Central centrifugal cicatricial alopecia

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ABSTRACT

Central centrifugal cicatricial alopecia is a common cause of progressive permanent apical alopecia. This unique form of alopecia includes entities previously know as "hot comb alopecia," "follicular degeneration syndrome," "pseudopelade" in African Americans and "central elliptical pseudopelade" in Caucasians. The etiology appears to be multifactorial and the condition occurs in all races.

Key words: Alopecia, central, centrifugal, cicatricial, pseudopelade

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Central centrifugal cicatricial alopecia (CCCA) is the term adopted by the North American Hair Research Society (NAHRS) to encompass the previous terms of "hot comb alopecia," "follicular degeneration syndrome," "pseudopelade" in African Americans and "central elliptical pseudopelade" in Caucasians.[1] CCCA is a subcategory of primary, inflammatory cicatricial alopecia, and is the most common form of scarring alopecia in many populations.[2] CCCA presents as progressive apical alopecia.[3] Certain hair care practices have been associated with increased risk for developing CCCA; in a review of 44 patients by McMichael,[4] more women with scarring alopecia had a history of hair weaving and long duration of chemical relaxer usage as compared to those unaffected by scarring alopecia.[5] However, there is developing evidence that the etiology of CCCA may be multifactorial, occurs in all races, and does not always relate to obvious hair care practices.[6]

CLINICAL FEATURES

The clinical presentation of CCCA appears to be similar between men and women.[5] However, clinically diagnosing CCCA may be challenging because it can resemble female pattern hair loss, alopecia areata, lichen planopilaris, or telogen effluvium.[7] A biopsy is often necessary to confirm the diagnosis. CCCA typically begins and remains most severe on the crown or vertex of the scalp, gradually expanding in a centrifugal fashion [Figure 1].[8] The affected scalp is in parts smooth and shiny and illustrates massive follicular dropout. Typically, a few short, brittle hairs remain within the scarred expanse.[9] Patients commonly complain of mild dysesthesia (pruritus, tenderness) in the affected area.[7,10] The disease generally progresses slowly, but longstanding or severe disease can result in hair loss covering the entire crown of the scalp.[11] This is in contrast to nonscarring (non-cicatricial) alopecia, which is reversible as the follicular epithelium remains intact.[9] CCCA, at its end stages, is irreversible, as the follicular epithelium has been replaced by connective tissues.[12] Additionally, pustules and crusting may be found in patients with superimposed folliculitis decalvans.

HISTOLOGICAL FEATURES

CCCA, in both men and women, will display similar histopathological features[6] including contraction of dermal collagen with loss of space between collagen bundles and broad hyalinized fibrous tracts that may contain naked hair shafts [Figure 2].[13] Premature desquamation of the inner root sheath has also been described as a characteristic feature, but is also present in lichen planopilaris and other forms of scarring alopecia. In elastic Van Geison-stained sections, thickened dermal elastic fibers are present.[14] The elastic sheath surrounding the fibrous tract is preserved [Figure 3] and often duplicated in contrast to lupus erythematosus, lichen planopilaris, and folliculitis decalvans, where there is loss of the elastic sheath.[15] Additionally, CCCA may demonstrate perifollicular mucinous fibrosis and sparse lymphocytic perifollicular inflammation, which primarily occurs at the level of the upper isthmus and lower infundibulum.[2] In advanced lesions, total destruction of the follicular epithelium with retained hair shaft fragments and granulomatous inflammation will be seen.

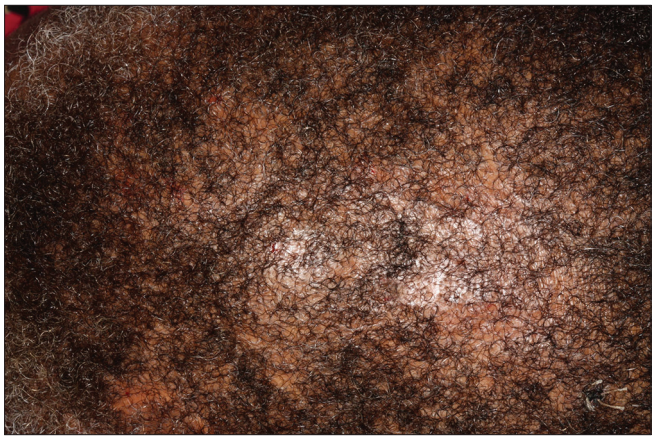

Figure 1: Central centrifugal cicatricial alopecia presents with progressive permanent apical alopecia

