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GRAHAM LITTLE PICARDI LASSUEUR SYNDROME IN A MALE

Medical Science

Dr. Sowmya Naga Dogiparthi* Shri Balaji Vidhyapeeth *Corresponding Author

Vijayakumar S Shri Balaji Vidhyapeeth

Jeyakumari Jeevan Shri Balaji Vidhyapeeth

ABSTRACT

A rare variant of lichen planopilaris; Graham Little Picardi Lassueur Syndrome (GLS) is characterized by the triad of patchy cicatricial alopecia of the scalp, noncicatricial alopecia of the axilla and groin, and keratotic follicular papules of the body. The exact pathogenesis is not known;but primarily involves an immune mediated inflammatory reaction against the bulge region of hair follicles resulting in cicatricial alopecia. The disease is most commonly seen in women 30 to 70 years of age. Hence we present a case of this rare syndrome in a 32 year-old male and provide a discussion about the disease and treatment options.

KEYWORDS

INTRODUCTION

In 1914; Piccardi described Graham Little syndrome in a patient with progressive cicatricial alopecia of the scalp, non-cicatricial alopecia of the axillae and groin, and follicular lichen planus (LP) on the trunk and extremities, to which he gave the name cheratosi spinulosa (keratotic spinulosa)[1]. Later, Ernst Graham-Little published a similar case study that had been referred by Lasseur of Lausanne in 1915[2]. GLS is an unusual type of called lichen planopilaris, a variant of Lichen Planus that affects the hair follicles. This rare lichenoid dermatosis is characterized by scarring alopecia, the loss of pubic and axillary hairs, and the progressive development of spinous or accuminate follicular papules on the trunk and extremities. GLS predominantly affects women, the duration of the illness varies from 6 months to 10 years. We present the case of a 47-year-old woman with GLS. Here we report the rarity of this case in a 32 year old male[3].

CASE REPORT

A 32 year old male came to the dermatology opd with complaints of skin lesions over the scalp, chest, back and groin and genitalia for six months duration. Its onset was noted on the skin of the back. After about three weeks the papules had intensified and spread to the trunk and genitalia. Two to three months after onset, the patient observed gradual hair loss, resulting later in the alopecia foci of scalp and axilla. His skin changes were accompanied by a marked and intensified cutaneous pruritus. His past medical history was non-contributory. There was no similar family history.

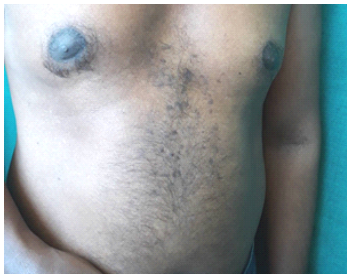

Figure 1.0: Discrete Lichenoid papules present over the chest wall.

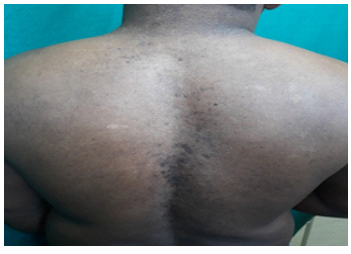

Figure 1.1: Discrete Lichenoid papules extending from the upper back till lower back.

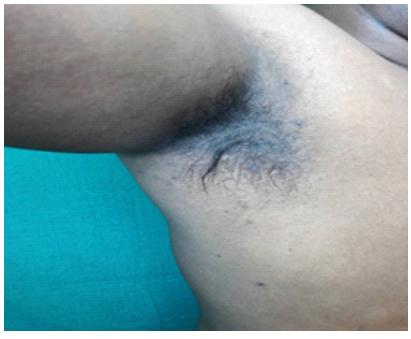

Figure 1.2: Non Scarring Alopecia of axillary hair along with lichenoid papules.

After obtaining consent from the patient; Upon examination multiple discrete multiple violaceous to hyperpigmented papules were present over both axilla , chest and lower back associated with non scarring alopecia of the axilla (*Fig1.0 -1.2*). Post inflammatory hyperpigmentation was seen over the skin of penis and scrotum. Erythematous to violaceous plaque was present over the groin and pubic area bilateral with non scarring alopecia. Further examination of the scalp showed scarring alopecia present more over the vertex and temporal region (*Fig1.3*). Nail and oral cavity examination was insignificant.

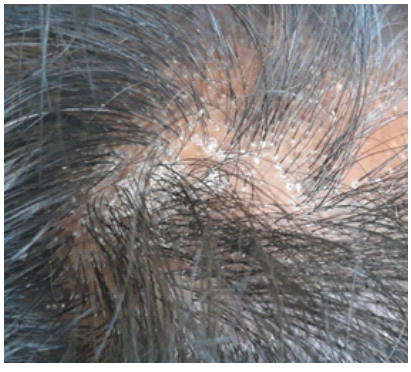

Figure 1.3: Scarring Alopecia of the scalp.

