doi: http://dx.doi.org/10.11606/issn.1679-9836.v99i6p626-628 Rev Med (São Paulo). 2020 Nov-Dec;99(6):626-8.

Graham-Little-Piccardi-Lassueur syndrome: a case report

Síndrome de Graham-Little-Piccardi-Lassueur: relato de caso

Lucas da Silva Madureira[1], Rafael Figueiredo Gatti[1], Giovanna Guzzo[2], Márcio Custódio Borges Júnior[2], Alessandra Martins de Souza[2], João Roberto Antônio[3]

Madureira LS, Gatti RF, Guzzo G, Borges Júnior MC, Souza AM, Antonio JR. Graham-Little-Piccardi-Lassueur syndrome: a case report / *Síndrome de Graham-Little-Piccardi-Lassueur: relato de caso*. Rev Med (São Paulo). 2020 Nov-Dec;99(6):626-8.

ABSTRACT: Graham-Little-Piccardi-Lassueur syndrome is a rare variant of lichen planopilaris, whose etiology is still unknown. It is characterized by progressive scarring alopecia of the scalp, non-scarring alopecia of the axilla and pubis, and follicular lichenoid eruption. This report presents a classic case of a 74-year-old female patient with hair thinning in the right frontotemporal region, axillary and genital hypotrichosis and keratosis pilaris in the lumbar region and flanks.

Keywords: Alopecia; Atrophy; Lichen planus; Hypotrichosis; Graham-Little-Piccardi-Lassueur syndrome.

RESUMO: A Síndrome de Graham-Little-Piccardi-Lassueur é uma variante rara do líquen plano pilar cuja etiologia é desconhecida. É caracterizada pela tríade alopécia cicatricial progressiva de couro cabeludo, alopécia não cicatricial de axilas e púbis e erupção liquenóide folicular. Relatamos caso de paciente feminina de 74 anos com quadro clássico de rarefação pilosa em região frontotemporal direita, hipotricose axilar e genital e queratose pilar em região lombar e flancos.

Palavras-chave: Alopecia; Atrofia; Líquen plano; Hipotricose; Síndrome de Graham-Little-Piccari-Lassueur.

INTRODUCTION

Graham-Little-Piccardi-Lassueur syndrome is a rare variant of lichen planopilaris, which was first reported by Piccardi[1] in 1914. It is characterized by progressive scarring alopecia of the scalp, non-scarring alopecia of the axilla and pubis, and follicular lichenoid eruption. This syndrome affects mainly postmenopausal women, and its etiology is still unknown, even though there is evidence of association with an autoimmune mechanism[2].

CASE REPORT

A female 74-year-old patient, in follow up at the dermatology outpatient clinic of *Faculdade de Medicina de São José do Rio Preto* given a personal history of epithelial neoplasms, came for an appointment with the complaint of hair loss and a hair thinning area on the scalp. The patient could not answer when the hair loss started, but reported that she had noticed the alopecic area in the frontal region a few months before.

1. Dermatologist, Faculdade Estadual de Medicina de São José do Rio Preto, Brazil. ORCID: Madureira LS - https://orcid.org/0000-0001-9412-1900, Gatti RF - https://orcid.org/0000-0003-2966-5674. Email: lucas.s.madureira@gmail.com, rafa_gatti@hotmail.com.
2. Dermatologist student, Faculdade Estadual de Medicina de São José do Rio Preto, Brazil. ORCID: Guzzo G - https://orcid.org/0000-0002-8444-3524; Borges Jr MC - https://orcid.org/0000-0002-6364-807X; Souza AM - https://orcid.org/0000-0002-2579-2708. Email: giovannaguzzo@yahoo.com.br.
3. Dermatologist, Head of the Dermatology Outpatient Clinic and Emeritus Professor at Faculdade Estadual de Medicina de São José do Rio Preto, Brazil. ORCID: https://orcid.org/0000-0002-0268-5934. Email: dr.joaora@gmail.com, marciocbj76@gmail.com, lelesouza_1@hotmail.com.
Correspondence: Lucas da Silva Madureira. Hospital de Base de São José do Rio Preto. Faculdade Estadual de Medicina de São José do Rio Preto. Av. Brigadeiro Faria Lima, 5416. Vila São Pedro, São José do Rio Preto, SP, Brazil. CEP: 15090-000. E-mail: lucas.s.madureira@gmail.com

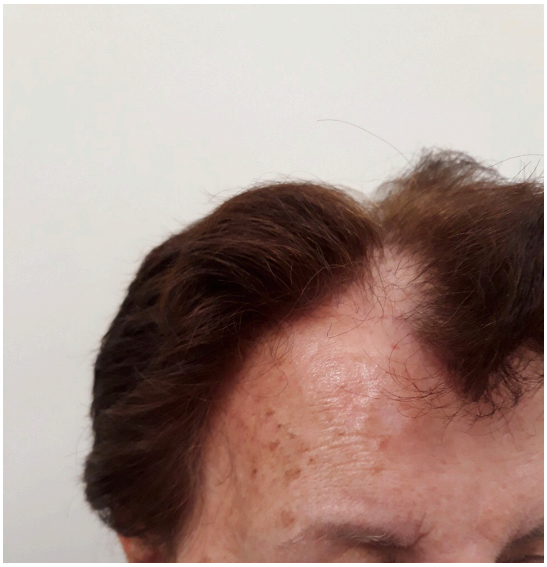

Figure 1. Alopecic area in the right frontoparietal region, with a recession of the capillary implantation line and absence of vellus hair

