Linear pustular psoriasis X ILVEN - Case report*

*Psoríase pustulosa linear X Nevil - Relato de caso**

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Abstract: Linear pustular psoriasis (LPP) and inflamamatory linear verrucous epidermal nevus (ILVEN) possess overlapping clinical and histopathological characteristics, which emphasizes the importance of the attempt to distinguish between these two pathologies, based on clinical and ancillary laboratory data. Here we report the case of a 24-year-old woman, who presented with erythematous-desquamative, pruritic lesions, with a linear unilateral distribution (left side of the body). Clinical examination and histopathological studies suggested the diagnosis of linear pustular psoriasis, with one year of evolution, and resistant to steroid therapy.
Keywords: Immunohistochemistry; Nevus; Psoriasis

***Resumo**: A psoríase pustulosa linear (PPL) e o nevo epidérmico verrucoso inflamatório linear (Nevil) possuem características clínicas e histopatológicas semelhantes, o que enfatiza a importância da tentativa de distinção entre essas duas patologias a partir de aspectos clínico e laboratorial auxiliares. Relata-se o caso de uma mulher com 24 anos, que apresentou lesões eritêmato-descamativas pruriginosas, com distribuição linear unilateral (lado esquerdo do corpo). O exame clínico e os estudos histopatológicos sugeriram o diagnóstico de psoríase pustulosa linear, com um ano de evolução e resistente à corticoterapia.*
Palavras-chave: Imuno-histoquímica; Nevo; Psoríase

INTRODUCTION

Linear pustular psoriasis (LPP) is accepted by some to be a distinct entity from inflamamatory linear verrucous epidermal nevus (ILVEN).[1-3] Others question its existence, considering it either as ILVEN or isomorphic effects of preexisting epidermal nevi.[4] LPP is described as having rare occurrence.[1,2]

Distinction between LPP and ILVEN is described as difficult in the literature, whether it is made clinically or on a histopathological basis, even claimed, by some authors, to be impossible relying solely on these two resources. Immunohistochemestry may have utility in the differentiation between the two entities.[5,6]

CASE REPORT

Twenty four-year-old female patient, who had been presenting with erythematous-desquamative, pruritic lesions on the left axilla for a year, being medicated with topic steroids and antifungals, with no improvements. Lesions progressed to hemithorax, abdomen and left upper limb, associated with intense pruritus, with occasional pustules, never going beyond the midline. There was a significant worsening of the clinical picture when treated with 20 mg of prednisone a day. Then she came to the service, presenting, on dermatological examination, erythematous-desquamative plaques with central pustules in the internal face of the arm, axilla, breast and

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abdomen, on the left, with occasional linear distribution, variable sizes, occasionally confluent, with temperature increase and pain on the lesions (Figures 1A and 1B).

The following complementary exams were performed: complete blood count 12.900 leukocytes/mm^3 (no left shift) and hemosedimentation rate = 40 mm/h (NV= 0 to 20 mm/h), with all the others – ANF, stool, urine I, calcium, sodium, potassium, BUN, creatinin, blood glucose, β-hCG, ALT, AST, gamma-GT, AP, PT, ASLO, bacilloscopy e pustular secretion culture – with no alterations.

Patient is now being followed up in an outpatient setting, having used dapsone 100 mg/day for the past two months, obtaining gradual improvement of the lesions, albeit with persistence of the linearly disposed lesions (Figures 2 and 3). The option was made for maintenance of this therapeutic regimen for yet a few months, with clinical and laboratorial follow-up. In case the linear lesions persist, new therapeutic proposals will be implemented with the consent of the patient.

Histopathological findings of biopsies carried out from the left breast and axilla were compatible with pustular psoriasis (Figures 4 and 5).

