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# Graham-Little-Piccardi-Lasseur Syndrome: A Case Report

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## ABSTRACT

Graham-Little-Piccardi-Lasseur syndrome is a rare dermatosis that affects the hair follicles throughout the body and often presents with a progressive cicatricial alopecia of the scalp that is unresponsive to medical therapy. While treatment options are limited, prompt recognition through a careful physical exam aided by dermoscopy can facilitate early intervention. Here we present a patient with GLPLS, discuss pertinent morphologic and dermoscopic findings, and review the current literature.

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## INTRODUCTION

Graham-Little-Piccardi-Lasseur Syndrome (GLPLS) is a rare clinical subtype of lichen planopilaris (LPP) that manifests as a triad of scarring alopecia of the scalp, nonscarring alopecia of the axillary and the pubic skin, and widespread lichenoid follicular papules.[1] GLPLS more commonly affects women (male-to-female ratio ≃ 1:4), with the classic patient being a middle-aged Caucasian woman.[2] While GLPLS is considered to be a manifestation of an immunologic disorder, predisposing factors are unclear and may be multifactorial. However immunosuppressive therapy confers limited and variable benefit, underscoring a longstanding need for therapeutic advances. The typical clinical course features progressive, scarring alopecia that may cause significant psychological distress to the patient.

## CASE

A 58-year-old woman presented to the outpatient dermatology clinic with a tender and severely pruritic scalp skin eruption that began a few weeks before presentation. She described initially noticing welt-like lesions on her neck and face, along with a burning sensation in her scalp accompanied by hair loss. A dermoscopic exam revealed numerous peripilar casts and erythema on the scalp and hyperkeratotic erythematous follicular papules throughout the lumbosacral area, abdomen, and thighs (Figures 1, 2). Perifollicular erythema was seen on the axillae. A scalp punch biopsy revealed a perifollicular lichenoid lymphocytic infiltrate with fibrosis and a decreased number of terminal anagen hair follicles, some of which lacked sebaceous gland lobules. An abdominal punch biopsy revealed a perifollicular lymphocytic infiltrate with interface changes, hypergranulosis, and follicular plugging (Figure 3). A colloidal iron stain failed to reveal increased mucin deposition, and a PAS-D stain showed no evidence of basement membrane thickening or fungal infection. Overall, the clinical and histopathological findings were consistent with a diagnosis of GLPLS. The patient was instructed to apply topical clobetasol propionate 0.05% solution and minoxidil 5% solution twice daily on the scalp and augmented betamethasone dipropionate cream 0.05% BID on the body.

**FIGURE 1.** Clinical image of superior scalp (A) showing marked erythema and mild alopecia along the central part line. Dermoscopy of the scalp (B) showed erythema, prominent peripilar casts, and background telangiectasia.

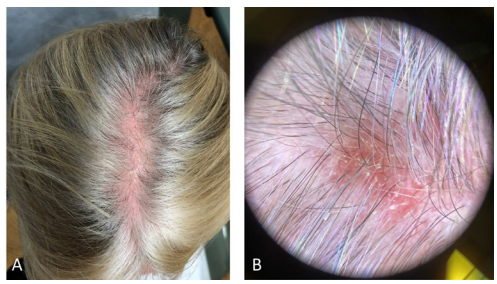
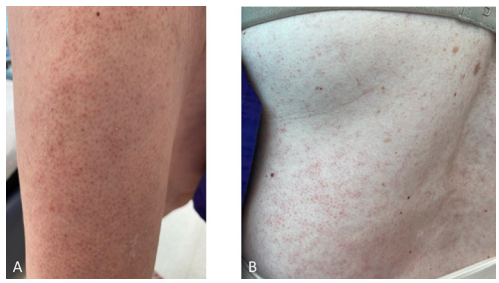
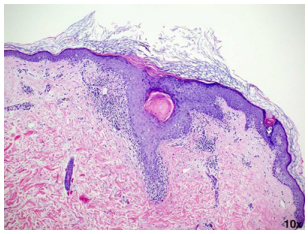



**FIGURE 2.** The thigh (A) and the back and left flank (B) showed pink patches with follicular prominence and occasional follicular scale.




