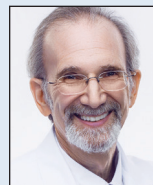




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## Central Centrifugal Cicatricial Alopecia

### THE PRESENTATION:

- A** Early central centrifugal cicatricial alopecia with a small central patch of hair loss in a 45-year-old Black woman.
- B** Late central centrifugal cicatricial alopecia with a large central patch of hair loss in a 43-year-old Black woman.



Photographs courtesy of Richard P. Usatine, MD.

Scarring alopecia is a collection of hair loss disorders including chronic cutaneous lupus erythematosus (discoid lupus), lichen planopilaris, dissecting cellulitis, acne keloidalis, and central centrifugal cicatricial alopecia (CCCA).<sup>1</sup> Central centrifugal cicatricial alopecia (formerly hot comb alopecia or follicular degeneration syndrome) is a progressive, scarring, inflammatory alopecia and represents the most common form of scarring alopecia in women of African descent. It results in permanent destruction of hair follicles.

### Epidemiology

Central centrifugal cicatricial alopecia predominantly affects women of African descent but also may affect men. The prevalence of CCCA in those of African descent has varied in the literature. Khumalo<sup>2</sup> reported a prevalence of 1.2% for women younger than 50 years and 6.7% in women older than 50 years. Central centrifugal cicatricial alopecia has been reported in other ethnic groups, such as those of Asian descent.<sup>3</sup>

Historically, hair care practices that are more common in those of African descent, such as high-tension hairstyles as well as heat and chemical hair relaxers, were implicated in the development of CCCA. However, the causes of CCCA are most likely multifactorial, including family history, genetic mutations, and hair care practices.<sup>4,7</sup> *PADI3* mutations likely predispose some women to CCCA. Mutations in *PADI3*, which encodes peptidyl arginine deiminase 3 (an enzyme that modifies proteins crucial for the formation of hair shafts), were found in some patients with CCCA.<sup>8</sup> Moreover, other genetic defects also likely play a role.<sup>7</sup>

### Key clinical features

Early recognition is key for patients with CCCA.

- Central centrifugal cicatricial alopecia begins in the central scalp (crown area, vertex) and spreads centrifugally.

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# Dx ACROSS THE SKIN COLOR SPECTRUM

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- Scalp symptoms such as tenderness, pain, a tingling or crawling sensation, and itching may occur.<sup>9</sup> Some patients may not have any symptoms at all, and hair loss may progress painlessly.
- Central hair breakage—forme fruste CCCA—may be a presenting sign of CCCA.<sup>9</sup>
- Loss of follicular ostia and mottled hypopigmented and hyperpigmented macules are common findings.<sup>6</sup>
- Central centrifugal cicatricial alopecia can be diagnosed clinically and by histopathology.

## Worth noting

Patients may experience hair loss and scalp symptoms for years before seeking medical evaluation. In some cultures, hair breakage or itching on the top of the scalp may be viewed as a normal occurrence in life.

It is important to set patient expectations that CCCA is a scarring alopecia, and the initial goal often is to maintain the patient's existing hair. However, hair and areas responding to treatment should still be treated. Without any intervention, the resulting scarring from CCCA may permanently scar follicles on the entire scalp.

Due to the inflammatory nature of CCCA, potent topical corticosteroids (eg, clobetasol propionate), intralesional corticosteroids (eg, triamcinolone acetonide), and oral anti-inflammatory agents (eg, doxycycline) are utilized in the treatment of CCCA. Minoxidil is another treatment option. Adjuvant therapies such as topical metformin also have been tried.<sup>10</sup> Importantly, treatment of CCCA may halt further permanent destruction of hair follicles, but scalp symptoms may reappear periodically and require re-treatment with anti-inflammatory agents.

## Health care highlight

Thorough scalp examination and awareness of clinical features of CCCA may prompt earlier diagnosis and prevent future severe permanent alopecia. Clinicians should encourage patients with suggestive signs or symptoms of CCCA to seek care from a dermatologist.

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