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Líquen Estriado Seguido de Líquen Plano Clássico: Estados dum Espectro de Doença?

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RESUMO – O líquen plano clássico e o líquen estriado representam duas patologias dentro do grupo das dermatoses liquenóides com características clinicas e histológicas bem estabelecidas. No entanto, a possibilidade de uma correlação entre elas tem sido proposto. Os autores apresentam o caso de uma doente do sexo feminino de 38 anos com diagnóstico consecutivo de líquen estriado e líquen plano clássico com apenas três meses de diferença. De facto alguns autores levantam a questão se existe uma forma intermédia entre estas duas patologias ou se representam extremos opostos da mesma doença. No entanto estas questões apenas serão resolvidas quando se identificar por completo a fisiopatologia destas doenças.
PALAVRAS-CHAVE – Dermatopatias Papuloescamosas; Líquen Plano.

Lichen Striatus Followed by Classical Lichen Planus: States of a Disease Spectrum?

ABSTRACT – *Classical lichen planus and lichen striatus represent separate entities within the group of lichenoid dermatoses with clinical and histologic well-defined differences. A possible relationship between both has been proposed. We present a case of a 38-year-old woman with a consecutive diagnosis of lichen striatus followed by classical lichen planus within three months. Some authors have questioned whether there is an intermediate form between these two entities or if they represent different states or opposite ends of the same disease spectrum. These questions are unlikely to be resolved until the pathophysiology of these two diseases is clarified.*
KEYWORDS – *Lichen Planus; Skin Diseases, Papulosquamous.*

INTRODUCTION

Lichen planus (LP) is an inflammatory dermatosis with an immunological pathogenesis that is not yet fully understood. Its classical form is characterized by pruritic, violaceous, polygonal, flat-topped papules and plaques distributed along the anterior aspects of the forearms, lower back and ankles.[1] A linear distribution is seen in linear lichen planus (LLP). It is usually unilateral, follows Blaschko lines and is a rare form of LP. The differential diagnosis of LLP includes other dermatoses following the Blaschko lines such as lichen striatus and linear lichen nitidus.[2,3]

On the other hand, lichen striatus (LS) is an asymptomatic dermatosis that generally occurs in childhood. It consists of small, scaly, erythematous papules distributed in a linear band along the Blaschko lines, usually unilaterally distributed on a limb.[4]

LP and LS represent separate entities within the group of lichenoid dermatoses. The simultaneous occurrence of lichen striatus and lichen planus is extremely rare and the resemblance of one dermatosis to the other is (in some instances) so pronounced that a differential diagnosis, based on clinical manifestations, is difficult.[5]

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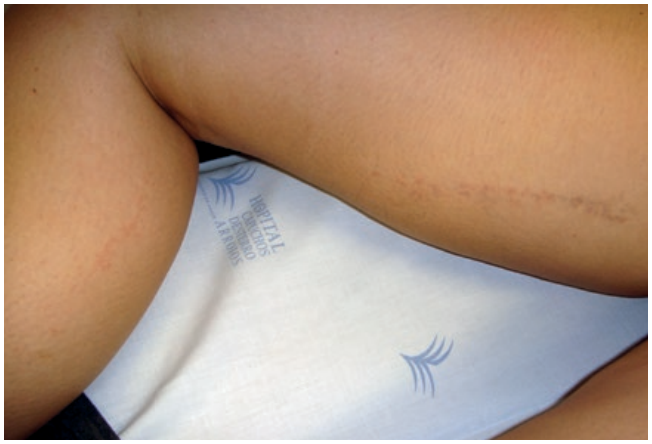


Figure 1 - Lichen striatus with erythematous papules in a linear distribution, extending from the right thigh to inner ankle.

