

Update on primary cicatricial alopecias

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The cicatricial alopecias encompass a diverse group of disorders characterized by permanent destruction of the hair follicle and irreversible hair loss. Destruction of the hair follicle can result from primary, folliculocentric disease or as a secondary result. This article focuses on the former, or primary cicatricial alopecias. The cause and pathogenesis of many of these disorders are largely unknown. Although unique clinicopathologic features allow for accurate diagnosis in some cases, diagnostic certainty is often elusive and reflects the limits of present understanding. Classification of the primary cicatricial alopecias on the basis of pathology provides a diagnostic and investigational framework and, it is hoped, will facilitate future enlightenment. Details of classification, etiopathogenesis, clinicopathologic features, differential diagnosis, and practical management of the primary cicatricial alopecias will be discussed. (*J Am Acad Dermatol* 2005;53:1-37.)

Learning objectives: Upon completion of this learning activity, participants should be familiar with the following aspects of the primary cicatricial alopecias: (1) the new, consensus-issued classification scheme, (2) current understanding about etiopathogenesis, (3) salient clinicopathologic features, (4) differential diagnosis, and (5) therapeutic management.

Scalp hair is a human characteristic that conveys aspects of self-image, identity, ethnicity, and health, among other attributes. Thus, it is not surprising that diseases that cause hair loss may result in disturbed self-perception and psychosocial interactions.¹ In disorders of permanent hair loss, such as the cicatricial alopecias, immediate diagnosis and therapeutic intervention are imperative.

The diagnostic hallmarks of all forms of cicatricial alopecia are both visible loss of follicular ostia (Fig 1) and destruction of the hair follicle on histopathologic examination.²⁻⁴ This can result from a primary or secondary process. In primary cicatricial alopecia, the hair follicle is the main target for destruction, evident microscopically as “preferential destruction of follicular epithelium and/or its associated adventitial dermis with relative sparing of the interfollicular reticular dermis.”⁴ Examples include discoid lupus erythematosus (DLE) and folliculitis decalvans. In secondary cicatricial alopecia, nonfollicular disease indirectly causes follicular destruction.²⁻⁴ Exogenous and endogenous factors, such as burns and

Abbreviations used:

AGA:	androgenetic alopecia
BMZ:	basement membrane zone
CCCA:	central centrifugal cicatricial alopecia
DIF:	direct immunofluorescence
DLE:	discoid lupus erythematosus
Ig:	immunoglobulin
KFSD:	keratosis follicularis spinulosa decalvans
LP:	lichen planus
MF:	mycosis fungoides
NAHRS:	North American Hair Research Society
SCC:	squamous cell carcinoma
SLE:	systemic lupus erythematosus
VVG:	Verhoeff–van Gieson stain

infiltrative and inflammatory diseases (eg, sarcoid, pemphigus vulgaris, tinea capitis), among several other disorders, can result in this outcome.

The primary cicatricial alopecias, which are the focus of this article, can be particularly challenging clinically. The reasons are multifactorial. Many of the conditions are nonscarring initially.⁵ Clinicopathologic correlation can be woefully lacking. Some entities are incompletely defined, reflecting a fundamental dearth of understanding regarding etiopathogenesis, as well as an imprecise definition of clinical and pathologic features. Because there are no placebo-controlled, double-blind, randomized studies, treatment options must be derived from small cases series, anecdote, and individual preference. Despite these limitations, new discoveries and conceptual advances continue to broaden our understanding of this complex subset of alopecia.

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Fig 1. Cicatricial alopecia demonstrating loss of follicular ostia (*center*), with intact follicular ostia seen peripherally. (Original magnification: $\times 16$).

OVERVIEW

Classification of primary cicatricial alopecias

Several classification schemes for primary cicatricial alopecia exist in the literature. Distinction has been based on age of onset, clinical features, and pathology, among other phenomena.^{6,7} In 2001, a group of leading hair clinicians, pathologists, and researchers, under the rubric of the North American Hair Research Society (NAHRS), issued a consensus opinion on classification of the primary cicatricial alopecias⁸ (Table I). Categorization is based on the principal inflammatory cell type (lymphocytic, neutrophilic) present in scalp biopsy specimens taken from representative, clinically active lesions. Disorders that cannot be categorized accordingly are designated as mixed or nonspecific. This classification system provides a standardized reference for the primary cicatricial alopecias that, it is hoped, will facilitate collaborative efforts among clinicians, pathologists, and researchers. It is considered provisional, with modifications expected as new insights develop. This article is organized according to the NAHRS classification system.

Epidemiology

The epidemiology of primary cicatricial alopecia in the general population is unknown. Two large clinicopathologic case series conducted at university outpatient hair clinics provide some insight. In a retrospective study by Whiting, cicatricial alopecia was diagnosed in 7.3% ($n = 427$) of all patients who underwent evaluation over a 10-year period.⁹ The majority affected were adult women (age range, 3-79 years; mean ages, 36 for males, 43 for females), who outnumbered males by 2.6:1. Of those who had scalp biopsies, primary cicatricial alopecias predominated (4:1).

In a 5-year retrospective study by Tan et al, 3.2% ($n = 112$) of patients who underwent evaluation for hair disorders had a primary form of

Table I. Proposed NAHRS working classification of primary cicatricial alopecia*

Lymphocytic

- Chronic cutaneous lupus erythematosus
- Lichen planopilaris
 - Classic lichen planopilaris
 - Frontal fibrosing alopecia
 - Graham-Little syndrome
- Classic pseudopelade (Brocq)
- Central centrifugal cicatricial alopecia
- Alopecia mucinosa
- Keratosis follicularis spinulosa decalvans

Neutrophilic

- Folliculitis decalvans
- Dissecting cellulitis/folliculitis (*perifolliculitis capitis abscedens et suffodiens*)

Mixed

- Folliculitis (acne) keloidalis
- Folliculitis (acne) necrotica
- Erosive pustular dermatosis

Nonspecific

*From reference 8.

cicatricial alopecia, the majority of which cases were characterized histopathologically by a lymphocytic infiltrate (4:1).¹⁰ Of the mutually recognized primary cicatricial alopecias evaluated in both series, a disparity in the frequency of observed types was noted. According to Whiting, pseudopelade predominated (40.6%), followed by lichen planopilaris (12.6%), and folliculitis decalvans (11.2%).⁹ Tan et al reported DLE as the foremost diagnosis (33.9%), followed by pseudopelade (24.1%) and lichen planopilaris (22.3%). Differences in patient demographics and criteria for clinicopathologic diagnosis of pseudopelade (a not uncommon point of contention) probably account for some of this disparity.

Pathophysiology of primary cicatricial alopecia

Familiarity with the basic anatomy and physiology of the hair follicle provides insight into the origin of primary cicatricial alopecia in humans. Although there is no human correlate, current animal models of primary cicatricial alopecia extend this understanding, demonstrating a variety of pathogenetic mechanisms by which this condition may develop.

Basic hair follicle anatomy and physiology. With each hair cycle, remarkable remodeling of the lower hair follicle (impermanent portion), below the isthmus or site of attachment of the arrector pili, occurs.¹¹ Unique epidermal and dermal follicular cell populations that largely reside in the

upper permanent portion of the hair follicle are thought to impart this regenerative capacity.^{11,12} Near the site of attachment of the arrector pili is a region of the outer root sheath called the *bulge*, within and perhaps somewhat below which a reservoir of slow-cycling, pluripotent cells exists.¹³⁻¹⁵ These stem cells are thought to produce secondary germ cells or transit amplifying cells that migrate in a bidirectional fashion, undergoing coordinated differentiation to (1) restore and renew the upper follicle, including the sebaceous gland, and adjacent epidermis; and (2) regrow the lower hair follicle during normal telogen-anagen cycling.^{12,16} Critical follicular dermal elements include the dermal papilla and dermal sheath, both of which approximate the bulge area during telogen and appear to have prime inductive and regulatory roles with onset of anagen.^{11,12} Langerhans cells, which are strikingly concentrated in the infundibular epithelium, bulge, and sebaceous epithelium, presumably initiate a first-line immune response to exogenous and endogenous antigenic threats to follicular viability.¹⁷

Clinical implications. In most cases of primary cicatricial alopecia, histopathologic examination reveals inflammation that affects the upper portion of the hair follicle wherein these vital components are housed. In some conditions, the localization of this inflammatory response may result from antigenic stimulation of the Langerhans cells that are positioned at the portal of the pilosebaceous unit.¹⁷ As will be discussed, examples of possible antigenic stimuli include ultraviolet light in those with scalp DLE; certain medications in lichen planopilaris; and *Staphylococcus aureus* in folliculitis decalvans. Whatever the initiating event, obliteration or permanent functional disruption of the critical elements required for follicular reconstitution is thought to ensue, resulting in permanent alopecia. Implicitly, the capacity for epidermal and sebaceous gland restoration after injury is also lost, which could account for the common observation of epidermal and sebaceous gland atrophy or destruction seen in several types of primary cicatricial alopecia. In contrast, when the inflammation is limited to the lower portion of the follicle, as occurs in non-cicatricial alopecias such as alopecia areata, these critical elements within the mid follicle are largely unperturbed and the potential to regrow hair with disease regression remains intact.

Animal models. The most extensively studied animal model for primary cicatricial alopecia is the Asebia mouse.¹⁸⁻²¹ Asebia is characterized by an autosomal mutation that results in rudimentary sebaceous glands and deficient production of stearyl coenzyme A desaturase 1, a sebaceous

gland-specific enzyme that affects the fatty acid content of sebum.²² On the basis of serial histopathologic examinations with disease progression, Sundberg et al have inferred that properly constituted sebum is required for normal desquamation of the inner root sheath and unhindered hair shaft egression; without it, the hair shaft is forced downward, ultimately perforating the hair bulb and inciting reactive inflammation and follicular destruction.²¹ It is intriguing that the Defolliculated²³ and Bareskin^{24,25} mouse models are phenotypically similar to Asebia but genotypically distinct. Other incompletely characterized mouse models for primary cicatricial alopecia attest to pathogenetic mechanisms that do not appear to depend on primary sebaceous gland pathology.²⁶⁻²⁹

Atrophy or destruction of sebaceous glands in early disease is a feature shared by many of the primary cicatricial alopecias that occur in humans and is a subject of active discussion.^{30,31} This feature may result from direct immune-mediated destruction of sebaceous glands, diminished or arrested supply of clonogenic cells from an injured bulge, or as suggested by the Asebia model, from an intrinsic defect in sebaceous gland function.

Approach to the patient with cicatricial alopecia

Scalp examination. Examination of the entire scalp, using a 3-fold or greater magnifying lens to view the scarred and adjacent hair-bearing areas, is recommended. Sidelighting and comparison to normal hair-bearing areas of the scalp may be necessary to appreciate subtle cues to the pathologic condition. Localization of symptoms, when present, can often direct the clinician to areas of active disease. Follicular and interfollicular stigmata (eg, erythema, hyperkeratosis, pigmentary alteration, atrophy), the pattern of alopecia (patchy, reticulate, central, etc.), and evidence of extracranial cutaneous and systemic features should be assessed. Confirmation of the diagnostic impression by means of scalp biopsy is suggested.

Scalp biopsy. The site chosen for scalp biopsy is crucial, as the yield of pathologic information is directly determined by it. Representative, early clinically active disease, with primary morphologic features and a positive pull test (when possible), should be captured. Symptomatic sites are often determinative. Sampling end-stage, bald areas of scarring alopecia is usually unproductive. In the absence of discernible inflammation, as may occur in entities such as pseudopelade of Brocq, biopsy of a hair-bearing site with a relative paucity of follicular ostia may suffice.

If only one biopsy is undertaken, it should be submitted for transverse sectioning and routine histologic examination.^{4,8,9,32-35} Compared with vertical sectioning, in which sampling error is significant, transverse sectioning allows visualization of most follicles and at multiple levels.³²⁻³⁵ Drawbacks include inexperience with specimen processing and interpretation, as well as loss of pathologic detail regarding the dermoepidermal junction, papillary dermis, and panniculus.³⁴ For this reason, combined use of transverse and vertical sectioning is advocated by some authors.^{2,34} When indicated, adjunctive use of direct immunofluorescence (DIF), particularly in cases of primary lymphocytic cicatricial alopecia,³⁶ and special stains such as periodic acid–Schiff and mucin can further strengthen diagnostic conviction. The Verhoeff–van Gieson (VVG) elastin stain may be of value in differentiating advanced cases of DLE, lichen planopilaris, and pseudopelade of Brocq, which often have overlap features on routine pathologic examination but display distinct patterns of elastic tissue staining.^{37,38} This fact presumably reflects disease-specific differences in follicular scar formation and remodeling of adventitial dermis with follicular destruction. Although less specific, fluorescent microscopic analysis of routinely stained sections is a rapid, comparable means of elastin pattern assessment.³⁹

At the University of British Columbia hair clinic, two 4-mm biopsy specimens are routinely obtained from patients with primary scarring alopecia: one is for transverse sectioning and the other is bisected vertically, with one-half designated for vertical sectioning and the remaining half for DIF.

LYMPHOCYTIC CICATRICAL ALOPECIAS

Although we largely agree with the NAHRS consensus–recognized list of primary cicatricial alopecias, there are some minor points of departure. For one, as will be discussed in the relevant section to follow, we are not convinced that “central centrifugal cicatricial alopecia” (CCCA) is a distinct form of primary cicatricial alopecia; it may represent a common morphologic pattern seen in different diseases. In addition, categorization of keratosis follicularis spinulosa decalvans (KFSD) as a *lymphocytic* rather than *mixed* primary cicatricial alopecia requires further investigation. Last, the lichen planus–lupus erythematosus overlap syndrome has been associated with cicatricial alopecia that is potentially primary in nature and will be addressed briefly.

Discoid lupus erythematosus

Background. *Discoid lupus erythematosus* and *chronic cutaneous lupus erythematosus* are terms

that are often used interchangeably. According to Gilliam and Sontheimer’s classification of cutaneous lupus erythematosus, DLE is one type of chronic cutaneous lupus erythematosus.⁴⁰ Lupus panniculitis and lupus tumidus are other examples. To avoid ambiguity, we will use the term *discoid lupus erythematosus* exclusively. DLE is the sole form of chronic cutaneous lupus that results in primary cicatricial alopecia.

The cause and pathogenesis of DLE are largely unknown. A complex interplay of genetic, environmental, and host factors determines expression of disease.⁴¹⁻⁵⁰ In susceptible individuals, ultraviolet light exposure is thought to be a key factor, inducing increased keratinocyte apoptosis and a reactive T-cell- or immune-complex-mediated response, eventuating in clinically apparent disease.^{41,51} Relevance of ultraviolet light to development of DLE affecting the hair-bearing scalp, a relatively sun-protected site, has yet to be fully explored. A small study showed that patients with coexisting androgenetic alopecia do not preferentially develop DLE in bald areas, nor do those with complaints of photosensitivity develop more severe scalp disease.⁵² Koebnerization is another consideration in the pathogenesis of scalp DLE. DLE is known to occur in areas of excoriation⁴² and in the ritual scalp “sorry cuts” of grieving Aboriginal women.⁴⁵ Moreover, a study in mice subjected to superficial wounding showed that keratinocytes adjacent to the site of trauma displayed cellular and molecular features that mirror those seen in DLE.^{51,53} In this paradigm, mundane trauma such as scratching or traumatic hair-care practices might elicit or aggravate scalp disease in affected individuals.

