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Lichen planopilaris in a Latin American (Chilean) population: demographics, clinical profile and treatment experience

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Summary

Background. Lichen planopilaris (LPP) is characterized by lymphocytic infiltrate, fibrosis and potential destruction of the hair follicle. Demographic and clinical studies in LPP are limited, and racial differences have not been thoroughly investigated.

Aim. To analyse epidemiological data and clinical profiles of Chilean adults with LPP, and report on the treatments used.

Methods. This was a retrospective review of medical records and clinical follow-up of Chilean adults with a clinical and histopathological diagnosis of LPP. Treatment response was categorized clinically as none (with progression of condition), mild or satisfactory.

Results. The study assessed 103 patients with LPP [67 women (mean age 54.1 years) and 36 men (mean age 39.1 years)]. Of the 103 patients, 41 women and 34 men were diagnosed with classic LPP (CLPP) and 26 women and 1 man with frontal fibrosing alopecia (FFA), while Graham–Little–Piccardi–Lassueur syndrome (GLPLS) was identified in 1 man. Men with CLPP had a significantly ($P < 0.001$) earlier age of onset than women. Scalp dysaesthesia, erythema and peripilar hyperkeratosis were common findings, and 51 (66%) of patients with CLPP had cicatricial patches, most of which were circumscribed in the vertex area. All patients with FFA had band-like scarring in the frontal and temporal hairlines. Morbidities associated with LPP were hypothyroidism, dyslipidaemia, hypertension and depression. For most patients, treatment halted or improved their inflammatory/scarring condition. A sustained combination of at least one topical (clobetasol, minoxidil and salicylic acid) and one systemic (cetirizine, hydroxychloroquine, finasteride, methotrexate and isotretinoin) medication was necessary in all of our patients with LPP.

Conclusion. This investigation is one of the first to describe the demographic, clinical and therapeutic features of LPP in a Latin American population. Similar profiles to previous reports may encourage research in larger multicentre international studies.

Introduction

Lichen planopilaris (LPP) is an uncommon chronic inflammatory hair disorder that may induce hair loss and scarring alopecia. Histologically, the hair follicle infundibulum and isthmus are seen to be surrounded by a lymphocytic infiltrate, with potential damage to resident stem cells and destruction of the pilosebaceous unit, resulting in permanent alopecia.[1] The pathogenesis of LPP remains unclear, but evidence points to an autoreactive inflammation to follicular antigens triggered by internal or external agents. Defects in the expression of nuclear transcriptional factors or regulators of cell mitosis as well as the role of androgens have also been suggested.[2]

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Clinically, LPP affects mostly middle-aged women, and follows a progressive course of hair loss, perifollicular erythema and scaling, often associated with itching and a burning sensation of the scalp, although other hair-bearing areas may be affected. Most patients eventually develop cicatricial patches, and the disease may have periods of activation or quiescence.[3] Three clinical variants of LPP have been identified: classic LPP (CLPP), frontal fibrosing alopecia (FFA) and Graham–Little–Piccardi–Lasseur Syndrome (GLPLS). Early diagnosis and treatment are paramount to prevent scarring and alleviate inflammatory symptoms. However, this can be challenging because the reports of effective therapies and assessment methods are inconsistent.[4] Epidemiological studies on LPP are limited (most data come from North America, Europe and Asia) and racial or ethnic differences have not been fully investigated.[5–7] In this study, we analyse demographic, clinical profiles and treatments used in Chilean adults with LPP.

Methods

Medical records of adult Chileans with a clinical and histological diagnosis of LPP from 2010 to 2014 were obtained from the Dermatology Department of the University of Chile Clinical Hospital and from a private practice. After obtaining informed consent from each case, data on demographics, medical background, clinical features and treatments were recorded, with follow-up every 3 months for 12 months in total. Treatment response was clinically recorded as: no response (with progression of condition), mild (≤ 50%) improvement or satisfactory (> 50%) depending on the intensity of reduction/absence of inflammation or hair loss. Statistical evaluation was performed using STATA® (v12; StataCorp LLC, College Station, TX, USA), with analysis of independent variables performed using Student *t*-test.

Results

The study assessed 103 patients [67 (65%) women (mean age 54.1 years and 36 (35%) men (mean age 39.1 years.)].

Demographic and clinical features

Table 1 shows the demographic and clinical features of Chilean adults with LPP. Of the 103 patients, 75 (72.8%) were diagnosed with CLPP [41 (54.7%) women (mean age 48.8 years) and 34 (45.3%) men (mean age 37.7 years)]. There was a statistically significant sex difference in age of onset, with men having onset at a significantly ($P < 0.001$) younger age. FFA was observed in 27 patients (26.2%) [26 (96.2%) women (mean age 62.9 years) and 1 (3.8%) man (aged 53 years)]. We found only one case (1%) of GLPLS, which was a 50-year-old man.