CASE REPORT

A 38-year-old Caucasian, otherwise healthy, female patient was observed for a linear eruption that had been present for four months on the right lower limb. Examination showed scaly erythematous papules in a strikingly linear distribution, extending from the lateral aspect of the right thigh down to the inner aspect of her right ankle. No other skin or mucous lesions were found and she had no pruritus or other clinical symptoms (Fig. 1). Lesions were clinically suggestive of LS and a skin biopsy confirmed this diagnosis: an interface lymphocytic infiltrate beneath a basal layer with vacuolar degeneration, and a deep perifollicular and glandular infiltrate were observed (Fig. 2). The patient was treated with topical corticosteroids (betamethasone 0.5% once daily). When she returned, three months later, the eruption on the right lower limb had undergone partial involution, leaving post-inflammatory hyperpigmented macules. However, new lesions occurred and spread irregularly over the limbs and trunk and she complained of significant pruritus. The lesions were shiny violaceous erythematous grouped papules with a similar lichenoid appearance (Figs.3 and 4). The skin biopsy showed a band-like lymphocytic dermal infiltrate and the diagnosis of LP was established (Fig. 5). The patient initiated prednisone 0.75 mg/kg/day, with no response after two weeks. Absence of response with corticosteroid therapy prompted start of cyclosporine 5mg/kg/day with complete remission in three months. After one year of follow-up, the patient remains asymptomatic.

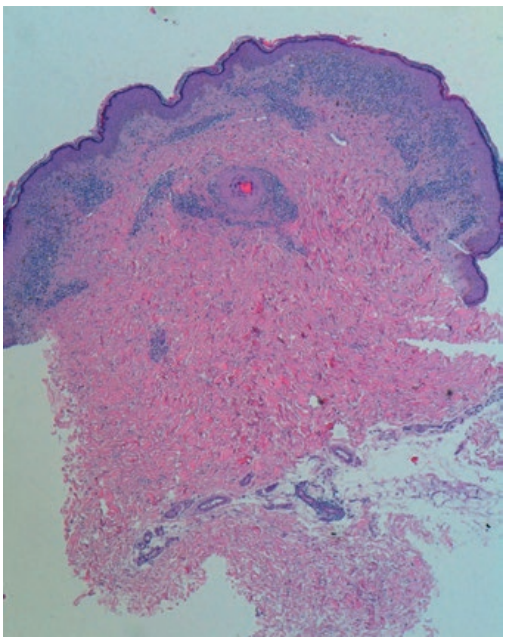
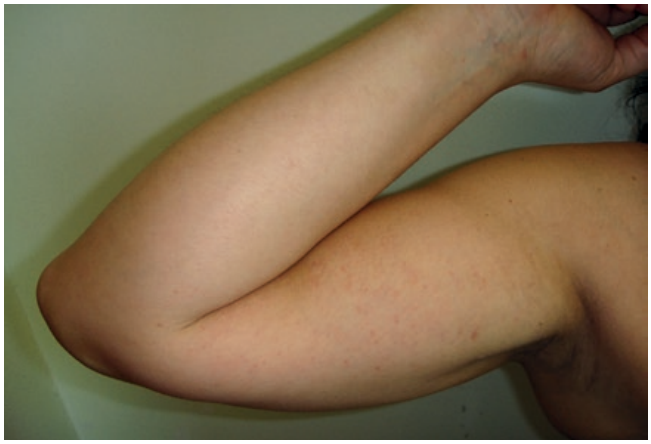


Figure 2 - A lichenoid reaction pattern spanning two adjacent dermal papillae with a deep perifollicular and glandular infiltrate (H&E, x40).



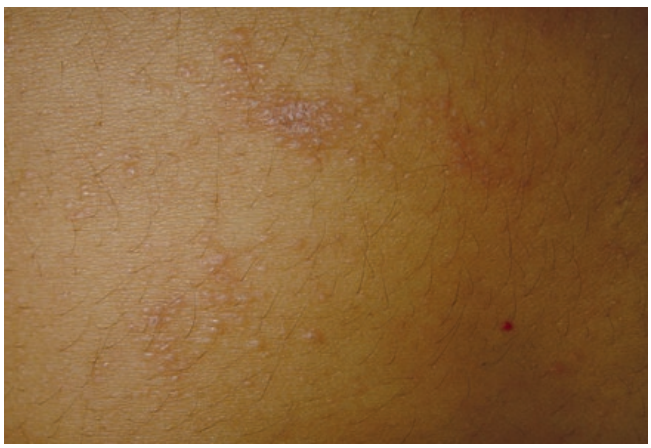


Figure 3 and 4 - Lichen planus with shiny violaceous erythematous grouped papules with a lichenoid appearance haphazardly distributed over the upper limbs (3), and lumbar area (4).

