Graham-Little-Piccardi-Lassueur Syndrome: A Case Report

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Abstract

Graham-Little-Piccardi-Lassueur syndrome is a variant of lichen planopilaris characterized by the triad of patchy cicatricial alopecia of the scalp, noncicatricial alopecia of the axilla and groin, and follicular spinous papules on the body, scalp, or both. The disease is most commonly seen in women 30 to 70 years of age. We present a case of this rare syndrome in a 68-year-old female with madarosis and provide a discussion about the disease and treatment options.

Introduction

Graham-Little-Piccardi-Lassueur syndrome (GLPLS) is a rare subtype of lichen planopilaris (LPP) that presents with the triad of multifocal cicatricial alopecia of the scalp, noncicatricial alopecia of the axilla and groin, and a follicular lichen planus (LP) eruption on the body, scalp, or both.[1] It is four times more likely to affect women and is characteristically seen in those who are middle-aged to post-menopausal.[1] Although the exact etiology of GLPLS is unknown, it is thought to be an immune-mediated disorder that causes an inflammatory reaction against the bulge region of hair follicles.[2] The disease is non-familial, although one case with a familial origin has been reported.[3]

Case Report

A 68-year-old female presented with a one-year history of a mildly pruritic, erythematous, scaly frontal scalp and alopecia involving her head, eyebrows, eyelashes, axillae, legs and arms. Her eyebrows and eyelashes experienced the most rapid progression of hair loss, with complete madarosis over six to eight months. However, her main areas of concern at the time of her initial visit were the frontal scalp and temporal regions. The patient reported that her hair was previously grey, but as her hairline receded, black pigmented hairs developed despite never coloring her hair. Previous treatment included triamcinolone ointment prescribed by her primary care physician for presumed scalp psoriasis, which reduced scaling but failed to arrest the hair loss. The patient then visited her beautician, who recommended over-the-counter selenium-sulfide shampoo for seborrheic dermatitis and tea tree oil shampoo and conditioner. These products caused mild reduction in scale, but she again noted an increasingly receding hairline. The patient had an otherwise unremarkable 12-point review of systems and had no known drug allergies except for gabapentin sensitivity, which caused nausea. Past medical history included hypothyroidism, aortic regurgitation, mitral regurgitation, hypertension, migraines, depression, seasonal allergies, and toxoplasmosis that had been treated 58 years prior. The patient's medications included levothyroxine, losartan, bupropion, sertraline, acetaminophen/butalbital/caffeine, sumatriptan, fish oil, and calcium with vitamin D. The only medication change in the past 18 months was from lisinopril to losartan. Family history revealed hypothyroidism in the patient's father and son. Social history revealed a 25 pack-year smoking history but no tobacco use in the last 30 years. She denied any alcohol or illegal drug use.


Figure 1

Dermatological exam revealed cicatricial alopecia of the frontal scalp and temples with associated perifollicular scalp erythema and hyperkeratotic follicular scaling; also noted were a few residual tufts of black, normal-looking terminal hair (**Figure 1**). Noncicatricial alopecia of the eyebrows, eyelashes, axillae, forearms, and legs was present in addition to multiple follicular, keratotic, and spinous papules over the remainder of the scalp (**Figures 2, 3**). Differential diagnosis included GLPLS, classical LPP, frontal fibrosing alopecia (FFA), lichen spinulosus, alopecia mucinosa, discoid lupus erythematosus, pityriasis rubra pilaris, pseudopelade of Brocq, and sarcoidosis. Two 4 mm punch biopsies taken from the frontal scalp revealed scarring alopecia with dermal fibrosis, a perifollicular lymphocytic infiltrate, and the absence of interface dermatitis in the overlying epidermis (**Figures 4, 5**). Histologic findings were consistent with a diagnosis of lichen planopilaris (LPP). The clinical picture of LPP, noncicatricial alopecia and keratotic papules is consistent with the rare variant of LPP known as GLPLS.