Clinical features. In adults, DLE affects females more often than males.^{42,54,55} There is no clear racial predilection, although increased prevalence among African Americans has been reported in the United States.^{42,56} Onset of disease is typically between 20 and 40 years of age,⁴² with less than 2% of those affected under age 10.⁵⁴ Approximately 5% to 10% of adults with DLE will develop systemic lupus erythematosus (SLE)⁴²; the likelihood is significantly higher in children and adolescents (26%–31%).^{54,55} The course of systemic disease is often severe, with renal or neurologic involvement not uncommon.^{42,57} Conversely, in adults with SLE that predates onset of DLE, systemic disease tends to be relatively mild.⁴²

According to most authors, involvement of the scalp is common in adults with DLE (34%–56%)^{52,56,58} but infrequent in children under age 10 (13%).⁵⁵ In more than half of affected persons, it is the first site of disease⁵² and remains exclusively involved in 11% to 20%.^{52,56} Females are 2 to 5 times more likely to

be affected,^{9,10,52} with a mean age of onset in the mid thirties.^{10,52} Scalp DLE is a marker of disease chronicity (21 years vs 11 years in those with non-scalp DLE).⁵² Between 4% and 14% of those with SLE have scalp DLE.⁵⁷

The majority of patients with scalp DLE present within 1 year of disease onset.¹⁰ Patients often complain of hair loss, increased shedding, and pruritus. Stinging, burning, and scalp tenderness may occur as well. There is no site predilection. The initial lesion is an erythematous papule or small plaque.^{2,43} With centrifugal spread, a coin-shaped (“discoid”) erythematous plaque forms, with follicular plugging and adherent scale that can be hyperkeratotic⁵⁹ (Fig 2). The “carpet tack” sign may be elicited with retraction of the scale, revealing keratotic spikes that correspond to follicular openings on the undersurface.² The pull test often yields anagen hairs. With continued disease progression, the plaque may enlarge. Thereafter, the erythema diminishes, and atrophy, telangiectases, hypopigmentation or depigmentation, and loss of follicular ostia become prominent.^{2,43} In darker-skinned persons and children under age 10 years, gross hyperpigmentation may delimit the scar margin.^{42,55} Mottled dyspigmentation throughout the lesion may also occur. Lesions can coalesce. Affected hair may be curly,² reflecting the torsional effect of follicular fibrosis on inner root sheath molding of the developing hair shaft.

Spontaneous remission occurs within 4 years in one-third to one-half of affected persons^{56,60} but is less likely in those with widespread disease.⁶⁰ Recurrences often present in the center of former lesions.⁹ Complications from scalp DLE include cosmetic disfigurement, ulceration,¹⁰ and squamous cell carcinoma (SCC).⁴² SCC arising in patients with DLE affects all skin types and is a potentially life-threatening occurrence, with Sulica and Kao reporting a metastatic rate of 31% and ensuing death in 10.5%.⁶¹ A low threshold for biopsy of persistent, treatment-resistant hyperkeratotic or ulcerated foci, classically arising in long-standing lesions of DLE, and prompt, aggressive management are mandatory. Other associated diseases include secondary verruciform xanthoma,⁶² papulonodular dermal mucinosis,⁶³ and alopecia areata.⁶⁴

Differential diagnosis. When extracranial DLE is present, the diagnosis is usually clear. Early scalp DLE can mimic psoriasis, tinea capitis, dermatomyositis, lichen planopilaris, alopecia mucinosa, and subacute folliculitis decalvans. In contrast to lichen planopilaris and folliculitis decalvans, the *center* of the lesion, rather than the hair-bearing periphery, is affected in active disease. Burnt-out DLE is often difficult to differentiate from other end-stage

primary cicatricial alopecias, although follicular plugging, atrophy, and dyschromia are suggestive remnants.

Pathology. Active scalp DLE is classically characterized by vacuolar interface alteration of the follicular epithelium, with a scattering of dyskeratotic keratinocytes, cytooid bodies, and a variably dense periadnexal and interstitial lymphocytic infiltrate with dermal mucin.^{4,7,9,65,66} Perifollicular inflammation usually affects the upper follicle, but a panfollicular pattern is also seen. Perivascular inflammation is superficial and deep. Sebaceous glands are atrophied or absent. Distention of follicular ostia with laminated keratin can be prominent. The adjacent epidermis may be involved, typically marked by atrophy, vacuolar interface change, and orthokeratotic hyperkeratosis. In more advanced lesions, a thickened periodic acid–Schiff-positive epidermal and follicular basement membrane zone (BMZ), pigmentary incontinence, and fibrosis of the papillary and reticular dermis develop. In end-stage lesions, concentric lamellar fibrosis is apparent around the upper follicle but can be panfollicular. When stained with VVG, advanced discoid lesions reveal diffuse dermal uptake that spares the fibrous tracts of extinct follicles.³⁸

Use of DIF for diagnosis and differentiation of scalp DLE from other primary lymphocytic cicatricial alopecias is often necessary. To optimize the yield from this technique, the lesion chosen for biopsy should be untreated for at least 3 weeks⁶⁷ and be at least 2 to 3 months old.^{67,68} Reported positive results from scalp specimens range from 63% to 100%.^{9,10,67-69} Diagnostic features of DLE on DIF are deposition of immunoglobulin (Ig) G or IgM and C3 in a granular or homogeneous bandlike pattern at the dermal interface with the follicular epithelium and epidermis.^{4,9,65} The presence of IgA is less common.^{4,65}

At times, differentiation of lichen planopilaris from DLE can be challenging. Perieccrine and deep perivascular inflammation, dermal mucin, and a relatively scant number of cytooid bodies are salient features of DLE that are not found in lichen planopilaris. In addition, the interface change is vacuolar and not lichenoid, often with a thickened basement membrane.

Therapeutic management. A complete history taking and thorough mucocutaneous examination with attention to signs and symptoms of SLE should be performed at each visit. A midstream urine sample and serum antinuclear antibody titer⁷⁰ should be obtained in all patients but especially in those with widespread lesions, progressive disease, or arthralgias and, arguably, in children.



Fig 2. Extensive acute and subacute DLE affecting the lateral (A) and occipital (B) scalp, with typical involvement of the conchal bowl of the ear (C).

A systematic, multimodal search for publications on the treatment of DLE for the period of 1956-2000 identified 54 studies, only 2 of which were randomized and controlled.⁷¹ Conclusions from these 2 studies were as follows: (1) class II corticosteroid creams are more efficacious than class VII preparations,⁷² and (2) marked improvement or resolution of disease is achieved equally in about half of subjects who undergo treatment with hydroxychloroquine or acitretin, but adverse effects from acitretin are more common.⁷³ Of the patients with scalp DLE in both studies, similar results were observed but were of insufficient power to draw statistically significant conclusions. With this caveat in mind, a tiered treatment approach will be proffered for the treatment of scalp DLE. It is not dissimilar to that suggested for treatment of DLE in general.⁴²

In patients with limited, active disease, first-line therapy is class I or class II steroids (lotion, gel, or foam twice daily) or intralesional triamcinolone acetonide (3-10 mg/mL every 4 to 6 weeks), or both.^{2,3,6,54} At the University of British Columbia hair clinic, patients over age 10 years with <10% of active scalp involvement undergo treatment with class I steroids (0.05% clobetasol propionate scalp solution twice daily) and intralesional triamcinolone acetonide used in conjunction. Every 4 to 6 weeks, serial 0.1 mL aliquots of triamcinolone acetonide, 10 mg/mL (maximum volume, 2 mL) are injected intradermally with a 0.5 inch 30-gauge needle into

hair-bearing sites with active disease. The predominant side effect observed is transient atrophy. Repeated treatment of the same site can result in permanent denting, hypopigmentation or depigmentation, and telangiectases. If an acceptable response is not achieved in 8 weeks, treatment with antimalarial medication is initiated.

Antimalarials are considered first-line, highly effective therapy in those with rapidly progressive or extensive active DLE.^{1,2,42,73,74} In general, adverse effects are less likely with hydroxychloroquine, and thus this agent is usually tried first. Prior to initiating therapy, a baseline ophthalmologic examination should be conducted, and patients who smoke should be encouraged to stop or at least decrease their habit, as cigarette smoking has been shown to reduce therapeutic responsiveness in a dose-dependent fashion.^{75,76} Hydroxychloroquine should be started at doses of 200-400 mg per day or 200 mg twice daily^{2,3,44,73,74} in adults or 4-6 mg/kg daily in children.^{54,75} Clinical improvement is often seen within 4-8 weeks, with the full extent of benefit not evident for several months.⁷⁴ Oral prednisone (1 mg/kg)^{2,3} tapered over 8 weeks may be required as a temporizing measure in those with severe disease.² Although unproved in those with scalp DLE specifically, combined use of different antimalarials can have a synergistic effect^{42,77} and should be entertained in those who continue to have active scalp disease despite sustained use of maximally dosed hydroxychloroquine for 3-6 months.^{2,3,6,75} In

general, treatment with antimalarials is long-term, with a 6-month relapse rate approaching 50%.⁷⁴ With remission, gradual reductions in dosage should be attempted.

In some patients with resistance to antimalarial medication, oral retinoids have been used with good to excellent results.⁶ Although, as mentioned, acitretin is as efficacious as hydroxychloroquine,⁷³ isotretinoin (40 mg twice daily or 1 mg/kg/d)^{2,78,79} should be tried initially. Compared with acitretin, isotretinoin-related telogen effluvium is less likely to occur and the drug half-life is shorter, with less risk to women of child-bearing age, who comprise a large portion of the affected cohort.⁸⁰ Response is often rapid and may not require bridge therapy with prednisone. Relapse usually occurs with cessation of therapy.^{6,81} Thus use of oral retinoids may be best reserved for stabilization of acute disease or as an adjunctive measure until control of disease is established. Alternatively, disease remission may be maintained with low-dose therapy (10–40 mg daily) instead.⁸¹

Several other systemic therapies have been advocated for treatment of DLE, with largely mixed outcomes. Relevance to scalp DLE is uncertain. Thalidomide, including low-dose regimens, is effective in recalcitrant disease, but a high risk-benefit profile makes its use relatively prohibitive.^{81–84} Other agents used include dapsone,^{74,81,85,86} mycophenolate mofetil,^{87,88} methotrexate,⁸⁹ azathioprine,⁹⁰ vitamin E and derivatives,^{74,91} clofazamine,^{74,81} gold,^{81,92} salicylate bismuth,⁵⁶ systemic⁹³ (and intralesional⁹⁴) interferon- α -2, and monoclonal anti-CD4 antibodies.⁹⁵

A few case reports suggest that topical tacrolimus,^{96–98} imiquimod,⁹⁹ and tazarotene¹⁰⁰ may prove to be effective therapeutic alternatives. Topical tacrolimus alone (0.1% ointment daily for 4 weeks) achieved marked improvement in 1 of 4 patients with facial DLE, working best in those with nonhyperkeratotic lesions.⁹⁶ When compounded with an ultrapotent corticosteroid ointment (0.3% tacrolimus in 0.05% clobetasol propionate ointment) a marked benefit was seen in 2 treatment-refractory patients after a few weeks of twice daily use,^{97,98} including one with scalp DLE⁹⁸; sustained use was required to maintain the effect. Isolated reports about resolution within weeks with imiquimod (5% cream daily for 3 weeks, for “2 cycles”) for scalp disease⁹⁹ and tazarotene (0.05% gel nightly for “months”) for facial disease¹⁰⁰ are also noteworthy. Use of these topical agents might be considered prior to initiation of nonconventional systemic therapies. Further investigation is required.

End-stage DLE can be surgically excised to improve cosmesis.⁵⁸ Risk of Koebnerization may be minimized with use of intercurrent therapy such as antimalarials.⁵⁸

Last, although scalp DLE has not been definitively linked to sun exposure, avoidance of the sun during peak hours, use of a tightly woven hat, and application of a broad-spectrum sunblock to bald areas is recommended. The role of Koebnerization from mundane trauma to the scalp in the evolution of disease is, as mentioned, a matter of speculation, but scratching, picking, and traumatic hair-grooming practices should be discouraged.

Lichen planopilaris

Background. Lichen planopilaris (also known as follicular lichen planus) is considered a follicular variant of lichen planus (LP).¹⁰¹ Three forms of lichen planopilaris are recognized: classic lichen planopilaris, Graham-Little syndrome, and frontal fibrosing alopecia. A tumid, plaque form of LP with prominent follicular involvement often affecting the retroauricular portion of the scalp has been described but does not cause cicatricial alopecia.^{102,103}

The pathogenesis of lichen planopilaris appears to resemble that of classic LP on the basis of shared histopathology and immunohistochemical staining patterns.¹⁰⁴ An antigenic trigger has been postulated to initiate disease, given the observation of increased numbers of Langerhans cells in affected epithelium early in disease^{104,105} and the occurrence of scalp lichen planopilaris or LP after ingestion of gold,¹⁰⁶ atabrine,¹⁰⁷ or quinacrine,¹⁰⁸ hepatitis B vaccination,¹⁰⁹ hepatitis C infection,¹¹⁰ arguably, and in association with microbial overgrowth in an epidermal nevus.¹⁰⁴ Hair is a necessary cofactor. In a case of concomitant alopecia areata and lichen planopilaris affecting the scalp, disease involvement with lichen planopilaris stopped abruptly at the border between the two conditions.¹⁰ Terminal hairs can be affected exclusively,¹⁰¹ in combination with vellus hairs,¹⁰³ or limitedly, as occurs in a condition called “fibrosing alopecia in a pattern distribution,” which has histopathologic features of lichen planopilaris and preferentially affects the miniaturized secondary vellus hairs in those with androgenetic alopecia (AGA).¹¹¹ Ultimately, these disparate etiologic stimuli are postulated to elicit lesional expression of common keratinocyte autoantigens¹⁰⁴ and an ensuing T-cell–mediated lichenoid response with destruction of follicular basilar epithelium by CD8⁺ activated T cells.^{104,112}

Classic lichen planopilaris

Clinical features. Lichen planopilaris is a disease of adults, with onset typically in middle

