Diffuse and patchy hair loss in occipital region of scalp

Hussein Hassab El-Naby, MD, Sadat Mosbeh, MD

Department of Dermatology, Al-Azhar University, Cairo, Egypt

CLINICAL FINDINGS

A 45-year-old female complaining of decreased number of hairs in occipital area of her scalp. She had a history of hair straightening using chemical products as well as hair styling that involved significant traction of the hair in her youth. She had hair loss for >5 years. Examination of her scalp showed diffuse hair loss as well as alopecic patches in the occipital region of scalp. Frontal region of scalp showed no hair loss (Fig. 1). No personal history of chronic diseases. Family history of the same condition was negative. General physical and systemic examination were within normal limits. Laboratory investigations including CBC, hepatic, renal, Vit D and B12, zinc, hormonal profile showed no abnormal findings.

Fig. 1 Diffuse and patchy Loss of hair in occipital area

What is your clinical diagnosis?

- Alopecia areata
- Traction alopecia
- Ophiasis
- Central centrifugal cicatricial alopecia
- Lichen planopilaris
- Disciod lupus erythematosus
- Tinea capitis

Trichoscopy

The trichoscopy revealed a honeycomb network, pinpoint white dots in an irregular distribution, white patches, and peripilar gray-white halos (Fig. 2)

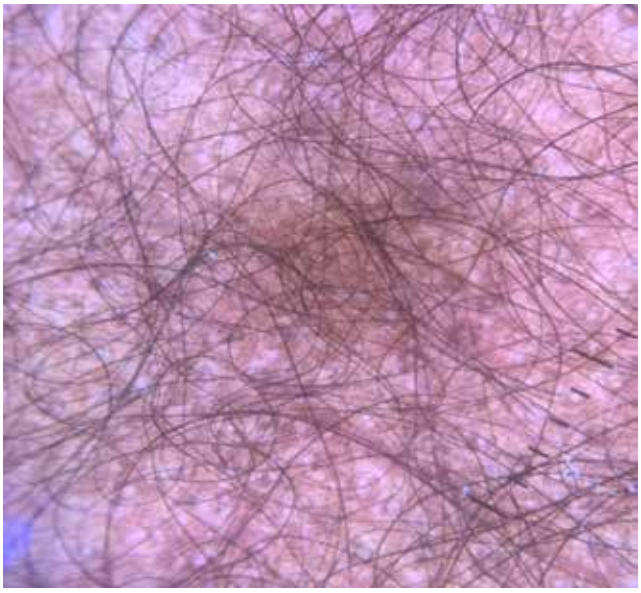

Fig. 2 The trichoscopy shows peripilar white halos, loss of follicular openings, disrupted pigmented network, honeycomb appearance, irregularly distributed pinpoint white dots

Correspondence: *Dr. Hassab El-Naby H, Department of Dermatology, Al-Azhar University, Cairo, Egypt*

HISTOPATHOLOGY

Microscopic examination form a skin biopsy showed perifollicular lymphocytic inflammatory infilterate, fibrosis and premature loss of inner root sheath. (Fig. 3)

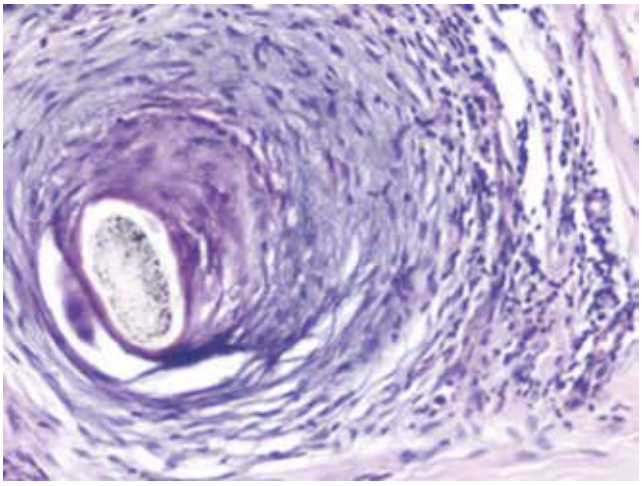

Fig. 3 Lymphocytic perifollicular inflammation, fibrosis and premature loss of inner root sheath

