Graham Little-Piccardi-Lassueur Syndrome: Case Report

Graham Little-Piccardi-Lassueur Sendromu

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ABSTRACT Graham-Little syndrome, also known as Graham Little-Piccardi-Lassueur Syndrome is characterized by the presence of cicatricial alopecia on the scalp, keratosis pilaris located to trunk and extremities, and non-cicatricial hair loss of pubis and axillae. Graham Little-Piccardi-Lassueur Syndrome is a relatively rare disease. It is four times more common in females in the age group of 30-70 years. Topical or systemic corticosteroids, retinoids or PUVA therapy are the treatments usually proposed and these have partial and temporary benefits. A 31-year-old woman presented with a 18-year history of scarring alopecia of the scalp and pruritic lichenoid papules on the trunk and extremities. On the basis of the clinical features and the histopathological findings, Graham Little-Piccardi-Lassueur syndrome (GLPLS) was diagnosed.

Key Words: Alopecia; keratosis pilaris; cicatrix

ÖZET Graham-Little sendromu, diğer adıyla Graham Little-Piccardi-Lassueur Sendromu saçlı deride ortaya çıkan skatrisyel alopesi, gövde ve ekstremitelerde keratozis pilaris ve pubis ile aksillada non-skatrisyel kıl kaybı ile karakterizedir. Graham Little-Piccardi-Lassueur Sendromu göreceli olarak nadir görülen bir hastalıktır. 30-70 yaş grubu kadınlarda dört kat daha sık görülmektedir. Tedavisinde topikal ya da sistemik kortikosteroidler, retinoidler veya PUVA tedavileri uygulanır ve bu tedavilerle genellikle kısmi ya da geçici bir düzelme sağlanır. 31 yaşında kadın hasta, 18 yıldır mevcut olan saçlı deride skatrisyel alopesi ve gövde ile ekstremitelerde lokalize pruritik likenoid papüller ile başvurdu. Klinik ve histopatolojik bulgular ışığında Graham Little-Piccardi-Lassueur sendromu (GLPLS) tanısı kondu.

Anahtar Kelimeler: Alopesi; keratozis; skatris

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Graham Little-Piccardi-Lassueur Syndrome is characterized by the presence of cicatricial alopecia on the scalp, keratosis pilaris located to trunk and extremities, and non-cicatricial hair loss of pubis and axillae. It is four times more common in females in the age group of 30-70 years. We here report a 31-year old woman with Graham Little-Piccardi-Lassueur Syndrome. A Pubmed search from 1951-2011 (all languages included) produced fewer than 20 cases of Graham Little-Piccardi-Lassueur Syndrome in the literature.

CASE REPORT

A 31-year-old woman presented with a 18-year history of scarring alopecia of the scalp and pruritic lichenoid papules on the trunk and extremities. There was a limited response to topical corticosteroids. She has no family history. In dermatologic examination nearly total scarring alopecia of the scalp and minute lichenoid papules on the back and follicular keratotic papules of the knees were observed Figure 1-3. There was no hair loss in the axillae, and a number of scarce and fine presence on the pubis. Systemic examination was normal. All routine laboratory tests including hemogram, erythrocyte sedimentation rate, lutenizing hormone (LH), testosterone, follicle stimulating hormone (FSH), oestradiol, androstenedione and DHEA-S were within normal limits. Skin biopsy from the follicular keratotic papules of knee showed an enlargement in the orifice of hair follicle, focal vacuolar degeneration of follicle epithelium and minimal lichenoid lymphocyte infiltration (Figure 4).

On the basis of the clinical features and the histopathological findings, Graham Little-Piccardi-Lassueur syndrome (GLPLS) was diagnosed.