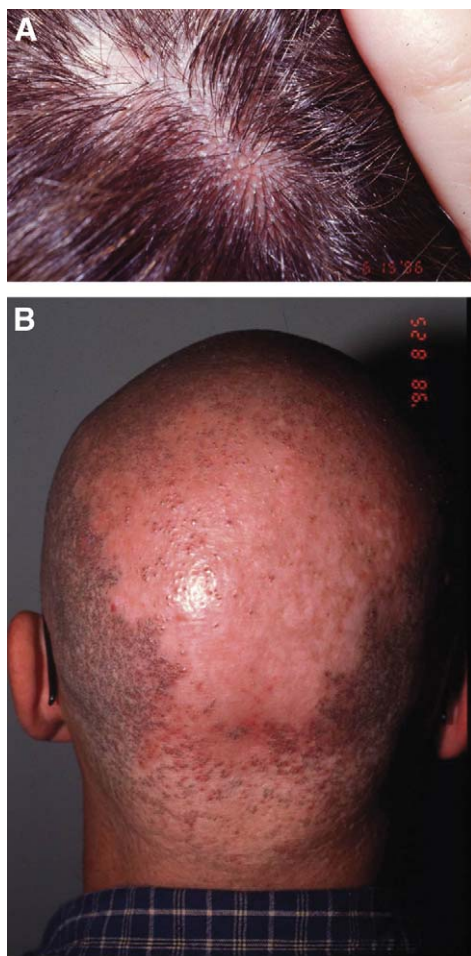


Fig 3. Biphasic classic lichen planopilaris. **A**, Active disease is marked by follicular hyperkeratosis and erythema (*center*), and burnt-out disease by depigmented shiny patches (*left superior*). **B**, Extensive cicatricial alopecia affecting the occipitoparietal scalp with perifollicular erythema at the hair-bearing margins. (Part B from Shapiro J. Hair loss: principles of diagnosis and management of alopecia. London: Martin Dunitz Ltd.; 2002.)

age.^{10,101,113} Females are particularly prone to the disease.^{9,10,101,103,113,114} Extracranial LP may be present in 17%-28% of patients at presentation^{10,115} and occurs at some time during the course of disease in 50%.¹⁰¹ Although children are rarely affected with LP,¹¹⁶ up to 9% of those affected develop this follicular form,¹¹⁷⁻¹¹⁹ with a high proportion of scalp involvement noted in one case series.¹¹⁷

LP affecting the scalp usually occurs as lichen planopilaris; typical lesions of LP are rarely observed.^{10,101,114} Most patients seek attention for their condition within 1 year of disease onset.^{10,113} Common presenting symptoms include shedding, hair loss, and pruritus.^{10,101,113} Scaling, pain, burning, and scalp tenderness may also occur.^{10,101} Clinical hallmarks of active disease include peri-

follicular erythematous or violaceous to brown papules and spinous follicular hyperkeratosis,^{10,101,103} associated with anagen hair release upon execution of the pull test¹⁰ (Fig 3). These inflammatory features are not always present contemporaneously.^{101,114} Multifocal disease is typical, although a central distribution is also common,^{10,101,113} and rarely the entire scalp may be affected.¹¹⁴ Involvement of hair in the peripheral portion of the scalp is uncommon but has been documented in those with AGA and frontal fibrosing alopecia.^{101,113,120} Months may lapse before the affected hair is shed and clinically evident scarring is observed.¹⁰³ Scars are not infrequently atrophic and depigmented, and measure millimeters to centimeters in breadth.^{10,101,113,121} Unaffected hairs are often contained within. Active disease is usually confined to the hair-bearing rim. Hair loss can be well masked by the remaining hair, resulting in shocked amazement when the examination is undertaken. Complications include ulceration.¹⁰

Disease can be self-limited or slowly progressive.^{101,114} Rapid onset is more likely in those with coexistent extracranial lichen planopilaris or bullous LP.¹²¹⁻¹²³ Associated extracranial forms of LP include lichen planopilaris,^{103,114} planar LP,¹¹⁴ atrophic LP,¹¹⁴ bullous LP of the soles with onychotrophy of the toenails,^{114,124,125} oral LP,^{101,113-115} and nail LP.^{10,101,113,114,126} Blood tests for autoantibodies and hepatitis B and C infection are routinely normal.¹¹³ There are sporadic reports of associated autoimmune disorders,^{10,103,127-131} erythema dyschromicum perstans,¹³² hyperthyroidism,¹¹³ and uterine neoplasia.¹¹³ In one study, 28% of patients with scalp lichen planopilaris had coexisting AGA.¹⁰

Differential diagnosis. Early scalp lichen planopilaris can be indistinguishable from early DLE, pseudopelade of Brocq, folliculitis decalvans, keratosis follicularis spinulosa decalvans (KFSD), and alopecia mucinosa. Unlike DLE and alopecia mucinosa, disease activity in lichen planopilaris is limited to the hair-bearing periphery of cicatrized alopecia. Pustules are conspicuously absent, making folliculitis decalvans unlikely. Distinction from incompletely expressed KFSD can be difficult, particularly when onset of disease occurs in childhood; however, a positive family history and photophobia should prompt this diagnostic consideration. Pseudopelade of Brocq can, at times, be impossible to differentiate from lichen planopilaris, and arguably may represent a variant of lichen planopilaris, as will be discussed.¹³³ As with DLE, end-stage lichen planopilaris can mimic other primary cicatricial alopecias, including those with a predilection for central scalp involvement.

Pathology. Biopsy of clinically active disease will often reveal the diagnostic features of lichenoid interface alteration.^{4,7,101,134,135} Not all follicles are affected.¹⁰¹ The upper follicle, and infundibulum in particular, is surrounded by a variably dense, bandlike array of lymphocytes that often obscures the follicular epithelial-dermal junction.^{4,7,101,134,135} Infundibular hyperkeratosis with underlying hypergranulosis can be seen.^{101,134,135} Cytoid bodies are prominently scattered along the follicular BMZ.^{4,101,134,135} Sebaceous glands are often atrophic or absent.^{7,134} Features of epidermal LP may co-exist.^{101,134} Pigmentary incontinence can be prominent, especially when the epidermis is affected.¹⁰¹ With disease evolution, follicular destruction occurs and foreign-body hair-shaft granulomas are seen.^{3,7,135} End-stage lichen planopilaris is marked by longitudinal tracts of fibrosis at the sites of former follicles. Adjacent epidermal atrophy and papillary fibrosis, unlike the extensive dermal fibrosis seen in DLE, may be observed.¹³⁴ Elastin staining with VVG reveals a superficial, wedged-shaped scar unlike the broad dermal scarring observed in developed DLE.³⁸

DIF can be valuable in ambiguous cases, although it is not uncommonly negative.^{9,10,101,134,136} "Shaggy" or "patchy" deposition of fibrinogen and clumped IgM or, less commonly, IgA and C3, is seen along the follicular BMZ.^{3,4,36,101,134,135} Although occasionally noted by others,^{101,136} Ioannides and Bystryn primarily observed a linear pattern of IgG and IgA deposition along the follicular BMZ in patients with scalp lichen planopilaris and suggested that this LP-discordant pattern reflects an unrelated disease process.¹³⁷ Race, disease activity, and duration of the lesion at the time of biopsy are variables postulated to account for this difference.¹³⁶

Therapeutic management. All patients should undergo assessment for a possible drug-related source for their disease.¹³⁸ Arguably, in patients with eroded or ulcerated scalp lesions, determination of hepatitis C status may be warranted.^{110,139}

There are limited reports about treatment of scalp lichen planopilaris, many of which do not provide details regarding the number of patients treated, dose and duration of treatment, and outcome measures. Several treatment regimens were incidentally discovered upon review of literature aimed at management of cutaneous LP.^{140,141} In general, local disease is managed with topical steroids,^{6,9,101,113} monthly intralesional triamcinolone acetonide (3-10 mg/mL),^{2,9,101} or combined use of these agents,^{2,6} with mixed outcomes. Use of moderate- or high-potency topical steroids should be considered first. According to one study involving 30 patients, this

approach can be quite effective, achieving remission in two-thirds, when a tapered dosing regimen for 12 weeks is used (twice daily for 3 weeks then daily for 3 weeks, and every other day thereafter; agent not specified).¹¹³ Others reported temporary benefit in 70%, but protocol details were not provided.¹⁰¹ An incidental report of one topical steroid-refractory patient who underwent successful treatment with topical cyclosporine (unknown concentration; twice daily for 20 days, then daily for 40 days) requires further investigation.¹¹³

Use of systemic medications should be reserved for those with local steroid-refractory, rapidly progressive, or extensive active or symptomatic scalp lichen planopilaris. Short, tapered courses of prednisone can often effectively gain control over intractable disease (1 mg/kg or 30-40mg daily for 2-4 months)^{2,6,101} and can be used as bridge therapy when use of retinoids or antimalarials is considered.² The assertion that acitretin should be considered first-line therapy for cutaneous LP owing to an extensive, evidence-based analysis of various therapeutic modalities,¹⁴² and the observance of marked improvement in 2 patients with scalp lichen planopilaris who underwent treatment with low-dose oral tretinoin (10 mg daily for 1-10.5 months),¹⁴³ suggests that trial of a low-dose oral retinoid should be considered initially in the appropriate patient. Antimalarials have also been recommended^{2,7,9,144} (200 mg twice daily²) as first-line treatment, but may only provide symptomatic relief.¹⁰¹ Some patients have been successfully managed with griseofulvin (250 mg twice daily for 7-10 months¹⁴¹).¹⁰¹ Mixed outcomes have been reported with low molecular weight heparin^{140,145} and thalidomide.^{146,147} Recently, a course of oral cyclosporine has been advocated for treatment-refractory scalp disease (3-5 mg/kg/d for 3-5 months¹⁴⁴; 5 mg/kg/d for 15 days, then 3 mg/kg/d for 30 days¹¹³). In the extended study of 3 patients with active disease, remission was achieved within 3 to 5 months and was sustained for a 12-month period in 2 patients, with recrudescence of mild pruritus in the remaining patient that was managed successfully with topical therapy.¹⁴⁴ However, others anecdotally reported no improvement with this approach in 2 patients.¹⁰¹

Frontal fibrosing alopecia

Background. First described by Kossard in 1994,¹⁴⁸ frontal fibrosing alopecia (also known as postmenopausal frontal fibrosing alopecia) is considered a variant of lichen planopilaris in a patterned distribution that primarily affects postmenopausal

women.^{120,148-159} The role of hormones in the pathogenesis of frontal fibrosing alopecia remains obscure. There are a few reports of the condition in premenopausal women^{120,154,159} and one in a man.¹⁵⁵ Onset can occur any time after menopause, whether surgically precipitated or occurring naturally, with the course of disease unaltered by introduction of hormone replacement therapy.^{120,157,159} Serum androgen levels are normal.^{120,148,152,153,157}

Clinical features. Postmenopausal women over age 40 years are typically affected.¹²⁰ Occasionally pruritic, cicatricial alopecia of the frontotemporal hairline is characteristic¹⁵⁹ (Fig 4). The affected area appears as a shiny, “uniformly pale,” bandlike zone of incomplete hair loss that is of variable width (1-8 cm)^{120,153,159} and contrasts with the photoaged skin of the superior forehead, allowing one to surmise the location of the original hairline.^{120,154} Loss of follicular ostia can be subtle. The new hairline is serrated and frequently contains hairs with perifollicular erythema and hyperkeratosis, indistinguishable from that seen in classic lichen planopilaris. The eyebrows are often strikingly thinned, if not absent, and in rare cases, erythematous.^{120,148,152,154,157,158} Nonscarring, noninflammatory symmetric axillary and extremity hair loss is a not uncommon feature.^{120,154,158} There are also rare reports of eyelash and abdominal hair loss.^{120,154} Frontal fibrosing alopecia is usually insidious but can be self-limited or rapidly progressive.¹²⁰

Classic scalp lichen planopilaris or extracranial LP may be present at the time of initial evaluation or thereafter.^{120,151,154,159} There has been 1 report of associated vulvar lichen sclerosus et atrophicus.¹⁴⁹ Results of routine blood tests are normal.^{120,148,152,153,154,157}

Differential diagnosis. Ophiasis, the frontotemporal variant of AGA, and familial recession are common misdiagnoses. Other mimickers of frontal fibrosing alopecia include KFSD, Graham-Little syndrome, and traction alopecia.

Pathology. Routine histopathologic, DIF, and immunohistochemical evaluation reveals features indistinguishable from that seen in classic scalp lichen planopilaris, although nondiagnostic findings may be seen in advanced disease.^{120,159}

Therapeutic management. There is no clearly effective treatment for frontal fibrosing alopecia.¹⁵⁹ Stabilization of disease may be achieved with twice-daily use of midpotency topical steroids^{154,158} but not predictably.^{120,157} If the condition is extending rapidly, oral prednisone or chloroquine may temporarily slow this progression.^{120,157} Limited reports on the use of intralesional triamcinolone acetonide,¹²⁰ isotretinoin,¹²⁰ acitretin,¹⁵⁹ griseofulvin,¹²⁰ 2% minoxi-

dil,^{120,157} hydroxychloroquine,¹⁵⁹ and topical retinoic acid¹²⁰ suggest that these agents are ineffective, but such a conclusion is far from proved.

Graham-Little syndrome

On the basis of variant clinical and pathologic findings, the so-called Graham-Little syndrome (also known as Graham-Little-Piccardi-Lassueur syndrome) has been alternately classified as a form of lichen planopilaris,¹⁶⁰⁻¹⁶² keratosis pilaris atrophicans,^{5,9} and a distinct nosologic entity.¹²¹ It is an uncommon condition of adults,^{9,10,121} characterized by patchy cicatricial alopecia of the scalp, nonscarring alopecia of the axillary and pubic areas, and grouped spinous follicular papules that resemble lichen spinulosus or keratosis pilaris on the trunk and extremities.¹²¹ The scalp alopecia may develop at any time during the course of the disease and is marked by follicular hyperkeratosis or erythematous, variably scaly patches.^{9,121,160,161} There are infrequent reports of face and eyebrow involvement.^{121,160} Features of lichen planopilaris¹⁶¹ or keratosis pilaris atrophicans^{5,9} may be found on pathologic examination. On the basis of limited reports, successful treatment modalities include high-potency topical corticosteroids, alone or in combination with intralesional triamcinolone acetonide (10 mg/mL),¹⁶¹ systemic corticosteroids,¹⁶³ and oral cyclosporine (4 mg/kg/d for 3 months).¹⁶⁰ In 1 case, combined use of psoralen and ultraviolet A light was not effective.¹⁶³

Lupus erythematosus—lichen planus overlap syndrome

An overlap syndrome with heterogeneous features of LP and lupus erythematosus on clinical, histologic, and immunopathologic analysis has been described,¹⁶⁴⁻¹⁷⁰ with a report of associated cicatricial alopecia.¹⁶⁴ It is unclear whether this is an autonomous disorder, a form of DLE with LP-like features, or a sign of coexisting LP and lupus erythematosus.¹⁶⁶⁻¹⁶⁹ Acral involvement with verrucous or annular livid red-violet plaques is typical and often affects the palmoplantar surfaces.^{168,169} The cicatricial alopecia is a poorly characterized feature^{164,167-169} that appeared as erythematous patches succeeded by follicular plugging in one report.¹⁶⁴ Awareness of the existence of this condition is important for management and prognostic purposes, since conversion to SLE has been reported and may not be evident for years.^{165,168,169} Different diagnostic laboratory techniques have been advocated to help distinguish those with covert LP, but none are readily feasible.^{167,168}

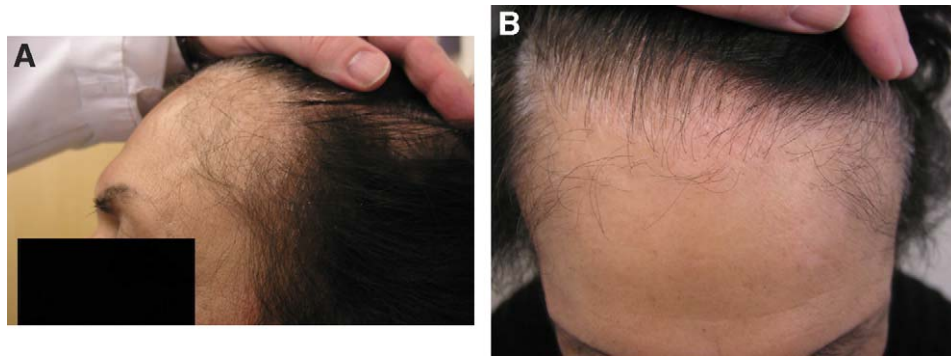


Fig 4. Lateral aspect (A) and overview (B) of frontal fibrosing alopecia showing incomplete recession of the frontotemporal hairline. Note associated lateral eyebrow hair loss (A).

