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Case Report

Graham-Little-Piccardi-Lassueur syndrome: a rare case report and review of literature

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Abstract Graham-Little-Piccardi-Lasseur syndrome (GLPLS) is a rare variant of lichen planopilaris comprising of a triad of multifocal cicatricial alopecia of scalp, non-cicatricial alopecia of axillae and pubic region and keratotic follicular papules over body. Its exact etiology is not known to date, but it primarily involves an immune mediated inflammation affecting hair follicles resulting in cicatricial alopecia. We report a case of 55 years old female diagnosed as having features of this syndrome with cicatricial alopecia of FFA and FADP.

Key words
Graham-Little-Piccardi-Lasseur syndrome, cicatricial alopecia, lichen planopilaris.

Introduction

Graham-Little-Piccardi-Lassueur syndrome (GLPLS) is a rare variant of follicular lichen planus characterized by a triad cicatricial alopecia of scalp, non cicatricial alopecia of axilla and groin, along with follicular lichen planus of limbs and trunk.[1] Only few cases have been described so far after first case description in 1914 by Piccardi *et al*.[2] Exact cause is unknown.[3] We describe a case of GLPS due to rarity of this syndrome.

Case report

A 55-year-old female, postmenopausal, with no known comorbidities, presented to us with mildly pruritic papules on limbs, and hair loss of 1-year duration. She had no personal or family history of any autoimmune disease or any drug intake. On examination, she was a healthy middle-aged lady having erythematous, lichenoid follicular papules present over forearms (**Figure 1**), back and temple area of face causing patches of hair loss. On scalp examination there was shiny atrophic skin with hair loss on vertex (**Figure 2**) and frontal scalp (**Figure 3**) over temples. There were perifollicular keratotic papules on the margins of patches. Examination of axilla showed noncicatricial alopecia with normal skin (**Figure 4**). Pubic hair was sparse. Oral cavity and nail examination was normal.

Histopathological findings from forearm showed follicular plugging, perifollicular infiltrate of lymphocytes, vacuolar degeneration of basal keratinocytes with pigmentary incontinence in dermis consistent with lichen planopilaris.

Therefore our patient fulfilled the triad of GLPS having cicatricial alopecia of scalp secondary to LPP, noncicatricial alopecia of axilla and follicular lichen planus of trunk and extremities.

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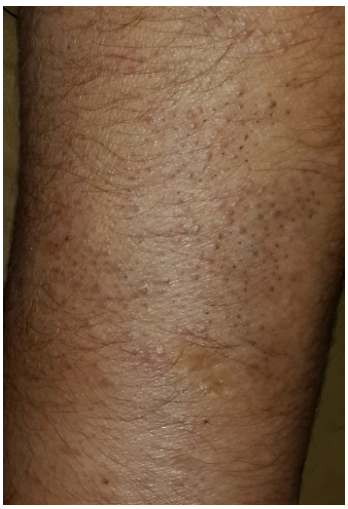

Figure 1 Follicular lichen planus on forearm.

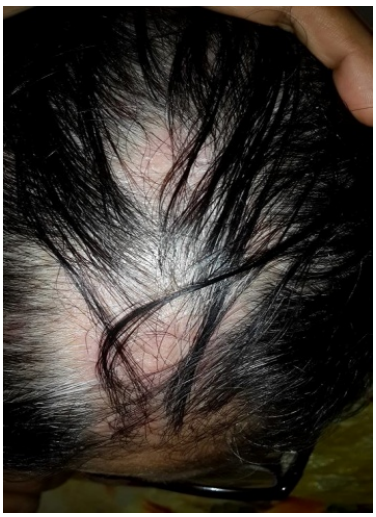

Figure 2 Fibrosing alopecia in pattern distribution.

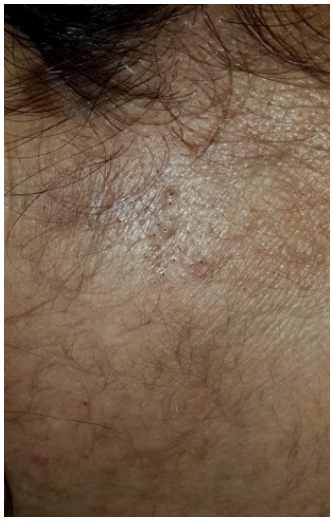

Figure 3 Frontal fibrosing alopecia.