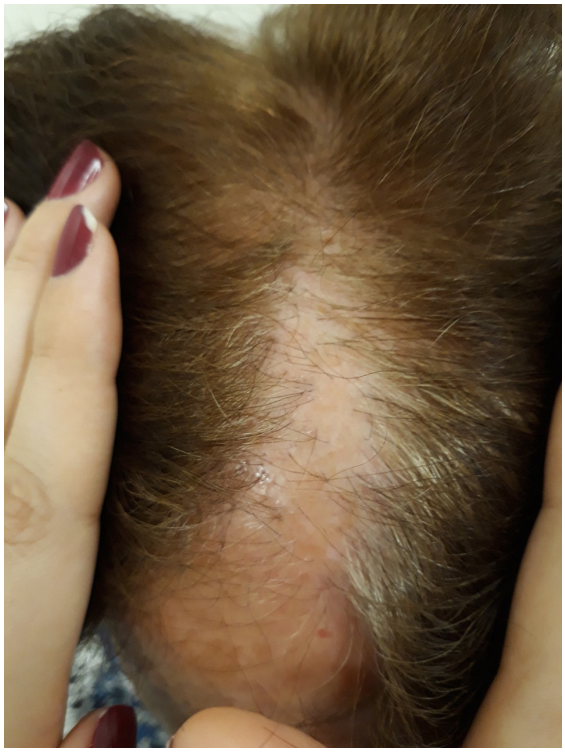

Figure 2. Extension of the alopecic area to the anterior region of the vertex, with centrifugal growth and a few terminal strands within the area

At physical examination the patient presented a well-marked area of hair thinning in the right frontoparietal region, with an extension to the vertex proximities. Dermoscopy showed perifollicular erythema and scaling, with absence of vellus hair and follicular openings. The patient also presented axillary and genital hypotrichosis, in addition to keratotic papules in the flanks, lower limbs and lumbar region, characterizing keratosis pilaris.

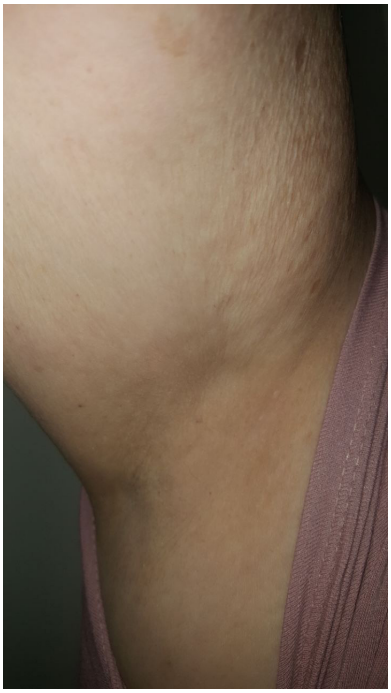

Figure 3. Axillary hypotrichosis

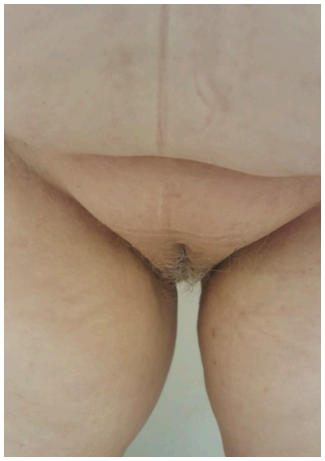

Figure 4. Genital hypotrichosis

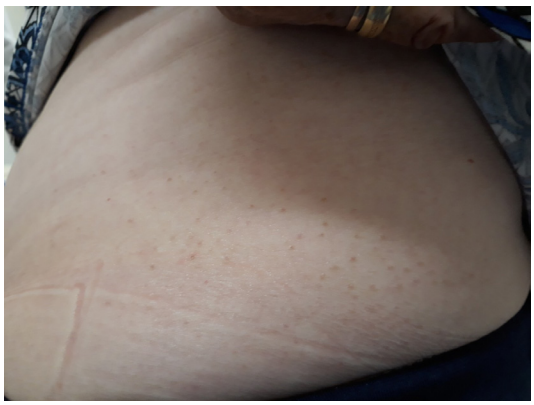

Figure 5. Keratosis pilaris lesions on the lateral surface of the gluteus and proximal third of the thigh

A histopathological examination of the scalp evidenced an interface dermatitis and fibrosis involving the hair follicle. After histopathological clinical correlation, the Graham-Little-Piccardi-Lassueur syndrome diagnosis was established. In the sequence, the patient was submitted to laboratorial tests, including for hepatitis B and C, which came negative.