Pseudopelade of Brocq

Background. This primary cicatricial alopecia was named “pseudo-pelade” by Brocq in 1888 for its likeness to *la pelade*, or hair loss of alopecia areata.¹⁷¹ In 1905 Brocq, Lenglet, and Ayrignac exhaustively detailed clinical features of the disorder based on a review of 29 published cases and 22 cases of their own.¹⁷² They concluded that pseudopelade is a unique nosologic entity, a notion that was disputed by several of their contemporaries.¹⁷² More than 100 years later, the debate continues. Many continue to maintain that pseudopelade of Brocq is distinct clinicopathologically.¹⁷³⁻¹⁷⁸ Others view it as a variant of certain primary cicatricial alopecias,^{66,133,179-182} namely, lichen planopilaris and DLE, or alternatively, as the common final stage of several cicatricial alopecias (*état pseudopeladique*).^{183,184} To confuse matters, the term *pseudopelade* alone is used indiscriminately in the literature to denote both pseudopelade of Brocq and other forms of cicatricial alopecia that simulate it; definition is required each time the term is used.^{5,8,177} Understandably, several authors have called for the term *pseudopelade* to be abandoned completely.^{5,177,185,186}

Attempts to define pseudopelade of Brocq clinicopathologically have been unsuccessful to date. Several factors have contributed to this lack of progress, including different clinical conceptions of the disorder that have, in turn, resulted in a broad range of histopathologic findings; limited data about the clinicopathologic evolution of disease, with little known, in particular, about early features; and the common experience of finding pseudopelade of Brocq-like histologic findings in biopsy specimens from patients with clinical diagnoses other than pseudopelade of Brocq (details provided in section on pathology). According to Brocq and others, atrophic, oval to round, white to ivory plaques classically lacking signs of inflammation are characteristic.^{171,172,178,187} Nayyar et al maintain that biopsy

of lesions that fit this description often reveals features of lichen planopilaris.¹³³ Braun-Falco et al, in their extensively cited 26-patient case series about the condition, in which lesions that were not diagnostic of other primary cicatricial alopecias were a priori defined as pseudopelade of Brocq, noted “erythema” at some time during the course of disease in a majority of patients (86%) and found features of DLE or lichen planopilaris on histology in 33%.¹⁷⁵ Amato et al, seeking to clarify this clinicopathologic disparity, analyzed the microscopic features of early, active disease in 36 patients diagnosed with pseudopelade of Brocq according to the clinical criteria of Braun-Falco et al and established a diagnosis of DLE or lichen planopilaris in an even higher proportion (69%).^{181,182} Last, mention should be made of the NAHRS consensus group’s clinical definition of pseudopelade of Brocq as “discrete, smooth, flesh-toned areas of alopecia without follicular hyperkeratosis or perifollicular inflammation.”⁸ Atrophy is omitted as a typical feature, and the descriptor flesh-toned is introduced presumably to include cases of pseudopelade of Brocq in dark-skinned individuals in whom the ivory-white color seen classically in whites is not applicable. Notably, reports of the condition in non-whites, both clinically and histopathologically, are lacking.

Among those who consider pseudopelade of Brocq an autonomous disease, acquired autoimmunity,¹⁸⁸ infection caused by *Borrelia*,^{189,190} and premature senescence of the follicular stem cell reservoir⁶⁶ have been posited as pathogenic factors.

For the purposes of this article, the term *pseudopelade* will be restricted to Brocq’s clinical conception of the condition.

Clinical features. Because of different clinicopathologic definitions of pseudopelade of Brocq, the true epidemiology is unknown. According to Brocq et al and Photinos, males are more commonly affected^{172,178,187}; however, in pseudopelade of

Brocq otherwise defined, a female predilection is reported.^{10,173,180} Onset usually occurs during adulthood.^{172,174,187} There are rare reports of familial disease presenting during childhood or adolescence.^{191,192}

Pseudopelade of Brocq is a chronic, insidious form of primary cicatricial alopecia. Symptoms are typically absent, but mild pruritus and diminished lesional sensation may be present.^{172,187} Brocq described 3 patterns of pseudopelade: scattered "petite plaques," large plaques, and a combination of these morphologies.¹⁷² The vertex is commonly involved. Initial lesions usually present as round to oval plaques that measure a few millimeters in diameter, similar to "une petite lentille," but on occasion are nickel-sized or slightly larger.¹⁷² In light-skinned whites, from review of the photographs and text by Brocq and Photinos, the lesions are ivory or pearly white ("blanc nacré") in color.^{172,174,178} Infrequently, diffuse or pale rose coloration can be seen. In our East Indian population with the same morphologic characteristics, lesions are typically hypopigmented but also can appear diffusely bronzed or flesh-colored, perhaps reflecting postinflammatory change or exposure of these bare areas to sunlight (personal observation of E.K.R., J.S.). In both patient subsets seen at the University of British Columbia, lesional skin is often slightly depressed and supple. Photinos likened the appearance to "les traces de la pulpe du doigt qu'on peut voir après l'avoir passé sur la neige," the canonical "footprints in the snow."¹⁷⁸ Uncommonly, the lesions can be slightly turgid,^{172,174,178} particularly in those who present with diffuse erythema (personal observation of E.K.R., J.S.). Rarely, scant fine scale is present. Evolution of the condition can result in the appearance of numerous discrete small plaques, in a confetti-like distribution; reticulate extension of lenticular disease; or coalescence of lesions into a large plaque with irregular or polycyclic borders that can span several centimeters, sometimes surrounded by small satellite plaques (Fig 5). Within large areas of cicatrized alopecia, isolated and grouped hairs may appear kinked.^{172,174,193} Active disease is marked by a positive pull test result for anagen hairs.^{172,174} There have been 2 reports of beard involvement with pseudopelade of Brocq.^{172,194} In one case, a man with biopsy-proved pseudopelade of Brocq of the beard and scalp¹⁹⁴ developed LP on the face 2 years later (personal observation of J.S.). Additional reports of associated extracranial LP continue to fuel the debate about the nature of pseudopelade of Brocq.^{133,173,183,184,195}

The course of disease is slowly progressive, with periods of quiescence and activity.^{172,194} Less often, it is inexorably progressive. Overt hair loss is usually not apparent for many years owing to this slow cadence of disease but also to remarkable camouflage by the remaining hair. Rapid progression is extremely uncommon.^{172,174}

Differential diagnosis. Pseudopelade of Brocq can mimic alopecia areata, lichen planopilaris, and DLE. In addition, central centrifugal cicatricial alopecia (CCCA), a noninflammatory condition of the central scalp that is seen primarily in black women¹⁹⁶ (see relevant section below), shares some features with pseudopelade of Brocq. However, CCCA usually presents as a symmetric patch, unlike the irregularly bordered, typically atrophic plaques seen in classic pseudopelade of Brocq. Moreover, coexisting lenticular disease is not seen in patients with CCCA. Indeed, it remains to be definitively determined whether blacks develop classic pseudopelade of Brocq at all. Other diagnoses that may be entertained include morphea, tinea capitis, secondary syphilis, and familial focal alopecia, an acquired form of pseudopelade of Brocq-like cicatricial alopecia of the scalp that is histopathologically distinct¹⁹⁷ but arguably represents a nondiagnostic form of pseudopelade of Brocq.¹⁹¹

Pathology. With the exception of the findings of Pierard-Franchimont and Pierard, who noted massive follicular sheath apoptosis in early disease,¹⁷⁶ no pathognomonic pathologic features of pseudopelade of Brocq have been described.^{3,185} Routine histologic examination of classic pseudopelade of Brocq usually shows nonspecific findings.^{3,4,133} A variably dense perifollicular lymphocytic infiltrate appears in early disease and is followed by eccentric atrophy of the follicular infundibular epithelium, concentric lamellar fibroplasia around the upper follicle, sebaceous gland loss, and ultimately, complete destruction of the pilosebaceous unit. End-stage disease is marked by follicular longitudinal fibrous tracts that extend into the subcutis and can often be found in association with hair-shaft granulomas and a sparse lymphohistiocytic infiltrate.^{4,7} Arrector pili remain intact. Epidermal atrophy, noted by Brocq et al,^{172,187} has not been remarked on by others.^{4,7} Interface changes are notably absent.⁴ Findings on DIF are usually negative but may reveal scanty IgM deposition along the follicular infundibular BMZ, distinct from the typical pattern seen in DLE and lichen planopilaris.⁴ In advanced disease, elastin stains reveal dense elastic tissue cuffing a broad, fibrotic follicular tract, unlike the characteristic pattern seen in advanced DLE and LPP.^{37,38}



Fig 5. Variants of pseudopelade of Brocq. Confetti-like, hypopigmented, oval to round areas of noninflamed cicatricial alopecia are shown in an adult white male with coexisting AGA (A) and an East Indian boy (B). Several years later, this same East Indian male, now a teenager, presented with coalescent lenticular disease affecting the central scalp as a shiny, flesh-colored atrophic irregularly defined plaque (C, right). His father, who also had long-standing classic pseudopelade of Brocq is shown next to him (C, left). (D) Large plaque disease affecting the central scalp in a white woman. (Parts A, B, and D from Shapiro J. Hair loss: principles of diagnosis and management of alopecia. London: Martin Dunitz Ltd.; 2002.)

Aspects of these nonspecific histologic features of classic pseudopelade of Brocq can also be seen in CCCA,^{3,4} in unrelated cicatricial alopecias that are subjected to biopsy in the midregion of bald areas, and in end-stage cicatricial alopecia of different origins.^{7,185} Longitudinal sectioning of biopsy specimens can also produce this result owing to limited follicle sample size and “missed” diagnostic pathology. As was mentioned in the introduction and deserving of reiteration in the context of pseudopelade of Brocq, to ensure optimal clinicopathologic correlation, the hair-bearing edge of an early lesion of cicatricial alopecia should be selected for biopsy. Signs of clinical activity aid in further refining the choice of scalp biopsy site.

Therapeutic management. Active disease, marked by a positive pull test result or extension of hair loss, should be treated. As with the majority of primary cicatricial alopecias, and perhaps more strikingly in the case of pseudopelade of Brocq because of ambiguous diagnostic determinants, no widely accepted treatment approach can be culled from literature. In addition, the task of objectively monitoring therapeutic efficacy in a condition that usually has no overt signs of inflammation and is

often asymptomatic can almost be impossible.¹⁰ Variable success has been reported with topical corticosteroids,^{2,7,10} intralesional triamcinolone acetonide (10 mg/mL),^{2,7,10,121} prednisone,¹⁰ hydroxychloroquine,^{2,7,10,198} and isotretinoin,¹⁰ with little detail on protocol. At the University of British Columbia hair clinic, pseudopelade of Brocq is managed similarly to lichen planopilaris with no single modality of exceptional benefit. Indeed, many authors consider the condition intractable.^{3,6,121}

Central centrifugal cicatricial alopecia

Background. *Central centrifugal cicatricial alopecia* is a new term adopted by the NAHRS consensus group to encompass the terms *hot comb alopecia*, *follicular degeneration syndrome*, *pseudopelade in African Americans*, and *central elliptical pseudopelade in Caucasians*.⁸ In general, it is a condition that presents with flesh-colored, noninflammatory cicatricial alopecia of the central scalp that, over time, enlarges centrifugally. As was mentioned, CCCA shares some clinicopathologic features with classic pseudopelade of Brocq, accounting for inclusion of the term *pseudopelade* in

the latter 2 descriptors of this condition. CCCA is not to be confused with central centrifugal *scarring alopecia* as promoted by Sperling et al.¹⁹⁹ Central centrifugal scarring alopecia refers to the frequent predilection of certain primary cicatricial alopecias to affect the central scalp, including follicular degeneration syndrome, pseudopelade (not Brocq's conception), and folliculitis decalvans, which the authors considered linked pathologically.^{5,35,199} Thus, the 2 terms CCCA and central centrifugal scarring alopecia, although essentially synonymous semantically, connote different clinical conceptions.

It remains to be determined whether CCCA is a unique nosologic entity or a common morphologic pattern shared by distinctly different disorders. Most of the literature, which relates to hot comb alopecia and follicular degeneration syndrome, upon review reveals differences in clinicopathologic features. Hot comb alopecia, as described in 51 African American women by LoPresti, Papa, and Kligman in their seminal 1968 article, was attributed to the "immediate" effects of liquefied "hot petrolatum" dripping onto the central scalp from vertically extended, well-greased hair as it was subjected to repeated passes of a heated comb used for straightening.²⁰⁰ Patients complained of soreness of the scalp during and immediately after the procedure. The slowly expanding cicatricial alopecia of the central part of the scalp, thought to result from long-term use of this procedure, commonly contained clusters of 4 to 7 hairs, emerging from dilated sunken ostia filled with horny debris. Histopathologic study revealed *outer root sheath* "degeneration" in association with superficial perifollicular lymphocytic infiltrate and "unusual" epidermal atrophy with elongated rete ridges and hypogranulosis, among other features. In contrast, Sperling and Sau cited an intrinsic defect in *inner root sheath* desquamation as causal owing to their observance of this histologic abnormality early on in affected persons.¹⁹⁶ This postulate was supported by the correlate finding that no single hair-care practice appeared to unify these patients, as was first appreciated by Price.^{196,201} Consequently, hot comb alopecia was renamed follicular degeneration syndrome.¹⁹⁶ Since then, the etiologic role of premature inner root sheath desquamation in follicular degeneration syndrome has been contested, given the observation of this feature in other primary cicatricial alopecias, and the involvement of only a few follicles in any one biopsy specimen.^{7,202-204}

Despite the continued debate as to the nature of this disorder, it is commonly acknowledged that some women develop an insidious, noninflammatory primary cicatricial alopecia of the central scalp that spreads centrifugally. Whether a primary,

inherited follicular defect or exogenous factors, such as culturally popular use of certain hair-care products or procedures, incite disease remains unknown.

