

Graham Little Piccardi Lasseueur Syndrome Presenting with Features of Frontal Fibrosing Alopecia: A Rare Presentation

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Abstract: *Graham-Little-Piccardi-Lasseueur Syndrome is characterized by multifocal, cicatricial alopecia over the scalp, non-cicatricial alopecia of the axillae and/or perineum and follicular hyperkeratotic papules typically over the trunk, thighs, arms and wrists with or without pruritus. Frontal fibrosing alopecia is typically seen in postmenopausal women presenting as asymptomatic, progressive recession of their fronto-parietal hairline with or without nonscarring alopecia of the eyebrows. Such a coexistence of these two variants of Lichen planopilaris may be a mere overlap or a phenotypic variation of the same entity. Here we report a case of a 34-year-old female with co-existing features of both the variants.*

Keywords: Graham-Little-Piccardi-Lasseueur Syndrome, Frontal fibrosing alopecia, cicatricial alopecia, Lichen planopilaris

1. Introduction

Graham-Little-Piccardi-Lasseueur Syndrome is a rare variant of Lichen Planopilaris characterized by cicatricial alopecia over the scalp, non-cicatricial alopecia of the axillae and/or groin and follicular hyperkeratosis of the body.¹ Frontal fibrosing alopecia is also considered a distinct variant of lichen planopilaris presenting as cicatricial alopecia of the frontoparietal hairline.² We report a rare case of a 34-year-old female with co-existing features of both the variants.

2. Case Report

A 34-year-old female presented to the dermatology OPD complaining of hair loss over the scalp, eyebrows and groin for the last six months. There was history of mild itching and scaling over the scalp. She also complained of asymptomatic, small pigmented lesions over the trunk and dark patches over the face. On examination, there was scarring alopecia with mild scaling over the frontoparietal area and vertex with recession of the frontal hairline. There

was thinning and patchy loss of the eyebrows which was concealed with eyebrow pencil and ill-defined patches of greyish brown pigmentation mainly localized over the forehead and malar region. There were numerous hyperpigmented follicular spinous papules distributed over the trunk. Non-scarring alopecia was present over the pubic region. The axillary hair, oral and genital mucosa and nails were healthy.

Histopathology from the scalp showed cicatricial alopecia in the several concentric fibrotic tracts in the upper and mid dermis with loss of hair follicles and fibroticstellae in upper subcutaneous zone and melanin pigment in the dermis consistent with cicatricial alopecia due to lichen planus. Histopathology of trunk showed dermal fibrosis with preserved arrector pili muscle. Based on the above findings, a diagnosis of Graham Little Piccardi Lasseueur Syndrome with Frontal fibrosing alopecia was made. The patient was started on treatment and counselled regarding the nature of the disease.



Figure 1, 2: showing frontal hairline recession, alopecia over the eyebrows and scalp



Figure 3, 4: showing normal axillary hair and non-scarring alopecia over the pubic area



Figure 5, 6: showing keratotic papules over the trunk.