DISCUSSION

The presence of cicatricial alopecia on the scalp, keratosis pilaris on the trunk and extremities, and non-cicatricial hair loss in the pubis and axillae was first described by Piccardi in 1914 and a year later by Graham Little in a patient referred by Lassueur.[1]

The follicular papules on the body in Graham-Little syndrome had been firstly defined as keratosis pilaris clinically but they were changed as lichen planopilaris on the basis of histopathologic findings. In some cases, non-scarring alopecia on axillae and pubis may accompany.[2,3] Although the non-scarring alopecia seen on pubic region, axillary regions were not observed hair loss, in our case. GLPLS is known as a non-familial disease. However, Viglizzo et al. reported cases of GLPLS in 47-year-old mother and her 19-year-old daughter.[2] Our case has no family history.

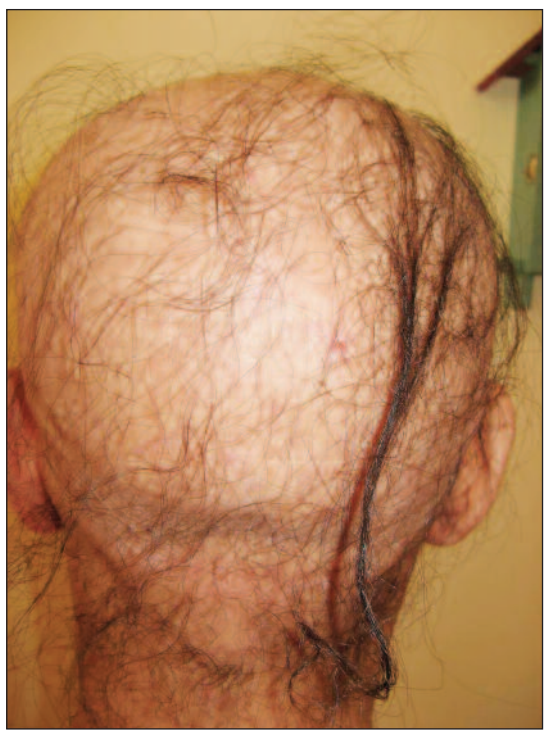

FIGURE 1: Cicatricial alopecia on the scalp.
(See for colored form http://dermatoloji.turkiyeklinikleri.com/)

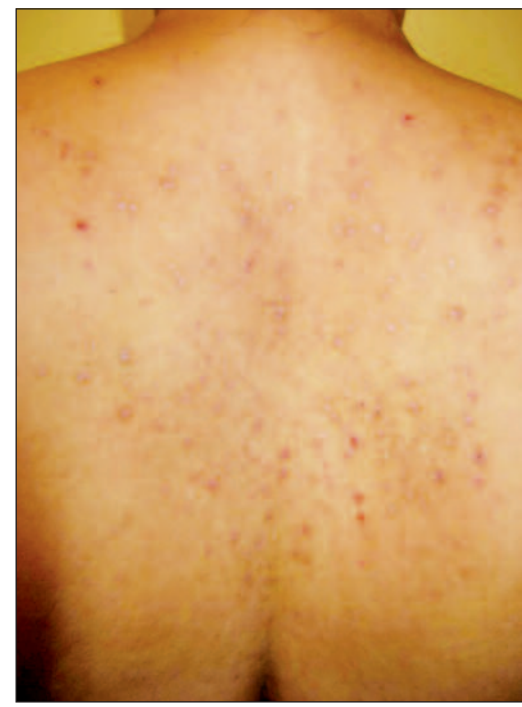

FIGURE 2: Pruritic lichenoid papules, postinflammatory hyperpigmented areas and excoriations in places on the back.
(See for colored form http://dermatoloji.turkiyeklinikleri.com/)

