

PhotoDermDiagnosis

A 12-year-old girl with scarring alopecia

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A 12-year-old girl was referred to the dermatology clinic of Aleppo University Hospital, Syria with the chief complaint of alopecia centred on the crown. Alopecia started on the apex a year ago and gradually there was obvious decrease in the density of hair which expanded in a centrifugal fashion (**Figure 1**). Physical examination of involved area of scalp revealed smooth and shiny surface with massive follicular dropout and a few short, brittle hair detectable within the scarred expanse. The patient had no other complaint such as dysesthesia or burning sensation on affected area. There was no evidence of systemic diseases and the patient was not taking any medicine.



Figure 1 Alopecia centred on the crown with centrifugally expansion.

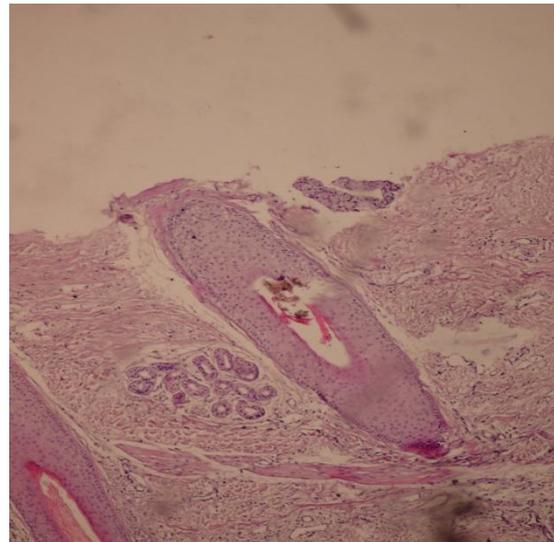


Figure 2 Premature desquamation of inner root sheath.

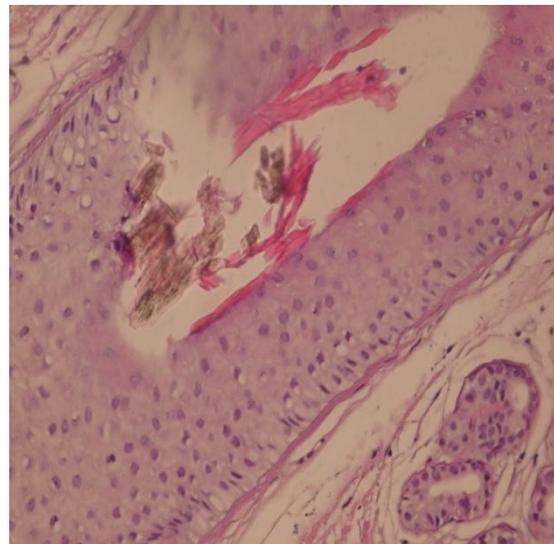


Figure 3 Mild concentric lamellar fibroplasia and a moderate degree of perifollicular, chronic inflammation.

What is the diagnosis?

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Diagnosis

Central centrifugal cicatricial alopecia

Microscopic findings

Histological examination of affected area showed premature desquamation of inner root sheath (**Figure 2**), and concentric lamellar fibroplasia of affected follicle (**Figure 3**), lymphocytic perifollicular inflammation.

Discussion

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,² and is the most common form of scarring alopecia in any population that includes a significant number of black patients. The majority of black patients with CCCA are women, with female: male ratio of about 3:1.^{3,4} The hypothesis of pathogenesis is that patients are predisposed to follicular damage because of an anatomic abnormality namely premature desquamation of the inner root sheath.^{3,4,5} All conditions that result in a progressive pattern of alopecia affecting the crown can mimic CCCA, these include male and female pattern alopecia, lichen planopilaris and even unusual cases of chronic cutaneous lupus erythematosus. A biopsy is often necessary to confirm the diagnosis.⁶ The disease begins and remains most severe on the crown or vertex of the scalp gradually expanding in a centrifugal fashion. Symptoms may be mild or absent. Most patients note only mild, episodic pruritus or tenderness of involved areas. Pustules and crusting may also be found in the minority of patients who suffer from rapidly progressive

disease or bacterial superinfection (follicular decalvans).^{7,8} For relatively non-inflammatory disease, a combination of a long-acting oral tetracycline (e.g. doxycycline or minocycline) plus a potent topical corticosteroid (e.g. clobetasol or fluocinonide) is usually sufficient to halt progression. Prolonged treatment for years is usually required for this chronic disease. For highly inflammatory cases (“folliculitis decalvans”), an initial 10-week regimen of oral rifampin and clindamycin (both at 300 mg twice daily) was very effective in a non-blinded, non-controlled study.⁸ Maintenance therapy (such as oral doxycycline plus topical clobetasol) should follow.

Conclusion

Whenever we come across patient presenting with progressive pattern of alopecia affecting the crown we should consider all conditions that can mimic CCCA. Although most often cases of CCCA found among black women of African descent, this case was not in this category. The patient was treated by oral tetracycline plus topical clobetasol and there was a noticeable response to treatment.

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