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Central Centrifugal Cicatricial Alopecia Following a Patchy Pattern: A New Form of Clinical Presentation and a Challenging Diagnosis for the Dermatologist

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ABSTRACT

Central centrifugal cicatricial alopecia (CCCA) is included among the primary lymphocytic cicatricial alopecias. The current nomenclature of CCCA suggested by the North American Hair Research Society refers to the traditional clinical presentation pattern of this type of alopecia, which begins in the central area of the scalp and has a progressive and symmetric centrifuge evolution. However, some exceptions should be highlighted, and a new clinical variety of CCCA presenting with patches of hair loss affecting the lateral and posterior scalp has been recently described. Here, we report a new case of CCCA presenting with a patchy pattern. In addition to the fact that this new patchy presentation of CCCA is not contemplated in the current terminology, it has a difficult differential diagnosis with other alopecias that have patches as their presentation. In these cases, both the trichoscopy and the histopathology are indispensable for diagnosis that will allow a targeted treatment and avoid an unfortunate prognosis.

Key words: Centrifugal, cicatricial alopecia, patchy pattern

INTRODUCTION

Central centrifugal cicatricial alopecia (CCCA) is included among the primary lymphocytic cicatricial alopecias. Its current term was established in reference to its clinical pattern of presentation, which begins in the central area of the scalp and has a progressive and symmetric centrifuge evolution.^[1,2] However, a new clinical variety of CCCA presenting with patches of hair loss affecting the lateral and posterior scalp has been recently described, which may delay and hinder its diagnosis.^[3]

CASE REPORT

We report the case of a 50-year-old female from Ghana with a previous history of hair straightening using chemical products as well as hair styling that involved important

traction of the hair in her youth. She referred hair loss for >10 years, and when examined, a central alopecic patch on the vertex and numerous interconnected alopecic patches in the occipital and both parietal areas were visible [Figure 1]. The trichoscopy revealed a honeycomb network, pinpoint white dots in an irregular distribution, white patches, and peripilar gray-white halos [Figure 2a].

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Figure 1: Central centrifugal cicatricial alopecia with a patchy pattern. Traditional central affection of the vertex as well as multiple interconnected alopecic patches with a maze aspect in the occipital and both parietal areas

A biopsy was performed on one of the parietal patches and confirmed CCCA diagnosis, with the presence of premature desquamation of the inner root sheath, broken hair shafts, compound follicular structures with perifollicular fibrosis forming goggles, and mild lichenoid perifollicular inflammation infiltrates around a follicle or compound follicular structures [Figure 2b].

DISCUSSION

The current nomenclature of CCCA suggested by the North American Hair Research Society (NAHRS) in 2001 refers to the traditional clinical presentation pattern of this type of alopecia. In the majority of patients with a clinical and/or histopathological diagnosis for this kind of alopecia, this pattern, which begins in the central area of the scalp and has a progressive centrifuge evolution conserving the occipital and parietal areas, is also described. However, some exceptions should be highlighted [Table 1].

Nicholson *et al.*^[4] point out to the possibility of a diffuse frontal-parietal affection, although they attribute it to possible concomitant traction alopecia (TA). In 2007, Khumalo's group reported cases of CCCA directly related to the use of hair relaxers, with involvement of vertex^[5] and both sides of the scalp or with "patterned hair loss"; however, the absence of histopathological information did not allow for the rejection of androgenetic alopecia.^[6,7] In 2008, Olsen *et al.* suggested a photographic scale to determine the severity of the CCCA, with two central affection patterns, namely frontal or vertex accentuation.^[9,13] Finally, in 2015, Miteva and Tosti describe a CCCA with

