The diagnosis and management of alopecia

Motswaledi MH, MBChB, MMED, FC Derm(SA), Head of Department Mathekga K, MBChB, FC Derm(SA), Specialist Department of Dermatology, University of Limpopo (Medunsa Campus) Correspondence to: Mojakgomo Motswaledi, e-mail: motswaledi1@webmail.co.za Keywords: alopecia, skin disorder, diagnosis, management

Abstract

Alopecia refers to loss of hair from the scalp or other hair-bearing areas of the body. Alopecia has a significant impact on quality of life. It is commonly associated with loss of self-esteem and emotional stress. It can be non-scarring (noncicatricial) and reversible, or scarring (cicatricial) and often permanent.

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Introduction

The hair follicle is a well-organised multicellular, cylindrical structure which produces the hair shaft. The hair follicle and its associated sebaceous gland form a pilosebaceous unit. The region of insertion of the arrector pili muscle is called the bulge. It is important in hair follicle regeneration. If this site is injured, hair regrowth will not occur.¹

Hair follicles undergo a repetitive sequence of growth and rest, known as the hair cycle, which comprises three phases, the anagen, catagen and telogen phases.²

Anagen is the period of active hair growth. Under normal circumstances, 80-90% of human scalp hairs are in the anagen phase at any one time.² At the end of the anagen phase, epithelial cell division declines and ceases. The hair follicle enters an involutionary phase called the catagen phase. During this phase, the lower part of the follicle involutes by apoptosis. The period between the completion of the follicular regression and beginning of the next anagen phase is called the telogen phase.

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Non-scarring (noncicatricial) alopecia

Alopecia areata

Alopecia areata is a common, autoimmune disease that can affect any hair-bearing area, but usually it is the scalp. The initial lesion is a circumscribed patch of hair loss. More patches may appear on the scalp. These may become confluent and result in diffuse loss of the entire scalp's hair² (Figures 1a and b). Short "exclamation mark" hairs with broad distal ends and thin proximal ends can often be seen, especially at the margins of the alopecia areas. Total loss of scalp hair is called alopecia totalis. Total loss of entire scalp and body hair is called alopecia universalis.³



Figures 1a and b: Alopecia areata

Alopecia areata may occur in association with atopic disease, autoimmune thyroiditis, inflammatory bowel disease and diabetes mellitus.² Alopecia areata is a clinical diagnosis. In active disease, in which alopecia patches continue to expand, a hair-pull test may be positive at the periphery of lesions.³

Treatment of alopecia areata is difficult, but spontaneous remission occurs in 80% of patients who have limited patchy alopecia.⁴ Several treatment modalities have been used with varying results. Various combinations of these therapies can be utilised. These include topical corticosteroids, intralesional corticosteroids and topical minoxidil.²

Systemic corticosteroids, oral cyclosporine and sulphasalazine have been used in recalcitrant cases, but the use of these drugs is limited by their side-effect profile.⁵

Telogen effluvium

Telogen effluvium is a disturbance of the hair cycle that is characterised by the shedding of normal club hairs following premature termination of the anagen phase and the start of the telogen phase of the hair cycle.² It can be precipitated by various forms of stress such as a difficult childbirth, a surgical operation, haemorrhage (including venesection), emotional stress, drugs (e.g. retinoids, beta blockers and anticoagulants), heavy metals, prolonged fever and a sudden reduction in food intake (crash dieting).² Clinically, diffuse shedding of hairs occurs, especially during combing. If shedding of hair is severe or prolonged, obvious diffuse baldness results.² If the precipitating factor is treated, spontaneous regrowth occurs. In some cases, only partial recovery occurs.² Post partum telogen effluvium tends to recur with subsequent pregnancies.

Trichotillomania

Trichotillomania is derived from the Greek words *thrix* (meaning "hair"), *tellerin* (meaning "pulling out") and *mania* (meaning "madness"). This refers to the compulsive practice of plucking hair from the scalp, eyebrows and eyelashes.⁶ It occurs more commonly in children than in adults, and is seen more often in females than males. Usually, affected individuals pluck their scalp hair the most, giving rise to patches of alopecia which often have bizarre shapes and irregular borders. Hairs of varying lengths result.⁶ Plucking is associated with hair shaft fractures. The emerging shafts have fractured ends that tend to feel rough.