His laboratory tests were within normal limits, including a complete blood-cell count with differential, routine chemistries, liver and thyroid function tests, serum testosterone and DHEA, chest X-ray, and serum antinuclear antibodies. Further evaluation of the scalp with a non polarised dermatoscope with 200x magnification, we were able to appreciate areas of fibrosed follicle with follicular and perifollicular hyperkeratosis and perihilar cast (*Fig1.4 – Fig 1.5)*.

Figure 1.4: Dermatoscope findings: Perifollicular hyperkeratosis.

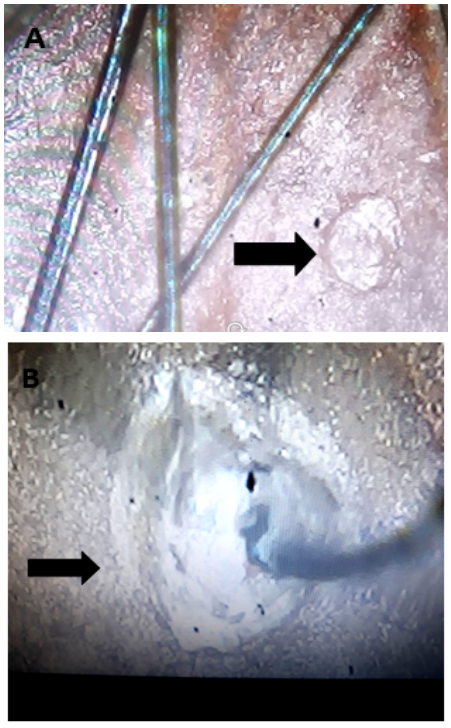

Figure 1. 5 : Dermatoscope finding : A. Scalp – Fibrosed follicle. B. Perihilar cast.

Skin biopsy specimens from the scalp revealed hyperkeratosis, acanthosis, granular layer hypertrophy, and vacuolar degeneration of the basal layer overlying both the follicular orifices and the interfollicular epithelium (*Fig 1.6*). A lichenoid infiltrate, predominately of the lymphocytes, was adherent to both the epidermis and hair follicles and the interfollicular epithelium. A lichenoid infiltrate, predominately of the lymphocytes, was adherent to both the epidermis and hair follicles (*Fig 1.7)*.

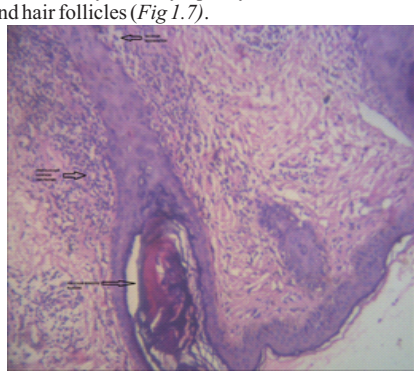

Figure 1.6: Histopathological findings: Follicular keratotic plugging, Perifollicular lichenoid; predominantly lymphocytic infiltration and Vacuolar degeneration.

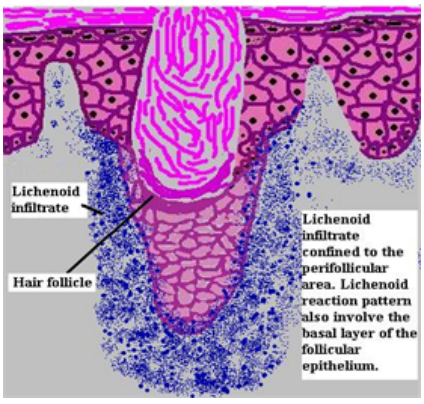

Figure 1.7: Diagramatic representation of LPP.

Based on the clinical, dermatoscopy and biopsy findings; a confirmatory diagnosis of GLS; a variant of LPP was made. Treatment included systemic prednisone in a dose of 30 mg each morning, which was decreased after approximately two weeks to 20 mg. Preparations containing corticosteroids were applied locally over the skin lesions.

DISCUSSION:

