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Graham-Little-Piccardi-Lassueur Syndrome: Two Case Reports

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Abstract

Graham-Little-Piccardi-Lassueur Syndrome (GLPLS) is a rare lichen planopilaris (LPP) subtype associated with scarring alopecia. It is thought to be an immune-mediated cause of alopecia. It is characterized by a triad of multifocal cicatricial alopecia of the scalp, noncicatricial alopecia of the axilla, and groin and a follicular lichen planus (LP) eruption. Herein, we present 2 cases of GLPLS. The first case is of a 33-year-old female presented with classical lichen planopilaris (LPP) of the scalp, follicular hyperkeratosis of bilateral arms, and noncicatricial alopecia of armpits. In the second case, a 17-year-old female presents with frontal fibrosing alopecia (FFA) of the scalp, follicular hyperkeratotic papules of the scalp, and noncicatricial alopecia of the bilateral armpits and genitalia. The physician can miss GLPLS due to the rarity of the condition.

Keywords: Frontal fibrosing alopecia; Graham-Little-Piccardi-Lassueur Syndrome; Lichen planopilaris

Introduction

Graham-Little-Piccardi-Lassueur Syndrome (GLPLS) is an uncommon subtype of lichen planopilaris (LPP) that is characterized by a triad of multifocal cicatricial alopecia of the scalp, noncicatricial alopecia of the axilla and groin and a follicular hyperkeratotic eruption on the body or 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 prognosis of GLPLS is variable, sooner or later resulting in irreversible cicatricial alopecia and follicular lesions over the body, inflicting substantial psychosocial distress.[3]

Case report

Case1

A 33-year-old female presented with slowly progressive patchy hair loss on the scalp for the last 11 years. On further inquiry, she complained of asymptomatic follicular keratotic and spinous papules in bilateral arms on the extensor area. She went to multiple institutions for treatment, where she was prescribed clobetasol propionate 0.05% cream, minoxidil 5% spray, oral anti-oxidants, intra-lesional triamcinolone acetonide (10 mg/ml), mometasone furoate 1% cream but without any improvement in the lesion. The patient denied any constitutional symptoms.

Examination revealed multiple patches of cicatricial alopecia present in the frontal area and vertex of the scalp the largest measuring 4 cm*3 cm (figure 1a). Dermoscopy of the lesion showed peripilar casts and reddish-whitish area with loss of follicular ostia (figure 1b). Multiple follicular keratotic papules were present in the extensor surface of bilateral arms (figure 2a). There was presence of noncicatricial alopecia of the bilateral armpit and genital area (figure 2b). There were no other relevant findings in the general physical

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and systemic examination. Baseline investigations were within normal limits.

Histopathological examination (HPE) with hematoxylin and eosin (H and E) stain of the scalp revealed perifollicular fibrosis, while few hair follicles were replaced by columns of fibrous tissue. Mild perivascular lymphocytic infiltration and minimal pigment incontinence.

On the basis of history, clinical examination, and histopathological examination, diagnosis of GLPLS was made, and the patient was treated with oral methylprednisolone 32mg twice weekly, topical tacrolimus 0.1% ointment twice daily, and intralesional triamcinolone acetonide (10 mg/ml) once a month.

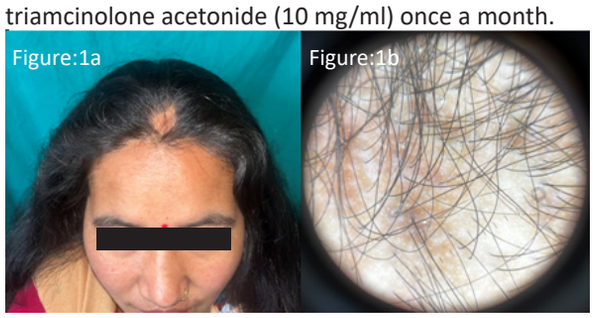

Figure:1a. Patchy alopecia in the scalp; **Figure: 1b**. Trichoscopy (magnification: 10X, polarized mode) of the scalp showing peripilar cast, loss of follicular ostia

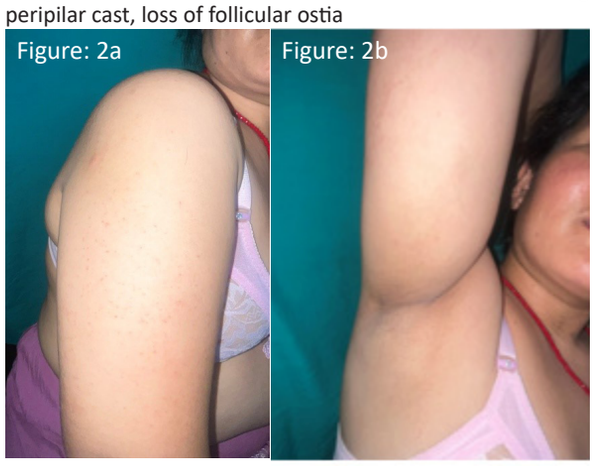

Figure: 2a. Multiple keratotic follicular papules of right arm; **Figure: 2b**. Noncicatricial alopecia of the armpit.