Treatment

The treatment protocol for LPP was carried out in a stepwise manner, beginning with topical formulations and moving up to systemic treatment if there was no response after 3 months. Overall, patients commonly required a combination of both. Table 2 summarizes the treatment regimens.

Regarding topical treatment, clobetasol 0.05% in foam or lotion was used by all patients, while minoxidil 2–5% lotion was used by 16 patients (21.3%) with CLPP and 15 patients (55.6%) with FFA. Salicylic acid 1% lotion was used by 52 patients (50.5%) with LPP.

Table 1 Demographic and clinical features of our cohort of Chilean adults with LPP.

	CLPP	FFA	GLPLS	Total
Patients, *n*, F/M	41/34	26/1	0/1	103
F/M ratio	1.2	26	–	1.9
Age of onset, years	44.5 F, 34.3 M	57.5 F, 52 M	50 M	
Morbidities, *n* (%)	40 (53.3): depression; insulin resistance; hypothyroidism; hypertension	23 (85.2); hypothyroidism; dyslipidaemia; hypertension	–	73 (70.9)
Scalp dysaesthesia, *n* (%)	60 (88)	8 (30)	Yes	69 (67)
Scalp erythema and peripilar hyperkeratosis, *n* (%)	53 (71) crown area; 22 (29) diffuse	27 (100)	Yes	27 (100)
Scalp scarring, *n* (%)	31 (41) small circumscribed; 20 (27) large coalescent	Yes: all cases band-like	Yes	–
Lichen planus other than scalp, *n* (%)	2 (2.7) face, axillae	0	–	2

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Table 2 Treatment regimens in study group.

Type of treatment	CLPPC	FFA	GLPLS	Total
Topical, *n* (%)				
Clobetasol shampoo/lotion	75 (100)	27 (100)	1 (100)	103 (100)
Minoxidil lotion 2% or 5%	16 (21.3)	15 (55.6)	1 (100)	32 (31.1)
Salicylic acid lotion 1%	41 (40)	10 (40)	1 (100)	52 (50.5)
Intralesional corticosteroid	4 (4)	0 (0)	0 (0)	4
Systemic, *n* (%)				
Cetirizine	75 (100)	27 (100)	1 (100)	103 (100)
Hydroxychloroquine	15 (20)	7 (26)	0 (0)	22 (21.4)
Finasteride	10 (13.3)	8 (29.6)	0 (0)	18 (17.5)
Methotrexate	5 (6.7)	0 (0)	0 (0)	5 (4.9)
Isotretinoin	4 (4)	0 (0)	0 (0)	4 (3.9)

CLPP, classic lichen planopilaris; FFA, frontal fibrosing alopecia; GLPLS, Graham–Little–Piccardi–Lasseur.

As for systemic treatment, all patients were treated with cetirizine 5 mg/day. In addition, 15 patients (20%) with CLLP and 7 (26%) with FFA were treated with hydroxyhydroxychloroquine 200–400 mg/day; 10 patients (13.3%) with CLLP and 8 (29.6%) with FFA were treated with finasteride 1–2.5 mg/day; and 9 patients with CLPP had other treatments: methotrexate 7.5–15 mg/day for 5 patients (6.7%) and isotretinoin 20 mg/day for 4 (4%).

After 12 months of treatment, none of the patients had achieved complete clinical remission of inflammatory signs and symptoms or of hair loss. Mild and satisfactory improvement results were recorded for 49 (65.3%) and 20 (74.1%) cases of CLPP and FFA respectively, and 26 (34.7%) patients with CLPP and 7 (25.9%) with FFA were categorized as progressive nonresponders.

Discussion

LPP is the most common cause of primary cicatricial alopecia, which is characterized by a chronic lymphocytic infiltrate and potential destruction of the hair follicles. Although recent clinical reports suggest a global increase in its incidence, there is a paucity of demographic and clinical studies in Latin America.

Updated epidemiological data shows that LPP is more common in women (ratios range from 1.8 to 9) and it usually starts in adulthood.[8] Overall, disease onset in all clinical variants is around the fifth decade, but interestingly, some reports indicate that onset of CLPP begins at a younger age in men.[9,10] In our study, we also found a female predominance (ratio of 1.9), but this differed according to clinical presentation (1.2 for CLPP and 26 for FFA), and men with CLPP had a statistically significantly earlier disease onset (34.3 vs. 44.5 years). This difference may be explained by the higher androgenic activity in men, which may influence the sebaceous gland function and local immune status. Additionally, scalp inflammation and hair loss in women may be attributed to other more common causes, delaying diagnosis.

