CASE REPORT

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

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ABSTRACT Graham Little-Piccardi-Lassueur syndrome is a rare dermatosis predominantly observed in middle-aged women. The syndrome is typically marked by a triad of cicatricial alopecia (scarring hair loss), non-scarring hair loss, and lichenoid skin or mucosal lesions. Affected individuals often present with distinct areas of scarring alopecia on the scalp, perifollicular papules on the body, and non-scarring hair loss. The diagnosis is usually clinical, supported by histopathological examination and dermoscopic evaluation to aid in differential diagnosis. Histopathology of follicular papules and affected scalp areas typically reveals perifollicular lymphocytic infiltrate and interface dermatitis, consistent with lichen planopilaris. Treatment remains challenging, with variable responses to corticosteroids, calcineurin inhibitors, retinoids, and systemic immunosuppressants.

Keywords: Lichen planoplaris; cicatricial alopecia; graham-little-piccardi-lasseur syndrome

Graham-Little-Piccardi-Lasseur syndrome (GLPLS) is a rare disease characterized by cicatricial alopecia with lichen planopilaris (LPP) of the scalp. In this syndrome, three different clinics are seen together: a) cicatricial alopecia of the scalp, b) non-cicatricial alopecia in the axillae and inguinal region, and c) keratotic spikes in the hair follicles.[1] The diagnosis of GLPLS is based on clinical findings and scalp biopsy findings consistent with LPP. In this case report, we describe the clinical, dermoscopic, and histopathologic findings and treatment of a patient with GLPLS, which is very rare.

CASE REPORT

A 47-year-old woman was admitted with the complaint of hair loss starting with itching. In addition, she also described hair loss in the axillae (Figure 1). Her complaints had been continuing for 6 months. She had no known comorbidities. The scalp showed focal cicatricial areas of hair loss accompanied by erythema. A dermoscopic examination revealed loss of follicular openings and perifollicular scaling (Figure 2). Her blood tests, renal and liver function tests were normal. The scalp biopsy showed lichenoid lymphocyte infiltration and basal vacuolar damage in the follicular infundibulum (Figure 3). The patient was diagnosed with GLPLS based on her clinical and histopathological findings. Since the patient had acute hair loss, methylprednisolone 32 mg (Prednol®, Gensenta, Türkiye) treatment was started, and it was planned to be gradually discontinued within 6 weeks. The patient was referred for a visual field examination for hydroxychloroquine. Hydroxychloroquine 400 mg/day (Plaquenil®, Sanofi, France) was started for the patient with a normal visual field simultaneously with steroid therapy. Informed consent was taken from the patient.

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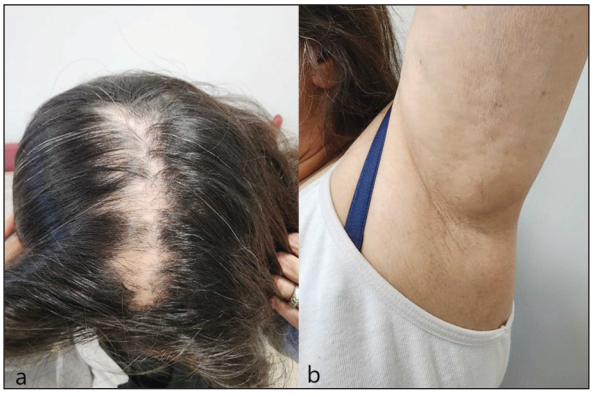

FIGURE 1: a) A patient with multifocal areas of cicatricial alopecia on the vertex of the scalp; **b)** A non-cicatricial alopecia in the axilla of the same patient

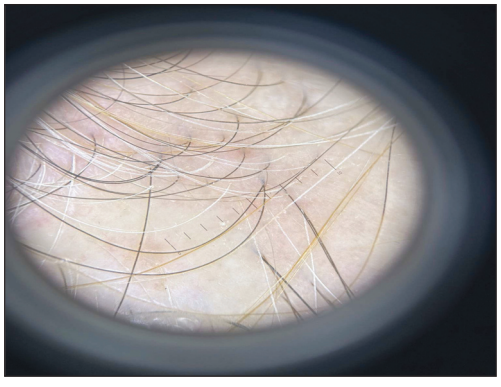

FIGURE 2: A scalp dermoscopy showing loss of follicular openings and perifollicular scaling