Graham-Little syndrome is characterized by the triad of multifocal cicatricial alopecia of the scalp, non-scarring alopecia of the axillae and/or groin, and keratotic follicular papules. In 1915, Graham-Little[2] described a patient's condition using the name lichen spinulosus and folliculitis decalvans, suggesting that it was a type of lichen planus. A similar case had been reported in 1914 by Piccardi[1]. The pathogenic mechanisms of GLS remain unexplained [4-110]. The disease's main target is the upper half of the pilosebaceous unit, where lymphohistiocytic infiltration begins and is associated with basal cell destruction. The follicles and sebaceous glands are gradually damaged by inflammatory infiltrates and are replaced by connective tissue, this change being represented by scarring. Damage to the follicles may be caused by pressure from the perifollicular infiltrations from one side and the keratinous plugs from the other. Follicular pressure may lead to reduced blood supply and gradual atrophy. A study report describes an autoimmune response against the INCENP centromere protein in one patient. The significance of this case remains uncertain[17]. GLS is treated topically or by systemic corticosteroids, retinoids, or PUVA therapy, each credited as having partial and temporary benefits[12-19]. Another effective treatment is the oral administration of prednisone in doses of 0.5 to 1.0 mg/kg body mass every morning with a simultaneous local application of corticosteroids and PUVA. Our patient was initially treated with 30 mg of prednisone once daily which was gradually tampered to 20mg over the weeks. The improvement noted was the gradual disappearance of lichenoid papules on the trunk and extremities. They resolved with only residual discoloration. Studies have shown that cyclosporin A was employed to a patient at a dosage of 4 mg/kg/day and produced a substantial reduction of both perifollicular erythema and follicular hyperkeratotic papules [13]. After 3 months of follow-up, some areas showed signs of hair re-growth in the scarring patches and a more consistent improvement of the follicular papules were noted. It is proven that cyclosporin A can be an effective treatment during the initial phases of this rare variant of lichen planopilaris, before the development of severe follicle damage has taken place, either by interfering with the acute inflammatory processes or by limiting the progression of the disease. The issue of alopecia merits considerable attention[12-16]. It can be challenging to distinguish lichen planopilaris from discoid lupus erythematosus, pseudopelade of Brocq, syphilis, and central centrifugal scarring alopecia (follicular degeneration syndrome). Hence it is mandatory to check a patient for signs of GLS, specifically for discrete follicular papules with no evidence of scarring on the trunk, non scarring alopecia of the axillary and pubic regions, and well-demarcated retroauricular plaques with prominent follicular papules. In our case; we are presenting this case for its rarity to occur in a 32 year old male.

REFERENCES

1. Piccardi G. Cheratosi spinulosa del capillizio a suoi rapporti con al pseudo-pelade di Brocq. Giornale Italiano della Malattie Veneree e della Pelle. 1914;49:416.
2. Graham-Little EG. Folliculitis decalvans et atrophicans. Br J Dermatol. 1915;27:183–5.
3. Horn RT, Goette DR, Odom RB et al. Immunofluorescent findings and clinical overlap in two cases of follicular lichen planus. J Am Acad Dermatol. 1982;7:203–7.
4. Matta M, Kibbi AG, Khattar J, Salman SM, Zaynoun ST. Lichen planopilaris: a clinicopathologic study. J Am Acad Dermatol. 1990;22:594–8.

5. Sachs W, De Oreo G. Lichen planopilaris: lichen planus et acuminatus atrophicans (Feldman) and lichen spinulosus and folliculitis decalvans (Little). Arch Dermatol Syph. 1942;45:1081–93.
6. Feldman S: Lichen planus et acuminatus atrophicans. Arch Dermatol Syph. 1922;5:102–13.
7. Silver H, Chargin L, Sachs PM. Follicular lichen planus (lichen planopilaris). Arch Dermatol Syph. 1953;67:346– 54.
8. Kostanecki W. Choroby wlosów. Warsaw: PZWL; 1979.
9. Wanicowa A. Przypadek zespołu Grahama-Little'a. Przegl Dermatol. 1970;57:535–8. 10. Kossard S, Lee MS, Wilkinson B. Postmenopausal frontal fibrosing alopecia: a frontal variant of lichen planopilaris. J Am Acad Dermatol. 1997;36:59–66.
11. Chew A-L, Bashir SJ, Wain EM, Fenton DA, Stefanato CM: Expanding the spectrum of frontal fibrosing alopecia: a unifying concept. J Am Acad Dermatol 2010; 63: 653-660.
12. Abbas O, Chedraoui A, Ghosn S. Frontal fibrosing alopecia presenting with components of Piccardi-LassueurGraham-Little syndrome. J Am Acad Dermatol. 2007;57(2 Suppl):S15–8.
13. Bianchi L, Paro-Vidolin A, Piemonte P, Carboni I, Chimenti S. Graham Little-Piccardi-Lassueur syndrome: effective treatment with cyclosporin A. Clin Exp Dermatol. 2001;26:518–20.
14. Lindmaier A, Jurecka W. Lichen ruber ulcerosus. Abgrenzung zum Graham-Little Syndrom. Pathogenese und Therapie. Z Hautkr. 1988;63:439–43.
15. Zinkernagel M, Trueb RM. Vernarbende Alopezien. Ther Umsch. 2002;59:243–50.
16. Zinkernagel M, Trueb RM: Fibrosing alopecia in a pattern distribution: patterned lichen planopilaris or androgenetic alopecia with a lichenoid tissue reaction pattern. Arch Dermatol. 2000;135:205–11.
17. Rodríguez-Bayona B, Ruchaud S, Rodríguez C, Linares M, Astola A, Ortiz M, Earnshaw WC, Valdivia MM. Autoantibodies against the chromosomal passenger protein INCENP found in a patient with Graham LittlePiccardi-Lassueur syndrome. J Autoimmune Dis. 2007;4:1.
18. Laymon CW. The cicatricial alopecias. J Invest Dermatol. 1974;8:99–122.
19. Sperling LC, Salomon AR, Whiting DA. A new look at scarring alopecia. Arch Dermatol. 2000;136:235–42.