Primary cicatricial alopecia

Other lymphocytic primary cicatricial alopecias and neutrophilic and mixed primary cicatricial alopecias

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Learning objectives
After completing this learning activity, participants should be able to describe effective strategies for treating each form of scarring alopecias.

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Primary cicatricial alopecias can be frustrating for both patients and physicians. Proper diagnosis guides more successful management of these challenging conditions. Part II will cover the remaining lymphocytic primary cicatricial alopecias, which include pseudopelade of Brocq, central centrifugal cicatricial alopecia, alopecia mucinosa, and keratosis follicularis spinulosa decalvans. It will also discuss the neutrophilic and mixed primary cicatricial alopecias, namely folliculitis decalvans, dissecting cellulitis, folliculitis keloidalis, folliculitis (acne) necrotica, and erosive pustular dermatosis. (J Am Acad Dermatol 2016;75:1101-17.)

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

PSEUDOPELADE OF BROCQ PATTERN OF CICATRICIAL ALOPECIA

Key points

- **Pseudopelade of Brocq has been described most often in middle-aged white women**
- **Pseudopelade of Brocq is described as a chronic, insidious, slowly evolving condition**
- **Little is known about the management of Pseudopelade of Brocq; the therapeutic approach tends to be similar to that of lichen planopilaris**

The concept of Pseudopelade of Brocq (PPB) has evolved, and there is no consensus yet as to whether it is a distinct entity[1-9] or a common final stage of a different primary cicatricial alopecia (PCA).[8,10-19] In this article, PPB will be discussed because the terminology persists in both the

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Abbreviations used:

AANS:	alopecic and aseptic nodules of the scalp
AM:	alopecia mucinosa
CCCA:	central centrifugal cicatricial alopecia
DC:	dissecting cellulitis
EPD:	erosive pustular dermatosis
FD:	folliculitis decalvans
FK:	folliculitis keloidalis
FM:	follicular mucinosis
FN:	folliculitis necrotica
ITA:	intralesional triamcinolone acetonide
FKSD:	keratosis follicularis spinulosa decalvans
LPP:	lichen planopilaris
MMF:	mycophenolate mofetil
PCA:	primary cicatricial alopecia
PDIRS:	premature desquamation of the inner root sheath
PDT:	photodynamic therapy
PPB:	pseudopelade of Brocq
TF:	tufted folliculitis

medical literature and clinical practice. However, PPB is probably best considered to be an unusual clinical pattern of cicatricial alopecia, sometimes representing the end-stage of lichen planopilaris (LPP) or other forms of inflammatory alopecia.[20,21] PPB presents with discrete, smooth, flesh-toned areas of alopecia without follicular hyperkeratosis or inflammation (Fig 1).[20] It most commonly affects middle-aged white women (30-50 years of age).[1,2,6,7,11,18,22,23] It is a chronic, insidious, slowly evolving condition. Plaques may be small, large, scattered, or reticulated, and are usually asymptomatic, but mild pruritus may occur. PPB may affect the beard and eyebrows.[24-26] The differential diagnosis includes alopecia areata, central centrifugal cicatricial alopecia (CCCA), other burned-out PCAs, syphilis, sarcoidosis, pattern hair loss, and morphea. The histologic features of PPB have nonspecific changes of an end-stage cicatricial alopecia. Follicular scars and loss of sebaceous glands, with variable amounts of residual chronic inflammation, are seen. Little is known about the effective management of PPB. The therapeutic approach tends to be similar to that of LPP. Topical and intralesional corticosteroids and topical tacrolimus 0.1% are used.[22,23,27-30] Hydroxychloroquine 200 mg twice daily is often used.[22,23,27,29-33] Response is usually seen within 3 to 6 months; most patients will require 1 to 2 years of treatment.[8] Oral prednisone 0.5 mg/kg has also been described.[22,23,27,29-33] Isotretinoin (1 mg/kg/day) and mycophenolate mofetil (MMF; initial dose, 1 g/day) have been reported to have some efficacy.[22,27,30,31]

CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA

Key points

- **Central centrifugal cicatricial alopecia is more common in middle-aged women of African ancestry**
- **Central centrifugal cicatricial alopecia most commonly affects the vertex of the scalp**
- **Hot comb and relaxers do not appear to be associated with central centrifugal cicatricial alopecia**
- **Little is known about the management of central centrifugal cicatricial alopecia; the therapeutic approach tends to be similar to that of lichen planopilaris**

The concept and terminology of CCCA have evolved. LoPresti et al[34] first described the condition as hot comb alopecia. The term follicular degeneration syndrome was subsequently proposed by Sperling and Sau.[35] Headington[29] suggested "scarring alopecia in African Americans." Finally, the term CCCA was chosen.[20,36] It is a descriptive term that includes follicular degeneration syndrome, pseudopelade in African Americans, and central elliptical pseudopelade in whites.[20] CCCA is insidious. It predominantly affects middle-aged women of African ancestry,[23,34,35,37,38] with a prevalence of 3% to 6% in that population.[39-42] It is uncommon in men[23,35] and children.[43] It most commonly appears on the vertex of the scalp and progresses centrifugally, often symmetrically[20,23,35,37] (Fig 2). The scalp is soft and pliable.[34] Mild perifollicular hyperpigmentation can be seen.[35] The affected area gradually blends with the surrounding normal scalp.[44] Polytrichia[34,35] and islands of unaffected hair may be present within affected areas.[31] Tenderness, itching, or burning are usually mild if present.[35,44-46] A considerable amount of hair is often lost before the alopecia is recognized.[35] Although not specific, hair breakage can be an early sign of CCCA.[47] Its etiology is likely multifactorial. Genetic factors have been suspected,[40,42,48,49] but this could be caused in part by similar hair care practices within families.[42] No correlation was found between suspected CCCA and male pattern hair loss in the father.[42] CCCA is not solely related to the unique shape of black hair because few cases have been described in black men.[50] Two studies used a questionnaire and standardized photographs[51] to assess CCCA in women of African ancestry (>800 women).[40,42,51] On a scale of 0 to 5, CCCA was suspected for central hair loss patterns 3 to 5, but was not confirmed histologically. No correlation was found with hot comb usage.[35,38,40,42,44,52] Relaxers had been used at least once in 90% of women,[40,42] but