INTRODUCTION

Frontal fibrosing alopecia (FFA) is increasingly considered as a distinct variant of lichen planopilaris (LPP), with scarring alopecia of frontotemporal hairline and nonscarring alopecia of the eyebrows. FFA and Graham–Little–Piccardi syndrome (GLPS) can be considered as spectrum of a disease as per the current literature due to the presence of overlapping features. Coexistence of these two uncommon presentations has rarely been reported in scientific literature.

CASE REPORT

A 49-year-old female presented with hair loss from both center and sides of the scalp for the last 1 year which did not improve following minoxidil therapy. She also complained of generalized pruritus and gritty sensation all over the body. She had completed her family gracefully with two healthy children. Her family history was unremarkable, and she did not suffer from any chronic disease. No history of chronic medication could be elicited. Personal and family history of photosensitivity was absent.

On examination, we found scarring alopecia with follicular papules and mild scaling over the vertex [Figure 1a and b]. She had definite recession of frontotemporal hairline with bilaterally symmetrical band-like scarring alopecia of the same region [Figure 2]. There were numerous violaceous follicular spinous papules distributed over the abdomen, back, arms, and legs [Figure 3a-c]. Her palms, soles, nail, and oral mucosa were not involved. Further examination revealed nonscarring alopecia over the axillae and pubis [Figure 4]. She did not allow us to take photograph of the pubic area.

Routine investigations with complete hemogram, blood sugar, liver function test including hepatitis profile and thyroid screening were within normal limit. Histopathological examination from scalp biopsy revealed...
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hyperkeratotic epidermis with acanthosis and follicular plugging. There was definite basal cell degeneration with band-like chronic inflammatory infiltrate around peril follicular region. Dermis showed occasional loss of hair follicle and perifollicular fibrosis. All these features were consistent with LPP [Figure 5a and b].

Based on above findings, a diagnosis of GLPS with FFA was made, and the patient was put on oral prednisolone (1 mg/kg/day) and oral retinoid (0.5 mg/kg/day) for initial 2 weeks. Dose of the steroid was tapered to 20 mg/day and continued along with the same dose of oral isotretinoin (20 mg) for another 2 weeks with strict monitoring of blood parameters without any alteration. At the end of 1 month, the patient had significant improvement of pruritus and hair loss with flattening of the follicular papules. Doses of steroid and isotretinoin

Figure 1: (a) Scarring alopecia with follicular papules and mild scaling over the vertex. (b) Close-up view

Figure 2: Recession of frontotemporal hairline with symmetrical band-like scarring alopecia

Figure 3: (a) Violaceous follicular spinous papules over the abdomen. (b) Violaceous follicular spinous papules over the back. (c) Violaceous follicular spinous papules over the leg

Figure 4: Nonscarring alopecia over the axilla

Figure 5: (a) Follicular plugging with band-like chronic inflammatory infiltrate around peril follicular region, occasional loss of hair follicles, and perifollicular fibrosis (H and E, x10). (b) Follicular plugging with definite basal cell degeneration with band-like chronic inflammatory infiltrate around peril follicular region (H and E, x40)

Figure 6: (a) Flattening of follicular papules with residual postinflammatory hyperpigmentation following treatment. (b) Posttherapy improvement of frontal fibrosing alopecia patch
were further reduced to 10 mg/day and 15 mg/day, respectively, for another 1 month with further flattening of follicular papules and improvement of FFA patch. Now, the patient is on only 15 mg oral retinoid every alternate day and with regular follow-up on a monthly basis. Follicular papules almost resolved with residual postinflammatory hyperpigmentation [Figure 6a and b].

**DISCUSSION**

The entity, GLPS was born with a lot of controversies as far as proper clinical definition is concerned.\[2-3\] Our observation of simultaneous presence of two variants may further add fire to the unsolved issues of this strange entity.

GLPS is an unusual variant of LPP characterized by multifocal cicatricial alopecia of the scalp, noncicatricial alopecia of the axillae, and/or pubis and follicular lichen planus (LP) involving the trunk and extremities. On the other hand, FFA is increasingly recognized as a distinct variant of LPP in recent years.\[8\] This progressive form of fibrosing alopecia has failed to show any significant difference from other forms of LPP as far as histopathology and immunophenotyping are concerned.\[5,6\] Only differentiating point is symmetrical distribution and predilection for frontotemporal and parietal regions. If we look into the pathogenesis, GLPS is generally considered of having an autoimmune origin.\[6,7\] In FFA, the lymphocytic infiltrate and fibrosis affect selectively the intermediate and the vellus-like follicles of the frontal margin and eyebrows. Although the reason for this selective involvement is still an enigma, it is hypothesized that it may represent a variety of LPP with selective involvement of certain androgen-dependent areas. The affected follicles may have typical biologic markers that could explain typical characteristic feature of FFA.\[8\]

There was several existing literatures where FFA was found to be associated with different forms of LP. In one report, authors concluded FFA as a phenotypic variation of GLPS where additional features are nonscarring alopecia of the eyebrow and axillae and follicular LP over the face.\[3\] Other authors described FFA in the association of cutaneous LP\[6\] and oral mucosal LP.\[6] To the best of our knowledge, like our case, no such case has been reported till date where FFA is found along with full blown GLPS with all features.

There are several treatment options available for LPP such as oral steroid, oral retinoid, cyclosporine, mycophenolate mofetil, hydroxychloroquine, and psoralen plus ultraviolet A therapy. Most of the recent and existing literatures say that treatment is difficult and challenging.\[10-12\] Surprisingly, our case responded very well in terms of symptomatic improvement, clearing of lesions, and halt of progressive alopecia.

**CONCLUSION**

The classical and extensive presentation of GLPS along with concomitant FFA in the index case portrays a rare and unique presentation of follicular LP. Such a coexistence of these two variants of LPP may be a mere overlap, a phenotypic variation of the same entity, or it may help in future to frame a newer nosology for LPP.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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