DISCUSSION

LP and LS represent clinically well-defined entities although in occasional cases the differential diagnosis is not really clear.[1] Generally, LS shows a linear distribution along Blaschko lines, while classical LP usually presents as papules or plaques with a typical distribution.[2-4] When LP appears with a linear

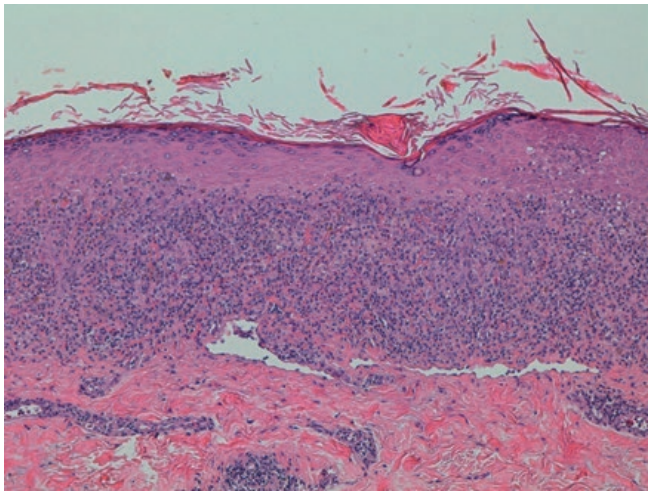


Figure 5 - LP histology with a band-like lymphocytic dermal infiltrate (H&E, x100).

distribution, the differential diagnosis between LLP and LS involves very subtle clinical distinctions and for this reason histology is necessary.[1]

Histologically, the most relevant feature in LS is the presence of a dense perivascular lymphohistiocytic infiltrate in the upper dermis and a deep perifollicular and glandular infiltrate. LP as the prototypical lichenoid dermatosis is, on the other hand, characterized by a band-like inflammatory dermal infiltrate with colloid bodies in the upper dermis and lowermost epidermis. However, the histologic variability of LS is considerable and the findings may sometimes be indistinguishable from those of LP.[6]

The pathophysiology of LS is not completely understood but environmental agents have been implicated, particularly virus.[4]

Clinical observations have also suggested a relationship between exposure to a number of exogenous agents and the development of LP. However, despite the similar pathophysiology, is still unclear the reason for clinical and histological differences between these two entities.[5]

Our patient initially presented with a linear lichenoid dermatosis clinically and histologically suggestive of LS. A few months later she had a generalized lichenoid dermatosis with similar lesions, yet the clinical distribution and histology were more suggestive of LP.

In the literature we found only four case reports that discuss the simultaneous or consecutive presence of LP and LS.[1,5,7,8]

Pulgar *et al* described a case of a 7-year-old girl, with a clinical and histological diagnosis of LS, that months later developed a generalized eruption that was compatible with LP.[1] Finkus described a case of simultaneous occurrence of LS and LP in a 37-year-old man. The LS lesions were located on his left leg and the LP lesions were on both sides of the abdomen and on the shaft of the penis. These two diagnoses were confirmed histologically.[5] These two cases had clinical and histologic characteristics of both LP and LS which is very uncommon. For this reason the authors consider that these cases support the hypothesis that LP and LS represent the opposite ends of the same disease spectrum.

Herd *et al* described the case of a 37-year-old man with an asymptomatic and spontaneously resolving linear eruption in his upper and lower left limbs. Histologically, the condition was consistent with LP and, in addition, showed occasional plasma cells in the upper dermal inflammatory infiltrate, which has been described in LS.[7] Rubio *et al* described a similar case of a 29-year-old woman with an asymptomatic and spontaneously resolving linear eruption that left residual hyperpigmentation. Histology was more consistent with the microscopic diagnosis of LP.[8] These two cases raise some unsolved diagnostic questions since several findings are compatible with LS, while others suggest a diagnosis of LLP. The clinical course, with an absence of itching and spontaneous clearance within a short period is suggestive of LS. However, the histopathological pattern strongly supports the diagnosis of LLP. With all these elements these authors suggest that some patients may have an intermediate form between these two entities.[7,8]

The overlap between LP and LS is very rare and only few cases have been reported in the literature. These cases prompt the question whether distinction between these two diseases is justified or whether they merely represent different states of a same disease spectrum. This question is unlikely to be resolved until the fundamental basis of both disease processes is elucidated.

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