Clinical features. As the use of hot-comb straightening has fallen out of favor, and little published information is available on CCCA in Caucasians, clinical features of CCCA related to follicular degeneration syndrome alone will be discussed. Follicular degeneration syndrome is largely a disorder of adult black women, with reported cases from Africa²⁰² and North America.^{2,196} African American men can also be affected.²⁰⁵ Sperling and Sau describe a localized "pins and needles" sensation, pruritus, and tenderness as common symptoms,¹⁹⁶ although in the authors' opinion, the condition is usually asymptomatic (E.K.R., J.S.). The midline crown or vertex of the scalp undergoes slowly progressive, symmetric, centrifugal scarring without overt inflammation. Islands of unaffected hair may be present within areas of scar. The scarred skin is supple, shiny, and flesh-colored^{196,200} (Fig 6). Perifollicular hyperpigmentation and polytrichia can be observed. According to Sperling et al, follicular degeneration syndrome in men is commonly marked by the additional feature of perifollicular firm, inflammatory papules within areas of hair loss; other forms of primary cicatricial alopecia (eg, acne keloidalis) may coexist.²⁰⁵ It remains unclear whether cessation of traumatic hair-care practices and chemical processing techniques alters the course of disease.^{196,202}

Differential diagnosis. Chronic traction alopecia, which we consider a separate entity on the basis of clinicopathologic features,²⁰⁶ AGA, and trichotillomania must be excluded.

Pathology. For reasons stated above, histopathologic findings in follicular degeneration syndrome alone will be relayed. Premature inner root sheath desquamation is a characteristic but nonspecific finding.^{7,199} Sperling et al maintain that observation of this feature *early* in the course of disease, in the absence of inflammation, is uniquely suggestive of follicular degeneration syndrome.^{5,199} In active disease, a perifollicular lymphocytic infiltrate surrounds the upper follicle and may be prominent opposite the area where the outer root sheath is most thinned. Lamellar fibroplasia encircles the follicle at this level. As mentioned before, not all hair follicles are affected. In advanced disease, perifollicular granulomatous inflammation and hair-shaft foreign-body giant cells are present.¹⁹⁶ End-stage disease is marked by follicular fibrosis with retention of arrector pili.^{7,196} The VVG staining pattern resembles that seen in pseudopelade of Brocq.³⁸

Therapeutic management. Some authors recommend complete cessation of traumatic hair grooming practices and use of potentially damaging chemical hair care products.^{3,7,200,202} Sperling et al have reported empiric success in treating active disease, marked by symptoms or histologic evidence of inflammation, with daily use of a potent topical corticosteroid and tetracycline (500 mg twice daily).²⁰⁵ A significant response is usually seen within a few months but may take up to 6 months. Once achieved, treatment frequency is gradually tapered and then discontinued when remission is sustained for a full year.

Alopecia mucinosa

Background. Alopecia mucinosa is an inflammatory condition of the pilosebaceous unit that can result in both nonscarring and scarring alopecia, which may reflect stage or severity of disease. Among other microscopic features, alopecia mucinosa is characterized by intrafollicular mucin deposition, a finding that is a nonspecific tissue reaction pattern^{207,208} and is arguably best termed *follicular mucinosis*.^{4,207-209}

Two distinct types of alopecia mucinosa are traditionally recognized: primary idiopathic and secondary lymphoma-associated disease.²¹⁰ Because of considerable epidemiologic and clinicopathologic overlap between these 2 categories,²¹¹⁻²¹⁶ and reports of development of lymphoma in a number of cases of primary disease,^{209,217,218} this distinction may be artificial and potentially misleading practically. Primary and secondary alopecia mucinosa may instead represent aspects of a disease spectrum, with primary alopecia mucinosa alternately viewed as a premalignant condition²¹⁹ or an indolent form of follicular mycosis fungoides (MF) outright, with an excellent prognosis.²¹²

The etiopathogenesis of alopecia mucinosa is incompletely understood. An antigenic stimulus originating in the hair follicle is thought to incite a T-cell-mediated folliculotropic response. Endogenous and exogenous causes,²²⁰⁻²²³ including superficial *S. aureus* infection,^{221,223} have been posited. Intrafollicular mucin deposition is thought to occur in response to the T-cell infiltrate.²²⁴ Follicular destruction by acantholysis,²²² cytolysis,²²⁵ or mucin deposition ensues.²²⁵

Clinical features. All ages are affected,^{213,226,227} with onset as early as infancy.^{226,228} Lesional pruritus,^{211,219} dysesthesia,²²⁹⁻²³¹ and anhidrosis²¹⁶ may be present. Expression of clear, mucinous fluid from follicular ostia may be elicited with application of pressure^{211,226} or from sites subjected to biopsy.²³²

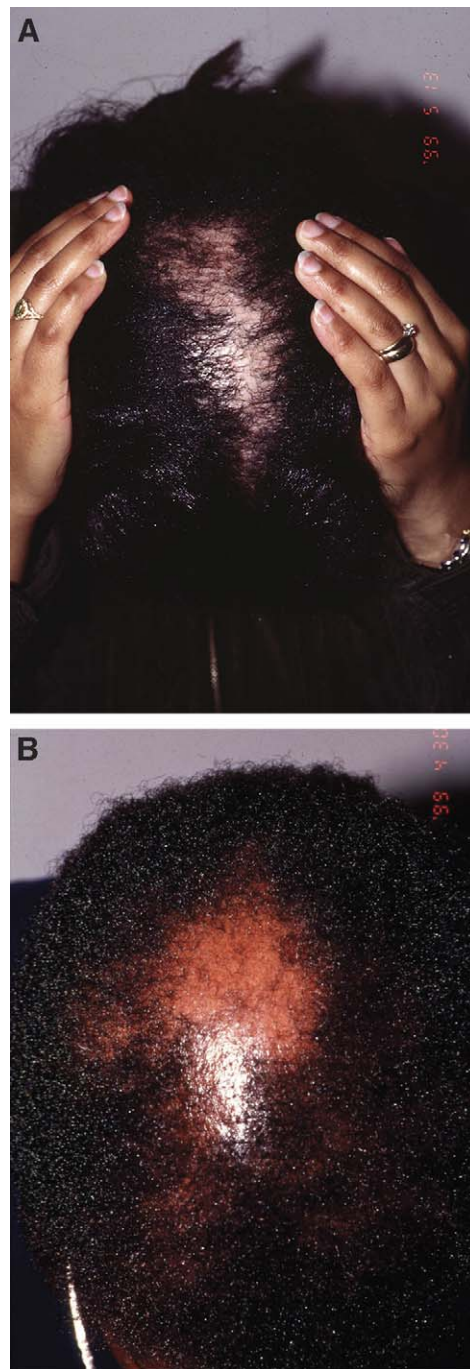


Fig 6. Overview of follicular degeneration syndrome in both an African American woman (A) and man (B). (From Shapiro J. Hair loss: principles of diagnosis and management of alopecia. London: Martin Dunitz Ltd.; 2002.)

The head and neck are commonly involved, but disease can be widespread. A predilection for the eyebrows^{207,211,226,233} and scalp^{211,226} has been noted by some authors. Monomorphous or polymorphous disease is observed.²¹¹ In nonscalp areas, 2 morphologies are commonly seen: well-demarcated,

indurated plaques with fine scale and patulous follicular ostia; and grouped or scattered folliculopapules.^{211,226} The lesions are usually pink or erythematous.^{211,226,229} Numerous other morphologies have been reported.^{211,226,232,234-240} Described scalp presentations include erythematous, finely scaled tumors with patulous pores that feel like lipomas,²²⁰ noninflamed alopecia areata-like patches^{213,226,228} and arciform alopecia,²⁴⁰ diffuse^{218,226} and complete hair loss,^{226,241,242} a scleroderma-like plaque,²²⁶ and an erythematous, indurated crusted plaque associated with *S aureus* infection.²²¹ Two of the authors (J.S., E.K.R.) have observed a case of primary alopecia mucinosa that mimicked both ophiasis and frontal fibrosing alopecia (Fig 7).

Partial or complete hair loss in the affected area is a cardinal feature but is not consistently observed.^{209,220,226,228,242} When the scalp is affected, the patient may complain of shedding²⁴² and results of the pull test may be positive.^{229,242} Because affected hair shafts are prone to breakage,^{229,242} alopecia mucinosa can present with a "black dots" sign resembling that seen in tinea capitis.²²⁹ Both scarring and nonscarring alopecia are seen.^{211,220,242} In nonscarring cases, hair regrowth may lag by months after disease resolution.²¹¹

In adults, MF is the most common malignancy associated with alopecia mucinosa, reported to occur in 9%-60%.^{213,226,227,243} Onset can precede, follow, or occur contemporaneously with the appearance of alopecia mucinosa.²²⁶ Malignant transformation can occur over months to years.^{217,219} Hodgkin's lymphoma is the predominant malignancy observed in children and young adults^{211,214-216,244,245} and, in the context of alopecia mucinosa, has a poor prognosis.^{211,216,244,245} Several other types of alopecia mucinosa-associated malignancies have been reported.^{209,246-251} Notably, with the exception of MF, alopecia mucinosa can present as a paraneoplastic phenomenon.^{248,250,251}

Other than clinical course, there appears to be no reliable clinical criteria for differentiation of patients with benign alopecia mucinosa from those with malignancy-associated alopecia mucinosa. Age, extent of disease, and duration are not consistently predictive of outcome.^{212,213,226,228,233,252} Although anecdotal and thus unproved, constitutional symptoms,^{218,233,241,246} rapid onset and extension,²¹⁸ ulceration,^{209,226,227} and leonine facies^{226,233,234} should heighten concern about associated malignancy. Notably, lesional T-cell clonality in primary alopecia mucinosa is common^{212,219} and does not connote malignancy de facto or predict progression to lymphoma.^{217,219}

Differential diagnosis. Scalp alopecia mucinosa can resemble other adnexal forms of MF,²⁵³⁻²⁵⁵ alopecia areata,^{213,242} telogen effluvium,²⁴² lichen planopilaris, morphea,²²⁶ lichen striatus,^{239,240} tinea capitis,²²⁹ subcutaneous panniculitis-like T-cell lymphoma,²⁵⁶ lichen planus follicularis tumidus,¹⁰² and dissecting cellulitis,²³⁶ among other possibilities.

Pathology. Early disease is marked by a variable amount of mucin in follicular epithelial interstices, which can be visually enhanced with the use of mucin counterstains.^{4,220,222} A perivascular and perifollicular lymphocytic infiltrate, often with eosinophils⁴ and histiocytes admixed, is present.²³³ The lymphocytes can appear banal, activated, or atypical.²⁵² Follicular lymphocytic exocytosis may be observed.²¹² In the wake of follicular destruction, a residual tract of mucin cuffed by inflammatory cells remains.²²⁰ Unlike many other primary cicatricial alopecias, concentric lamellar fibrosis is reportedly absent.⁹

Akin to the clinical situation, there appears to be no consistently reliable histopathologic criteria for differentiation of benign from malignancy-associated alopecia mucinosa.^{212,243} Certain features, especially when seen in combination, are suggestive but by no means decisively diagnostic of lymphoma-related disease^{213,233,243,252,257}: (1) Pautrier microabscesses in the follicular epithelium²⁵²; (2) a diffuse, nodular dermal infiltrate not confined to adnexae²⁵²; (3) atypical cells with cerebriform nuclei seen in a bandlike distribution^{213,233,243,252}; and (4) perieccrine or epidermal features of malignant infiltration.²⁵⁷ The significance of these findings can only be fully interpreted when related to the clinical context. Multiple biopsies may be required before a definitive diagnosis can be made,^{213,243} except in cases of paraneoplastic alopecia mucinosa.^{216,244,245}

Therapeutic management. Identification of malignancy-associated alopecia mucinosa requires malignancy-directed therapy. In those with chronic benign alopecia mucinosa, long-term follow-up with regular examinations, including lymph node palpation, is essential in children and adults alike.²¹⁷⁻²¹⁹ The patient's primary care physician should be enlisted in this endeavor. Serial biopsies may be indicated, particularly in those with evidence of disease progression. Observance of lesional clonality in those with primary alopecia mucinosa should be interpreted with caution but arguably should heighten vigilance.

There are no controlled studies on the treatment of benign alopecia mucinosa and little commentary about the effect of treatment on scalp disease specifically. Several, variably successful modalities



Fig 7. Polymorphous primary alopecia mucinosa. **A**, In the right, midfrontal hairline, a noninflamed area of cicatricial alopecia is shown. **B**, In the same patient, juicy pink follicular papules with perifollicular collarettes are present within an area of cicatricial alopecia affecting the left temporal hairline. Features of this case resemble alopecia areata and frontal fibrosing alopecia.

have been employed, including topical,^{211,226,228,239,258-260} intralesional,^{9,222,226,258} and oral²⁴⁹ corticosteroids, antibiotics,^{237,249,260,261} topical and oral retinoids,^{237,261} dapsone,²⁶² topical and oral indomethacin,^{258,263} topical nitrogen mustard,²³⁷ phototherapy,^{259,260} excision,²²¹ and superficial x-ray radiation,^{220,226,260} among many others.^{237,249,258,264} In *S aureus* culture—positive alopecia mucinosa, treatment with an oral anti-staphylococcal agent may result in lesion clearance.²²¹ For localized scalp disease without signs of infection, an initial trial of topical corticosteroids or intralesional triamcinolone acetonide (10 mg/mL)⁹ may be worthwhile. Although based on a small number of cases, alternatives include minocycline, with which complete remission was seen in 5-8 weeks (100 mg twice daily,²⁴⁹ 200 mg daily²⁶²) and sustained with lowered dosing (50 mg twice daily,²⁴⁹ 100 mg daily²⁶²), and isotretinoin (0.5 mg/kg daily for 11 weeks, then tapered over 5 weeks) with which no recrudescence of disease was observed for years after treatment.²⁶¹ Others corroborated this beneficial effect of isotretinoin with 40 mg daily, but the response may not have been as dramatic (details not provided).²³⁷ Variable periods of remission also have been reported with the combined use of psoralen and ultraviolet A light,²⁶⁰ and ultraviolet A1 cold light phototherapy.²⁵⁹

Keratosis follicularis spinulosa decalvans

Background. Keratosis follicularis spinulosa decalvans (also known as keratosis pilaris decalvans) is characterized by widespread follicular hyperkeratosis variably succeeded by atrophy, cicatricial alopecia of the scalp, and photophobia. Inherited X-linked

disease²⁶⁵⁻²⁶⁷ and sporadic onset can occur. Two morphologically similar disorders, keratosis pilaris atrophicans faciei (also known as ulerythema ophryogenes) and atrophoderma vermiculata, distinctly involve the eyebrows and cheeks, respectively, and lack scalp involvement.^{268,269} A fourth entity, folliculitis spinulosa decalvans, has been distinguished from KFSD by autosomal dominant inheritance and development of scalp pustules after puberty. However, as KFSD is genetically and clinically heterogeneous, with features that overlap those seen in folliculitis spinulosa decalvans, this distinction may be contrived.^{265,270-272} In fact, some authors consider all three conditions—KFSD, keratosis pilaris atrophicans faciei, and atrophoderma vermiculata—aspects of a disease spectrum, encompassed by the term *keratosis pilaris atrophicans*.^{269,273} Unraveling the underlying genetic defect will probably settle the issue.