COMMENT

Central centrifugal cicatricial alopecia (CCCA) is a rare type of scarring alopecia that clinically presents as patches of permanent hair loss on the vertex or crown of the scalp and spreads centrifugally.[1] This type of hair loss is associated with signs and symptoms of inflammation. CCCA is a common condition that usually affects women of African descent, and it runs in families.[2,3]

There are several different theories on the origin of CCCA. According to one theory, there is a direct correlation between CCCA and the hair care products used by Black women, such as heated combs, chemical relaxers, and different haircuts that cause traction. Over time, this idea was abandoned when it became apparent that women's non-usage of these hair products was a problem. The etiology may also be influenced by genetic and environmental factors, according to the literature.[5] Some etiologic ideas have centered on the early desquamation of the inner root sheath, which makes it possible for outside elements to penetrate the hair shaft or follicular unit and irritate the outer root sheath, triggering an inflammatory cascade. Examples of these elements include chemicals and bacteria.[6-8] Pathologic evidence of early desquamation of the inner root sheath is observed by microscopic examination from the lesion of CCCA. Another explanation proposes that CCCA might be a fibroproliferative condition that results in low-grade inflammation, follicular scarring, and ultimately decreased hair growth.[9-11] Nevertheless, none of these different explanations is supported by the available data. This suggests that CCCA has a complex etiology. Other hypothesized causes include hereditary factors, autoimmune illnesses, and infections. Further research is required to elucidate the possibility that CCCA is idiopathic.[12]

The most prevalent kind of cicatricial alopecia in middle-aged African American women is called central centrifugal alopecia; the prevalence of this condition varies between 2% and 7% according to different research, and it is most common in those with tightly curled or kinked hair.[13] Men and children rarely experience CCCA, yet certain case studies have mentioned it.[14-16] The condition typically manifests in women at the age of 36.[5] These are the established features of the illness process; further population-level research is required because the majority of studies rely on data from small patient samples collected in urban areas. As of right now, there is a lack of solidly published data regarding the role of CCCA in different populations.[17-19]

Premature desquamation of the inner root sheath is the histologic finding that is discovered in early instances (PDIRS).[7] Perifollicular fibroplasia and a lymphocytic infiltration are visible in the early

histologic alterations. Usually, these infiltrates of inflammation spread from the upper portion of the isthmus to the lower follicular infundibulum.[19] Reduced terminal hair follicles are seen in a small percentage of patients. Subsequent phases are linked to granulomatous inflammation, retention of hair shaft fragments, and destruction of the follicular epithelium. This is followed by the tufting or polytrichia of hair follicles (fusion of infundibulum) and the replacement of follicular epithelium by connective tissue. Histologically, this is similar to both the advanced stage of lichen planopilaris and the early stages of folliculitis keloidalis (FK).[20]

The following details from the patient's medical history and clinical examination aid in the diagnosis of central centrifugal cicatricial alopecia: Usually begins at the scalp's vertex.[21] Palpably sensitive scalp; mild hyperpigmentation surrounding hair follicles; Centrifugally progressive, typically symmetrical.[22] The alopecia area gradually becomes blended with the surrounding healthy scalp. A white peripilar halo or the disappearance of follicular ostia were noted on dermoscopy (a particular marker).[23-25] Historical findings associated with CCCA (e.g., chemical use, African descent, tight hairstyles, hot comb use). Mild burning, soreness, or itching can be limited to the area of hair loss.Islands of unaffected hairs with polytrichia within affected areas. Our case was presented with a rare site in occipital area of scalp.[26,27]