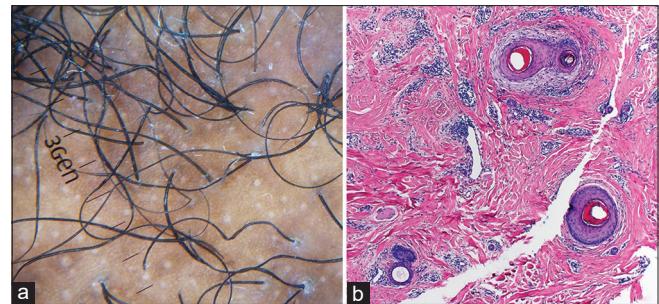


Figure 2: (a) Trichoscopy: A honeycomb network, pinpoint white dots in an irregular distribution, and peripilar gray-white halos were observed in trichoscopy. (b) Histopathology H and E, $\times 10$: We can see premature desquamation of the inner root sheath, compound follicular structures with perifollicular fibrosis forming goggles, and mild lymphocytic lichenoid perifollicular inflammation

a patchy pattern, manifesting with alopecic patches in the parietal or occipital areas in addition to the traditional central affection.^[3] Here, we report a new case of CCCA presenting with a patchy pattern.

In addition to the fact that this new patchy presentation of CCCA is not contemplated in the current terminology suggested by the NAHRS, this alopecia has a difficult differential diagnosis with other alopecias that have patches as their presentation such as alopecia areata, lichen planopilaris, discoid lupus erythematosus, tinea capitis, or TA. The last one is also highly prevalent in African and Caribbean women or high phototype women, and even though they have predilection for marginal frontal-parietal areas, they can also show a patchy pattern in the rest of the scalp. The characteristic trichoscopy findings of the CCCA such as the peripilar gray-white halos, the irregular interfollicular white dots over a honeycomb network, or cicatricial white areas, and at a histological level, the inflammation and compound follicular structures with perifollicular fibrosis or the destruction of sebaceous glands, which will be absent in the TA, will allow the differential diagnosis.^[12,14]

Knowing the existence of this new presentation of the CCCA is essential to perform an accurate diagnosis that will allow a targeted treatment and avoid an unfortunate prognosis. Both the trichoscopy and the histopathology are indispensable for confirmation. We suggest a reflexion regarding the current CCCA terminology due to the fact it does not comprise all the cases of this pathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

Table 1: Main series of cases of central centrifugal cicatricial alopecia with their clinical presentation

Authors	Years	Number of cases	Sex and origin	Diagnosis	CCCA clinical pattern
LoPresti et al. ^[1]	1968	51	African-American women	Clinical and histopathological in 23 cases	Hair loss in the geometric center of the head, eventually reach the frontal hairline
Sperling and Sau ^[2]	1992	10	African-American Women	Clinical and histopathological	Hair loss on the crown of the scalp with centrifugal and symmetrical spreading
Nicholson et al. ^[4]	1993	8	African-Caribbean women	Clinical and histopathological in 6 cases	Hair loss over the vertex, although diffuse and frontal-parietal involvement was also present in four cases
Nnoruka ^[5]	2005	6	African women	Clinical and histopathological	Hair loss centered on the vertex
Khumalo et al. ^[6]	2007	5	African women	Clinical and histopathological	Hair loss centered on the vertex except two cases affecting sides of the scalp
Khumalo et al. ^[7]	2007	16	African women	Exclusively clinical	Hair loss centered on the vertex except two cases with patterned hair loss
Gathers et al. ^[8]	2009	51	African American Women	Clinical and histopathological	Hair loss centered on the crown or vertex of the scalp
Olsen et al. ^[9]	2011	61	African-American women	Exclusively clinical	Central hair loss with frontal accentuation or vertex accentuation
Kyei et al. ^[10]	2011	52	African-American women	Exclusively clinical	Central hair loss with frontal accentuation or vertex accentuation
Callender et al. ^[11]	2012	9	African-American women	Clinical and histopathological	Area of hair breakage and decreased hair density on central crown or vertex
Miteva and Tosti ^[12]	2015	14	African-American women	Clinical and histopathological	Patches of hair loss on the parietal and posterior scalp in addition to the central involvement

CCCA – Central centrifugal cicatricial alopecia

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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