Sir,
A 56-year-old postmenopausal woman, Fitzpatrick's skin Type III, presented with a 5-year-history of diffuse hair loss on the vertex with mild pruritus. She did not have received previous treatments. She had no family history of hair loss. Her mother was a Spanish Caucasian woman and her father was a Dominican male of African descent.

Clinical examination showed decreased hair density on the vertex without significant hair loss patches [Figure 1]. Trichoscopy showed a diffuse loss of follicular openings with patches of perifollicular erythema. Hair shaft variability with vellus hairs was observed, and moderate peripilar casts were present. It also revealed a notable peripilar white halo and a honeycomb pigmented network on the interfollicular area [Figure 2a and b]. A 4-mm punch biopsy was performed. Histological examination showed a mild perifollicular lymphocytic inflammation at the level of the upper isthmus and concentric fibrosis with an absent inner root sheath. Compound follicles with fusion of the outer root sheaths (ORSs) were present [“Google-like” structures, Figure 3]. The diagnosis of central centrifugal cicatricial alopecia (CCCA) Grade II-B was established.[1]

CCCA is a common and progressive form of lymphocyte-predominant scarring alopecia, seen more commonly in women of African descent.[2]

To our knowledge, this is the first report of CCCA in a fair phototype patient. Hair characteristics are typical of African descendants but with a blond color due to her Caucasian miscegenation. African hair has less tensile strength and breaks easily, and it has fewer elastic fibers anchoring the hair follicles to the dermis.[3] PADI3 encodes a peptidylarginine deiminase that is essential to proper hair-shaft formation. Mutations in PADI3 that leads to a decreased enzymatic expression are strongly associated with CCCA.[4]

Trichoscopy of CCCA shows a peripilar gray-white halo, which is a sensitive sign for the diagnosis of CCCA. This corresponds on histopathology to the ORS of the affected follicles with a surrounding zone of lamellar perifollicular fibrosis.

Figure 1: Diffuse hair loss on the vertex in a 56-year-old woman with blond curly hair

Figure 2: Trichoscopy revealed peripilar white halos, honeycomb pigmented network, perifollicular scaling, and diffuse loss of follicular openings (a and b). The images were obtained using a Heine iC1 polarized dermatoscope attached to an iPhone 7 (magnification ×20)

Figure 3: Central centrifugal cicatricial alopecia. Histopathologic examination revealed reduced follicular density and mild lichenoid lymphoid infiltrate with perifollicular fibrosis. Compound follicular structures with perifollicular fibrosis and the absence of inner root sheath (“Google-like structures”) were present.
Pigment-related trichoscopic features of CCCA are pigmented asterisk-like blotches (related to postinflammatory hyperpigmentation) and a preserved honeycomb pigmented network.[3] We hypothesize that pigment-related trichoscopic features are less frequent in fair phototype patients with CCCA. Other main features of CCCA include hair shaft variability, perifollicular erythema, scattered white patches, and pinpoint white dots with an irregular distribution.

CCCA diagnosis is made by histologic examination. It shows common features with other lymphocytic cicatricial alopecias. In active stages, there is a follicular lichenoid inflammation that progresses into follicular fibrosis. The main diagnostic keys are reduced follicular density with altered follicular architecture due to areas of follicular dropout, absent or focally preserved sebaceous glands in a “hugging pattern” distribution, premature desquamation of the inner root sheath, and finally, individual or compound follicular structures surrounded by perifollicular fibrosis. This hallmark is known as “Google-like structures” and represents fusion in the ORS of adjacent follicles, entrapped in mild inflammation and marked fibrosis.[3]

CCCA should be always suspected, despite skin phototype. Trichoscopy is a useful tool that can help clinicians to select the most rentable site to obtain scalp biopsy sample.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that their name 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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