Central centrifugal cicatricial alopecia in Black men: A case series highlighting key clinical features in this cohort

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Key words: alopecia; androgenetic alopecia; central centrifugal cicatricial alopecia; male pattern baldness; scarring alopecia.

INTRODUCTION

Central centrifugal cicatricial alopecia (CCCA) is the most common cicatricial alopecia in Black patients and results in progressive hair loss and profound impact on well-being, that is associated with depression and poorer quality of life.[1,2] Given the progressive nature of this condition, early and accurate diagnosis and treatment initiation are essential. Although CCCA has been well studied in Black women, there is a dearth of information regarding its manifestations in Black men. Furthermore, the history, physical examination, and trichoscopy patterns have been well described in female cohorts with CCCA; however, there is less information on special considerations for males with this condition. Given the predilection for CCCA to occur in female patients, providers may have a delayed diagnosis of CCCA in males with hair loss in the vertex scalp or may misdiagnose their condition for more common forms of hair loss such as androgenetic alopecia (AGA). Although AGA may exist concomitantly with CCCA, especially in male patients, and therapies, such as minoxidil, can be used to increase hair density in both conditions, it is important to identify CCCA correctly to initiate additional anti-inflammatory therapy as soon as possible.

Herein, we present 3 cases of biopsy-supported CCCA in Black men, along with a review of pertinent clinical, trichoscopic, and histopathologic findings of CCCA. All patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

Abbreviations used:

AGA: androgenetic alopecia
CCCA: central centrifugal cicatricial alopecia

CASE 1

A 41-year-old man presented to a hair clinic with a chief complaint of hair loss for 1.5 years. He reported hair breakage and balding on the crown of his scalp, with associated hair shedding. He wore his hair in waist-length locs in a mid-ponytail for >18 years and reported washing it every 2 weeks with various shampoos. He had a family history of progressive hair loss in his maternal grandmother. Physical examination revealed a normal frontal hairline but decreased hair density and obliteration of the follicular ostia on the vertex scalp (Fig 1). A honeycomb pigmented pattern, decreased hair density, and peripilar gray halos were observed using trichoscopy of the vertex (Fig 2). A 4-mm punch biopsy revealed a decreased number of terminal anagen follicles, with fibrosis, a lymphocytic infiltrate, and loss of sebaceous glands. Clinicopathologic correlation was consistent with CCCA. Treatment with twice-daily minoxidil solution 5%, daily metformin

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Funding sources: None.

IRB approval status: Not applicable.

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JAAD Case Reports 2023;38:27-31.

2352-5126

https://doi.org/10.1016/j.jdcr.2023.05.026

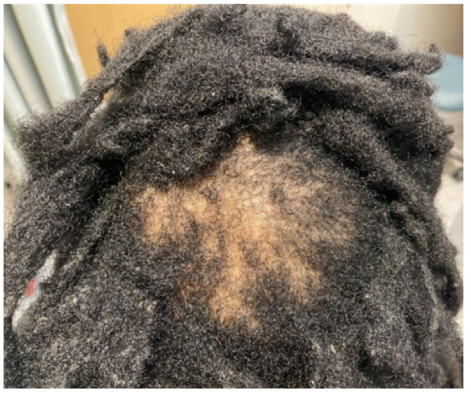

Fig 1. Patient 1, at initial presentation, with a hair loc style, exhibited hair loss in the central portion of the scalp/vertex region.

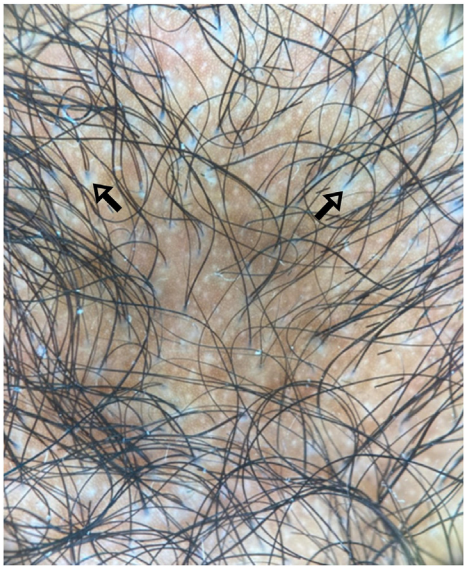

Fig 2. Peripilar white/gray halos (*arrows*) with mild surrounding scale (*white arrows*), honeycomb pattern with small white dots in between, and increased distance between hair follicles, with decreased follicular ostia.

compounded solution 10%, and clobetasol solution 0.05% was initiated. Later in the treatment course, he started 1 mg of oral finasteride daily and received regular 5-mg/mL intralesional triamcinolone acetonide injections.

