Case for diagnosis*

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CASE REPORT

A 60-year-old woman reports a 5-year history of violaceous and intensely pruritic lesions on the dorsum and scalp, associated with a 2-year history of hair loss. She also reports decreased hair growth in the axillary and inguinal regions in the same period.

Dermatological examination shows small, scaly, erythematous-violaceous, flat papules on the dorsal region; multifocal scarring alopecia areas, with smooth, bright and atrophic surface; discrete hair rarefaction in the axillary and inguinal regions; presence of longitudinal grooves and some depressions on the surface of the nail plate; no oral lesions (Figures 1 and 2). The histopathology of the dorsal lesion is shown in figure 3A and that of the scalp is shown in figure 3B.

The treatment was performed using high-potency corticoids and resulted, after three months, in an improvement of pruritus and a slight lightening of the lesions.

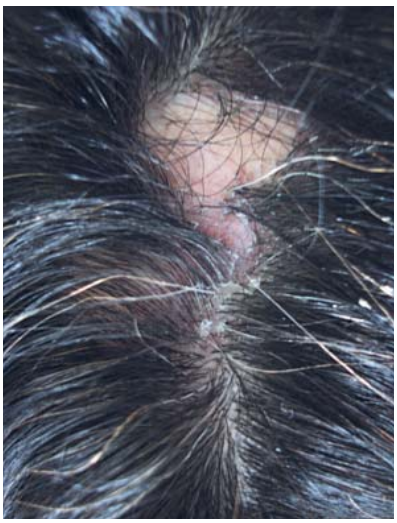

FIGURE 2: Perifollicular erythema with desquamation at the vertex of the scalp; cicatricial alopecia and smooth, bright and atrophic surface

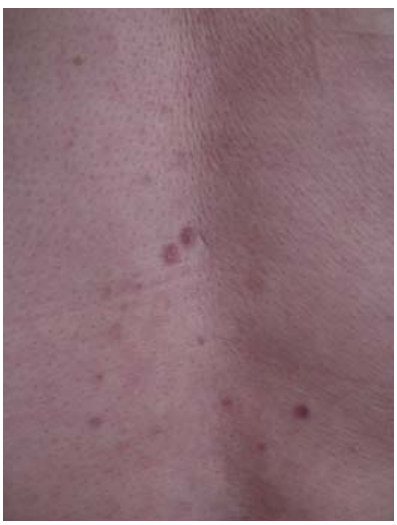

FIGURE 1: Cutaneous, erythematous-purpuric lesions on the dorsal region

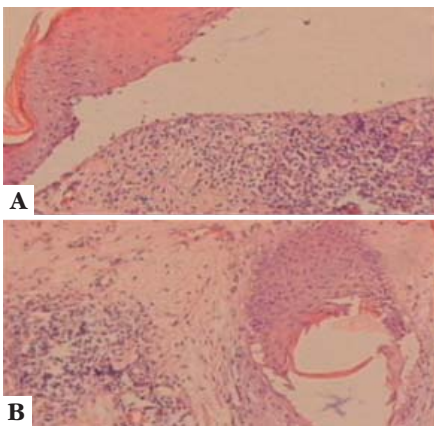

FIGURE 3: A. HE 200x. Interface dermatitis with lichenoid pattern associated with dermo-epidermic detachment and lymphocytic infiltrate in band-like pattern in the upper dermis. **B.** HE 200x. Detail of partially destroyed follicle, with perifollicular fibrosis and perivascular lymphocytic infiltrate

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DISCUSSION

Graham-Little-Piccardi-Lassueur (GLPL) syndrome is a rare form of Lichen Planus (LP), characterized by the triad: non-scarring hair loss in the inguinal and axillary regions, and follicular spinous or disseminated acuminate papules; typical, cutaneous or mucous LP; and scarring alopecia of the scalp with or without atrophy. These features do not have to be present simultaneously.[1-3]

Its etiopathogenesis is attributed to emotional factors, endocrine disorders, infections, neurological disorders, enzymatic changes, drug use and immune changes. However, its etiology remains unknown.[4,5] There is a growing body of evidence that the dermatosis represents an autoimmune damage mediated by T cells against basal keratinocytes expressing autoantigens altered on their surface.[1,4] Particularly in the case of GLPL, studies have shown the role of inner centromere proteins (INCENP) functioning as autoimmunity stimulating antigens; and Antigenic Mimicry or scattering of epitopes to stimulate a transition from LP to an ulcerative, exanthematous or other rare forms of the disease.[6-,9]

Diagnosis is clinical and histopathological. Histological findings, however, are not always specific and conclusive. The most characteristic consists of interface dermatitis with vacuolar degeneration of the basal cell layer and perivascular lymphocytic inflammatory infiltrate in the middle dermis. The striking pigmentary incontinence favors the diagnosis of LP. In the **LP of the hair,** from the early stages, an inflammatory infiltrate appears around the hair follicles, especially in their upper half; and the follicular destruction occurs at a late stage.[2,4,9]

Differential diagnosis should be made with other causes of acquired scarring alopecia, such as pseudopelade of Brocq, discoid lupus erythematosus, sarcoidosis, follicular mucinosis, folliculitis decalvans, atrophic keratosis pilaris, graft-versus-host disease, eosinophilic cellulitis, dermatomyositis, lichen sclerosus et atrophicus, scleroderma, mastocytosis, pyoderma gangrenosum, necrobiosis lipoidica and cicatricial pemphigoid.[3,5]

The classic treatment is done with the use of systemic and topical corticosteroids, retinoids, phototherapy, tacrolimus, hydroxychloroquine and cyclosporine, with partial and temporary results.[3,8] ❑

Abstract: Graham-Little-Piccardi-Lassueur Syndrome is a rare form of Lichen Planus, characterized by the presence of the triad: non-scarring hair loss in the inguinal and axillary regions and follicular spinous or disseminated acuminate papules; typical, cutaneous or mucous LP; and scarring alopecia of the scalp with or without atrophy. These features do not have to be present simultaneously.
Keywords: Alopecia; Dermatitis; Lichen planus

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