At a follow-up visit one month later, the patient reported resolution of her scalp pain and improvement of involved skin on the trunk and extremities. Tacrolimus ointment 0.1% for the scalp and the body twice daily on weekends and oral doxycycline 20 mg BID twice daily were added to the medication regimen, and topical corticosteroid treatments were restricted

**FIGURE 3.** The abdominal biopsy revealed a perifollicular lichenoid lymphocytic infiltrate with apoptotic keratinocytes and overlying hyperkeratosis.



to weekdays. However, the patient did not tolerate doxycycline and had scalp irritation with tacrolimus. The patient was then started on hydroxychloroquine 200 mg twice daily. Four months after initiating this regimen, she reported minimal scalp pruritus and significant improvement in her hair loss. The remainder of the skin exam showed residual perifollicular erythema on the trunk and thighs, which had completely resolved by her 12-month follow-up visit.

## DISCUSSION

In this report, we presented the case of a patient with GLPLS, a clinical subtype of LPP, which classically features progressive scarring alopecia, axillary alopecia, and lichenoid follicular papules diffusely distributed on the trunk and extremities.[3] In conjunction with these findings, patients often report pruritus, burning, or tenderness. A confirmatory skin biopsy is required to make the diagnosis.[4] While there are no established therapeutic guidelines specific to GLPLS, the first-line approach for LPP often includes topical or intralesional corticosteroids, albeit with variable success.[5] In addition, there are reports of improvement with doxycycline or hydroxychloroquine.[6] Our patient experienced limited benefit from topical corticosteroids, which improved scalp tenderness and diffuse follicular erythema but did not halt the progression of alopecia. However, she achieved a significant symptomatic improvement on hydroxychloroquine.

Early recognition of GLPLS, before the development of scarring alopecia, is paramount for therapeutic trials to relieve symptoms and attenuate or potentially halt disease progression. Evaluation of involved skin by dermoscopy can facilitate diagnosis and aid in biopsy site selection. Nonspecific dermoscopic features of cicatricial alopecia include the loss of follicular ostia and the presence of white patches indicative of fibrosis.[7] More specifically for LPP, peripilar casts and perifollicular scales can be prominent. A subset of patients with LPP may also present with blue-gray dots, which may correspond to the presence of melanophages.[7] Dermoscopic evaluation of our patient revealed decreased hair density, marked erythema with background telangiectasia, peripilar hair casts, and scaling. These findings correlated histopathology with an inflammatory lymphocytic infiltrate, perifollicular fibrosis, and follicular plugging.

It is important to distinguish GLPLS from other forms of scarring hair loss. The anatomic distribution of skin lesions in GLPLS is distinct from most other forms of inflammatory cicatricial alopecia including other variants of LPP, discoid lupus erythematosus (DLE), central centrifugal cicatricial alopecia (CCCA). Dermoscopically, findings in GLPLS overlap with features of other types of LPP and depend on the stage of the lesion and degree of scarring.[8] In contrast to findings in LPP, DLE may feature hyperpigmentation and interfollicular white patches corresponding to interfollicular fibrosis, whereas peripilar white halos are relatively specific to CCCA.[9,10] The histologic differential likewise comprises other causes of lymphocytic cicatricial alopecia including LPP variants, DLE, CCCA, and pseudopelade of Brocq (which may represent end-stage scarring of other processes).[11]

Treatment approaches and prognoses vary among scarring alopecias, and accurate diagnosis is essential for proper counseling and treatment of GLPLS. Early diagnosis may be critical for the timely implementation of interventions prior to the onset of significant scarring, and both dermoscopic and histologic findings are instrumental in supporting a diagnosis of GLPLS. As GLPLS is typically progressive and may have a minimal or variable response to current treatments, patients would benefit from additional treatment options.

## DISCLOSURES

The authors have no conflicts of interest to declare.

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