In order to prevent or stop the disease's progression and promote hair growth, hair follicles that have been irreversibly damaged cannot produce new hair. Despite the lack of definitive standards for the management of CCCA and the primarily empirical nature of therapeutic options, prompt action can mitigate the risk of disease progression.[28] Treatment approaches for CCCA elicit a sluggish response. Many times, topical steroids or intralesional triamcinolone acetonide are used in anti-inflammatory therapy, which is thought to be the first line of treatment. When topical steroids are used at lower concentrations, individuals with darker skin types are less likely to get hypopigmentation. Since antibiotics contain anti-inflammatory qualities, they should be used continuously until symptoms resolve (at least two to six months), such as tetracyclines like doxycycline.[29] After the drug has been in a quiescent condition for at least a year, the dose may be lowered and then gradually stopped. In instances of comparable scarring alopecias, systemic anti-inflammatory therapies like as mycophenolate mofetil, hydroxychloroquine, and cyclosporine have also been employed.[30] When there is active inflammation, short doses of oral corticosteroids are recommended. Additionally, vitamin D might be useful in management.[2] Other small-scale studies with tacrolimus, topical metformin, and minoxidil have been conducted.[3,31,32] It is advised to groom hair little, however the evidence is insufficient to support this. Haircare routines must be shortened or changed to be more concise. It is preferable to refrain from excessive hair traction. Mild shampooing is used once a week to alleviate symptoms.[3]

Clinically, central centrifugal cicatricial alopecia is similar to the following conditions: Female pattern hair loss, a nonscarring alopecia that develops around adolescence or after menopause and is linked to hyperandrogenism. The main characteristics that set it apart from CCCA are the obvious follicular openings and lack of scarring.[33] Lichen planopilaris: In certain situations, this form of scarring alopecia is similarly indistinguishable. As

opposed to central centrifugal cicatricial alopecia, follicular keratosis and perifollicular erythema are the symptoms of lichen planopilaris. An other lichen planopilaris variant is frontal fibrosing alopecia. Face papules and gradually worsening scalp baldness are the hallmarks of this. Alopecia also affects other body parts, such as the eyebrows and eyelashes.[34] Tinea capitis: A wood lamp examination can be used to distinguish between this fungal infection of the scalp, which produces intense green fluorescence when microsporum is present and weak blue fluorescence when Trichophyton schoenleinii species is present. Alopecia both with and without scarring is a common presentation of the illness. Discoid lupus erythematosus: This type of alopecia that leaves scars typically affects the scalp and manifests as erythematous, scaly plaques with pigmentary alterations and follicular plugging. The histology results distinguish this illness from CCCA. Histopathology reveals interface dermatitis along with perivascular and periadnexal lymphohistiocytic infiltrates.[34] Degenerative alterations might be seen in the basal layer. Pseudopelade of Brocq: This condition usually affects middle-aged and older women and commonly presents as irregular patches of hair loss that usually begin at the vertex. The bald areas look like "footprints in the snow". Histopathology of the lesion shows a thin epidermis with sclerotic dermis and streamers of fibrosis that go up to the fat layer.[35] Similar to central centrifugal alopecia, lichen planopilaris manifests as superficial perifollicular fibrosis, infundibular inflammation, and destruction, resulting in liberated hair shafts in the dermis. Nevertheless, peri-infundibular hypergranulosis and vacuolar lichenoid dermatitis with epidermal cytoid bodies distinguish it. One common characteristic is dyskeratosis with perifollicular lymphocytic inflammation. Frontal fibrosing alopecia is characterized by a slow, progressive loss of hair follicles, lamellar fibrosis around the isthmus and lower infundibulum, and lymphohistiocytic infiltration.[2]

The disease's stage affects the prognosis for CCCA. Hair regrowth may be enhanced by regular treatment and early diagnosis.[37] Due to delayed identification and treatment, patients with severe disease have a poor prognosis and may have minimal chance of hair regeneration due to scarring.[1] patient education is crucial in reducing the anxiety related to central centrifugal cicatricial alopecia. It is important to emphasize that while there is no known cure for the scarring stage, it can be managed and controlled. Patients should refrain from using heat or friction in their hairstyles. Regular use of hair relaxers raises the risk of disease, so it should be avoided. Participation in different support groups and hair advocacy organizations should be encouraged for patients.