DISCUSSION

In 1986, the term inflamamatory linear verrucous epidermal nevus (ILVEN) was proposed by Unna.[7] From there, reports of linear inflammatory pruritic verrucous lesions followed.[8,9] In 1971, Altman & Mehregan[4] studied 25 cases compatible with the diagnosis of ILVEN and proposed clinical criteria (Table 1). The psoriasis-like histopathologic aspect described by this authors was revised by Dupré & Christol,[10] identifying specific and non-specific histopathological features. As specific features, they described: alternation between hypergranulosis associated with orthokeratotic areas and agranolosis associated with parakeratotic areas. As non-specific features the described: papillomatosis, acanthosis, chronic perivascular and dermal lymphomonocytic infiltrate. These latter findings can also be seen in psoriasis. Unfortunately, proposed clinical and anatomopathological criteria fail to meet situations in which there is an important overlapping of ILVEN and LPP. ILVEN findings can be found in psiriasis; on the other hand, Munro's microabcesses, considered as being characteristic of psoriasis, can occasionally be seen in ILVEN and other pathologies.[6]

Hence, new attempts have been implemented in the search for diagnostic ellucidation. Adrian and Baden,[11] by means of electrophoretic analysis of epidermal proteins in lauryl-polyacrylamide sulfate gel (SLS-PAGE), demonstrated differences in the electrophoretic patterns of patients with hard-to-diagnose psoriasis. Later on, using this same principle, Bernhard et al.[3] demonstrated, in four ILVEN cases, distinct patterns from those found in normal and psoriatic skin. When the electrophoretic patterns of the ILVEN patients were analyzed, no single characteristic pattern was found, demonstrating that there may be case-to-case variations.

In an immunohistochemical study made by Jong et al.,[6] differences were also observed between ILVEN and psoriasis. Such differences were demonstrated by using specific markers directed against elastase, present in polymorphonuclear cells; against keratin 16 present in hyperproliferating epidermis; against keratin 13 absent from adult human skin; and

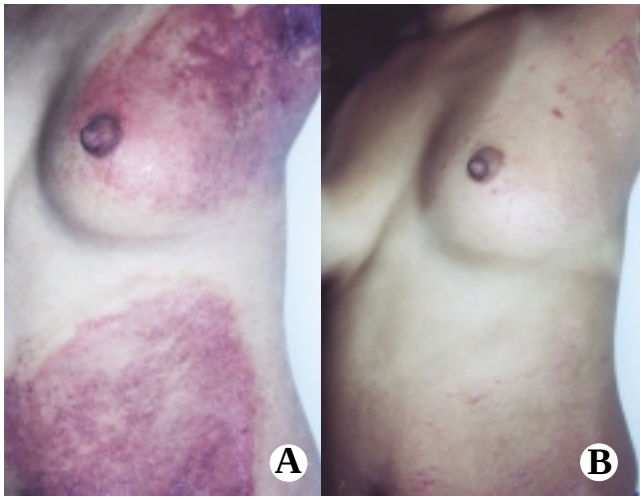

FIGURE 1: (A) Erythematous-desquamative plaques with innumerous pustules, restricted to the left sid of the body (B) To the left, linear lesions extending from the mammary region to the arm