Studies have reported an association between LPP and morbidities, especially thyroid disease.[11] Although in our study, the incidence of concomitant disease was high (mostly hypothyroidism, hypertension and metabolic disorders), age-matched controlled studies are necessary to clarify this result. Depression was also a common finding in patients with CLPP, but it was not clear if this acted as a triggering factor or a secondary event.

LPP subtypes have common clinical features, but they can differ in location and extent, which may reflect disease activity. In our investigation, scalp dysaesthesia (itching, tenderness and burning sensation) was more common in CLPP (88%) than FFA (30%). In a multicentre study[12] of 335 FFA Spanish patients, 30% reported pruritus and 20% trichodynia. Erythema, follicular hyperkeratosis and alopecic scarring (and nonscarring) patches are key features of LPP (Fig. 1a) In CLPP, these signs may be localized or diffuse and may become interconnected, but in FFA, there is a particular distribution on the frontal, frontotemporal and temporal areas, together with eyebrow loss (Fig. 1b). In our study, the majority of patients with CLLP had inflammatory signs in the vertex area but almost 30% had a more disseminated condition. Additionally, 41% had small localized cicatricial patches and 27% had large and coalescent patches at the time of diagnosis. In FFA, all of the patients had band-like scarring on the hairline margins of the scalp. These findings may reflect the delayed diagnosis factor

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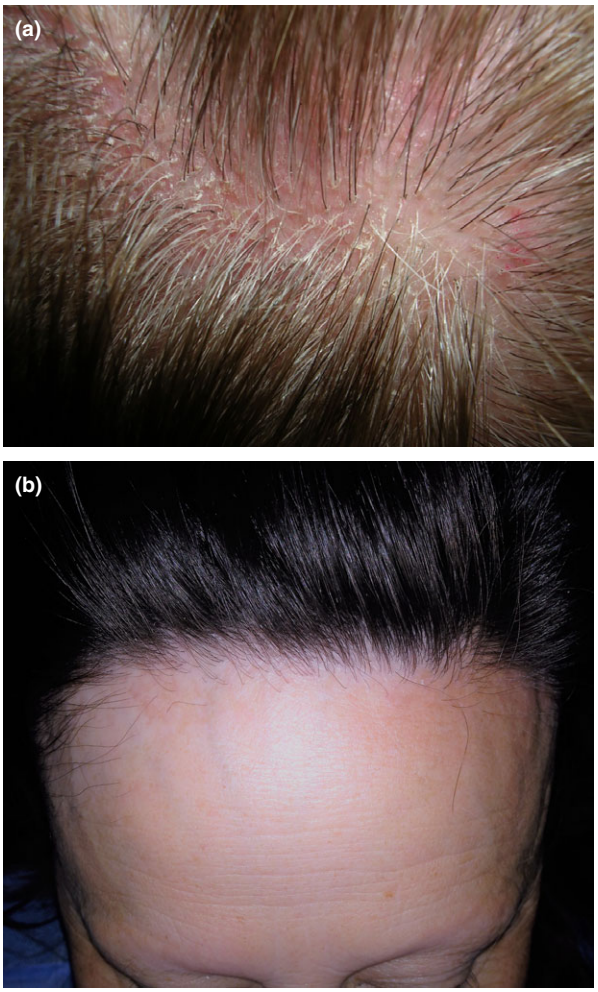

Figure 1 (a) Scalp erythema, perifollicular scaling and cicatricial patches; (b) cicatricial hair loss in frontal and temporal areas and eyebrow loss.

of FFA, which reaffirms the importance of early detection and treatment of this condition. Unlike previous reports, only 2.7% of our patients had lichen planus-like lesions in other body areas, mostly on the face and axillae.