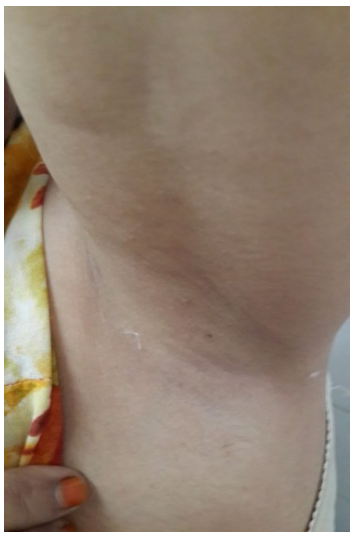

Figure 4 Noncicatricial alopecia of axilla.

Discussion

Lichen planopilaris is a rare inflammatory follicular variant of lichen planus which causes cicatricial alopecia of scalp.[1] Its cause is unknown but autoimmunity seems to play a role. It has three subtypes: classic lichen planopilaris, frontal fibrosing alopecia and Graham-Little-Piccardi-Lassueur syndrome.

Graham-Little-Piccardi-Lassueur syndrome, a rare variant of lichen planopilaris, was first described by Piccardi in 1914. Graham-Little in 1915 described another patient of GLPLS and named it as folliculitis atrophicans decalvans.[2] About 50 cases have been described since then.

Majority of cases described are middle-aged postmenopausal females, age ranges from 30-70 years.[4] Only few cases of GLPLS are reported in males.[5] One case was reported in a patient of androgen insensitivity syndrome.[6] It usually occurs sporadically however a familial case of GLPLS syndrome has been described in a mother and daughter, both were positive for HLA DR1.[7] T cell-mediated autoimmunity seems to play a role.[8] Associated conditions described are hyperthyroidism, HBV vaccination, and autoimmunity.[9] A preceding or

concomitant history of typical LP may be present.[10]

Recently, researches have tried to unveil the mystery of its pathogenesis. Tchernev *et al*.[11] have proposed the phenomenon of antigen mimicry and epitope spreading as a likely mechanism of progression of disease. Rodriguez-Bayona *et al*.[12] found autoantibodies against the chromosomal passenger protein INCENP in a patient with Graham-Little-Piccardi-Lassueur syndrome.

It is clinically characterized by a triad of cicatricial alopecia of scalp, axillary and pubic hypotrichosis of nonscarring type and keratotic follicular papules over body consistent with follicular LP.[2] LPP causing cicatricial alopecia may be classic LPP, frontal fibrosing alopecia (FFP) or fibrosing alopecia in a patterned distribution (FADP).[13,14] In our patient hair loss was consistent with FFA and FAPD. FFA has been described in literature but cicatricial alopecia of FAPD type has not been described so far.

Various modalities have been described for treatment including topical and intralesional and systemic corticosteroids, hydroxychloroquine, cyclosporin A, thalidomide and PUVA but results of treatment have been unsatisfactory.[15] The main effect of disease is psychological disturbance caused by cicatricial alopecia.[16]

To conclude Graham Little-Piccardi-Lassueur syndrome is a rare variant of lichen planopilaris. We report this case due to rarity of syndrome and the unique feature of presence of both FFA and FAPD of cicatricial alopecia which have not been described so far.

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ERRATUM

In the original article titled *'**Prevalence of polycystic ovaries among patients with hirsutism**'* authored by ***Drs. Shazia Rasool*** *(Corresponding author),* ***Lubna Riaz*** and ***Abdul Hameed,*** published in the July-September, 2011 issue of the *Journal of Pakistan Association of Dermatologists* (*J Pak Assoc Dermatol.* 2011;**21 (3)**: 174-178), the name of Corresponding Author was misspelt as ***Shazia Rasool*** in the Article, Contents of July-September, 2011 issue of *JPAD* and Index of Volume 21. The correct spelling of her name are ***Shazia Rasul.***