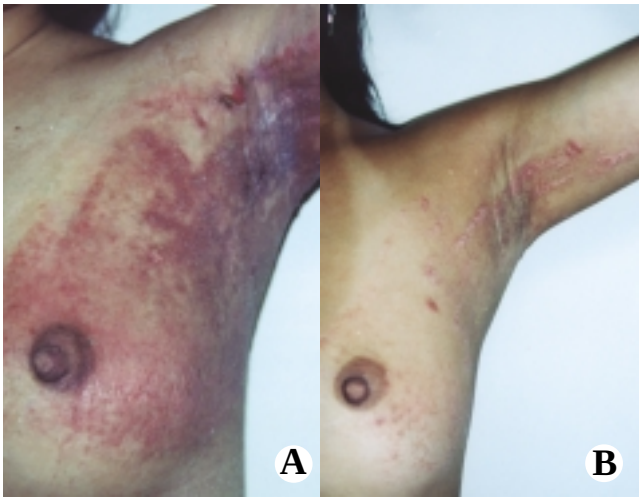

FIGURE 2: Improvement of lesions after introduction of dapsone

against keratin 10 present in differentiated epidermal cells. In ILVEN a relative decrease in polymorphonuclear neutrophyls, a focal pattern in anti-keratin 16 and homogenous distribution of anti-keratin 10 were observed. In psoriasis, the occurrence of elastase-positive cells was more significant, with a homogenous pattern of anti-keratin 16 and a focal or diminished pattern of anti-keratin 10 thus demonstrating immunhistochemical differences between these two entities. The authors even discussed the possibility that lack of response to antipsoriatic treatment delivered to ILVEN patients is due to the relative absence of polymorphonuclear neutrophyls found in the latter.

Another more recent immunohistochemical study, made by Ginarte et al.,[5] using involucrin, because it is a marker for epidermal differentiation, being one of the first proteic precursors of the hornified envelope, serving as a possible auxiliary method for the distinction between ILVEN and LPP. In the normal skin, involucrin is present in the superior portion of the epidermis, including granular layer, whereas in psoriasis it is detected in deeper levels, i.e., suprabasal. In ILVEN, invlucrine is increased in orthokeratotic regions, but lacks in regions of parakeratosis, unlike what is seen in psoriatic parakeratosis, in which most suprabasal keratonocytes express involucrin.[5,12,13] Differential diagnosis between ILVEN and LPP is very important, for it allows a better definition of therapeutics. While in LPP a certain degree of response is described, in ILVEN the rule is persistence of lesions and resistance to the most diverse treatment modalities. [4]

A possible explanation for the linear arrangement found in psoriasis has been suggested by Happle,[14] who proposed that linearity is due to the phenomenon of crossing over (chromosomal recombination) during an early phase of embryogenesis, with exchange of sequential genetic material in somatic cells, allowing a genetic rearrangement that, in its turn, predisposes to psoriasis, In that context, cutaneous cells which derive from this altered precursor could present a pattern of embryologic development that would follow a linear arrangement, by a mechanism similar to that of Blaschko's lines.[14] Moreover, understanding psoriasis as a poligenetically-influenced disease, for its expression to occur it is fundamental that other unknown factors be present, including possible environmental determinants. By proposing this concept, the author suggests that it may serve as a feasible explnation for the following features found in linear psoriasis: (I) non-heredity; (II) linear distribution patte correspondent to many other observed mosaic disorders; (III) possibility of coexistence of common and linear forms; and (IV) this would be the reason why psoriatic lesions are much more prominent in the linear rather than in the common form.[15]

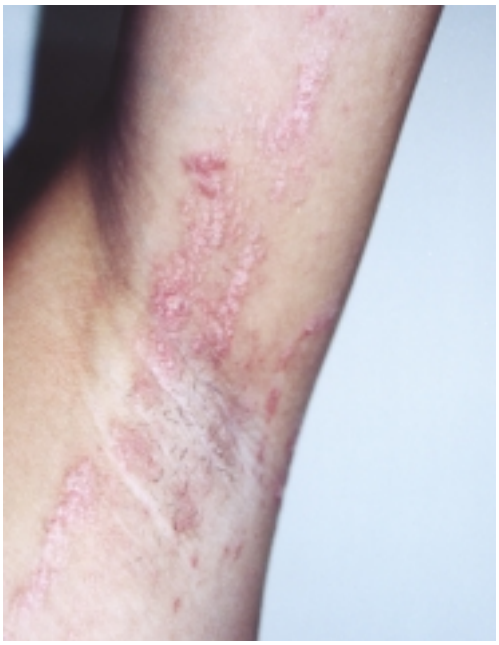

FIGURE 3: In greater detail, zoster-like aspect of plaques on left axilla

In the case described here, components favoring diagnosis of LPP were: patient age, unilateral and preferentially left side distribution, absence of previous nevic lesions, present of other non-linear pustules, typical histological findings of pustular psoarisis in the linear lesions, worsening with oral steroid therapy and considerable improvement with the use of dapsone.