CASE 2

A 36-year-old man presented to a hair clinic with a chief complaint of asymptomatic hair loss along the crown of his scalp for 3 years. He had a history of seborrheic dermatitis, which he managed with antifungal shampoos and supplements. Physical examination revealed significant thinning along the vertex scalp, with a retained frontal hairline (Figs 3 and 4).

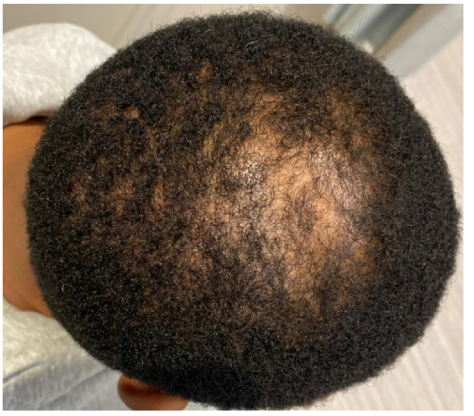

Fig 3. Patient 2 with hair loss in the central portion of the scalp/vertex area at initial presentation.

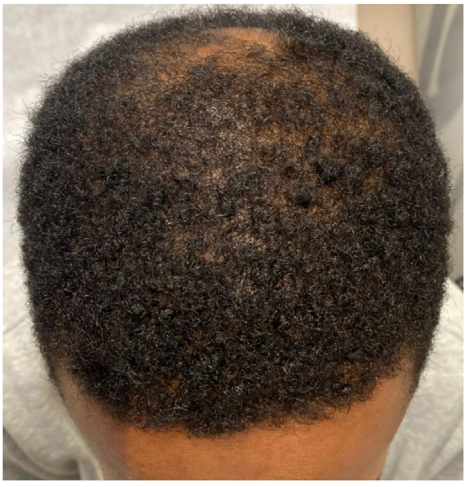

Fig 4. Patient 2 with hair loss on the vertex scalp, with retention of the frontotemporal hairline, at initial presentation.

Trichoscopy demonstrated follicular miniaturization, a honeycomb pigmented pattern, and peripilar white halos. A scalp biopsy revealed a decrease in the size and number of terminal hair follicles, prominent sebaceous lobules, and foci with prominent perifollicular fibrosis and inflammation. Given the clinicopathologic correlation, the patient was diagnosed with CCCA and started on twice-daily minoxidil solution 5%.

CASE 3

A 37-year-old man presented to a hair clinic with a chief complaint of hair loss on the scalp for 8 years. His initial symptoms of scalp flaking and pruritus gradually progressed to generalized hair loss and a focal alopecic patch on the vertex scalp (Fig 5). He reported a history of high-tension hairstyles, including locs and braids. He had a strong family

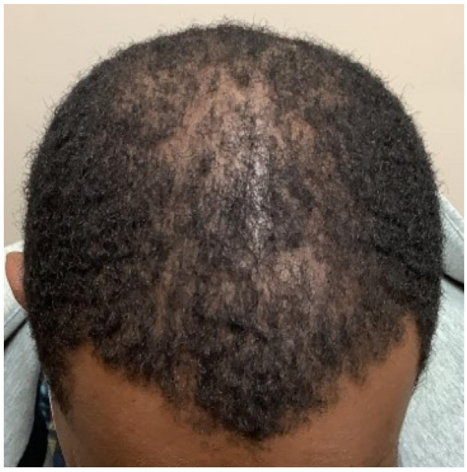

Fig 5. Patient 3 with hair loss on the vertex scalp, with retention of the frontotemporal hairline, at initial presentation.