REFERENCES

1. Flamm A, Moshiri AS, Roche F, Onyekaba G, Nguyen J, James AJ, Taylor S, Seykora JT. Characterization of the inflammatory features of central centrifugal cicatricial alopecia. J Cutan Pathol. 2020; 47(6):530-34.
2. Aguh C. Updates in our understanding of central centrifugal cicatricial alopecia. Cutis. 2019; 104(6):316:40.
3. Araoye EF, Thomas JAL, Aguh CU. Hair regrowth in 2 patients with recalcitrant central centrifugal cicatricial alopecia after use of topical metformin. J Amer Acad Dermatol Case Rep. 2020; 6(2):106-08.
4. Sundberg JP, Hordinsky MK, Bergfeld W, Lenzy YM, McMichael AJ, Christiano AM, McGregor T, Stenn KS, Sivamani RK, Pratt CH, King LE. Cicatricial Alopecia Research Foundation meeting, May 2016: Progress towards the diagnosis, treatment and cure of primary cicatricial alopecias. Exp Dermatol. 2018; 27(3):302-10.

5. Malki L, Sarig O, Romano MT, Méchin MC, Peled A, Pavlovsky M, *et al.* Variant PADI3 in Central Centrifugal Cicatricial Alopecia. N Engl J Med. 2019; 380(9):833-41.
6. Tan T, Guitart J, Gerami P, Yazdan P. Premature Desquamation of the Inner Root Sheath in Noninflamed Hair Follicles as a Specific Marker for Central Centrifugal Cicatricial Alopecia. Am J Dermatopathol. 2019; 41(5):350-54.
7. Miteva M, Tosti A. Pathologic diagnosis of central centrifugal cicatricial alopecia on horizontal sections. Am J Dermatopathol. 2014; 36(11):859-64; quiz 865-67.
8. Miteva M, Tosti A. 'A detective look' at hair biopsies from African-American patients. Br J Dermatol. 2012; 166(6):1289-94.
9. Samrao A, Lyon L, Mirmirani P. Evaluating the association of central centrifugal cicatricial alopecia (CCCA) and fibroproliferative disorders. Dermatol Online J. 2021; 27(8).
10. Aguh C, Dina Y, Talbot CC, Garza L. Fibroproliferative genes are preferentially expressed in central centrifugal cicatricial alopecia. J Am Acad Dermatol. 2018; 79(5):904-12.e1.
11. Jamerson TA, Talbot CC, Dina Y, Aguh C. Presence of Uterine Leiomyomas Has No Significant Impact on Gene Expression Profile in the Scalp of Patients with Central Centrifugal Cicatricial Alopecia. J Inves Dermatol Innov. 2022; 2(1):100060.
12. Anzai A, Wang EHC, Lee EY, Aoki V, Christiano AM. Pathomechanisms of immune-mediated alopecia. Int Immunol. 2019; 31(7):439-47.
13. Herskovitz I, Miteva M. Central centrifugal cicatricial alopecia: challenges and solutions. Clin Cosmet Investig Dermatol. 2016; 9:175-81.
14. Eginli AN, Dlova NC, McMichael A. Central Centrifugal Cicatricial Alopecia in Children: A Case Series and Review of the Literature. Pediatr Dermatol. 2017; 34(2):133-37.
15. Davis EC, Reid SD, Callender VD, Sperling LC. Differentiating central centrifugal cicatricial alopecia and androgenetic alopecia in african american men: report of three cases. J Clin Aesthet Dermatol. 2012; 5(6):37-40.
16. Sperling LC, Skelton HG, Smith KJ, Sau P, Friedman K. Follicular degeneration syndrome in men. Arch Dermatol. 1994; 130(6):763-69.
17. Akintilo L, Hahn EA, Yu JMA, Patterson SSL. Health care barriers and quality of life in central centrifugal cicatricial alopecia patients. Cutis. 2018; 102(6):427-32.
18. Sperling LC, Hussey S, Sorrells T, Wang JA, Darling T. Cytokeratin 75 expression in central, centrifugal, cicatricial alopecia--new observations in normal and diseased hair follicles. J Cutan Pathol. 2010; 37(2):243-48.
19. Heath CR, Usatine RP. Central Centrifugal Cicatricial Alopecia. Cutis. 2022; 109(4):235-36.
20. Fernandez-Flores A. Use of Giemsa staining in the evaluation of central centrifugal cicatricial alopecia. J Cutan Pathol. 2020; 47(5):496-99.
21. Miteva M, Tosti A. Central Centrifugal Cicatricial Alopecia Presenting with Irregular Patchy Alopecia on the Lateral and Posterior Scalp. Skin Appendage Disord. 2015; 1(1):1-5.
22. Griggs J, Trüeb RM, Gavazzoni Dias MFR, Hordinsky M, Tosti A. Fibrosing alopecia in a pattern distribution. J Am Acad Dermatol. 2021; 85(6):1557-64.
23. Miteva M, Tosti A. Dermatoscopic features of central centrifugal cicatricial alopecia. J Am Acad Dermatol. 2014; 71(3):443-49.
24. Su HJ, Cheng AY, Liu CH, Chu CB, Lee CN, Hsu CK, Lee JY, Yang CC. Primary scarring alopecia: A retrospective study of 89 patients in Taiwan. J Dermatol. 2018; 45(4):450-55.
25. Wu S. Hair and Nail Conditions: Alopecia Evaluation. FP Essent. 2022; 517:11-16.
26. Jiang VS, Hawkins SD, McMichael A. Female pattern hair loss and polycystic ovarian syndrome: more than just hirsutism. Curr Opin Endocrinol Diabetes Obes. 2022; 29(6):535-40.
27. Carmina E, Azziz R, Bergfeld W, Escobar-Morreale HF, Futterweit W, Huddleston H, Lobo R, Olsen E. Female Pattern Hair Loss and Androgen Excess: A Report From the Multidisciplinary Androgen Excess and PCOS Committee. J Clin Endocrinol Metab. 2019; 104(7):2875-91.
28. Bolduc C, Sperling LC, Shapiro J. Primary cicatricial alopecia: Other lymphocytic primary cicatricial alopecias and neutrophilic and mixed primary cicatricial

