Trichoscopy as an Additional Diagnostic Tool for Monitoring of Lichen planopilaris

Trichoskopie als diagnostische Hilfe in der Beobachtung eines Lichen planopilaris

Authors J. Andziukeviciute[1], J. Makstiene[2], S. Valiukeviciene[1]

Institutions
[1] Department of Skin and Venereal Diseases, Lithuanian University of Health Sciences, Medical Academy, Kaunas, Lithuania
[2] Department of Pathology, Lithuanian University of Health Sciences, Medical Academy, Kaunas, Lithuania

Bibliography
DOI http://dx.doi.org/10.1055/s-0042-108009
Published online: 24.5.2016
Akt Dermatol 2016; 42: 280–282

ISSN 0340-2541

Corresponding author
Skaidra Valiukeviciene
Professor and Head
Department of Skin and Venereal Diseases
Medical Academy, Lithuanian University of Health Sciences, Kaunas
Eiveniu 2
50009 Kaunas
Litauen
skaidra.valiukeviciene@kaunoklinikos.lt

Abstract

Lichen planopilaris (LPP) is a rare inflammatory disorder selectively involving hair follicles and leading to scarring alopecia. This disease is clinically characterised by follicular hyperkeratosis, perifollicular erythema and permanent hair loss mainly on the scalp. A 34-year-old woman was admitted with hair loss and itching of the scalp. Based on clinical symptoms, trichoscopy and histological findings we diagnosed classic LPP. The patient was treated with topical corticosteroids, but this therapy was ineffective. The remission of the disease was achieved after 2 months treatment with hydroxychloroquine 200 mg twice daily. Trichoscopy can be used as an additional LPP diagnostic tool for the monitoring of inflammation associated with this disease.

Case report

A 34-year-old woman was presented with complaints of increased hair shedding and episodic itching of the scalp. Symptoms appeared 3 years ago. Physical examination revealed areas of 1 – 2 cm in diameter with cicatricial alopecia in temporal and occipital regions (**Fig. 1**). Hair pull test was negative. Trichoscopy showed zones of extinct hair follicules, follicular hyperkeratosis and erythema (**Fig. 2**). Patch test with 29 European standard allergens identified contact allergy to paraphenylenediamine. Punch 5 mm biopsy was accomplished from two sites of affected skin. Dermatohistopathology revealed dilated hair follicles with perifollicular lymphocytic infiltrate. There were no mucine deposits in dermis. Normal basal membrane without vacuolar degeneration was seen (**Fig. 3 a, b, c**).

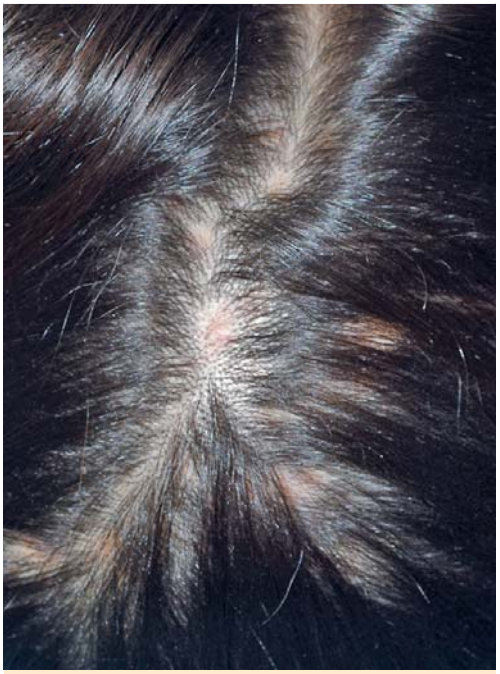

Fig. 1 Macroscopic view of the scalp before treatment.

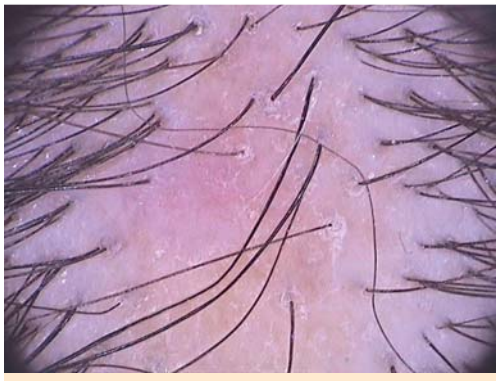

Fig. 2 Trichoscopy (×20 magnification): the bald area with zones of extinct hair follicules, follicular hyperkeratosis and perifollicular erythema.

