is elevated in orthokeratotic regions and deficient in areas of parakeratosis. In psoriasis, most suprabasal keratinocytes express involucrin, findings that distinguish the two diseases[9]. In addition, other markers such as ki-67, elastin and anti-antikeratin 10 also seem to be useful in differentiating these two entities[11].

Histopathologically, the reported case was compatible with ILVEN, however, the pattern of involucrin expression in the epidermis, as well as the test of other markers mentioned above, were not performed due to the difficulty of accessing them routinely.

Several treatment modalities have been proposed for ILVEN: topical corticosteroids under occlusion or intralesional combined or not with 0.1% tretinoin, 5% fluororacil, anthralin, vitamin D analogues, cryotherapy with liquid nitrogen, surgical excision, laser of carbon dioxide (CO2) or erbium garnet of aluminum yttrium (Erbium yag), however the disease tends to be refractory[9]. Due to the child's age and difficulty accessing laser therapy, we chose to use steroids associated with salicylic acid and dexpanthenol, with little clinical response.

Clinical and evolutionary disease characteristics bring physical and psycho-emotional repercussions, negatively affecting the life quality of patients[12]. The risk of malignancy, although low, has been mentioned for this condition[8].

In conclusion, this report aimed to disclose a little-known dermatosis called ILVEN, emphasizing an unusual location and focusing on analyzing clinical and laboratory criteria that make its diagnosis possible, regarding the differential with linear psoriasis. Due to the possibility, albeit minimal, of malignant ILVEN, long-term monitoring of patients with this disease is necessary.

REFERENCES

1. Trejo-Acuña JR, Ramos-Garibay A, Rosas-Cano M, Hernández-Ramírez H, Chávez-Alonso J. Nevo epidérmico verrugoso inflamatorio lineal. Comunicación de un caso de aparición tardía. Dermatología Revista Mexicana. 2015;59(6):570-5.

2. Gon AdS, Minelli L, Franzon PGU. Case for diagnosis. Anais brasileiros de dermatologia. 2010;85(5):729-31.

3. Burgos LG, Di Martino Ortiz B, Valdovinos G, Masi MR, Knopfelmacher O, de Lezcano LB. Nevus epidérmico lineal con hiperqueratosis epidermolítica. Descripción de caso. Pediatría (Asunción): Organo Oficial de la Sociedad Paraguaya de Pediatría. 2010;37(3):195-8.

4.Osman MAR, Kassab AN. Carbon dioxide laser versus erbium: YAG laser in treatment of epidermal verrucous nevus: a comparative randomized clinical study. Journal of Dermatological Treatment. 2017;28(5):452-7.

5.Tanita K, Fujimura T, Sato Y, Lyu C, Aiba S. Widely spread unilateral inflammatory linear verrucous epidermal nevus (ILVEN). Case reports in dermatology. 2018;10(2):169-74.

6.Saifaldeen RH, Fatani MI, Baltow B, Khan AS. Successful Treatment of Inflammatory Linear Verrucous Epidermal Nevus with Concomitant Psoriasis Using Etanercept. Case reports in dermatology. 2018;10(1):29-34.

7.Gianfaldoni S, Tchernev G, Gianfaldoni R, Wollina U, Lotti T. A case of "inflammatory linear verrucous epidermal nevus"(ILVEN) treated with CO2 laser ablation. Open access Macedonian journal of medical sciences. 2017;5(4):454.

8.Morrison JE, Bonham A. Massive Vulvar Linear Verrucous Epidermal Nevus Presenting at Menarche. Journal of pediatric and adolescent gynecology. 2017;30(3):429-30.

9 .Paixão MP, Machado CDA, Ito LM, Enokihara MMS. Psoríase pustulosa linear X Nevil: relato de caso. Anais brasileiros de dermatologia. 2005;80(6):607-10.

10.Ferreira FR, Chiacchio NGD, Alvarenga MLd, Mandelbaum SH. Involucrin in the differential diagnosis between linear psoriasis and inflammatory linear verrucous epidermal nevus: a report of one case. Anais brasileiros de dermatologia. 2013;88(4):604-7.

11 . Vissers W, Muys L, Van Erp P, De Jong E, Van de Kerkhof P. Immunohistochemical differentiation between inflammatory linear verrucous epidermal nevus (ILVEN) and psoriasis. European Journal of Dermatology. 2004;14(4):216-20.

12.Nakai N, Ohshita A, Katoh N. A case of inflammatory linear verrucous epidermal nevus on the upper eyelid. Indian journal of dermatology. 2015;60(3):323.

Conflicts of Interest:
Nothing to declare..

Financial support:
The present study was financed by the researchers.

Research Ethics Committee:
This study was sent to the Research Ethics Committee and has an approval number: 36631520.1.0000.5174.

Corresponding Author:
Maria Amélia Lopes dos Santos
dermatoamelia@gmail.com

Editor:
Prof. Dr. Felipe Villela Gomes

Received in: oct 22, 2020
Approved in: may 07, 2021