alopecias. J Am Acad Dermatol. 2016; 75(6):1101-17.
29. George EA, Matthews C, Roche FC, Taylor SC. Beyond the Hot Comb: Updates in Epidemiology, Pathogenesis, and Treatment of Central Centrifugal Cicatricial Alopecia from 2011 to 2021. Am J Clin Dermatol. 2023; 24(1):81-88.
30. Fagan N, Meah N, York K, Bokhari L, Fletcher G, Chen G, *et al.* Shedding light on therapeutics in alopecia and their relevance to COVID-19. Clin Dermatol. 2021; 39(1):76-83.
31. Lobon K, Pinczewski J, Bhoyrul B. Significant hair regrowth in a Middle Eastern woman with central centrifugal cicatricial alopecia. Clin Exp Dermatol. 2022; 47(1):136-38.
32. Umar S, Kan P, Carter MJ, Shitabata P, Novosilska M. Lichen Planopilaris Responsive to a Novel Phytoactive Botanical Treatment: A Case Series. Dermatol Ther (Heidelb). 2022; 12(7):1697-1710.
33. Rogers N. Imposters of androgenetic alopecia: diagnostic pearls for the hair restoration surgeon. Facial Plast Surg Clin North Am. 2013; 21(3):325-34.
34. Callender VD, Wright DR, Davis EC, Sperling LC. Hair breakage as a presenting sign of early or occult central centrifugal cicatricial alopecia: clinicopathologic findings in 9 patients. Arch Dermatol. 2012; 148(9):1047-52.
35. Fernandez-Nieto D, Saceda-Corralo D, Jimenez-Cauhe J, Pindado-Ortega C, Erana I, Moreno-Arrones OM, *et al.* Central Centrifugal Cicatricial Alopecia in a Fair Phototype Patient. Int J Trichology. 2019; 11(6):251-52.
36. Fung MA, Sharon VR, Ratnarathorn M, Konia TH, Barr KL, Mirmirani P. Elastin staining patterns in primary cicatricial alopecia. J Am Acad Dermatol. 2013; 69(5):776-82.
37. Eginli A, Dothard E, Bagayoko CW, Huang K, Daniel A, McMichael AJ. A Retrospective Review of Treatment Results for Patients With Central Centrifugal Cicatrical Alopecia. J Drugs Dermatol. 2017; 16(4):317-20.
38. Pärna E, Aluoja A, Kingo K. Quality of life and emotional state in chronic skin disease. Acta Derm Venereol. 2015; 95(3):312-16.