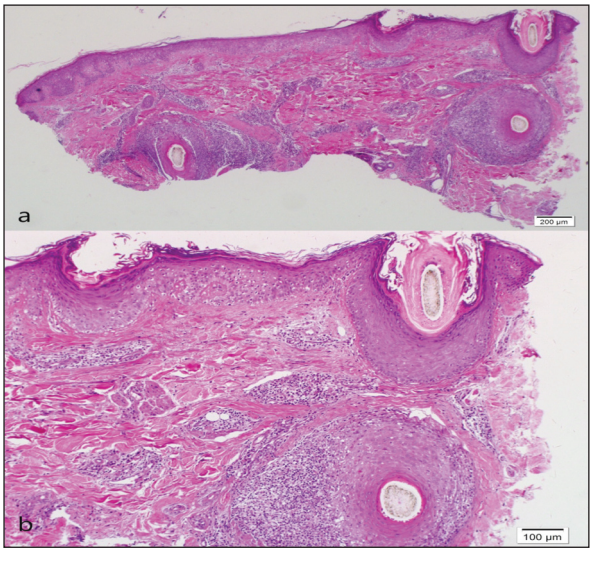

FIGURE 3: A dense lymphocytic infiltration around hair follicles **(a)** (H&E, x20) and **(b)** (H&E, x40).

DISCUSSION

Additional studies on GLPLS with varying clinical presentations have been reported in the literature. Cases lacking follicular papules but maintaining other hallmark features have been described, reinforcing the clinical spectrum of GLPLS. Notably, László reported a similar case where follicular papules were absent, yet histopathological findings confirmed the diagnosis.

GLPLS is a subtype of lichen planopilaris, first described by Piccardi in 1913 and later by Ernst Graham Little and Lasseur 2 years later.[2] Lichen planoplaris may present with 3 different clinical form: classic lichen planoplaris, frontal fibrosing alopecia and GLPLS.[3] Although the etiology of GLPLS is unknown, T-cell-mediated immune system dysregulation is thought to be responsible for the pathogenesis of the disease.[1] Our patient was a 47-year old woman with cicatricial alopecia compatible with LPP on the scalp and non-cicatricial alopecia on the axilla. However, hyperkeratotic papules in the hair follicles were not observed on her body.

Hystopathologically, early stage lesions of lichen planopilaris show basement membrane damage accompanied by perifollicular lymphocytic infiltration and basal vacuolar degeneration at the level of infundibulum and isthmus, while late stage lesions show perifollicular fibrosis and atrophy over time.[4] In our case, intense lymphocytic infiltration was observed around the hair follicles, which was consistent with the early stage histopathologic findings of LPP.

Differentiation from alopecia areata is crucial. Unlike alopecia areata, GLPLS exhibits perifollicular lymphocytic infiltration targeting the infundibulum and isthmus, leading to irreversible follicular destruction. Dermoscopy further aids in differentiation, as alopecia areata classically presents with exclamation-mark hairs, which were absent in our case.[5]

There is no single treatment modality that stands out in the treatment of GLPLS. However, it is important to start early treatment for LPP before follicular atrophy develops. Although systemic steroid treatment is effective in preventing shedding during an acute attack, its long-term use is unfavorable due to its side effects. Other preferred treatment modali-

ties include topical steroids, intralesional steroid therapy, cyclosporine, systemic retinoids and anti- malarial drugs.[6]

This case contributes to the literature by documenting an atypical presentation of GLPLS. Given the rarity of the syndrome, variability in clinical presentation should be recognized to avoid misdiagnosis. Furthermore, our study underscores the importance of histopathology in cases where clinical findings are incomplete.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Sera Nur Yücesoy, Güllü Gencebay;* ***Design:*** *Sera Nur Yücesoy;* ***Control/Supervision:*** *Sera Nur Yücesoy, Güllü Gencebay;* ***Data Collection and/or Processing:*** *Sera Nur Yücesoy, Güllü Gencebay, Koray Temiz;* ***Analysis and/or Interpretation:*** *Sera Nur Yücesoy, Güllü Gencebay, Koray Temiz;* ***Literature Review:*** *Sera Nur Yücesoy, Güllü Gencebay, Koray Temiz;* ***Writing the Article:*** *Sera Nur Yücesoy, Hümeyra Günel;* ***Critical Review:*** *Sera Nur Yücesoy, Hümeyra Günel;* ***References and Fundings:*** *Sera Nur Yücesoy, Koray Temiz;* ***Materials:*** *Sera Nur Yücesoy, Hümeyra Günel.*

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