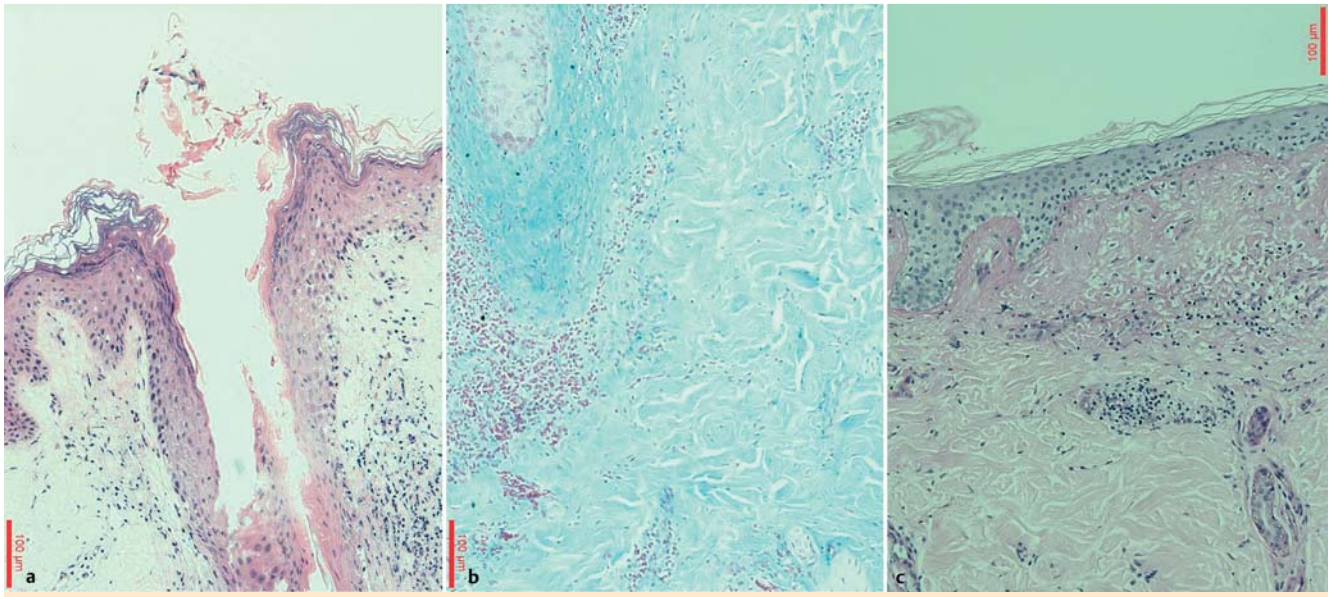

Fig. 3 Dermatohistopathology: a dilated hair follicle with perifollicular lymphocytic infiltrate (HE stain) (**a**), mucine deposits in dermis were not found (Alcian blue stain) (**b**), normal basal membrane without vacuolar degeneration (PAS staining) (**c**).

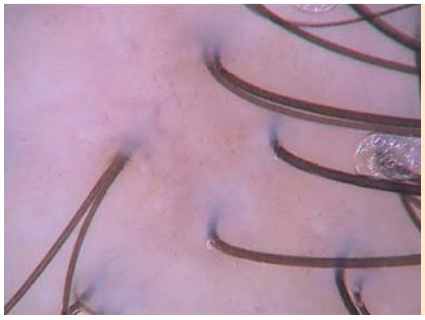

Fig. 4 Trichoscopy (×70 magnification) after treatment: white areas without ostia of hair follicules.

Laboratory test results, including a complete blood count, liver enzymes, creatinine, glucose tests, showed no pathological changes. The diagnosis of classic LPP was confirmed by clinical evaluation, trichoscopy and histopathological findings. The patient was treated with clobetasol propionate solution for 3 months without significant improvement, even progression of the disease was observed. The systemic therapy with hydroxychloroquine 200 mg twice daily started. Two months after treatment, the development of new lesions has been stabilized. Trichoscopy showed white areas without ostia of hair follicules in the affected areas of scalp, which is characteristic to late stage of classic LPP (**Fig. 4**).

Discussion

Lichen planopilaris (lichen follicularis) was described for the first time by Pringle in 1895 [1]. LPP is a rare disorder of the skin that leads to hair follicle destruction mainly on the scalp [2]. In early stage of the disease inflammation with an increase of $CD8^+$: $CD4^+$ T lymphocytes ratio around the infuldibuloisthmic area appears. Cytotoxic immune response affects stem cells of the hair follicule, that leads to destruction of hair follicules and development of scarring alopecia [3].

LPP is a primary scarring alopecia and is divided into three clinical types: classic lichen planopilaris (presented in this case report), frontal fibrosing alopecia and Graham-Little-Piccardi-Lassueur syndrome [4]. All types of the disease progress lead untreated to irreversible hair loss. It is very important to diagnose LPP on time and to treat it adequately.