Case 2

A 17-year-old female came with the complaints of progressive hair loss from the frontal region of the scalp associated with loss of hair from eyebrows for the last 7 years, which was associated with itching. She took methylprednisolone pulse dose and minoxidil lotion for 5 weeks, which did not show any improvement in the condition. There was no similar family history. There were no other constitutional symptoms.

On physical examination, cicatricial alopecia was seen on the scalp, with a recession of the fronto-temporal hairline and loss of eyebrows (figure 3a). The patient had keratotic papules on the scalp (figure 3b), back, and arms. Noncicatricial alopecia was noted in the pubic and axillar regions (figure 4). Dermoscopy of the lesion showed loss of follicular ostia, peri-pilar cast, and round-to-oval yellowish areas with keratotic follicular plugs surrounded by mild erythema (figure 5). There were no other significant systemic findings. Baseline investigations were within normal limits.

Histopathology with hematoxylin and eosin stain (H and E) (figure 6) showed orthokeratosis, follicular plugging, and hypergranulosis of epidermis. Perivascular infiltration of lymphocytes and macrophages. Mucinous fibroplasia with perifollicular lymphocytic infiltration with hourglass-like narrowing at follicular infundibulum.

Based on history, physical examination, and histopathological examination, the diagnosis of GLPLS associated with frontal fibrosing alopecia (FFA) was confirmed. Treatment was started with monthly intralesional triamcinolone acetonide (10mg/ml) with hydroxychloroquine 100mg twice daily.

Figure: 3a. Receding fronto-temporal region of the scalp; Figure: 3b. Multiple erythematous follicular papules present in occipital region of scalp.

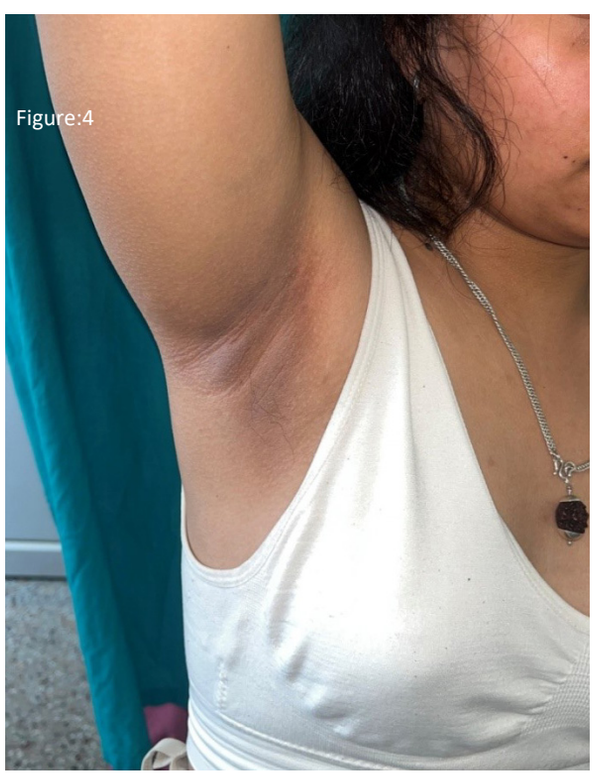

Figure 4: Noncicatricial alopecia of the armpit

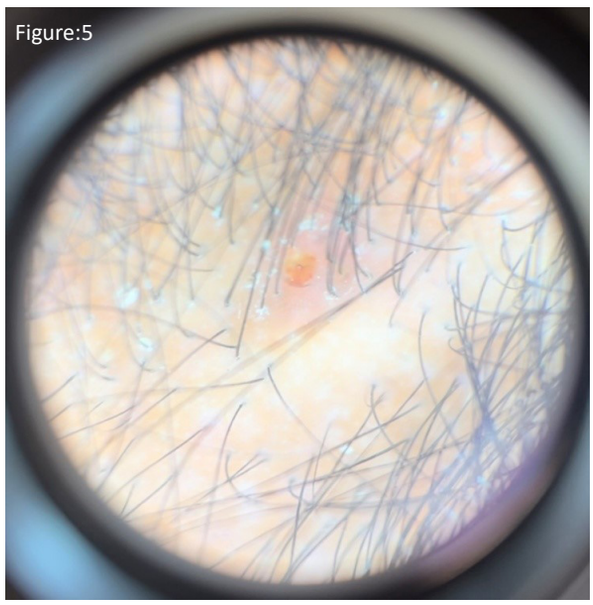

Figure 5: Dermoscopy of scalp lesion (magnification: 10X, polarized mode) showing loss of follicular ostia, peri-pilar cast and keratotic follicular plugs.

