Dermatology Practical & Conceptual

Graham-Little-Piccardi-Lassueur Syndrome With Concomitant Mucocutaneous Lichen Planus: Rare Presentation in a Man

Avita Dhiman[1], Priyanka Sangwan[1], Neirita Hazarika[1], Prashant Durgapal[2]

1 Department of Dermatology, Venereology and Leprosy, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India
2 Department of Pathology, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India

Key words: alopecia, cicatricial, lichen planopilaris, male, non-cicatricial

Citation: Dhiman A, Sangwan P, Hazarika N, Durgapal P. Graham-little-piccardi-lassueur syndrome with concomitant mucocutaneous lichen planus: rare presentation in a man. *Dermatol Pract Concept*. 2022;12(1):e2022042. DOI: https://doi.org/10.5826/dpc.1201a42

Accepted: July 14, 2021; **Published:** January 2022

Funding: None.

Competing interests: None.

Authorship: All authors have contributed significantly to this publication

Corresponding author: Neirita Hazarika, MD, Additional Professor, Department of Dermatology, Venereology and Leprosy, All India Institute of Medical Sciences, Rishikesh Uttarakhand, India. E-mail: neirita.derma@aiimsrishikesh.edu.in

Introduction

Graham-Little-Piccardi-Lassueur syndrome (GLPLS) described by Piccardi in 1913 is a rare variant of lichen planopilaris, characterized by a triad of multifocal cicatricial alopecia of the scalp, non-cicatricial alopecia of axillae, pubis and lichenoid follicular eruption [1]. The exact etiology is unknown, but cell-mediated immunity may play a role. Over 50% of patients with GLPLS presents with at least 1 episode of cutaneous or mucosal lichen planus (LP) [1]. The progression of GLPLS is variable, and results in irreversible cicatricial alopecia causing significant psychosocial distress [1]. Herein we report a case of GLPLS in a male with mucocutaneous LP and incidental findings of hidradenitis suppurativa (HS) and fixed drug eruption (FDE).

Case Presentation

A male in his late forties presented with progressive loss of scalp hair over thirty years and history of loss of axillary and pubic hair, along with multiple, pruritic, violaceous lesions over face, trunk for the last 6 years. He also complained of pain and burning sensation in his mouth for the past 6 months; associated history of recurrent, multiple, painful swellings in bilateral axillae, groin in the last 2 years; 3 of such episodes occurred in the previous 3 months. He also reported history of 3 episodes of itchy, burning reddish lesion over abdomen, each time after taking painkillers.

Cutaneous examination revealed scarring alopecia of whole scalp with few patches of uninvolved hair in occipital area (Figure 1A) and non-scarring hypotrichosis of axillary, beard area (Figure 1B,1C) and pubic region. Multiple polyporous comedones and bridging scars were also seen in axillae, (Figure 1B) and groin. Multiple violaceous, flat-topped papules over face (Figure 1C), trunk with lichenoid follicular papules over upper back (Figure 1D) were noted. Buccal mucosa showed whitish, reticular plaques (Figure 1E). Single, well-defined, oval hyperpigmented macule was noted over abdomen (Figure 1F).

Investigations revealed positive serology for hepatitis C virus (HCV) with aspartate aminotransferase level of 131U/L,