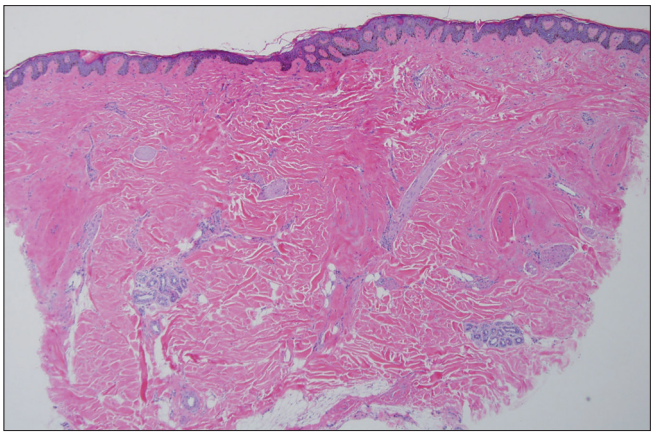

Figure 2: Central centrifugal cicatricial alopecia is characterized by hyalinization of dermal collagen with broad tree trunk-like fibrous tracts (H and E, ×20)

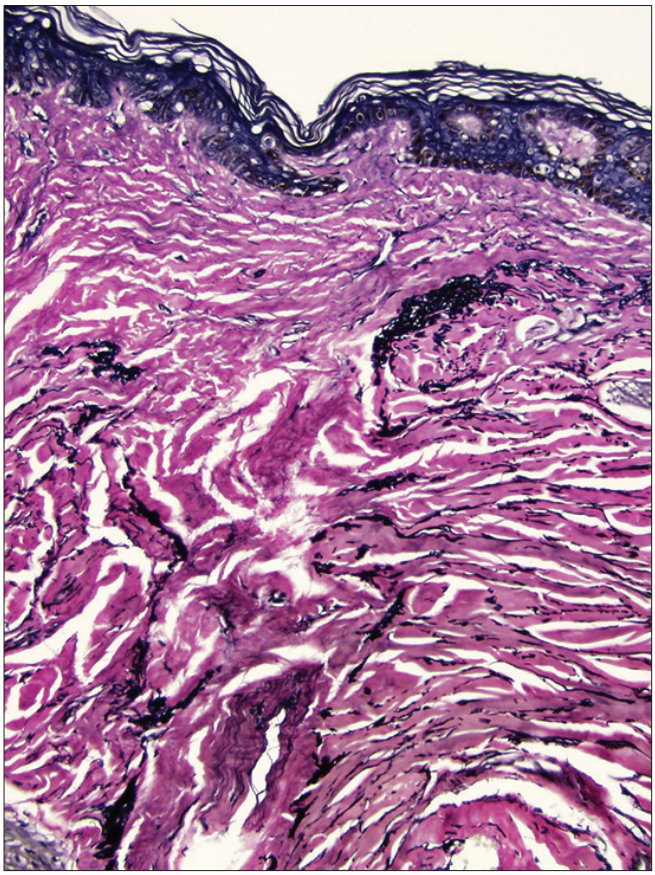

Figure 3: Central centrifugal cicatricial alopecia is characterized by preservation of the elastic sheath surrounding the fibrous tracts (H and E, ×100)

TREATMENT

Therapeutic options are limited. Hair styles that require heat treatment or produce traction should be avoided. Ceasing the use of relaxers is also suggested, as this practice correlates with increased incidence of disease.[1] For those patients who have crops of pustules, suggesting superimposed folliculitis decalvans, topical corticosteroids, and oral tetracyclines can be of benefit. Hair transplantation can be useful in patients with advanced disease; however, CCCA presents a unique challenge for hair transplantation as the presence of scarring can decrease the transplanted graft survival rate.[4]

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