Treatment was initiated with topical high-potency steroids with consideration for systemic steroids or antimalarials pending punch-biopsy results and clinical course. At two-week follow-up, our patient demonstrated noticeable improvement in scalp erythema, scaling and pruritus. At one-month follow-up, scalp erythema was no longer present, scaling had improved, and hair loss had ceased. Systemic medications were not initiated given the significant clinical improvement, and she will continue to be monitored regularly.

Discussion

The name GLPLS comes from the names of the physicians who first described this condition. The disease was originally defined in 1913 by Piccardi, who described a case of progressive cicatricial scalp alopecia, noncicatricial alopecia in the axillae and pubic area, and follicular spinous papules on the trunk and extremities, to which he gave the name *cheratosi spinulosa*, or keratotic spinulosa.[4] In 1915, Graham-Little published a case of a similar condition in a 55-year-old woman who was referred by Lassueur, describing it as "folliculitis decalvans et atrophicans."[5] In addition to the classical triad of cicatricial alopecia of the scalp, noncicatricial alopecia of the axillae and groin, and a follicular keratosis eruption, GLPLS can affect the eyebrows and lateral face.[6,7]

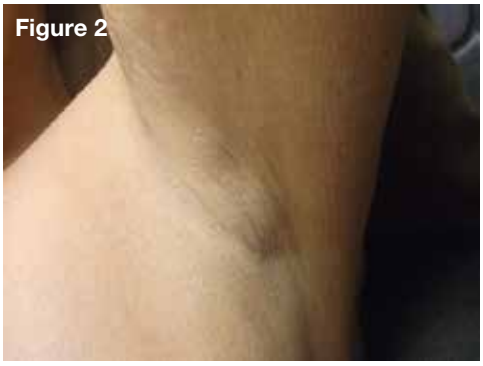


Figure 2

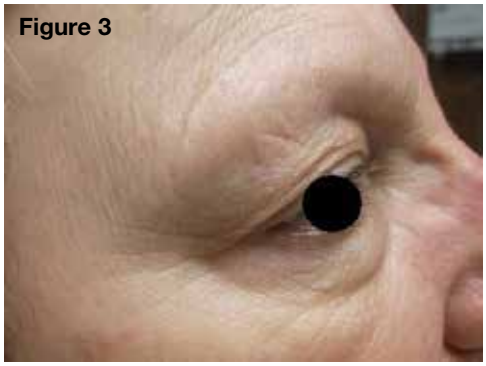


Figure 3

Figure 4

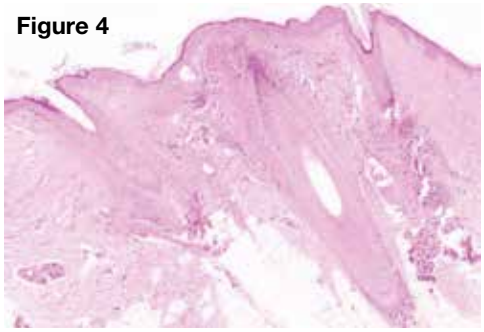
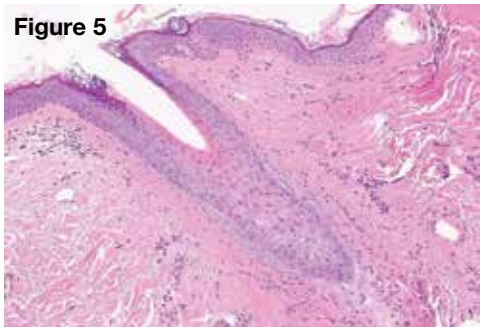


Figure 5



GLPLS is a rare type of LPP that typically presents in women who are 30 to 70 years old, although the condition has been reported in males and younger individuals.[1] LPP can be subdivided into three clinical variants: classical LPP, FFA, and GLPLS.[8] GLPLS may have a positive pull test for anagen hairs due to the same altered integrin expression seen in active LPP.[8] Histopathological findings of GLPLS are similar to those seen in LPP, but the absence of interface dermatitis of the overlying epidermis can help differentiate the two. Early lesions of LPP reveal a perifollicular lymphocytic infiltrate at the level of the infundibulum and the isthmus, along with vacuolar changes of the outer root sheath.[10] More advanced cases show perifollicular fibrosis and epithelial atrophy at the level of the infundibulum that give rise to a characteristic hourglass configuration.[10] As the disease progresses, vertically oriented elastic fibers replace the destroyed hair follicles.[10]

