Cicatricial (scaring) alopecia

Definition: follicular epithelium been replaced by scarring tissue.

Permanent injury of the follicular stem cell region has occurred.

Primary cicatricial alopecia:
Idiopathic folliculocentric inflammatory process destroys the hair follicle
Secondary cicatricial alopecia:
Any cutaneous inflammatory process of the scalp skin or by physical trauma
討論內容:

# Primary cicatricial alopecia
Idiopathic folliculocentric inflammatory process destroys the hair follicle. Alopecic patches show atrophy and lack of follicular ostia with inflammatory change.

# Secondary cicatricial alopecia
Any cutaneous inflammatory process of the scalp skin or by physical trauma.

# Lymphocytic cicatricial alopecia
This group of scarring alopecia is characterized by a lymphocyte infiltrate that always affects the superior segment of the follicle around the bulge. There is loss of the sebaceous glands and fusion of hair follicles in numbers of two to a maximum of three. This fusion must be differentiated from that frequently observed in normal hair follicles at the infundibular level.

# Lichen Planopilaris
A follicular variant of lichen planus
- Epidemiology: Female > Male, early 50 Caucasians > dark-skinned individuals
- Course: slowly progressive or stable.
- Usually, only the scalp is affected. Extracranial lichen planus may occur in up to 28% of patients: Cutaneous lichen planus; Nails and mucosa are affected in <10% of patients
- Starts at vertex area; perifollicular erythema and follicular hyperkeratosis
- “Reticulated” clinical patterns: irregularly shaped and interconnected alopecia
- Itching, burning sensations and sensitivity of the scalp
- Frontal, band-like, or circumferential scarring alopecia
- Follicular hyperkeratosis and perifollicular erythema in the frontal hairline
- Graham–Little syndrome: Triad: Cicatricial alopecia on scalp; nonscarring alopecia of axillae, pubic area, and eyebrows; keratosis pilaris of the trunk and extremities
- The diagnosis of LPP cannot be based on clinical features alone.
- Microscopically-Lymphocytic infiltrate limited in upper part-Typical findings: lichenoid tissue reaction with formation of apoptotic bodies within the follicular epithelium but with little involvement of the intervening epidermis.
- The interface changes are characterized by focal loss of attachment between the follicular epithelium and the surrounding dermis; perifollicular fibrosis; grouped globular immunofluorescence (usually IgM), especially when found adjacent to the follicular epithelium.
- Diagnose LPP: Clinical + Pathology.
- Dermoscopy can help!! Most common dermoscopic findings: perifollicular scales and milky-red fibrotic pattern. Correlate with LPPAI.
- Most common skin comorbidity: rosacea (OR, 4.43; 95% CI, 2.34-8.39; P<0.0001) and skin cancer, LPP with AA is possible but rare.
- Early stage alopecia areata is associated with inflammation in the upper dermis and damage to the hair follicle infundibulum
- Treatment: limit form: IL steroid or topical steroid; extensive: HCQ→MTX→other immunosuppressive
- Other developing treatment: low-level laser, oral minoxidil, PRP, hair transplant.
Hydroxychloroquine and lichen planopilaris: Efficacy and introduction of Lichen Planopilaris Activity Index scoring system

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Background: Lichen planopilaris (LPP) and its variant frontal fibrosing alopecia (FFA) are primary lymphocytic cicatricial alopecias for which there is no evidence-based therapy.

Objective: We assessed the efficacy of hydroxychloroquine in active LPP and FFA using the LPP Activity Index (LPPAI), a numeric score that allows quantification of the symptoms and signs of the condition for statistical comparison. In addition, we determined with the LPPAI if any improvement (reduction) in the numeric score pretreatment and posttreatment reached statistical significance.

Methods: This was a retrospective, single-center chart review of 40 adult patients with LPP, FFA, or both who were treated with hydroxychloroquine for up to 12 months from 2004 to 2007 at the University of California, San Francisco Hair Center. Symptoms, signs, activity, and spreading were scored at each visit in the standardized cicatricial alopecia flow chart. A numeric score was assigned to these markers of disease activity and a numeric score was calculated at each visit.

Results: There was significant reduction (P < .001) in the LPPAI at both 6 and 12 months. After 6 months, 60% had improved (reduced) symptoms and signs. At 12 months, 85% had improvement (reduction) in symptoms and signs.

Limitations: Retrospective analysis and uncontrolled study are limitations.

