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

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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 syndrome affects mainly postmenopausal women, and its etiology is still unknown, even though there is evidence of association with an autoimmune mechanism.

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

INTRODUCTION

Graham-Little-Piccardi-Lassueur syndrome is a rare variant of lichen planopilaris, which was first reported by Piccardi¹ 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 mechanism².

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.

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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.
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. It can be subdivided into three variants: classic lichen planopilaris, frontal fibrosing alopecia and Graham-Little-Piccardi-Lassueur syndrome. 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. 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.

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.

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.

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. Intralional 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.

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 RF - 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; Gazzo 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


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