The exact etiology of GLPLS is unknown, but it is likely similar to the T-cell-mediated immunological mechanism that triggers the clinical expression of LP.[2] GLPLS has not been associated with underlying systemic disease, but it may cause stress and anxiety due to its presentation. Isolated cases describing a familial pattern (HLA DR1), association with hepatitis B vaccination, and a female (genetically XY) patient with complete androgen insensitivity syndrome have been reported.[3,9,10]

Unless GLPLS is recognized early, treatment is usually only mildly effective. Once scarring occurs, hair regrowth will not occur. Treatment is directed at halting the progression of disease, preventing further alopecia, and providing symptomatic relief. Various therapies including intralesional and systemic corticosteroids, retinoids, PUVA therapy, topical tacrolimus, and antimalarials have all produced varying results.[8] Isolated reports have demonstrated anecdotal success with cyclosporine and thalidomide[11,12] The disease is often progressive, with little potential for hair regrowth once complete destruction of the follicle occurs. Early, accurate diagnosis is imperative to prevent progression.

Conclusion

GLPLS is an uncommon entity that has been reported fewer than 50 times in the literature. It has a classical clinical presentation and is not associated with systemic disease. Although its pathogenesis is unknown, the T-cell-mediated immune response in GLPLS is similar to that in LP. There is no universally effective treatment, so therapy should be directed at halting disease progression.

References

1. Pai VV, Kikkeri NN, Sori T, Dinesh U. Graham-Little Piccardi Lassueur syndrome: an unusual variant of follicular lichen planus. Int J Trichology. 2011 Jan;3(1):28-30.

2. Tchernev G, Nenoff P. Antigen mimicry followed by epitope spreading: a pathogenetic trigger for the clinical morphology of lichen planus and its transition to Graham Lassueur Piccardi Little Syndrome and keratosis lichenoides chronica - Medical hypotheses or reality? An Bras Dermatol. 2009 Nov-Dec;84(6):682-8.

3. Viglizzo G, Verrini A, Rongioletti F. Familial Lassueur-Graham-Little-Piccardi syndrome. Dermatology. 2004;208(2):142-4.

4. Piccardi G. Keratosis spinulosa of the scalp in its relations with the pseudo-pelade of Brocq. Ital J Skin Vener Dis. 1914;49:416.

5. Graham-Little EG. Folliculitis decalvans et atrophicans. Br J Dermatol. 1915;27:183–5.

6. Camacho F. Alopecias cicatriciales. In: Camacho F, Montagna W, eds. Tricología. Madrid: Grupo Aula Médica; 1996. p. 537–551.

7. Morillo M, Rodríguez Pichardo A, Herrera A, Camacho F. Síndrome de Piccardi-Lassueur-Graham Little. Estudio de seis casos. Actas Dermosifiliogr. 1998;89:392–395.

8. Assouly P, Reygagne P. Lichen planopilaris: update on diagnosis and treatment. Semin Cutan Med Surg. 2009 Mar;28(1):3-10.

9. Bardazzi F, Landi C, Orlandi C, Neri I, Varotti C. Graham Little-Piccardi-Lasseur syndrome following HBV vaccination. Acta Derm Venereol. 1999 Jan;79(1):93.

10. Vega Gutiérrez J, Miranda-Romero A, Pérez Milán F, Martínez García G. Graham Little-Piccardi-Lassueur syndrome associated with androgen insensitivity syndrome (testicular feminization). J Eur Acad Dermatol Venereol. 2004 Jul;18(4):463-6.

11. George SJ, Hsu S. Lichen planopilaris treated with thalidomide. J Am Acad Dermatol. 2001 Dec;45(6):965-6.

12. Kubba R, Rook A. Graham Little syndrome: follicular keratosis with cicatricial alopecia. Br J Dermatol. 1975;93(suppl 11):53-55.

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