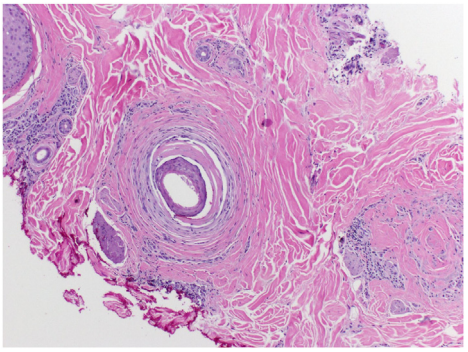

Fig 6. Follicle at center exhibiting premature desquamation of the inner root sheath. The inner root sheath of this follicle has desquamated below the level of the isthmus and shows perifollicular concentric fibrosis. There is an adjacent fibrous follicular scar to the right, a sequela of follicular dropout. Hematoxylin-eosin stain; original magnification: ×10.

history of total baldness in his brother, father, and mother. Trichoscopic examination revealed perifollicular erythema, peripilar gray halos, and perifollicular scale in the focal alopecic patch. A 4-mm scalp biopsy was notable for perifollicular fibrosis, with loss of sebaceous glands, a lymphohistiocytic inflammatory infiltrate, and focal areas of premature loss of the inner root sheath (Fig 6), which was most consistent with CCCA. The patient was diagnosed with CCCA, given the biopsy findings and clinical corollary, and started on a compounded cream consisting of 12% minoxidil with 0.05% clobetasol in a petrolatum emulsion 3 times weekly, 100-mg doxycycline hyclate capsules once daily, and regular 5-mg/mL intralesional triamcinolone acetonide injections.

DISCUSSION

CCCA is a primary scarring alopecia that disproportionately impacts adult women of African ancestry. It classically begins at the vertex scalp and expands centrifugally with loss of follicular ostia as the disease progresses. Most cases of CCCA have been reported in the late second or third decade of life in Black women,[2] whereas few cases are known in men, which could result from underdiagnosis in the male population. Hair grooming practices, such as chemical relaxers, hot combs, and tight hairstyles, have been linked to the etiology of CCCA, although multiple studies have not shown consistent findings.[3]

The pathophysiology of CCCA is not entirely understood; however, it is thought to be multifactorial. In some families, the inheritance pattern is similar to that of an autosomal dominant trait.[4] Additionally, missense mutations leading to decreased expression of peptidyl arginine deiminase type III (PADI3) have been found in scalp biopsies of women with CCCA.[5] Along with fibroproliferative disorders, type 2 diabetes mellitus, hyperlipidemia, and breast cancer have also recently been shown to be associated with CCCA.[3,4]

CCCA clinically manifests as a patch of hair thinning at the midline portion of the vertex scalp, progressing over time in a centrifugal pattern (Figs 1, 3, and 5). Atypical presentations, with irregular patchy alopecia in the occipital or temporal scalp, have also been reported.[6] Our patients demonstrated the classic presentation with thinning at the vertex scalp and a retained frontal hairline (Figs 4 and 5). In early disease, unexplained hair breakage on the crown can be one of the first signs and can occasionally be accompanied by a burning sensation, pain, tenderness, dysesthesia, or pruritus of the scalp.[6,7] Clinical manifestations of inflammation are highly variable among patients; it can be apparent with follicular papules, pustules, perifollicular erythema, and scaling.[8] Notably, 2 of the 3 patients in our case series reported ongoing or prior locs, a high-tension styling practice, which may have potentially propagated inflammation and progression of the disease, although more studies are needed to confirm this observation. As with our patients, most people with CCCA do not seek medical care for several months or even years as hair loss progresses, making an early diagnosis of the disease less common.[7]

Clinical examination often reveals loss of follicular ostia between hair shafts, which is characteristic of cicatricial alopecias.[8] The most common and specific trichoscopy finding is peripilar white or gray halos,

Table I. Comparison of androgenetic alopecia and central centrifugal cicatricial alopecia in Black men