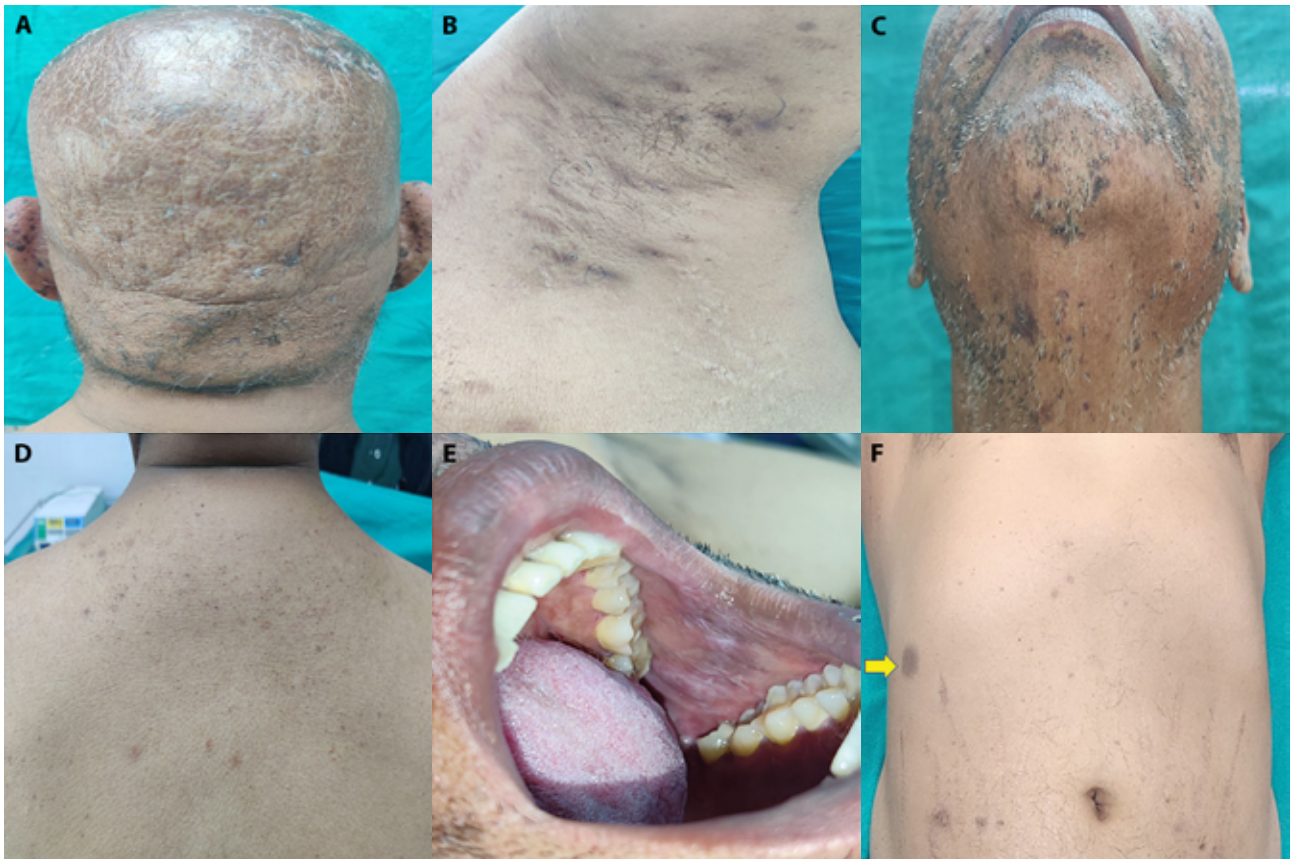

Figure 1. (A) Scarring alopecia of the scalp with few hair follicles at occipital hair line. (B) Non scarring alopecia of axilla with polyporous comedones and scarred tracts. (C) Non scarring alopecia of beard area. (D) Multiple pinpoint hyperpigmented follicular papules over upper back. (E) Reticulate whitish plaque over left buccal mucosa with surrounding hyperpigmentation. (F) Single well-defined oval hyperpigmented macule over right hypochondrium region (arrow).