LPP is associated with various endogenous triggers such as hypothyroidism [5], and a medication such gold or para-amino-salicylic acid [4] or TNF-α inhibitors [6, 7]. There are several reports of LPP onset after hair transplantation [8, 9]. Classic LPP affects 5 % of patients suffering from alopecia [10, 11] and 10 % to 28 % of subjects with cicatricial alopecia [4, 11, 12]. It is more common in females at adult age [10, 13]. More than half of patients with LPP complain of pruritus or pain, some mention burning of the scalp (6 % to 21 %) [12 – 14]. According to reports, hair loss is asymptomatic in 40 % of cases [13]. All parts of the scalp might be affected including frontal and occipital scalp areas, but most commonly parietal area [14]. A positive hair pull test shows active disease [2].

The Cleveland Clinic Foundation's retrospective review of histological samples and medical records showed that follicular hyperkeratosis (69 %) and perifollicular erythema (62 %) are most common symptoms in the active stage of LPP [14]. Histopathological findings, such as lichenoid interface inflammation at infundibulum and isthmus of hair follicules and destruction of sebaceous epithelium are important for differential diagnosis at the beginning of LPP. In late stage of the disease perifollicular lamellar fibrosis appears [4, 12]. Direct immunofluorescence reveals IgM and IgA in cytoid bodies and patchy or linear fibrinogen deposition along the basement membrane of hair folllicules [15]. Habashi-Daniel A. et al. reported that in active LPP a decrease or extinction of catagen and telogen phase of hair follicules is detected [16].

Trichoscopy is used as an additional LPP diagnostic tool, which was also important in our case. According to expert recommendation [17], trichoscopy of LPP in early stage shows a tubular perifollicular scaling, violaceous areas and elongated linear blood vessels. Irregular white dots, white and milky red areas without hair follicules reflect late stage of the disease.

Treatment of LPP is very controversial and based on published case reports, recommendations of experts or monocentric clinical studies with small numbers of cases. Chieregato C. et al. reported a study with 30 patients, who received treatment with topical steroids for 12 weeks. Stabilization of the disease was seen in 66% of patients, 20% of patients achieved mild improvement [18]. However in other studies monotherapy with topical corticosteroids was less effective. Lyakhovitsky A. et al. reported a retrospective study with 46 patients, that showed only 4,8% of patients who reached remission of the disease. This study also presented 86,7% of patients with partial remission after injections of corticosteroids [13]. Chiang C. et al. presented encouraging results of treatment with hydroxychloroquine in a retrospective study. Authors observed significant improvement (89%) of the disease after 12 months treatment with hydroxychloroquine 200 mg twice daily [19]. Oral retinoids may be effective for patients with LPP [11,20]. L. A. Spencer et al. retrospective analysis shows that oral acitretine 25 mg daily for 3 – 6 months can relieve the symptoms. Authors also suggested systemic therapy with doxycycline [11]. Doxycycline besides its anti-infectious effects has immunomodulating properties [21]. It was reported as an effective treatment in retrospective studies using 100 mg twice daily for 3 – 6 months [11]. Several studies showed successful treatment with mycophenolate mofetil [11,22]. B. K. Cho et al. observed good response for treatment with mycophenolate mofetil 0,5 g twice daily for 4 weeks, then 1 g twice daily for 20 weeks and longer [11,22]. However, this treatment should be discussed critically because of side effects. Mirmirani P. et al. presented treatment with oral cyclosporine 3 – 5 mg/kg 3 – 5 months, for those patients who did not respond to therapy with hydroxycholoroquine [23].

When the remission of the disease is confirmed histologically, there is the opportunity to perform hair transplantation, but not during the active stage of LPP [2].

In our case, the patient did not respond to treatment with high-potency topical corticosteroids. However our report demonstrates a positive course of LPP under treatment with hydroxychloroquine 200 mg twice daily. Trichoscopy can be used as an additional LPP diagnostic tool for monitoring of inflammation associated with this disease.

Acknowledgment

We would like to express sincere gratitude for Prof. Ulrike Blume-Peytavi and her team from Charité – Universitätsmedizin Berlin, Department of Dermatology and Allergy, Clinical Research Center for Hair and Skin Science, for the consultations and education in the field of trichology.

Conflict of Interest

The authors declare no conflict of interest.