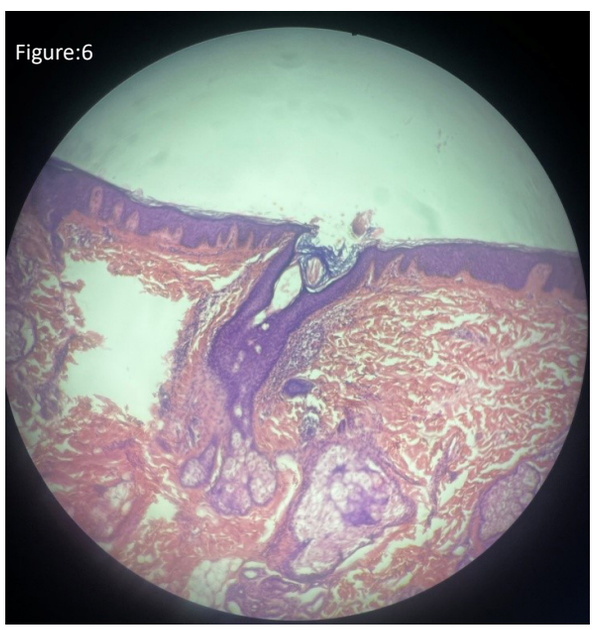

Figure: 6. Hematoxylin and eosin stain, 40X. Histopathology of scalp lesion shows hour-glass narrowing at infundibum of hair.

Discussion

In 1913, Piccardi first reported a case of follicular spinous papules on the trunk and extremities, noncicatricial alopecia in the axilla and pubic area and progressive scalp cicatricial alopecia, which he named cheratosi spinulosa (keratotic spinulosa).[4] Graham Little published a similar case of a 55-year-old lady in 1915, and her condition was defined as "folliculitis decalvans et atrophicans", which was further followed by similar case reports later.[4]

GLPLS is a variant of LPP seen predominantly in the female population, usually aged between 30 and 70.[5] Three clinical variations of LPP are classical LPP, FFA, and GLPLS.[6] The triad of findings in GLPLS may not necessarily be required to manifest simultaneously.[7] Alopecia of GLPLS with scarring frequently occurs clinically months or years before any follicular emergence.[7] The illness is frequently chronic, progresses slowly, and might take months or years.[8] Even though the exact cause of LPP is unknown, autoimmune mechanisms have been proposed.[2] Recent studies have shown decreased expression of peroxisome proliferator-activated receptor (PPAR). Also, interferon and JAK singling are upregulated in LPP.[9] Few cases have reported its association with hepatitis B vaccination and androgen insensitivity syndrome (testicular feminization).[10]

Classic LPP should be differentiated from other causes of patchy alopecia like alopecia areata, tractional alopecia, discoid lupus erythematosus, and folliculitis decalvans. In contrast, FFA should be differentiated from androgenetic alopecia and tractional alopecia. A high risk of suspicion should be made to rule out those conditions.

In an extensive literature review, less than 50 case reports of GLPLS were found, and only a few of its associations with frontal fibrosing alopecia were found. Therapy for GLPLS and other scarring alopecia aims to stop the progression of hair loss; hence, early detection and treatment are essential. Clinical responses can be found with topical or systemic steroids, retinoids, PUVA (Psoralen plus Ultraviolet-A radiation) therapy, cyclosporine, or thalidomide.[8]

This case report's restriction stems from the inability to obtain the histopathological imaging of the initial case due to the lesion's biopsy being performed at a different institution and the patient's contact being lost. Due to one patient being on follow-up and another being lost to follow-up, it was not possible to evaluate the response to the treatment.

Conclusion

GLPLS is an uncommon condition affecting the follicular appendages seen most commonly in middle-aged females. We report an unusual correlation between GLPLS and FFA and the association of GLPLS with classic LPP. Patients with follicular hyperkeratotic lesions on the scalp, trunk and extremities, alopecia of the pubic and axillary areas, and progressive cicatricial scalp alopecia of the scalp should be suspected of GLPLS. Early diagnosis of the case by a physician helps to prevent progression and provide early treatment of the condition.

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