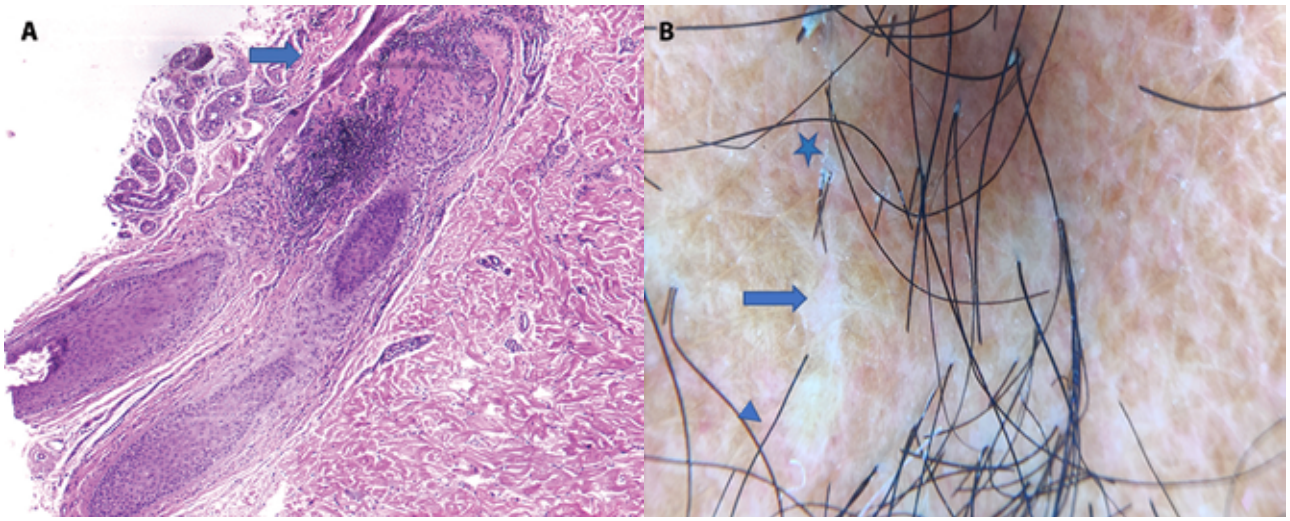

Figure 2. (A) Hisotopathological examination of a 3-mm skin punch biopsy from scalp showing perifollicular lymphocytic infiltrate (arrow) (H&E, 100x). (B) Trichoscopy from occipital area showing perifollicular casts (star), homogenous white area suggestive of scarring (arrow), and faint perifollicular erythema (triangle) (Dermlite DL3, polarized mode, ×10 magnification).

alanine aminotransferase 76 U/L, alkaline phosphatase of 301 U/L; other lab values were within normal ranges. Scalp biopsy showed perifollicular lymphocytic inflammation (Figure 2A). Trichoscopy of occipital scalp revealed loss of hair follicles with perifollicular scales, erythema, hyperpigmentation, and peri-pilar casts (Figure 2B). Based on clinical, dermoscopic and histopathological features, a diagnosis of GLPLS with quiescent HS, FDE and concurrent HCV infection was made.

Oral drug provocation test to ascertain cause of FDE was refused by the patient. Dermatological treatment was initiated

with topical tacrolimus for scalp and facial lesions, topical triamcinolone acetonide for oral lesions and clindamycin gel for axillae and groin. Patient was counseled about the permanent hair loss in scalp and to use painkillers with caution. HCV infection was managed by gastroenterology department as per protocol. Monthly follow-up showed moderate improvement in oral and cutaneous lesions of LP, no flare in HS lesions and no further episodes of FDE.

Conclusions

GLPLS is a rare entity. Associations with Hepatitis B virus vaccination, androgen insensitivity, and HLADR-1 genetic susceptibility syndrome have been described [2]. Concomitant HCV infection associated with classical LP was seen in our case. GLPLS predominantly affects middle-aged women [1]; male cases of GLPLS are rare. This case of GLPLS is reported because of extreme rarity of presentation in a male with concomitant HCV infection and incidental findings of HS and FDE.

Informed consent: Written informed consent for publication of clinical details and clinical images was obtained from the patient.

References

1. Brar B, Khanna E, Mahajan BB. Graham Little Piccardi Lasseur Syndrome: A Rare Case Report with Concomitant Hypertrophic Lichen Planus. *Int J Trichology*. 2013;5:199–200. DOI: 10.4103/0974-7753.130403. PMID: 24778531. PMCID: PMC3999651.
2. Pai VV, Kikkeri NN, Sori T, Dinesh U. Graham-little piccardi lassueur syndrome: an unusual variant of follicular lichen planus. *Int J Trichology.* 2011;3:28–30. DOI: 10.4103/0974-7753.82129. PMID: 21769233. PMCID: PMC3129120.