Zusammenfassung

Trichoskopie als diagnostische Hilfe in der Beobachtung eines Lichen planopilaris

Der Lichen planopilaris (LPP) ist eine entzündliche Hauterkrankung, die vorwiegend die Haarwurzeln betrifft, lokalisiert am behaarten Kopf, und führt zur vernarbender Alopezie. Der LPP ist charakterisiert durch einen progessiven Haarausfall, ein perifollikuläres Erythem und Hyperkeratose. Eine 34-jährige Frau stellte sich mit den Beschwerden Haarausfall und Jucken am behaartem Kopf vor. Gemäß der klinischen Symptome, der trichoskopischen und histologischen Untersuchung wurde eine klassische Form des LPP diagnostiziert. Die topische Steroidtherapie zeigte kein Ansprechen. Eine Verbesserung des Krankheitsverlaufs wurde nach der 2-monatigen Behandlung mit Hydroxychloroquine 200 mg 2-mal pro Tag erreicht. Die Trichoskopie wurde als hilfreiche Diagnostik für den Verlauf der Entzündung eingesetzt.

References

1 *Pringle JJ*. Cited by Adamson HG. Lichen pilaris spinulosus. Br J Dermatol 1905; 17: 78 – 79
2 *Assouly P, Reygagne P*. Lichen planopilaris: update on diagnosis and treatment. Semin Cutan Med Surg 2009; 28: 3 – 10
3 *Mobini N, Tam S, Kamino H*. Possible role of the bulge region in the pathogenesis of inflammatory scarring alopecia: lichen planopilaris as the prototype. J Cutan Pathol 2005; 32: 675 – 679
4 *Whiting DA*. Cicatricial alopecia: clinico-pathological findings and treatment. Clin Dermatol 2001; 19: 211 – 225
5 *Mesinkovska NA, Brankov N, Piliang M* et al. Association of lichen planopilaris with thyroid disease: a retrospective case-control study. J Am Acad Dermatol 2014; 70: 889 – 892
6 *Fernandez-Torres R, Paradela S, Valbuena L* et al. Infliximab-induced lichen planopilaris. Ann Pharmacother 2010; 44: 1501 – 1503
7 *Garcovich S, Manco S, Zampetti A* et al. Onset of lichen planopilaris during treatment with etanercept. Br J Dermatol 2008; 158: 1161 – 1163
8 *Chiang YZ, Tosti A, Chaudhry IH* et al. Lichen planopilaris following hair transplantation and face-lift surgery. Br J Dermatol 2012; 166: 666 – 670
9 *Donovan J*. Lichen planopilaris after hair transplantation: report of 17 cases. Dermatol Surg 2012; 38: 1998 – 2004
10 *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 – 235
11 *Spencer LA, Hawryluk EB, English JC3rd*. Lichen planopilaris: retrospective study and stepwise therapeutic approach. Arch Dermatol 2009; 145: 333 – 334
12 *Tan E, Martinka M, Ball N* et al. Primary cicatricial alopecias: clinicopathology of 112 cases. J Am Acad Dermatol 2004; 50: 25 – 32
13 *Lyakhovitsky A, Amichai B, Sizopoulou C* et al. A case series of 46 patients with lichen planopilaris: demographics, clinical evaluation, and treatment experience. J Dermatolog Treat 2015; 26: 275 – 279
14 *Cevasco NC, Bergfeld WF, Remzi BK* et al. A case-series of 29 patients with lichen planopilaris: the Cleveland Clinic Foundation experience on evaluation, diagnosis, and treatment. J Am Acad Dermatol 2007; 57: 47 – 53
15 *Mehregan DA, Van Hale HM, Muller SA*. Lichen planopilaris: clinical and pathologic study of forty-five patients. J Am Acad Dermatol 1992; 27: 935 – 942
16 *Habashi-Daniel A, Roberts JL, Desai N* et al. Absence of catagen/telogen phase and loss of cytokeratin 15 expression in hair follicles in lichen planopilaris. J Am Acad Dermatol 2014; 71: 969 – 972
17 *Rudnicka L, Olszewska M, Rakowska A*. Atlas of trichoscopy: dermoscopy in hair and scalp disease. London: Springer; 2012
18 *Chieregato C, Zini A, Barba A* et al. Lichen planopilaris: report of 30 cases and review of the literature. Int J Dermatol 2003; 42: 342 – 345
19 *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 – 392
20 *Spano F, Donovan JC*. Efficacy of oral retinoids in treatment-resistant lichen planopilaris. J Am Acad Dermatol 2014; 71: 1016 – 1018
21 *Thong YH, Ferrante A*. Inhibition of mitogen-induced human lymphocyte proliferative responses by tetracycline analogues. Clin Exp Immunol 1979; 35: 443 – 446
22 *Cho BK, Sah D, Chwalek J* et al. Efficacy and safety of mycophenolate mofetil for lichen planopilaris. J Am Acad Dermatol 2010; 62: 393 – 397
23 *Mirmirani P, Willey A, Price VH*. Short course of oral cyclosporin in lichen planopilaris. J Am Acad Derm 2003; 49: 667 – 671