History/Features	AGA	CCCA
Clinical history		
Hair care practices		History of high-tension hairstyle practices (cornrows, braids, extensions, weaves with sewn-in or glued-on hair), use of hot combs, and frequent use of hair relaxers[3]
Family history	Family history of hair loss consistent with Norwood scale	Family history of hair loss in similar pattern
Clinical features		
Distribution	Bitemporal or frontal hairline recession in addition to thinning of temporal and vertex scalp	Focal, vertex/crown of the scalp, with retained frontal hairline
Inflammatory features	Inflammatory features not present	Pain, burning sensation, tenderness, dysesthesia, and pruritus[8]
Trichoscopic features	Follicular miniaturization[10,11]	Peripilar white-gray halo[8]
	Peripilar sign (brown halo)[11]	Follicular dropout[8]
	Yellow dots (empty follicles)[10]	Follicular papules and pustules[8]
	Anisotrichosis[10]	Perifollicular erythema and scale[8]
	Increased proportion of vellus hairs[11]	Follicular miniaturization[8]
	Focal atrichia[10]	Honeycomb pigmented network, often interrupted by white patches[8]
	Honeycomb pigmented network (normal)[8]	
Histologic features	Progressive miniaturization of terminal hair follicles[11]	Lymphocytic infiltrates[8,9]
	Increased number of vellus and telogen hairs[11]	Perifollicular fibrosis[8,9]
	Reduction of anagen-to-telogen ratio (<5:1)[11]	Follicular dropout[8,9]
		Premature degeneration of the inner root sheath[8,9]

AGA, Androgenetic alopecia, *CCCA*, central centrifugal cicatricial alopecia.

which was seen in all our patients (Fig 2). Other trichoscopy findings include a honeycomb pigmented network interrupted by irregular white patches, follicular papules and pustules, perifollicular erythema, and scale.[8] The peripilar white-gray halos seen using trichoscopy are often correlated with the histologic finding of perifollicular fibrosis surrounding the outer root sheath.[9] Additional histologic features of CCCA include lymphocytic infiltrate, follicular dropout, and premature degeneration of the inner root sheath (Fig 6).[9]

CCCA and AGA have overlapping clinical and trichoscopic features that can lead to misdiagnosis (Table I).[8-11] Both CCCA and AGA can present initially as hair thinning on the vertex scalp; however, the limited extent of hairline recession in CCCA contrasts the more extensive recession of the frontotemporal scalp in AGA (Figs 4 and 5). Unlike the Norwood Hamilton scale for male pattern hair loss, wherein patients demonstrate progressive bitemporal thinning prior to notable vertex scalp hair loss, our series of men had relative retention of the temporal hairline. This may be a useful clue for distinguishing CCCA from AGA clinically, considering their potential overlap and concurrent existence, because AGA is the leading form of alopecia in men. Patients with CCCA may also present with symptoms of itch, pain, or burning. Perifollicular erythema, follicular papules, and scaling are also more suggestive of a scarring process, with deep-red or violaceous hues often seen in darker skin types. The lack of inflammatory features in AGA can prompt clinicians to consider other types of alopecia, such as CCCA, while formulating a comprehensive differential. These 2 forms of alopecia can also be distinguished based on trichoscopy findings given the absence of follicular ostia is key to the scarring process in CCCA.[7] There are no established diagnostic criteria for CCCA[12]; therefore, clinicopathologic correlation is often necessary to differentiate AGA and CCCA or confirm the diagnosis in a concomitant case of both and aid in individualized management options for better outcomes. Although AGA may exist concomitantly with CCCA, especially in male patients, and therapies, such as minoxidil, can be used to increase hair density in both conditions, it is important to identify CCCA correctly to initiate additional anti-inflammatory therapy as soon as possible.

Our 3 cases reveal that although CCCA has been predominant in Black women, it should also be considered in Black men. Our cases highlight a key clinical distinguishing feature of male CCCA, demonstrating notable thinning on the vertex scalp with

relative maintenance of the hairline, a feature that is yet to be noted in previous literature and can help clinicians more easily differentiate this form of scarring alopecia in Black men from AGA. To our knowledge, there have only been 12 reported cases of CCCA in Black men in the literature.[13-15] The paucity of published evidence of CCCA cases in Black men raises the following question: how common is misdiagnosis of CCCA for other alopecias? Our case series demonstrates the need for clinicians to thoroughly review symptoms as well as clinical and trichoscopic findings and have an appropriate threshold for scalp biopsy when there is concern for a scarring process. Our cases add to the limited, but now growing, evidence of CCCA in Black men and are a call for future studies to explore the clinical characteristics of CCCA and diagnostic outcomes in Black men.

Conflicts of interest

None disclosed.

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