



# Graham-Little Piccardi Lassueur Syndrome - Case report \*

## Síndrome de Graham-Little Piccardi Lassueur - Relato de caso

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**Abstract:** A 33-year-old woman presented with a 3-year history of progressive alopecia of the scalp. Past treatment with hydroxycloquine did not show improvement. Physical examination revealed multiple areas of alopecia with atrophic aspect of the scalp, and axillary and pubic hypotrichosis. Dermoscopy showed hyperkeratosis and accentuation of follicular ostia. Anatomopathological examination revealed decrease in the number of hair follicles, upper perifollicular infiltrate and areas with fibrosis. The Piccardi-Lassueur-Graham-Little syndrome is a rare disorder, characterized by the triad of multifocal scarring alopecia of the scalp, keratotic follicular eruption and hypotrichosis of axillary and pubic regions. Management is a challenge and many medications tried have controversial results. We report a case of this rare syndrome which improved with corticoids.

**Keywords:** Alopecia; Hypotrichosis; Lichen planus

**Resumo:** Mulher, 33 anos, apresenta quadro de alopecia progressiva do couro cabeludo há 3 anos. Tratamento com hidroxycloquina há 12 meses, sem apresentar melhora. Ao exame físico exibe múltiplas áreas de alopecia cicatricial no couro cabeludo, além de hipotricose axilar e pubiana. A dermatoscopia evidencia hiperqueratose folicular e acentuação dos óstios foliculares. O exame anatomopatológico revela diminuição do número de folículos pilosos, infiltrado perifollicular e fibrose. A síndrome de Graham-Little Piccardi Lassueur é uma dermatose rara, caracterizada pela tríade de alopecia cicatricial multifocal do couro cabeludo, ceratose folicular disseminada e hipotricose das regiões axilares e pubianas. A terapêutica desta dermatose é um desafio, muitas medicações relatadas tem resultados controversos. Relatamos o caso desta síndrome rara que apresentou melhora com corticoterapia.

**Palavras-chave:** Alopecia; Hipotricose; Líquen plano

### INTRODUCTION

Reported initially by Piccardi, in 1913, Piccardi Lassueur Graham-Little syndrome (PLGLS) is a rare skin condition, that primarily affects women between 30 and 60 years old.<sup>1,2,3</sup> This syndrome is characterized by a triad of multifocal cicatricial alopecia of the scalp, lichenoid follicular eruption and noncicatricial alopecia of the axillae and pubis. The etiology remains unknown and controversial.<sup>4,5</sup>

### CASE REPORT

We report a case of a 33-year-old woman with

progressive alopecia of the scalp of three years duration. The patient was treated with hydroxychloroquine for a year without improvement. Physical examination revealed multiple areas of alopecia with atrophic aspect of the scalp (Figure 1). Dermoscopy of the scalp showed perifollicular hyperkeratosis and accentuation of follicular ostia (Figure 2). There was axillary and pubic hypotrichosis without skin atrophy (Figure 3). On the upper limbs and trunk there was widespread follicular keratosis (Figure 4). The patient displayed lichenoid lesions on the pretibial region, soles of feet, shoulders,

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FIGURE 1: Areas of alopecia with atrophic aspect of the scalp



FIGURE 3: Pubic hypotrichosis without skin atrophy

inter-mammary region and oral mucosa (Figure 5). Histopathologic examination showed overall decrease in the number of hair follicles, areas with perifollicular infiltrate and fibrosis, which was suggestive of lichen planopilaris (Figure 6). Treatment with systemic corticosteroids resulted in improvement of the lesions and stabilization of alopecia.

### DISCUSSION

The essential features in PLGLS are progressively multifocal cicatricial alopecia of the scalp associated with disseminated follicular hyperkeratosis and non-cicatricial axillary and pubic hypotrichosis. The clinical findings of this syndrome must be present simultaneously, even though scalp alopecia often precedes the others by months or years.<sup>1,2,4,6</sup>

Histology of the scalp lesions usually shows a pseudopelagic stage of cicatricial alopecia. The axillary and pubic alopecia is regularly described as non-

cicatricial, because there is no clinical atrophy. Histology of the papules is frequently lichenoid.<sup>1</sup>

It is estimated that over 50% of patients with PLGLS present at least one episode of cutaneous or mucous lichen planus in the course of the disease. PLGLS as lichen planus would stem from an immune response mediated by T cells against unknown antigens, determining destruction of keratinocytes.<sup>1,5</sup> The etiology of PLGLS remains unknown and controversial.<sup>3,4,6,7</sup> Most authors, based on clinical, histological and immunofluorescence consider PLGLS as a variant of lichen planus.<sup>3,6</sup> There have been cases of familial PLGLS where both patients demonstrated HLA-DR1. This HLA type is also seen in familial cases of lichen planus and provides further evidence that PLGLS is etiologically related to lichen planus.<sup>4,5,7</sup>