The best approach and treatment regimen for LPP are still uncertain, owing to the lack of high-grade evidence from designed trials, consistent response-assessment methods and the intrinsically variable course of the disease.[13] The principal therapeutic goals are to lessen symptomatology, and prevent hair loss and scarring. It is common for multiple treatments to be needed to obtain effective and long-term stabilization. Overall, treatment options include local, systemic and cosmetic (camouflaging) measures. Expert recommendations for CLLP and FFA are similar, and include topical steroids and/or tacrolimus, intralesional steroids, or minoxidil as first-line therapy for acute, small and circumscribed lesions. For patients who respond poorly to these or have an extensive rapidly progressive condition, systemic drugs (corticosteroid, doxycycline, hydroxychloroquine, ciclosporin, mycophenolate mofetil, oral retinoids and peroxisome proliferator-activated receptor-γ agonists) may be indicated. Additionally, benefit from 5α-reductase inhibitors (finasteride and dutasteride) has been reported in FFA. In our local treatment protocol, we prefer topical clobetasol 0.05% in shampoo or lotion over intralesional corticosteroid as it is better tolerated and shown to be effective. In almost a third of the cases (mostly FFA), 2% or 5% minoxidil lotion was also required. All of the cases received cetirizine to relieve pruritus, and around 20% of the patients (similar numbers with CLPP or FFA) were prescribed hydroxychloroquine when there was little clinical improvement with corticosteroids. Only 17.5% of the patients received finasteride, but interestingly, the percentage of FFA was higher than CLPP (29% vs. 13%). This may reflect differences in the role of androgens in the physiopathology of LPP clinical variants. We prescribed methotrexate and oral isotretinoin in few cases of CLLP; the first in nonresponders to topical corticosteroids and hydroxychloroquine, and the latter in men with scalp hyperseborrhoea. Previous studies demonstrate that the natural history of LPP is variable; some patients stabilize over time while others progress despite therapeutic interventions. Although none of our patients achieved full remission after 1 year of treatment, only 34.7% of patients with CLPP and 25.9% of patients with FFA were categorized as nonresponders.

Conclusion

This investigation is one of the first to analyse the demographic, clinical and treatment features of LPP in a Latin American population. In our group of Chilean adults, there was a higher female predominance in FFA than in CLLP, whereas men had an earlier disease onset with CLLP. Inflammatory signs, symptoms and cicatricial alopecic patches were common in our patients, and the majority needed continuous topical and systemic treatment. In agreement with previous publications, early diagnosis and new multicentre therapeutic trials are necessary to manage this disabling condition.

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What's already known about this topic?

• LPP is an uncommon hair disorder that may cause scarring alopecia.
• International clinical studies in LPP are limited, and racial differences have not been investigated extensively.

What does this study add?

• There were demographic and clinical similarities between a Latin American (Chilean) study group and previous reports, including a higher prevalence in women and an earlier age of disease onset in men.
• Inflammation and cicatricial alopecic patches were common in our patients.
• The majority of patients needed both topical and systemic treatment.

References

1 Olsen E, Stenn K, Bergfeld W *et al.* Update on cicatricial alopecia. *J Investig Dermatol Symp Proc* 2003; **8**: 18–19.
2 Harries MJ, Paus R. The pathogenesis of primary cicatricial alopecias. *Am J Pathol* 2010; **177**: 2152–62.
3 Mubki T, Rudnicka L, Olszewska M, Shapiro J. Evaluation and diagnosis of the hair loss patient: part I. History and clinical examination. *J Am Acad Dermatol* 2014; **71**: 415.e1–15.
4 Siah TW, Shapiro J. Scarring alopecias: a trichologic emergency. *Semin Cutan Med Surg* 2015; **34**: 76–80.
5 Ochoa BE, King LE Jr, Price VH. Lichen planopilaris: Annual incidence in four hair referral centers in the United States. *J Am Acad Dermatol* 2008; **58**: 352–3.
6 Qi S, Zhao Y, Zhang X *et al.* Clinical features of primary cicatricial alopecia in Chinese patients. *Indian J Dermatol Venereol Leprol* 2014; **80**: 306–12.
7 Meinhard J, Stroux A, Lünnemann L *et al.* Lichen planopilaris: epidemiology and prevalence of subtypes – a retrospective analysis in 104 patients. *J Dtsch Dermatol Ges* 2014; **12**: 229–36.
8 Assouly P, Reygagne P. Lichen planopilaris: update on diagnosis and treatment. *Semin Cutan Med Surg* 2009; **28**: 3–10.
9 Chiang C, Sah D, Cho BK *et al.* Hydroxychloroquine and lichen planopilaris: efficacy and introduction of Lichen Planopilaris Activity Index scoring system. *J Am Acad Dermatol* 2010; **62**: 387–92.
10 Lyakhovitsky A, Amichai B, Sizopoulou C, Barzilai A. A case series of 46 patients with lichen planopilaris: demographics, clinical evaluation, and treatment experience. *J Dermatolog Treat* 2015; **26**: 275–9.
11 Atanaskova Mesinkovska N, Brankov N *et al.* Association of lichen planopilaris with thyroid disease: a retrospective case-control study. *J Am Acad Dermatol* 2014; **70**: 889–92.
12 Vañó-Galván S, Molina-Ruiz AM, Serrano-Falcón C *et al.* Frontal fibrosing alopecia: a multicenter review of 355 patients. *J Am Acad Dermatol* 2014; **70**: 670–8.
13 Harries MJ, Messenger A. Treatment of frontal fibrosing alopecia and lichen planopilaris. *J Eur Acad Dermatol Venereol* 2014; **28**: 1404–5.

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