Clinical features. The epidemiology of KFSD is unknown. Onset of follicular hyperkeratosis usually occurs during infancy or early childhood, first on the face, variably affecting the eyebrows, cheeks, forehead, and nose.^{121,274,275} The lesions are classically flesh-colored but can be erythematous. Red-brown telangiectases may also be seen.²⁷⁶ Widespread involvement usually ensues, affecting the scalp, neck, trunk, and extensor extremities. Mild pruritus and tenderness may be present. Patchy scalp, eyebrow, and eyelash alopecia is often evident soon thereafter, followed by scarring^{121,269,273-275} (Fig 8). Residual follicular plugs with surrounding erythema, patulous follicular ostia, and punctate atrophy can be pronounced, particularly on the face.^{269,273,275} Inconstant features include the development of scalp pustules, often associated with *S aureus* infection and

acute worsening of the cicatrizing process,^{271-273,277} focal palmoplantar keratoderma,^{269,270,274,275} high periungual cuticles,^{269,274} and ichthyotic xerosis.²⁶⁹ Onset of photophobia typically coincides with cutaneous disease^{270,274} and is marked by corneal dystrophy with punctate defects on ophthalmologic examination.^{268,269,273} There are rare reports of other ocular abnormalities.^{270,273,275,278}

The triad of photophobia, widespread keratosis pilaris-like lesions, and scarring alopecia is variably expressed, probably reflecting incomplete penetrance and random X inactivation.^{267,269,270,275,279,280} In general, disease is often more severe in males.^{267,269,270,274,275} Postpubertal remission or improvement can occur but is not predictable.^{269,270,274} KFSD is associated occasionally with atopy^{269,270,272,273} and rarely with other syndromes,^{276,278,281-283} some of which may be different genodermatoses unknown at the time of publication.^{121,276}

Differential diagnosis. With the rare exception of atrophoderma vermiculata, which is characterized by honeycomb atrophy of the cheeks, scarring alopecia of the scalp is absent in other forms of keratosis pilaris atrophicans. Of the several genodermatoses characterized by keratosis pilaris-like lesions and alopecia, only KFSD is routinely associated with cicatricial alopecia of the scalp. Graham-Little syndrome in adults, lichen planopilaris, and folliculitis decalvans in those with pustules are additional diagnostic considerations.

Pathology. Data about the histopathologic findings in scalp biopsy specimens are limited but presumably are consonant with those seen in non-scalp specimens and, arguably, other forms of keratosis pilaris atrophicans. The primary defect appears to be abnormal keratinization, marked by compact hyperkeratosis and hypergranulosis of the upper follicular epithelium in early disease.^{269,273} In acutely inflamed lesions, superficial intrafollicular and perifollicular edema and neutrophils are seen. With advancing disease, a sparse perivascular and perifollicular mononuclear cell infiltrate is present, in association with mucin and loose connective tissue around the upper follicle. Plasma cells may be seen. Granulomatous inflammation with follicular destruction, concentric perifollicular and horizontal adventitial lamellar fibrosis, and scarred follicular tracts characterize late-stage disease. The fate of the sebaceous gland in disease remains unclear.^{269,274,276}

Therapeutic management. It is important to intervene therapeutically when disease is active, often during childhood. Unfortunately, there are limited data about the treatment of scalp KFSD. Mid- to high-potency topical corticosteroids and intralesional triamcinolone acetonide (3 mg/mL)

can be of some benefit and usually require sustained use.^{269,273} Oral retinoids have been used with wide-ranging results,^{269-271,278,280,284} which may reflect differences in stage or activity of disease at the time of treatment. Notably, in 2 patients with active disease, treatment with etretinate (0.8 mg/kg) or isotretinoin (0.5 mg/kg) daily for 12 weeks achieved remission.²⁸⁴ Careful risk-benefit analysis is particularly warranted when treating a child or adolescent, and high-dose, prolonged therapy should be avoided.⁷⁸ Laser-assisted hair removal in those with treatment-refractory scalp disease has been advocated by Chui et al, who reason that by destroying the target of disease, progression will be halted.²⁸⁵ In an isolated case, this approach resulted in significantly diminished inflammation.

Pustular flares associated with *S aureus* usually can be controlled with antistaphylococcal agents.^{269,271-273,286} Treatment-resistant pustular disease may respond to dapsone (100 mg/d).²⁷⁷ Rifampin is reportedly ineffective, but details were not provided.²⁸⁵

Baseline and routine ophthalmologic examinations are suggested.

NEUTROPHILIC CICATRICIAL ALOPECIAS

Folliculitis decalvans is a commonly encountered form of primary cicatrizing alopecia (10.7%-11.2%), unlike perifolliculitis capitis abscedens et suffodiens (1.4%-4.5%).^{9,10}

Folliculitis decalvans

Background. This condition is characterized by a destructive, suppurative folliculitis.²⁸⁷ *S aureus* is usually isolated from primary lesions and has been implicated in the genesis of folliculitis decalvans by some authors.^{174,199,288-292} Host predisposition to infection due to systemic or local immune deficits and *S aureus* strain-related properties have been cited as causal factors.^{288-290,292} Several acquired and inherited immune disturbances have been associated with folliculitis decalvans^{203,292-299}; however, the majority of affected patients have no demonstrable systemic^{289,290,300,301} or lesional^{289,290} immune abnormality. It may be that strain-specific virulence factors of *S aureus*, including superantigen production^{288,290} and intracellular persistence after phagocytosis,^{288,302} account for expression of disease.

Clinical features. Young and middle-aged adults of both sexes are typically affected.^{6,9,10} Familial disease has been described in identical twins²⁸⁹ and in 2 families with inherited disorders in neutrophil bacteriolytic function²⁹⁸ and cellular immunity,²⁹⁹ respectively. The initial lesion is a pinpoint

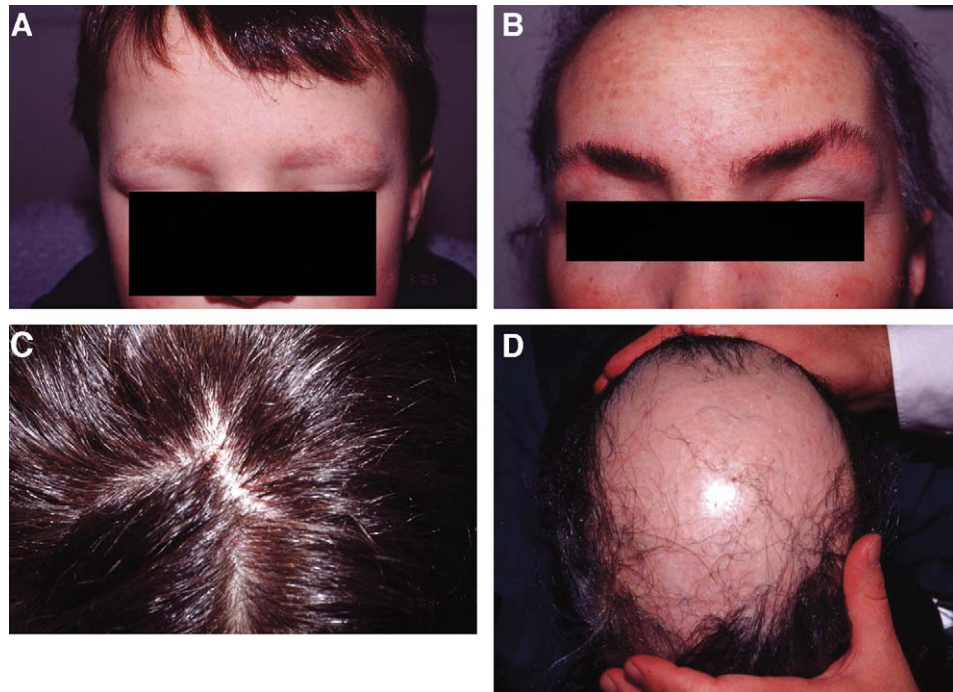


Fig 8. Cutaneous facial and scalp features of KFSD in a male child (**A, C**) and his mother (**B, D**). Cicatricial alopecia prominently affects the lateral portions of the eyebrows of the child (**A**) and is less evident on the scalp (**C**). His mother, who has long-standing disease, has red- to rust-hued telangiectases and macules on the face and nonscarring erythema of the eyebrows (**B**), with near complete cicatricial alopecia of the scalp (**D**).

erythematous follicular pustule or papule that may be painful or pruritic.^{174,290} Neighboring papulopustules soon appear and may evolve into “miliary abscesses.” Crusting ensues. Eventually round to irregularly shaped, atrophic flesh-colored or ivory-white areas of scarring alopecia develop.^{2,3,174,203} Pinpoint erythema may transiently mark the site of former hair follicles. Active disease often continues peripherally, appearing as a marginated “zone of folliculitis.”²⁹¹ Involvement is often multifocal, with occasional coalescence of lesions.^{6,174,291} There is no site predilection, but the crown is not uncommonly affected.¹⁰ In rare cases, folliculitis decalvans can affect the beard, face, and nape of the neck.^{300,303} Reports of extremity, axillary, and pubic involvement²⁹¹ may represent coincident or alternative diagnoses. The disease course is usually chronic and slowly progressive.^{174,300} Intranasal carriage of *S aureus*,^{7,290,304} and an elevated serum erythrocyte sedimentation rate^{289,300} and antistaphylococcal antibody titer^{291,301} occasionally are noted.

Tufted hair folliculitis, marked by *S aureus*-positive suppurative folliculitis of multiple hairs (5 to 20) that emerge from a common dilated follicular orifice³⁰⁵⁻³⁰⁹ and gross resemblance to “dolly

hair”³⁰⁷ when extensive, is occasionally coexistent (Fig 9). Although some authors consider tufted hair folliculitis a pathognomonic feature of folliculitis decalvans,^{290,307} the prevailing opinion is that it is a nonspecific form of cicatricial alopecia^{2,3,35,305,306,310,311} because of its occurrence in a wide range of unrelated primary and secondary cicatrizing disorders.

Differential diagnosis. The differential diagnosis of folliculitis decalvans includes classic folliculitis, acne necrotica, lichen planopilaris, DLE, and perifolliculitis capitis abscedens et suffodiens, particularly in the early stages. Grouped follicular pustules typically are not seen in ordinary folliculitis or acne necrotica. Perifolliculitis capitis abscedens et suffodiens uniquely affects black men, and early papulopustules are promptly succeeded by the development of distinctive dermal nodules.

Pathology. The earliest pathologic feature of folliculitis decalvans is acneiform infundibular dilatation. A variably dense, intrafollicular and perifollicular neutrophilic infiltrate affects the upper and middle parts of the follicle.^{3,4,7,9} With disease progression, the infiltrate becomes mixed with neutrophils, lymphocytes, and plasma cells and extends into the adventitial dermis. Abscess formation is less



Fig 9. Folliculitis decalvans with clusters of marginated perifollicular pustules and tufted folliculitis in a bandlike area of cicatricial alopecia.

prominent than in perifolliculitis capitis abscedens et suffodiens. Granulomatous inflammation occurs, with foreign-body giant cells surrounding ectopic bits of hair shaft.^{3,6,7} Late-stage disease is characterized by follicular and adventitial dermal fibrosis.^{4,7} Sinus tract formation is absent.⁹

Therapeutic management. The presence of folliculitis decalvans in the setting of chronic infections or other features of immunodeficiency should be evaluated accordingly. Intact pustules should be cultured and antibiotic sensitivities determined. Until recently, folliculitis decalvans was a notoriously difficult condition to treat, with a poor prognosis.^{288,290,301} Although Whiting has reported success with sulfamethoxazole-trimethoprim (details not provided),⁹ in general, antistaphylococcal, antineutrophilic, and broad-spectrum antibiotics achieve at best temporary relief, with frequent relapse upon cessation.^{186,288,290,294,303,312} The addition of prednisone can improve efficacy, but adverse effects with long-term use are prohibitive.^{186,304,312} Corticosteroids alone have a variable effect.^{9,290,295,313} Limited data on oral isotretinoin^{312,313} suggest that it is ineffective.²⁹⁶ Isolated reports of improvement with shaving³¹³ and oral L-tyrosine administration³¹⁴ require further study. Intranasal eradication of *S aureus* with topical antibacterial agents has been empirically advocated.⁷

With the introduction of rifampin and combined use of fusidic acid and zinc to the therapeutic arena, successful suppression and sustained disease-free remission, lasting from months to years after treatment withdrawal, are possible in some patients.^{288-291,296,301,304,312,313} Bactericidal action with excellent intracellular penetration and pathogen eradication by rifampin and fusidic acid,^{288,290,300,312,315} and immunomodulatory effects noted with all 3 agents^{288-290,300,312} probably underlie this remarkable effect. Rifampin can also eliminate the carriage state of *S aureus*.³¹⁶

Reports about the use of rifampin outnumber those about fusidic acid. Rapid emergence of resistance prohibits use of rifampin as monotherapy.^{290,316} Typically, high-dose rifampin (300 mg twice daily) and clindamycin (300 mg twice daily) are administered for 10 weeks.^{288,290,296} Sustained remission for months is seen in a significant number of patients after 1 course, although a second or third course may be required to achieve this effect.^{288,290} Reported complications are uncommon and include diarrhea and rash, both of which were attributed to clindamycin and resolved with its discontinuation.²⁸⁸ In those with the inability to tolerate clindamycin, oral ciprofloxacin or clarithromycin may be substituted.²⁹⁰ Alternatively, topical mupirocin ointment twice daily³⁰⁴ or topical 2% erythromycin with oral zinc sulfate (dosing unknown)³¹² may be used in combination with oral rifampin, apparently with similar success, based on isolated case reports. Patients should be forewarned about the expected orange-red discoloration of urine and permanent staining of soft contact lenses with rifampin. Although not observed in this circumstance, development of a hypersensitivity syndrome with flulike symptoms, rash, and eosinophilia can occur with repeated high-dose therapy.³¹⁶

Efficacy with oral fusidic acid, used alone or in combination with other agents, has been mixed but remarkable, particularly when zinc is co-administered.^{288,289,291,300,301} Fusidic acid is not available for routine use in the United States. In 3 patients, triple therapy with oral fusidic acid (1,500 mg daily for 3 weeks), zinc sulfate (400 mg daily for 6 months), and topical 1.5% fusidic acid cream (2 weeks; frequency not specified) resulted in complete remission of disease that was sustained over a 1-year follow-up period with daily zinc sulfate (200 mg).³⁰¹ Rapid relapse can occur with discontinuation of maintenance therapy. Others were unable to duplicate this result with oral zinc and fusidic acid at similar doses, but the treatment duration was not disclosed.²⁸⁹ Interestingly, in 1 patient, combined use of rifampin and fusidic acid resulted in only short-term improvement.²⁸⁸ In another case, combined use of high-dose zinc sulfate and oral erythromycin was ineffective.³¹² Caution should be exercised when prescribing maintenance therapy with zinc. Diminished intestinal uptake of copper resulting in potentially irreversible severe sideroblastic anemia and neutropenia can occur.³¹⁷ The most common adverse effect with oral fusidic acid is gastrointestinal distress, which can be mitigated with food intake.³¹⁵ Avoidance of use in those with liver disease is recommended, and liver functions tests should be performed regularly.

