Primary cicatricial alopecia

Lymphocytic primary cicatricial alopecias, including chronic cutaneous lupus erythematosus, lichen planopilaris, frontal fibrosing alopecia, and Graham-Little syndrome

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Learning objectives
After completing this learning activity, participants should be able to identify the individual characteristics of each form of scarring alopecias and possible coexisting extracranial features.

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Both primary and secondary forms of cicatricial alopecia have been described. The hair follicles are the specific target of inflammation in primary cicatricial alopecias. Hair follicles are destroyed randomly with surrounding structures in secondary cicatricial alopecia. This 2-part continuing medical education article will review primary cicatricial alopecias according to the working classification suggested by the North American Hair Research Society. In this classification, the different entities are classified into 3 different groups according to their prominent inflammatory infiltrate (ie, lymphocytic, neutrophilic, and mixed). Part I discusses the following lymphocytic primary cicatricial alopecias: chronic cutaneous lupus erythematosus, lichen planopilaris, frontal fibrosing alopecia, and Graham–Little syndrome. (J Am Acad Dermatol 2016;75:1081-99.)

Key words: alopecia; cicatricial; fibrosis; follicles; hair; hair loss; lymphocytes; neutrophils; permanent.

INTRODUCTION AND GENERAL ASSESSMENT

Key points

- **Hair loss may progress subclinically**
- **Diagnosis is often delayed**
- **All hair-bearing areas should be examined**
- **Perifollicular accentuation and isolated hairs are precious clues**
- **Unless there is a true primary infection, cultures are usually negative in patients with primary cicatricial alopecia**
- **Laboratory testing should be performed in accordance with the clinical setting**

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Abbreviations used:

CCLE:	chronic cutaneous lupus erythematosus
DIF:	direct immunofluorescence
DLE:	discoid lupus erythematosus
FFA:	frontal fibrosing alopecia
ITA:	intralesional triamcinolone acetonide
LE:	lupus erythematosus
LPP:	lichen planopilaris
MMF:	mycophenolate mofetil
NAHRS:	North American Hair Research Society
PDIRS:	premature desquamation of the inner root sheath

Primary cicatricial alopecias (PCAs) represent a group of poorly understood conditions in which the destruction of follicular structures leads to permanent hair loss. Although some familial cases have been described, most cases of PCA are acquired. A working classification for PCA was proposed by the North American Hair Research Society[1] (Tables I and II). This classification may evolve as more data become available. PCAs account for approximately 5% of cases in specialized hair clinics,[2-4] and the ratio of lymphocytic to neutrophilic or mixed PCA is 4:1.[2,3]

Hair loss may progress subclinically,[5] and scalp biopsy specimens obtained from clinically "normal" areas may have evidence of disease.[6,7] A significant amount of hair is usually lost before the alopecia is apparent, making it difficult to precisely determine its onset.[8,9] Patients are often aware of their alopecia for >1 year before consulting a dermatologist.[10,11] Inflammation frequently extends well beyond the alopecic area(s). Assessment is done by parting the hair over the entire scalp and looking for signs of alopecia and inflammation. Hair-bearing areas, the skin, mucosa, and the nails should be examined (Table III). Subtle clues to PCA may be easily overlooked. Discrete, millimeter-wide alopecic patches and perifollicular accentuation may be the only signs present. Patients may present without obvious areas of hair loss but rather diffuse hair thinning and discrete perifollicular erythema and scaling. They are often misdiagnosed with pattern hair loss and seborrheic dermatitis (Fig 1). Long-standing plaques without a history of regrowth and the presence of isolated hair(s) (Fig 2) within the plaques should raise suspicion for PCA. Female pattern hair loss (FPHL) or male pattern hair loss (MPHL) may be present concomitantly. The pull test (Table IV) is a good indicator of hair loss activity. Even though ≤10% of hairs are in the telogen phase, not all of them are ready to shed at the same time (the telogen phase lasts 2-4 months). In our experience, pulling 50 to 100 hairs (in 7-10 pulls) in a normal individual will usually yield few telogen hairs (approximately 2-5 hairs).[12] One or more hair(s) coming out at each pull is considered positive. A false-positive pull test occurs if the hair is not washed for several days. Active hair shedding usually yields a positive pull test even if the hair was washed and groomed shortly before examination. One should assess the surface area (Table IV) affected by hair loss and the surface area affected with inflammation.[13,14] Serial photographs may not detect slowly evolving hair loss, but a baseline photograph is useful for long-term follow-up. Taking and classifying photographs can be time consuming. An alternate option is to use the patient's smartphone or other mobile device to take pictures that can then be used during subsequent visits. Dermoscopy can help differentiate non-cicatricial alopecia from cicatrical alopecia and can improve biopsy site selection[4,15-25] (Table V). A scalp biopsy specimen[26-28] (Table IV) helps confirm the diagnosis in clinically ambiguous cases and identifies the nature and density of the inflammatory infiltrate. One biopsy specimen usually suffices, and it should be processed with horizontal, transverse sections rather than vertical sections.[3,28,29] In cases of lupus erythematosus (LE), a second biopsy specimen should be obtained; 1 half is used for direct immunofluorescence (DIF) study and the other is processed with vertical sections. Fungal and bacterial cultures should be performed when there is suspicion of tinea capitis or bacterial infection and to guide the choice of antibiotic, but most cultures are negative or lead to nonspecific pathogens.[30-41] Many series and case reports have reported normal or nonspecific laboratory tests, including complete blood cell count, thyroid function tests, iron, zinc, ferritin levels, and many others.[1,9,10,30,34,36,38-40,42-62] A recent study reported a higher prevalence of hypothyroidism in 355 patients with frontal fibrosing alopecia (FFA; 15%) compared to the general population (4%).[62] Although some biochemical or nutritional anomalies have been inconsistently reported,[50] there is no blood test specifically recommended for PCA. Laboratory studies should be dictated by the clinical context and to monitor potential side effects of treatment. Antinuclear antibody levels should be assessed in patients with LE. Syphilis should be ruled out if suspected.[63] Low ferritin levels have been a cause for debate.[64] It is controversial to treat iron deficiency without anemia, and there is no clear proof that it reduces hair loss.[50,65-67]

GENERAL MANAGEMENT OF PRIMARY CICATRICIAL ALOPECIA

Key points

- **Evidence supporting therapy is generally poor**