Central Centrifugal Scarring Alopecia

Getting familiar with this newly named condition. Plus, how CCSA differs from other alopecias.

By Heather Woolery-Lloyd, M.D.

Central centrifugal scarring alopecia (CCSA) is a new term that encompasses the same clinical presentation originally described with hot comb alopecia and follicular degeneration syndrome (FDS). It also serves to include other related diseases with slightly different epidemiology (pseudopelade) and extent of inflammation (folliculitis decalvans).

A LOOK BACK

This pattern of alopecia, which describes a pattern of progressive, symmetric scarring alopecia on the vertex of the scalp, was first described in African-American women in 1968 and was termed hot comb alopecia. Clinically, patients had a history of hot comb use and presented with progressive scarring alopecia on the crown of the scalp. Hot comb styling, common at that time, involved applying petrolatum to the hair while using a heated metal comb to straighten the hair in sections. It was proposed that, as the sections of hair on the crown were straightened, the extremely hot petrolatum traveled down to the scalp causing burns, significant inflammation, and scarring.

It was thought the damaged hair follicles were localized to the crown due to gravity. The lateral and posterior scalp were spared since the hairs in those sections were stretched horizontally preventing the hot petrolatum from contacting the scalp. With time, however, the diagnosis of hot comb alopecia became controversial because many women who presented with this pattern of hair loss denied a history of hot comb use.

In 1992, hot comb alopecia was renamed follicular degeneration syndrome by Sperling and Sau. Patients included women and men with and without a history of hot comb use. Clinically, patients had a slowly progressive, symmetric alopecia on the crown of the scalp. The disease process usually began in patients in their twenties and progressed over 20 to 30 years. Early in the disease, patients complained of tenderness, dysesthesia or pruritus of the scalp. The hair loss spread peripherally and sometimes involved the frontal hairline. The lateral and posterior scalp were spared. Erythema, scaling and induration were not present; however, mild perifollicular hyperpigmentation was described. In end-stage disease, the skin on the affected scalp appeared thin and shiny with few follicular ostia present. The remaining hairs often demonstrated polytrichia with multiple hairs growing from a common orifice.

Pathology revealed the following:
1. desquamation of the inner root sheath well below the isthmus
2. thinning of the outer root sheath
3. a mononuclear cell infiltrate
4. lamellar fibroplasia
5. fibrosing columns replacing follicles.

A SHIFT IN THOUGHT

At that time, the focus on the cause of this scarring alopecia shifted from hot comb use to the specific histological findings in this disease. Premature degeneration of the inner root sheath was considered the earliest and most important finding in FDS.

One theory proposed that the tightly curled hair shaft in predisposed African-Americans exerted pressure on the follicular wall. In follicles with premature desquamation, this hair shaft could lead to inflammation, fibroplasia and resulting scarring alopecia. Others suggested that in predisposed populations, FDS might be initiated by puberty, haircare practices or breakdown after years of subclinical follicular injury.
Although a specific cause for FDS wasn’t asserted at that time, the authors did conclude that this form of alopecia wasn’t solely related to hot comb use.

PROPOSING A NEW NAME
In 2001, a new schema was proposed to simplify the classification of scarring alopecia and to eliminate redundant nomenclature. The term central centrifugal scarring alopecia was suggested to describe the clinical entity of progressive, symmetric, scarring alopecia on the crown of the scalp.

Clinical presentation was identical to that of FDS with the characteristic inflammation and other histological findings. However, the term CCSA included more than FDS. The broader terminology was aimed to group FDS with pseudopelade and folliculitis decalvans because of their similar clinical and histological presentations. Pseudopelade, as described by the authors, differed from FDS primarily by its presence in both African Americans and Caucasians. Folliculitis decalvans, the authors proposed, was a more acute, inflammatory presentation of the same condition. Although inflammation was absent in the original description of hot comb alopecia and FDS, current consensus is that inflammatory papules may be present in early disease.

DISTINGUISHING CCSA FROM OTHER ALOPECIAS
CCSA should be distinguished from other forms of alopecia seen in African-American women and men such as the following conditions below.

- **Traction alopecia.** This condition is a non-scarring alopecia characterized by temporal scalp thinning. It’s most common in patients who wear tight braids or ponytails. You’ll also see this condition in the areas between tight cornrows, rollers and multi-tufted braids. With early intervention and modification of hairstyling techniques, this alopecia is reversible.

- **Androgenetic alopecia.** Also seen in African-American women, this condition clinically presents with diffuse hair thinning that’s most prominent on the frontal scalp. This is a non-scarring alopecia, and the follicular ostia are preserved. These patients often have a family history of androgenetic alopecia and can be treated with topical minoxidil (Rogaine).

- **Acne keloidalis nuchae.** Although this is a common condition in African Americans, it has a clinically distinct presentation on the posterior scalp that’s not likely to be confused with CCSA.

- **Other forms of scarring alopecias.** Conditions such as lichen planopilaris, discoid lupus, and morphea have different clinical presentations and can be distinguished from CCSA by their histologies, which demonstrate an interface dermatitis and epidermal involvement.

TACKLING THE CHALLENGES OF TREATMENT
CCSA is easily diagnosed by the classic clinical presentation described; however, biopsies help in confirming the diagnosis and ruling out other causes of scarring alopecia. Once diagnosed, treatment is challenging.

Treatment options have primarily focused on topical and intrallesional steroids, but they haven’t provided dramatic results. High-potency topical steroids may halt the process in those patients with early disease. Oral antibiotics have sometimes been utilized for their anti-inflammatory effects. The newer topical immunomodulators such as tacrolimus (Protopic) and pimecrolimus (Elidel) may offer novel options to patients with early inflammation. Hair transplantation was reported successful in a patient with scarring alopecia with retention of the transplanted hair after two years. Temporal transposition flaps of hair-bearing scalp have also been reported.

In general, the management of CCSA has been a therapeutic challenge. Patients most often utilize hairstyles that disguise this central alopecia, in addition to hair pieces and wigs.

CONTINUED DEBATE
The cause of CCSA continues to be a topic of debate. In 2000, Ackerman asserted that FDS was traction induced and was end-stage traction alopecia. Others have said that extensive chemical processing leads to chronic inflammation in the scalp and scarring alopecia.

Since many different haircare practices are used by African Americans to improve hair manageability, it’s difficult to assume one particular cause of CCSA. Likely, the etiology of this scarring alopecia is multifactorial. Both haircare practices and a genetic predisposition may play a role. Further research is needed to pinpoint the cause and improve treatment options for patients who have this disfiguring dermatologic condition.

Dr. Woolery-Lloyd is the director of ethnic skin research for the Division of Cosmetic Dermatology in the Department of Dermatology at the University of Miami.