The treatment of the patient was initiated with topical corticoids in a hair solution associated with hydroxychloroquine 400 mg per day, after ophthalmological assessment. The patient was followed up by the dermatology team of Faculdade de Medicina de São José do Rio Preto.

DISCUSSION

Lichen planopilaris, the follicular form of lichen planus, is an inflammatory dermatosis mediated by lymphocytes[3]. It can be subdivided into three variants: classic lichen planopilaris, frontal fibrosing alopecia and Graham-Little-Piccardi-Lassueur syndrome[3]. The syndrome is clinically characterized by axillary and genital hypotrichosis, keratosis pilaris and progressive scarring alopecia of the scalp.

Initially, there may be only follicular papules or perifollicular erythema. However, progressive hair loss may lead to the development of irregular atrophic patches of scarring alopecia on the scalp.[4] Scarring alopecia of the scalp is the earliest change for most patients. In general, alopecia of the scalp precedes keratosis pilaris for months or years[5].

Histologically, most of the early lesions to the lichen planopilaris show a band-like perifollicular lymphocytic infiltrate, focally thick, at the level of the pituitary gland and the isthmus. In addition, orthokeratosis and follicular obstruction can be observed. In more advanced lesions, perifollicular fibrosis and epithelial atrophy at the level of the pituitary gland and the isthmus are characteristically found[4].

Differential diagnoses include other forms of scarring alopecia, such as discoid lupus erythematosus, folliculitis spinulosa decalvans, pseudopelade of Brocq, follicular mucinosis, sarcoidosis, keratosis pilaris atrophicans[6].

The disease course is chronic and progressive, and its treatment aims at stabilizing the progression of the disease. Topical corticosteroids are recommended to control mild inflammations. Intralesional corticosteroids can be regularly applied. In case of inflammation aggravation, systemic therapy with hydroxychloroquine, doxycycline or oral retinoids must be associated with topical therapy. In severe cases, with severe hyperkeratosis and/or erythema, pruritus or pain, systemic treatment with corticosteroids or immunosuppressant drugs (cyclosporine, mycophenolate, methotrexate) is recommended[7].

Ethical aspects: The authors hereby state that the patient's personal information was not disclosed in this article.

Author contributions: All authors actively participated in the patient's follow up appointments, which was the study object of this case report. *Madureira LS* - was responsible for the first outpatient appointment, literature review on the disease (Graham-Little-Piccardi-Lassueur syndrome), which served as theoretical ground, and structuring of the report; *Gatti R*F - was responsible for the first outpatient appointment, literature review on the disease (Graham-Little-Piccardi-Lassueur syndrome), which served as theoretical ground, and structuring of the report; *Guzzo G* - was responsible for collecting data, interviewing the patient, and looking up the patient's medical and photographic records to build the case report, in addition to conducting the literature review on the disease (Graham-Little-Piccardi-Lassueur syndrome), which served as theoretical ground, and structuring the report; *Antonio JR* - was responsible for guiding the first appointment of the patient at the dermatology outpatient clinic of FAMERP; *Borges Jr MC* - was responsible for collecting data, interviewing the patient, and looking up the patient's medical and photographic records to build the case report; *Souza AM* - was responsible for collecting data, interviewing the patient, and looking up the patient's medical and photographic records to build the case report.

REFERENCES

1. Piccardi G. Cheratosi spinulosa del capillizio a suoi rapporti con al pseudo-pelade di Brocq. Giornale Ital Mal Veneree Pelle (Milano). 1914;49:416.
2. 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 Little-Piccardi-Lassueur syndrome. J Autoimmune Dis. 2007;4:1. doi:10.1186/1740-2557-4-1.
3. Assouly P, Reygagne P. Lichen planopilaris: update on diagnosis and treatment. Semin Cutan Med Surg, 2009; 28:3-10. doi:10.1016/j.sder.2008.12.006.
4. Elder DE, Elenitsas R, Johnson BL, Ioffreda MD, Miller J, Miller OF. Atlas and synopsis of Lever's histopathology of skin. 2nd ed. Philadelphia: Lippincott William e Wilkins; 2007. p.415-16.
5. Messenger AG, Sinclair RD, Farrant P, Berker DAR. Acquired disorders of hair. In: Rooks's textbook of dermatology. 9th ed. New York: Wiley-Blackwell; 2016. p.39-40.
6. Viglizzo G, Verrini A, Rongioletti F. Familial Lassueur-Graham-Little-Piccardi syndrome. Dermatology. 2004;208:142-4. doi: 10.1159/000076489.
7. Vañó-Galván S, Saceda-Corralo D, Pirmez R. Scarring alopecias. In: Tosti A, Asz-Sigall D, Pirmez R. Hair and scalp treatments: a practical guide. Switzerland: Springer Nature; 2020. p.140-42.

Received: 2020, April 24
Accepted: 2020, December 07