Given the considerations here approached, all based in previous studies, immunhistochemical examination, if proper markers were easily accessible, was considered as an important complementary resource for differntial diagnosis between ILVEN and LPP. For this case, major laboratories in the country,

CHART 1: ILVEN diagnostic criteria according to Altman and Mehregan

1. Early age of onset (75% of cases with onset before 5 years of age, and 50% before 6 months)
2. Predominance in women in a 4:1 proportion
3. Left side more commonly affected
4. Pruritus
5. Psoriasis-like aspect
6. Persistence and resistance to treatment

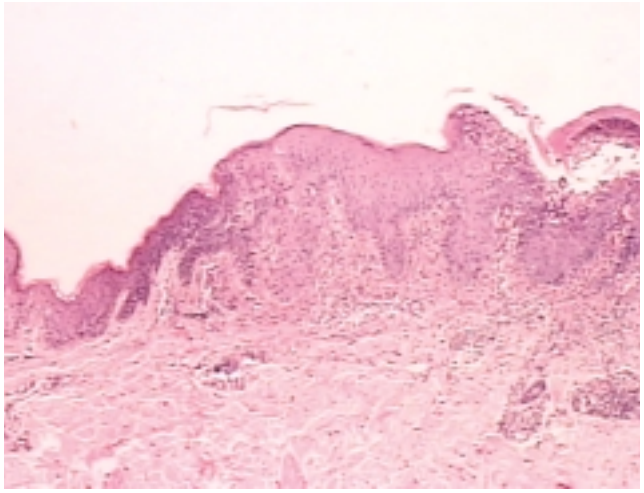

FIGURE 4: Psoarisis-like dermatitis with Kogoj's spongiform pustule (HE, 40x)

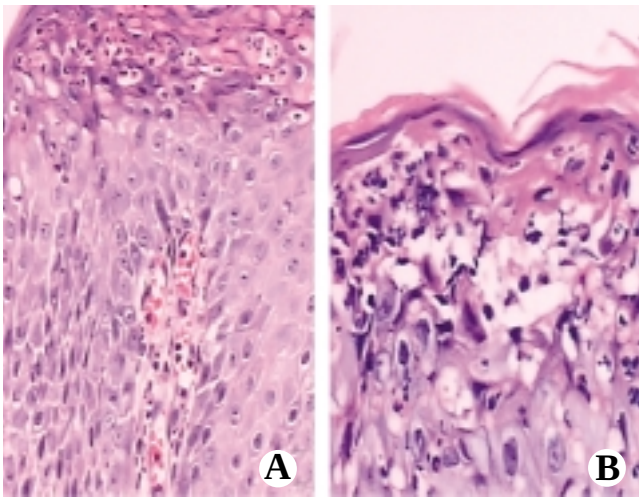

FIGURE 5: (A) Psoarisis-like dermatitis with Kogoj's spongiform pustule with polymorphonuclear neutrophyls in the midst of keratinocytes in the epidermal surface. Dilated and tortuous capillaries are seen touching papillary cupola. (HE, 100x) (B) Detail of Kogoj's spongiform pustule (HE, 400x)

public or private (these latter also have agreements with foreign laboratories), linked or not to teaching institutions, were contacted in order to attempt to perform an immunohistochemical study. Pathologists who were responsible for immunohistochemistry in these laboratories informed us that they did not have the specific markers. The costly acquisition of these markers, as we were informed, did not justify the purchase as an attempt to elucidate a single case.

Yet, the clinical presentation and the proper summary of new diagnostic methods have been judged to supersede the impossibility of carrying out an immunhistochemical exam. The preparation of this report is justified by the rarity of the diagnosis and by the brief literature review regarding auxiliary diagnostic possibilities. ❑

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