Perifolliculitis capitis abscedens et suffodiens

Background. Perifolliculitis capitis abscedens et suffodiens is also known as dissecting cellulitis, dissecting folliculitis, dissecting perifolliculitis, perifolliculitis capitis, and Hoffman disease. Because of the association of this disease with acne conglobata and hidradenitis suppurativa, together comprising the follicular occlusion triad—or tetrad, if one includes pilonidal cysts—abnormal follicular keratinization leading to obstruction, secondary bacterial infection, and follicular destruction is postulated to result in disease.^{318,319} A primary defect in the host response to bacterial pathogens also has been proposed but has been largely unsubstantiated by bacterial cultures and response of disease to antibiotics.^{2,318-323}

Clinical features. More than 80% of those affected are black men³²² age 18 to 40 years.^{322,324} Reports of disease in white males are not uncommon and may approximate 10%.^{10,319,322} Perifolliculitis capitis abscedens et suffodiens can also occur in women³¹⁸ and rarely in children.³²¹ There is one report of familial perifolliculitis capitis abscedens et suffodiens.³²⁵

The initial lesion is a follicular pustule often found on the occipital or vertex scalp³¹⁸ that then transforms into an often painful, bulbous, firm or fluctuant nodule soon thereafter.^{6,318,322} Multifocal, variably contiguous disease is common and may impart a cerebriform appearance to the scalp³²⁶ (Fig 10, *A, B*). Infrequently, the whole scalp is affected.³²⁷ Lesional scalp skin may be freely mobile.³²² Pressure on one nodule can result in expression of seropurulent exudate directly or from an adjacent interconnected nodule.^{318,322} Spontaneous exudation of purulent material can also occur.⁹ A nonscarring alopecia initially develops over the nodules.^{318,326} The pull test result is positive in areas of inflammation.³¹⁸ Long-standing, inadequately treated disease results in cicatricial alopecia³¹⁸ marked by depressed,³²⁴ hypertrophic, or keloidal scars.^{6,327} Although spontaneous remission can occur, chronic relapsing disease is characteristic.^{318,322} Cervical or occipital lymphadenopathy may be present. An elevated serum erythrocyte sedimentation rate occasionally is noted.^{322,326}

Coexisting acne conglobata or hidradenitis suppurativa is present in about one-third of cases^{318,322} and is a risk factor for development of human leukocyte antigen-B27—negative spondyloarthropathy, particularly in black males. Asymmetric peripheral and axial joint involvement is characteristic. Active skin disease usually precedes onset of the arthritis and mirrors acute worsening.^{328,329} There are also sporadic reports

of perifolliculitis capitis abscedens et suffodiens—associated polyarticular arthritis with sternoclavicular hyperostosis,³³⁰ SAPHO syndrome (synovitis, acne, palmoplantar pustulosis, hyperostosis, osteitis),³²⁹ marginal keratitis,³³¹ life-threatening secondary SCC,³³² and *S aureus* osteomyelitis.³³³

Differential diagnosis. The somewhat exclusive occurrence of this condition in adult black men and the distinctive clinical appearance of perifolliculitis capitis abscedens et suffodiens are usually diagnostically determinative. Nonetheless, two of the authors (J.S., E.K.R.) and others have observed inflammatory tinea capitis (kerion celsi) in children and adolescents^{334,335} that remarkably resembled perifolliculitis capitis abscedens et suffodiens. In the former case, antecedent topical steroid use may have played a role in the pathogenesis. A high index of suspicion and a low threshold for biopsy in those with potassium hydroxide-negative scrapings is essential to early diagnosis. Biopsy culture results are usually positive, whereas routine histopathologic findings and periodic acid—Schiff staining may be negative.³³⁴ In addition to kerion, there is an isolated report of fatal folliculotropic MF with follicular mucinosis and large-cell transformation mimicking perifolliculitis capitis abscedens et suffodiens.²³⁶

Pathology. Early disease is characterized by infundibular acneiform distension with intrafollicular and perifollicular neutrophilic infiltration.⁴ With follicular perforation, abscesses composed of neutrophils, lymphocytes, and numerous plasma cells become apparent in the perifollicular middle to deep dermis and superficial fat. The abscesses become partially lined by squamous epithelium that will define the sinus tracts of well-developed disease. Sebaceous glands persist until late-stage disease.³³⁶ With end-stage disease, pilosebaceous units are obliterated and extensive fibrosis is present in the adjacent dermis and subcutis, surrounding sinus tracts.^{4,336}

Therapeutic management. Until recently, perifolliculitis capitis abscedens et suffodiens was a notoriously treatment-resistant condition.³¹⁹ Oral isotretinoin is now arguably regarded as first-line therapy. In a small number of cases, long-standing (up to 2.5 years) remission of disease has been reported with its use.^{320,325,326} Treatment with isotretinoin, 1 mg/kg/d, for a minimum of 4 months, followed by 0.75-1 mg/kg/d for an additional 5-7 months, is advocated.³²⁶ Flattening of the nodules may take months and should ward against premature discontinuation of treatment for lack of an immediate response (Fig 10, *B, C, and D*). Suboptimal dosing and inadequate treatment duration can result in an unsatisfactory outcome.^{285,326,337,338}

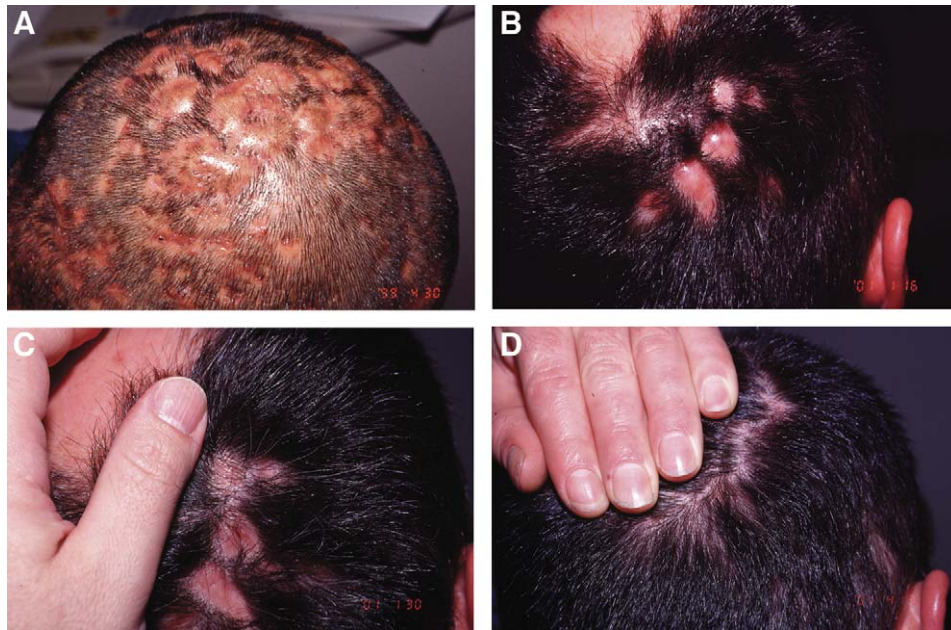


Fig 10. Extensive (A) and localized (B) examples of perifolliculitis capitis abscedens et suffodiens. The latter patient (B) was treated with oral isotretinoin (40 mg twice daily) for 7 weeks without benefit. Intralesional triamcinolone acetonide (10 mg/mL) was given for symptomatic relief and likely accounts for the flattening of the nodules 2 weeks later (C). Thereafter, the isotretinoin was increased to 40 mg thrice daily and, after 4 additional months of therapy, resulted in regrowth of hair and prevention of significant scarring (D). (Part A from Shapiro J. Hair loss: principles of diagnosis and management of alopecia. London: Martin Dunitz Ltd.; 2002.)

In isolated case reports, prolonged remission of disease for 1 year or more has been reported with isotretinoin gel (0.05%, used with 1% clindamycin gel for 8 weeks, followed by isotretinoin gel alone for 8 months, frequency not specified³³⁹), zinc sulfate^{326,340,341} (400 mg by mouth three times daily for 12 weeks, then 200 mg by mouth three times daily for 10 weeks³⁴¹), and oral steroids with low-dose alternate-day maintenance therapy.³²³ Other options include oral and topical antibiotics (eg, tetracyclines, antistaphylococcal agents), antibacterial soaps and intralesional triamcinolone acetonide, which are variably effective when used as single agents or in combination^{6,318,319,322-324}; dapson, reportedly effective in severe, treatment-refractory disease (regimen unknown) by some³⁴² but not by others^{285,343}; and colchicine (0.6 g twice daily), which was moderately effective in 1 case.³⁴³ Hydroxychloroquine,²⁸⁵ methotrexate,³⁴³ and ketoconazole³⁴³ are ineffective on the basis of isolated reports. Successful treatment of the follicular occlusion triad in a woman, with minocycline and cyproterone acetate, an oral antiandrogen not available in the United States, has also been reported.³⁴⁴

Incision and drainage of painful nodules³¹⁹ or excisional carbon dioxide laser with secondary intention healing³⁴⁵ are among several surgical tech-

niques presently advocated. Laser epilation using nonpulsed, non-Q-switched ruby²⁸⁵ and 800-nm pulsed diode³⁴³ modalities recently has been promoted as a therapeutic alternative in those with severe, treatment-refractory disease, but long-term follow-up is lacking and resignation of the patient to permanent hair loss and possible dyschromia is a concern.

MIXED CICATRICAL ALOPECIAS

In this category, acne keloidalis, acne necrotica, and erosive pustular dermatosis of the scalp are discussed. We view classification of acne necrotica varioliformis as a mixed inflammatory primary cicatricial as somewhat controversial, given the infiltrate is predominantly lymphocytic. In addition, it is unclear whether erosive pustular dermatosis of the scalp results from primary, folliculocentric disease or from nonfollicular events such as exogenous trauma. Only acne keloidalis is seen with any regularity in general dermatologic practice.³⁴⁶⁻³⁴⁸

Acne keloidalis

Background. Acne keloidalis, also known as acne keloidalis nuchae, dermatitis papillaris capillitii, sycosis nuchae, and folliculitis keloidalis, is an inflammatory condition that predominantly affects

the nuchal hairline of young, black postpubertal males. The term *acne keloidalis* is a misnomer, as the condition is neither acneiform nor keloidal in nature.³⁴⁹ A unique race-related property of the pilosebaceous unit, hair shaft, or scalp skin has been postulated to underlie evolution of disease.^{346,347,350-352} Proposed disease precipitants include mechanical trauma from shirt collars,³⁵³ excoriation,³⁵⁴ and seborrhea³⁴⁶; infection with *Demodex*^{351,355} or bacteria³¹¹; and autoimmunity.³⁴⁷ Acne mechanica from sports helmets,³⁵² hair length,^{352,356} and hair impaction ("pseudofolliculitis nuchae"³⁵²)^{351,355} are largely dispelled notions. Illustrative aspects of pathogenesis may be suggested by the reported occurrence of drug-induced acne keloidalis in white males who were treated with the anticonvulsants diphenylhydantoin and carbamazepine together (1 case)³⁵⁷ and cyclosporine (4 cases).^{358,359} Common to these drug regimens are cutaneous adverse effects that may simulate aspects of acne keloidalis that occur de novo. Histopathologic analysis of different stages of disease suggests that in predisposed persons, a progressive, locally destructive folliculitis incites reiterative cycles of acute and granulomatous inflammation with reparative fibrosis, eventuating in scar formation and transepithelial elimination of hair.^{351,355,360}

Clinical features. Onset of disease occurs after adolescence, predominantly in black males.³⁵⁰ Black females and Caucasians may also be affected.^{348,356} The occipital portion of the scalp and the nape of the neck are typically affected, although involvement of the vertex and parietal scalp may also occur^{346,350,351} (Fig 11). Pinpoint, soft to firm, flesh-colored to reddish-brown, smooth follicular papules are evident early in disease. The papules may be crusted, umbilicated, or pustular and contain hair. Patients may complain of pruritus and burning. In some individuals, these papules coalesce to form nodules or broad keloidal plaques that can be disfiguring. Pustules, abscesses, sinuses, polytrichia, tufted hair folliculitis, foul-smelling discharge, and pain are not uncommon features of this form of disease. In rare cases, large sclerotic tumors result and can significantly impact daily living. Acne keloidalis is a chronic condition, unlikely to remit without treatment. Women may have a more complete response to treatment.³⁵⁶

Differential diagnosis. Acne mechanica, early folliculitis decalvans, conventional folliculitis, and molluscum contagiosum occasionally must be differentiated from acne keloidalis.

Pathology. Early lesions are characterized by a perifollicular and intrafollicular lymphoplasmacytic infiltrate that is pronounced at the level of the

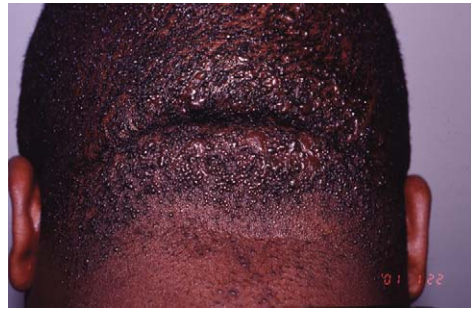


Fig 11. Acne keloidalis affecting the occipitoparietal scalp.

sebaceous gland,^{351,355} although others report features similar to early folliculitis decalvans.⁴ The isthmic follicular epithelium may be thinned and lamellar fibroplasia evident. With ongoing disease, focal or complete follicular destruction occurs and a granulomatous reaction or microabscesses form around extruded hair-shaft fragments. Sebaceous glands are absent. Chronic inflammation, with numerous plasma cells and significant dermal fibrosis, ensues.^{351,355} Notably, keloidal collagen is not seen.^{351,355}

Therapeutic management. Early, mild disease may be controlled with class I or II topical steroids alone³ or in combination with topical antibiotics.^{347,361} For papular disease, there are empiric reports of success with monthly intralesional triamcinolone acetonide^{350-352,356,361} (10 mg/mL³⁵⁶) alone or combined with topical (eg, 2% clindamycin) or oral (eg, tetracyclines) antibiotics.^{3,9,350,351,356,361} Adjunctive use of antibacterial soaps has also been promoted.⁹ On the basis of limited studies, topical^{350,352,362} and oral^{350,362} retinoids, and cryotherapy alone³⁶³ or combined with rifampin,³¹¹ appear to have little impact. Vaporization of lesions with carbon dioxide laser is associated with a high rate of recurrence.³⁵³

In those patients with extensive keloidal plaques or symptomatic treatment-refractory disease, surgical excision may be indicated. Popular approaches include excision with primary closure or secondary intention healing. Fair to excellent outcomes have been reported^{349,350,362,364,365} and are arguably optimized with the use of cold-steel scalpel blades and, when possible, inclusion of the occipital hairline in the excision, which should be exacted down to muscle fascia.^{350,362,364,365} With massive lesions, excellent outcomes have been reported with staged excision and primary closure³⁶⁵ and may surpass that obtained with healing by secondary intention.³⁴⁹ Excision with skin grafting offers no real cosmetic benefit.³⁵⁰ Successful treatment with carbon dioxide laser excision and secondary

intention healing has been reported³⁵³ but is based on a small sample size.