The differential diagnosis of PLGLS includes other causes of cicatricial alopecia, such as pseudopelade of Brocq, discoid lupus erythematosus, sarcoido-



FIGURE 2: Dermoscopy of the scalp with perifollicular hyperkeratosis and accentuation of follicular ostia



FIGURE 4: Follicular keratosis

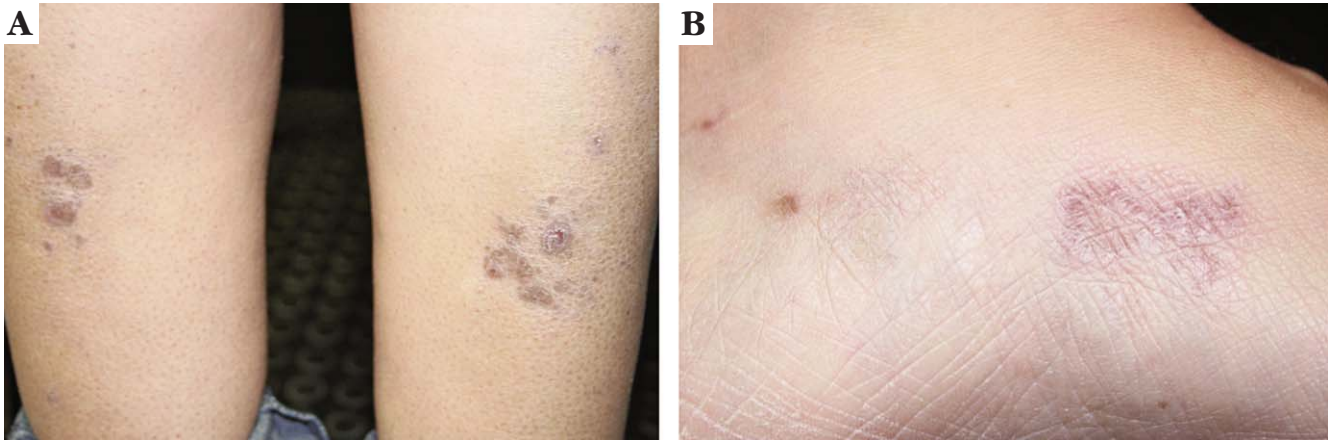


FIGURE 5: Lichenoid lesions on the pretibial region and soles of feet

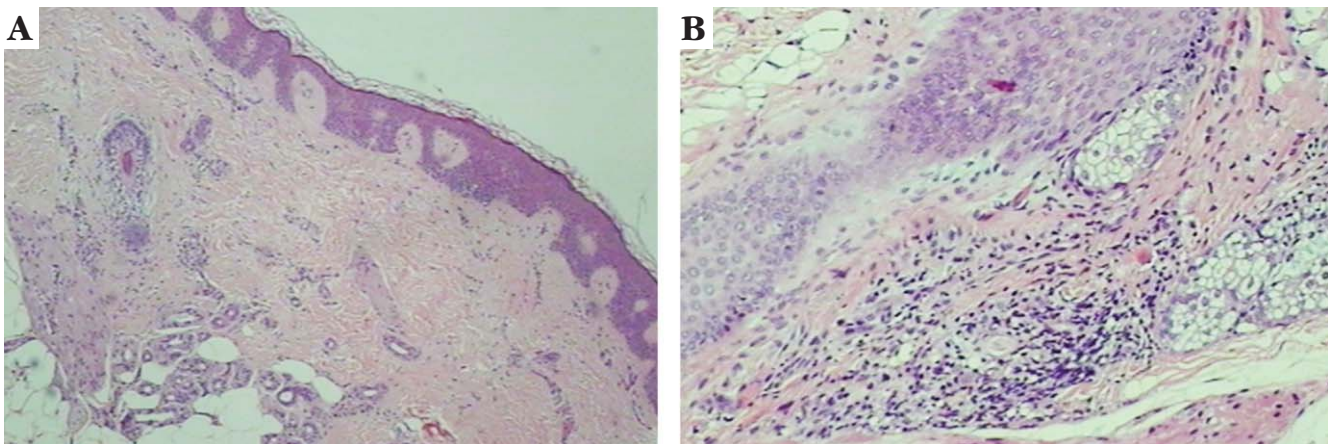


FIGURE 6: Histopathologic examination: overall decrease in the number of hair follicles, areas with perifollicular infiltrate and fibrosis

sis, follicular mucinosis, folliculitis spinulosa decalvans and keratosis pilaris atrophicans.<sup>4,7</sup>

Treatment of PLGLS is difficult with respect to both the scarring alopecia and the follicular, keratotic eruption. The scalp changes tend to be refractory to established therapies. Among the first-line therapies

are the high-potency topical corticosteroids, as well as intralesional injections of corticoids. The second-line therapy includes systemic glucocorticoids, retinoids, methotrexate, cycloporine, thalidomide and PUVA photochemotherapy, with inconsistent results.<sup>1,4</sup> □

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