Conclusions: Hydroxychloroquine is effective in decreasing symptoms and signs in LPP and FFA as shown by significant reduction in the LPPAI in 69% and 85% of patients after 6 and 12 months of treatment, respectively. (J Am Acad Dermatol 2010;62:887-92)

Key words: cicatricial alopecia; frontal fibrosing alopecia; hydroxychloroquine; lichen planopilaris; Lichen Planopilaris Activity Index.

Add abbreviations used:

LPP: Lichen planopilaris
LPPAI: Lichen Planopilaris Activity Index
UCSF: University of California, San Francisco

Lichen planopilaris (LPP) is a primary cicatricial alopecia caused by chronic lymphocytic inflammation around the upper portion of the hair follicle. The origin of LPP, and the other primary cicatricial alopecias, remains poorly understood, but they all have in common a targeted folliculocentric attack, which leads to irreversible follicular destruction and permanent hair loss. In 2001, the North American Hair Research Society proposed a working classification of the primary cicatricial alopecias and separated them into lymphocytic, neutrophilic, and mixed groups based on the predominant inflammatory cellular infiltrate. LPP has become the prototype for the lymphocytic group.
REVIEW ARTICLE

Treatment of frontal fibrosing alopecia and lichen planopilaris: a systematic review

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Abstract
Frontal fibrosing alopecia (FFA) is a primary lymphocytic cicatricial alopecia with characteristic clinical pattern of progressive frontotemporal hairline recession, perifolliculitis etma and hyperkeratosis and symptoms of itch and burning, occurring mainly in post-menopausal women. FFA is considered a subtype of lichen planopilaris (LPP), based on their identical histopathology. Currently, no evidence-based treatment is available for FFA. Our aim was to determine the effectiveness of available treatment options for FFA, and to identify promising treatment options for future studies. For this, literature search was conducted to find all primary studies on the treatment of FFA and LPP. From the primary studies, data were structured and analyzed. No randomized controlled trials were found, and one controlled trial of 114 patients is described in the literature. They received 10 different regimes, of which oral 5-alpha-reductase inhibitors were provided most often, resulting in good clinical response in 45% of them. Hydroxychloroquine resulted in good clinical response in 30% of the 29 treated patients. Topical corticosteroid preparations are ineffective in FFA. The remaining treatments were all reported in less than 10 patients. For the treatment of LPP, topical corticosteroid preparations are the first line of treatment, followed by oral cyclosporine and systemic corticosteroids, although they are characterized by a high relapse rate. Summarizing, there is currently no effective treatment of FFA, the most effective being oral 5-alpha-reductase inhibitors that possibly affect the accompanying androgenetic alopecia. We argue that oral cyclosporine might be a good candidate for future studies on the treatment of FFA.

Conflicts of interest
The authors declare to have no conflict of interest.

Funding sources
Institutional

Introduction
Frontal fibrosing alopecia (FFA) is a primary lymphocytic cicatricial alopecia with a characteristic clinical pattern of progressive frontotemporal hairline recession, perifolliculitis etma and hyperkeratosis accompanied by subjective symptoms of itch, burning or pain. Progressive loss of the eyebrows is often associated with the disease, as well as loss of body hair.1 It occurs predominantly in post-menopausal women, although multiple cases are also described in pre-menopausal women and men.1,6,7 Untreated, the disease is slowly progressive over many years, although spontaneous stabilization is also described.

Histologically, a dense lymphocytic infiltrate and fibrosis are seen around the infundibulum and isthmus of the hair follicle, often with lichenoid interface dermatitis involving the upper follicle, and loss of sebaceous glands. The histological presentation of FFA cannot be distinguished from that of lichen planopilaris (LPP), which is regarded as a follicular variant of lichen planus, presenting with multifocal patches of alopecia, pruritus, burning and tenderness in the affected hair bearing skin. FFA is considered a clinical variant of LPP.1

The pathogenesis of FFA is not fully understood. A key element seems to be destruction of the epithelial hair follicle stem cells that are located in the so called bulge region of the hair follicle. This is the region where the inflammatory cell infiltrate is primarily located in FFA.1 Destruction of the stem cells leads to permanent hair loss. Contributing factors to this stem cell destruction might be an ongoing inflammatory response triggered by proinflammatory cytokines such as interferons, increased apoptotic response, collapse of the relative immune privilege of the hair follicle.1 Recent gene expression studies identified deficiency in peroxisome proliferator-activated receptor (PPAR)-gamma-mediated signalling in LPP, indicating a defective