Acne necrotica

Background. In the words of Plewig and Kligman, "awareness of this bizarre disease is a prerequisite for an accurate diagnosis."³⁶⁶ Two forms of acne necrotica, also known as folliculitis necrotica, exist: acne necrotica *varioliiformis* and acne necrotica *miliaris*. Acne necrotica *varioliiformis* is a rare but clinically distinctive necrotizing disorder of the hair follicle that heals with varioliform scars. Proposed causes include an abnormal host response to *S aureus* or *Propionibacterium acnes* folliculitis,^{367,368} neurotic excoriation of an underlying folliculitis,³⁶⁸ and a rosacea-like genesis.³⁶⁹ The older literature emphasizes coexistence with seborrheic dermatitis.³⁶⁷ Acne necrotica *miliaris* is a nonscarring superficial folliculitis that may be caused by the same bacterial pathogens^{367,370} and arguably represents a variant of the same disease process, with individual host response determining ultimate clinical expression.³⁶⁷

Clinical features. Acne necrotica *varioliiformis* is a chronic, relapsing disorder that usually affects adults. The anterior hairline is commonly involved.^{367,369} Seborrheic regions of the face and trunk can also be affected.^{367,369,371} Crops of variably pruritic, tender, pinhead to pea-sized, reddish-brown papules or papulopustules indolently appear, umbilicate, and then slowly undergo central necrosis, leaving round hemorrhagic crusts that are shed a few weeks later. Punched-out, depressed scars result, appearing as focal areas of cicatricial alopecia when terminal hair-bearing areas are impacted.^{367,369,371} A few lesions typically appear with each outbreak.³⁶⁷ Aggravation in the summer has been reported.³⁶⁹ With chronicity, cribriform scars can develop and can be disfiguring.³⁶⁹

Differential diagnosis. Neurotic excoriations, acne necrotica *miliaris*, folliculitis decalvans, eczema herpeticum, conventional folliculitis, and molluscum contagiosum should be considered in the diagnostic differential of acne necrotica *varioliiformis*.^{368,369} Close inspection for primary lesions can be diagnostically determinative. Acne necrotica *miliaris* more completely mimics neurotic excoriations, as the follicular vesicopustules that characterize the disorder are so intensely pruritic that observance of an intact lesion is rare, with all lesions typically showing signs of excoriation upon examination.^{367,371} Unlike acne necrotica *varioliiformis*, acne necrotica *miliaris* is not associated with exclusive involvement of the anterior portion of the scalp and extracranial disease.

Early acne necrotica *varioliiformis* and conventional folliculitis can be indistinguishable.

Pathology. Early disease is marked by lymphocytic exocytosis, spongiosis, and individual cell necrosis of keratinocytes in the upper pilosebaceous unit and is associated with a dense perifollicular and perivascular lymphocytic infiltrate.³⁶⁹ Subepidermal edema is usually prominent. With advanced disease, confluent necrosis of the follicular epithelium and adjacent epidermis and dermis occurs, leaving a zone of destruction within which is contained fragmented bits of hair. Neutrophils can be seen in the superficial dermis, often beneath a bacteria-laden stratum corneum.

Therapeutic management. Fastidious culture of an intact pustule under both anaerobic and aerobic growth conditions can aid in directed therapy,³⁷² although empiric use of oral tetracyclines and anti-staphylococcal agents, as well as antibacterial shampoos, can be effective.^{9,368,369,372} The benefit can be transient or prolonged with sustained use.^{367-369,372} In those with an incomplete response, intralesional triamcinolone acetonide may be substituted (5 mg/mL, frequency not stated)⁹ or a high-potency topical steroid may be added.³⁶⁹ Institution of isotretinoin (1-2 mg/kg for 20 weeks) in cases of culture-proved *Propionibacterium acnes*³⁷² infection or empirically (30 mg daily) in intractable cases,³⁶⁹ can lead to prolonged remission. Adjunctive treatment of potential bacterial carriage sites with topical antibiotics has also been advocated.³⁶⁸

Erosive pustular dermatosis

Background. Erosive pustular dermatosis (also known as erosive pustular dermatosis of the scalp) is an idiopathic chronic, relapsing amicrobial pustular dermatosis of that scalp that results in cicatricial alopecia. A history of antecedent accidental or iatrogenic trauma to the affected scalp site is often elicited.³⁷³ Specific precipitants include minor lacerations,^{373,374} contusions,³⁷⁴ accidental scalping,^{373,375} sunburn,³⁷³ varicella,^{373,376,377} zoster,^{373,376,377} skin grafting,^{378,379} radiation,³⁸⁰ synthetic hair fiber implantation,³⁸¹ cryotherapy,³⁸² topical 5% fluorouracil,³⁸³ and topical tretinoin.³⁸² The predominance of disease in the elderly has led some to postulate that chronic actinic damage to the scalp may be a predisposing factor for disease.^{373,384}

Clinical features. Erosive pustular dermatosis is an uncommon disorder that largely affects the elderly,^{373,385,386} with an apparent female predominance.³⁷⁹ In cases with known preceding trauma, onset of disease can occur contemporaneously or months to years thereafter.^{373,379,387} The characteristic lesion is a large asymptomatic, well-demarcated,

boggy, superficially crusted plaque that is easily unroofed to reveal a beefy red, exudative erosion with discrete or coalescent flaccid pustules beneath. Moist erosions or crusts in the absence of pustules have also been described. Untreated lesions undergo episodic pustular flares, with slow enlargement over years. Cicatricial alopecia is a cardinal feature of advanced disease, the extent of which may not be fully appreciated until the lesion is healed with treatment.^{373,374,379} Wounds may be colonized by staphylococcal species and, less frequently, *Candida*.^{373,374,385} Aggravation of disease has been reported with attempts at reparative skin grafting and treatment of surrounding actinic keratoses.^{373,383} Development of secondary carcinoma with squamous and basal cell features has been reported in a long-standing case.³⁷⁵ Reports of a variant of erosive pustular dermatosis affecting the leg³⁸⁸ have been attributed to other diseases by some.³⁸⁹

Differential diagnosis. The differential diagnosis is extensive and includes amicrobial pustulosis associated with autoimmune disease, pustular ulcerative dermatosis of the scalp, pyoderma gangrenosum, pustular psoriasis, kerion, bacterial folliculitis, cicatricial pemphigoid, pemphigus vulgaris, blastomycosis-like pyoderma, erosive candidiasis of the scalp, and temporal arteritis, among other possibilities. Scalp involvement in amicrobial pustulosis associated with autoimmune disease, a newly recognized amicrobial chronic, relapsing intertriginous follicular and nonfollicular pustular eruption that affects young women with autoimmune disorders, is poorly characterized.^{390,391} However, scant reports suggest that there is some clinical, histopathologic, and treatment-response overlap with erosive pustular dermatosis that, given the added observation of erosive pustular dermatosis of the scalp in 2 patients with autoimmune disease,^{392,393} arguably could reflect a common disease process. Pustular ulcerative dermatosis of the scalp is a rare, noncrusted, ulcerative rather than erosive dermatosis of the vertex scalp that occurs in malnourished young male Africans, a cohort distinctly different from those with erosive pustular dermatosis.³⁹⁴

Pathology. Histopathologic features of erosive pustular dermatosis are nonspecific.³⁹⁵ Characterization of early disease is lacking. Observed epidermal changes include erosion, atrophy, acanthosis, parakeratosis, and subcorneal pustules. A dense, chronic mixed inflammatory cell infiltrate and occasional foreign-body giant cells occupy the dermis and do not appear folliculocentric. Pilosebaceous units are variably diminished in number or are absent. Remnants of arrector pili may be seen.^{373,374} Findings on DIF are routinely negative.^{373,374,396}

Therapeutic management. In general, erosive pustular dermatosis is steroid-responsive. Rapid improvement with twice-daily use of class I and class II topical steroids is typical^{373,374,379,396} but requires maintenance therapy to sustain the effect.^{373,374} Relapses are particularly common in those with postzoster erosive pustular dermatosis.³⁷³ Combined use of antibiotics and topical steroids results in a similar outcome.^{373,374,379} Although based on isolated case reports, potentially efficacious steroid alternatives include topical 0.005% calcipotriol cream (twice daily for 2 months), which induced remission that was sustained upon follow-up 9 months later³⁸⁶; and zinc sulfate (30 mg by mouth three times daily for 1 week), which required maintenance therapy (60 mg by mouth three times daily).³⁸⁷ Oral and topical antibiotics alone are at best transiently beneficial.^{373,385,396} Oral isotretinoin^{379,397} and dapsone^{374,379} appear to be ineffective.

ADJUNCTIVE TREATMENT OPTIONS

In addition to disease-specific treatment, a few adjunctive therapies may be of generic benefit. Suggested medical options have yet to be subjected to rigorously controlled studies.

In cases of primary cicatricial alopecia that coexist with AGA, a trial of topical minoxidil may improve cosmesis by enlarging miniaturized hairs.¹⁰ At the University of British Columbia hair clinic, topical 5% minoxidil solution (1 mL twice daily) is prescribed for 1 year and is continued in those with improvement. Thickening of vellus hairs may be appreciated 4 months after initiation of treatment.³⁹⁸ This approach may also have merit in those with early, treated disease³ by accelerating the rate of hair regrowth and prolonging anagen duration of unaffected hairs.³⁹⁹

In patients with active, extensive cicatricial alopecia in whom adequate scalp coverage cannot be achieved with the remaining hair, use of cosmetic aids such as a hairpiece or hair color-matched powder that camouflages visible scalp skin (eg, Toppik, Spencer Forrest, Westport, Conn) should be sensitively broached. Once the condition is completely burnt out, surgical correction of the defect can be offered to eligible candidates. Hair transplantation and scalp reduction are accepted approaches.⁴⁰¹⁻⁴⁰³ At the University of British Columbia hair clinic, a minimum 2-year period of disease-free remission is required before a surgical remedy is undertaken. Intercurrent disease-specific therapy may be needed to minimize the potential for recrudescence of disease,⁴⁰⁰ or Koebnerization in DLE.⁵⁶

Last, although life-threatening secondary SCC is uncommon, with reports limited to cases of DLE

Table II. Primary cicatricial alopecias: Salient clinical features and working treatment regimens for management of scalp disease*

Entity	Epidemiology	Clinical		Therapeutics [†]
		Scalp	Non-scalp	
Lymphocytic				
DLE	Women >	Symptomatic, erythematous scaly plaques with follicular plugs; telangiectases, atrophy and depigmentation with time; activity in <i>center</i> of alopecic patch	May be present; rule out SLE	1. Topical steroid ± ILTAC 2. HCQ, prednisone, <i>topical tacrolimus, tazarotene, imiquimod</i> 3. Isotretinoin
Lichen planopilaris				
1. Classic	Women >	Pruritic, multifocal or central alopecic patches with follicular hyperkeratosis and erythema at hair-bearing <i>margin</i>	May be present	1. Topical steroid ± ILTAC 2. <i>Oral retinoid</i> , prednisone, HCQ, <i>topical cyclosporine</i> 3. <i>Oral cyclosporine, griseofulvin</i>
2. Frontal fibrosing alopecia	Postmenopausal >	Frontotemporal recession often with classic LPP at hair-bearing <i>margin</i>	May be present (eyebrows>)	See text
3. Graham-Little syndrome	Adults	Patches with follicular hyperkeratosis	May be present (nonscarring in axillae, pubic area)	See text
Pseudopelade of Brocq	Adults >	Asymptomatic, <i>noninflamed</i> ivory-white or flesh-colored small oval-round confetti-like, reticulate, or large, irregular patches, ± atrophy	Rarely present (beard)	1. <i>Topical steroid</i> ± ILTAC 2. <i>Prednisone, HCQ, isotretinoin</i>
CCCA	Black women >	Central scalp; <i>noninflamed</i> , flesh-colored symmetric patch	Absent	1a. <i>Cease traumatic/chemical hair-care practices</i> 1b. <i>Topical steroid + oral tetracycline</i>
Alopecia mucinosa	All ages	<i>Polymorphous</i> disease: eg, erythematous plaques with patulous ostia; alopecia areata-like, diffuse or complete alopecia, etc	May be present; rule out malignancy	For primary form: 1. <i>Topical steroid, ILTAC</i> 2. <i>Minocycline, isotretinoin</i> 3. <i>Phototherapy</i>
KFSD	Onset in childhood	Patchy, follicular hyperkeratosis ± perifollicular erythema	Present; photophobia	1. <i>Topical steroid, ILTAC</i> 2. <i>Isotretinoin</i>

Table II. Cont'd

Entity	Epidemiology	Clinical		Therapeutics [†]
		Scalp	Non-scalp	
Neutrophilic				
Folliculitis decalvans	Adults	Central scalp >; grouped follicular pustules, miliary abscesses at hair-bearing margin	Absent	1. Antibiotic ± steroid 2. Rifampin + 2nd antibiotic 3. Fusidic acid + zinc
Perifolliculitis capitis abscedens et suffodiens	Black men >	Painful, boggy, contiguous dermal alopecic nodules that can spontaneously suppurate; sinus tracts	Follicular occlusion triad; arthritis	1a. Oral isotretinoin, topical isotretinoin + clindamycin 1b. <i>ILTAC</i> , <i>I&D</i> if painful, localized 2. <i>Antibiotics</i> , <i>zinc</i> 3. <i>Prednisone</i> , <i>dapsone</i>
Mixed				
Acne keloidalis	Black men >	Occipital scalp; firm red-brown papules, papulopustules, nodules and keloidal plaques	Absent	1a. <i>ILTAC</i> ± antibiotics 1b. Excision (plaque form)
Acne necrotica varioliformis	Adults	Anterior scalp; pruritic, tender umbilicated papules, punched-out crusts, varioliform scars	May be present (seborrheic areas)	1. Antibiotics ± topical steroid 2. Isotretinoin, <i>ILTAC</i>
Erosive pustular dermatosis	Elderly women >	Asymptomatic, crusted purulent plaque	Absent	1. Topical steroid, <i>calcipotriol</i> 2. <i>Zinc</i>

*See text for details.

>, More commonly affected; *ILTAC*, intralesional triamcinolone acetonide; *HCO*, hydroxychloroquine; *I&D*, incision and drainage.

[†]Therapeutics in italics relate to regimens that have been tried in one to a few cases with moderate to excellent results.

and perifolliculitis capitis abscedens et suffodiens, it seems prudent to advise all patients with bald cicatrized areas, regardless of skin type, to apply a broad-spectrum sunblock to the area and to wear a tightly woven hat during exposure to the sun.

CONCLUSION

The initial phase of many of the primary cicatricial alopecias is nonscarring and should be managed as a "trichologic emergency"²: prompt diagnosis and therapeutic intervention are key to thwarting permanent hair loss and a potential lifelong struggle with the psychosocial sequelae. Choice of treatment is dependent on diagnosis, age, severity of disease, and extent. In general, local treatment should be used in limited disease. Systemic modalities should be reserved for rapidly advancing, extensive, local treatment-refractory, and intractably symptomatic disease; however, given the lack of controlled studies regarding efficacy, an initial short-term

course of an empirically effective, relatively safe topical agent may be warranted. Table II depicts a working formulation for the management of primary cicatricial alopecias, based on the aforementioned referenced details, that, it is hoped, will stimulate further discussion, investigation, and understanding.

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