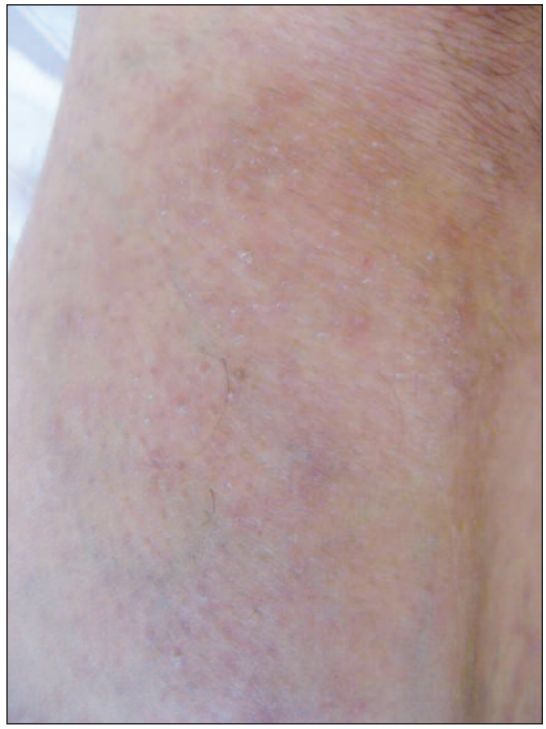

FIGURE 3: Follicular keratotic papules on the knee.
(See for colored form http://dermatoloji.turkiyeklinikleri.com/)

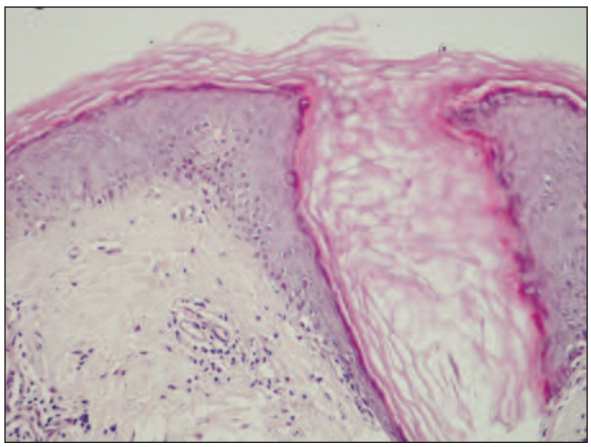

FIGURE 4: Enlargement in the orifice of hair follicle, focal vacuolar degeneration of follicle epithelium and minimal lichenoid lymphocyte infiltration (HE, x40).
(See for colored form http://dermatoloji.turkiyeklinikleri.com/)

The course of disease is slowly progressive and often chronic. Ghislain et al. reported a 50-year-old woman who initially presented with disseminated lichen planus and then progressed to the classic triad of GLPLS over a 20-year period.[3] The etiology of GLPLS is unknown; however, several hypotheses have been proposed. Viglizzo et al. documented a familial pattern of GLPLS correlated with the presence of HLA-DR1 in a mother and her daughter. Hormonal influences, stress and deficiency of vitamin A are other proposed theories.[4]

Vega-Gutiérrez et al. reported a case of GLPLS with androgen insensitivity syndrome. While the significance of these findings is unknown, the authors implied that a hormonal etiology may be associated with the non-cicatricial alopecia of the axilla and groin observed in persons with GLPLS.[4] Hormonal tests including thyroid hormones, lutenizing hormone (LH), testosterone, follicle stimulating hormone (FSH), oestradiol, androstenedione, DHEA-S, free testosterone and total testosterone were in normal limits in our patient. Topical, intralesional, and systemic corticosteroids; psoralen plus ultraviolet light A (PUVA); retinoids; and antimalarials have been used with limited success. In recent years, case reports have documented successful treatment of GLPLS with cyclosporine A, thalidomide.[5-8] Corticosteroids (topical, intralesional or systemic) have not been shown to be effective in the treatment of cicatricial alopecia associated with GLPLS, although they are moderately effective for follicular LP.[2,5]

Thalidomide at 50-150 mg/day for up to two months has been associated with significant hair regrowth of cicatricial scalp alopecia in two case reports.[5,6] However, thalidomide titrated up to 200 mg/day (from an initial 1-month dose of 100 mg/day) was reported to be ineffective in a case series of 6 patients (4 with LPP and 2 with pseudopelade of Brocq) during a 6-month open-label trial.[7] We did not give any medication to the patient because of her end-stage disease.

GLPLS is a relatively rare disease and quite a few number of cases has been reported. When the faced with similar clinical and histopathologic findings like this case, GLPLS should be suspected.

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