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. 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.

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].
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 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
Diagnosis

Hair loss on the crown of the scalp with centrifugal and symmetrical spreading.

Area of hair breakage and decreased hair density on the frontal-parietal region.

Clinical and histopathological findings in 9 patients. Arch Dermatol 2012;148:1047‑52.

Hair loss centered on the vertex except two cases affecting sides of the scalp.

Clinical and histopathological in 6 cases.

Hair loss centered on the vertex except two cases with patterned hair loss.

Diagnosis

Clinical and histopathological in 23 cases.

Hair loss in the geometric center of the head, eventually reach the frontal hairline.

Clinical and histopathological in 8 cases.

Hair loss over the vertex, although diffuse and frontal-parietal involvement was also present in four cases.

Clinical and histopathological in 6 cases.

Hair loss centered on the vertex.

Clinical and histopathological in 5 cases.

Hair loss centered on the vertex except two cases affecting sides of the scalp.

Clinical and histopathological in 6 cases.

Hair loss centered on the vertex except two cases with patterned hair loss.

Clinical and histopathological in 5 cases.

Central hair loss with frontal accentuation or vertex accentuation.

Clinical and histopathological in 5 cases.

Area of hair breakage and decreased hair density on the central crown or vertex.

Clinical and histopathological in 16 cases.

Hair loss centered on the vertex.

Clinical and histopathological in 14 cases.

Patches of hair loss on the parietal and posterior scalp in addition to the central involvement.

Clinical and histopathological in 10 cases.

Hair loss on the crown of the scalp with centrifugal and symmetrical spreading.

Clinical and histopathological in 8 cases.

Hair loss centered on the vertex.

Clinical and histopathological in 6 cases.

Hair loss centered on the vertex except two cases affecting sides of the scalp.

Clinical and histopathological in 5 cases.

Hair loss centered on the vertex except two cases with patterned hair loss.

Clinical and histopathological in 5 cases.

Central hair loss with frontal accentuation or vertex accentuation.

Clinical and histopathological in 5 cases.

Area of hair breakage and decreased hair density on the central crown or vertex.

Clinical and histopathological in 5 cases.

Hair loss centered on the vertex except two cases with patterned hair loss.

Clinical and histopathological in 5 cases.

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