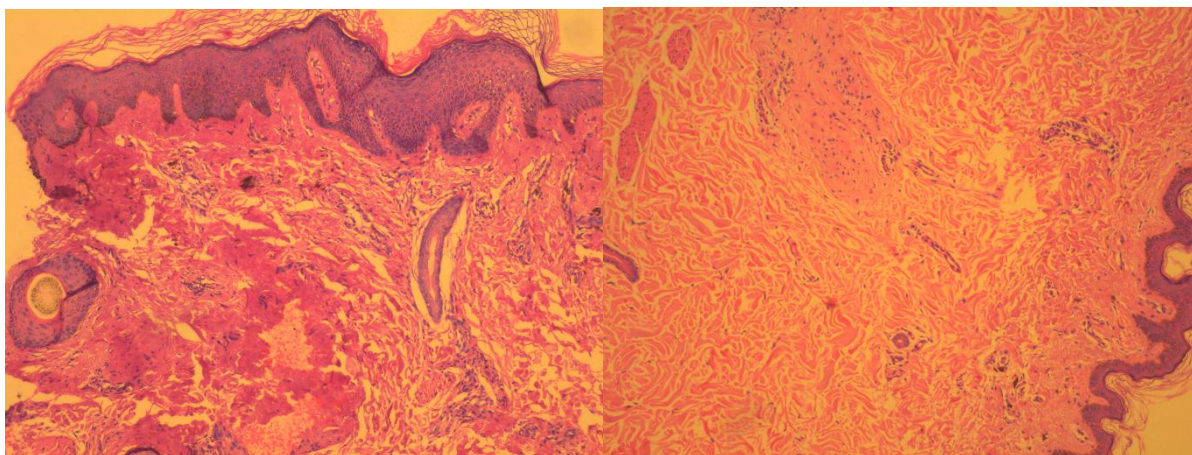


Figure 7, 8: showing melanin incontinence with perifollicular and dermal fibrosis over scalp and dermal fibrosis over the trunk respectively (10x)

3. Discussion

Lichen planopilaris refers to the follicular involvement of lichen planus. There are three variants of lichen planopilaris: classic lichen planopilaris, Graham-Little-Piccardi-Lasseur Syndrome and Frontal Fibrosing Alopecia.¹

Graham-Little-Piccardi-Lasseur Syndrome was first discovered by Piccardi in 1913 and two years later by Ernst Graham Little and Lasseur in a mutual patient.^{1,3} It is characterized by multifocal, cicatricial alopecia over the

scalp, non-cicatricial alopecia of the axillae and/or perineum and follicular hyperkeratotic papules typically over the trunk, thighs, arms and wrists with or without pruritus.^{1,4} In addition to the triad, a positive pull test for anagen hair can be present.^{4,5} There is no set chronological order of the appearance of symptoms.¹ It predominantly affects middle-aged females.^{1,3,4} The exact etiology is unknown. T-cell-mediated immunity may play a major role in triggering the clinical expression of disease.¹ GLPLS is found to be associated with HLA-DR1.^{3,6} An autoimmune link with

autoantibody directed to INCENP protein, a component of centromere has also been described.⁶

FFA was first described in 1994 by Kossard.^{7,8} It is typically seen in postmenopausal women and presents as asymptomatic, progressive recession of their frontoparietal hairline.^{7,8,9} It is commonly associated with non-scarring alopecia of the eyebrows. In this condition, disease process selectively affects the intermediate and the vellus-like follicles of the frontal margin and eyebrows but the reason for this selective involvement is unknown.⁹ These variants have failed to show any histopathological and immunophenotypical differences from lichen planopilaris.^{6,7,9} Histopathology in early lesions show perifollicular lymphocytic infiltrate at the level of the infundibulum and the isthmus. Advanced cases show alopecia with vertically oriented elastic fibers that replace the destroyed hair follicles.⁷ Similar findings were seen in our case.

There have been a few reports in literature describing patients with overlapping features of FFA and GLPLS.⁷ Saha et al⁹ described a case of classical and extensive presentation of GLPLS along with concomitant FFA. Abbas et al² described report a 37-year-old premenopausal woman with FFA associated with non-scarring alopecia of the eyebrows and axillae and follicular lichen planus-like lesions of the face. Such a coexistence of these two variants of LPP may be a mere overlap or a phenotypic variation of the same entity.

It is estimated that over 50% patients with GLPLS present with at least one episode of cutaneous or mucosal LP in the course of the disease.⁴ The greyish brown pigmentation over the forehead and malar area seen in our patient clinically resembles lichen planus pigmentosus. However, confirmation by biopsy could not be done.

Various treatment options include oral prednisolone, chloroquine, topical glucocorticoid and intralesional glucocorticoids, topical retinoic acid, minoxidil 2% solution, oral isotretinoin, and ultramicrozoned griseofulvin. Treatment remains challenging and there is little potential for regrowth after follicular inflammation and destruction.^{7,9}

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