Trichotillomania is a manifestation of an obsessivecompulsive disorder, but may be associated with depression, anxiety or psychosis. There is no specific treatment for it. Behavioural modification, psychotherapy and psychopharmacologic medication may be helpful.⁶

Alopecia syphilitica

Alopecia syphilitica is loss of hair on the scalp, eyebrows and beard area that may occur with other cutaneous manifestations of secondary syphilis. The alopecia is irregularly distributed. The result is that the scalp looks moth-eaten⁷ (Figure 2). The usual treatment of secondary syphilis with benzathine penicillin and doxycycline reverses the condition.

Traction alopecia

Traction alopecia is a frequent cause of alopecia in women. It results from the tension that is caused by procedures that are intended to straighten kinky hair, such as hair weaving and braiding.⁸ Traction alopecia most commonly affects the periphery of the scalp, especially the temples and above the ears.

Scarring (cicatricial) alopecia

Cicatricial alopecia is the generic term that applies to alopecia that accompanies or occurs after the destruction of hair follicles by a disease that affects the follicles primarily or secondarily.²

Primary cicatricial alopecia refers to a rare group of disorders in which the hair follicle is the main target of destructive inflammation. This results in hair loss and scarring. They include central centrifugal cicatricial alopecia (CCCA), frontal fibrosing alopecia, lichen planopilaris (LPP) and many others.⁹ In the case of secondary cicatricial alopecia, the hair follicle is a "bystander" during more global damage to the scalp. Hair loss is a secondary event.⁹

Central centrifugal cicatricial alopecia

CCCA is a progressive vertex-centred alopecia that usually occurs in women of African descent.¹⁰ The cause is unknown, but it is thought that in the case of CCCA, the use of hot combs or chemical relaxers results in follicular damage. Most women with CCCA use, or have used, chemical hair relaxers for styling purposes. Clinically, scarring alopecia occurs. It starts as a small patch at the vertex and then enlarges and spreads centrifugally (Figure 3). Follicular destruction results, together with scarring and loss of follicular ostia. The lateral and posterior aspects of the scalp are spared.

Inflammatory changes such as erythema and scaling are absent. The affected scalp is soft and pliable with obvious loss of follicular ostia. Treatment of CCCA is challenging. The goal of treatment is to stop progression of the disease, rather than regrowth of hair. This is achieved by administering anti-inflammatory agents.¹¹ In the early stages of the disease, topical and intralesional corticosteroids may halt disease progression and save a significant amount of hair.¹⁰ Oral antibiotics, hydroxychloroquine and cyclosporine have been used. There is no treatment for the late stages of the disease.

Frontal fibrosing alopecia

Frontal fibrosing alopecia is a form of cicatricial alopecia that is characterised by a band of frontal or frontoparietal hair recession and a marked decrease in loss of eyebrows. This is typically seen in postmenopausal women.¹² It is characterised by destruction of the hair follicles by a lymphocytic inflammatory infiltrate that is localised around the upper portion of the hair follicle. Clinically, a band of symmetric recession of the frontoparietal hairline manifests and extends to the preauricular areas. There is associated loss of follicular orifices, mild skin atrophy and perifollicular erythema at the scalp margin.¹²

Treatment modalities include systemic steroids and topical minoxidil, as well as finasteride. The response to finasteride suggests some hormonal basis for this disease.¹²

Lichen planopilaris

LPP is a variant of lichen planus. It affects the scalp. It rarely occurs on its own. It is commonly associated with lichen planus elsewhere on the body.13 LPP affects women more than men and usually, Caucasians are more affected than dark-skinned people. The clinical course and pattern of hair loss in LPP may be insidious or fulminant. The pattern of scalp hair loss is highly variable. Clinically, follicular papules with perifollicular erythema and loss of hair result. Patients with indolent scalp disease may be asymptomatic, but pruritus and tenderness are often present. Affected hairs are easily pulled out with minimal traction.⁶ Progressive scarring occurs. There is an absence of follicular ostia in the areas that are affected by the alopecia. If treated early, LPP responds to oral and intralesional corticosteroids. Patients may respond to topical corticosteroids. The disease waxes, wanes and progresses slowly. In some instances, it can be aggressive and result in extensive hair loss within a few months.



Figure 2: Alopecia syphilitica. Note the moth-eaten appearance of the scalp.



Figure 3: Central centrifugal cicatricial alopecia. Note the scarred areas.

Discoid lupus erythematosus

Discoid lupus erythematosus is a form of lupus erythematosus. It affects the skin, usually the face and the scalp. A typical lesion of discoid lupus erythematosus is a well circumscribed, depigmented lesion with atrophy, scaling, telangiectasia and scarring. There is often hyperpigmentation around the lesion (Figure 4). There is hair loss, follicular plugging and permanent scarring on the scalp.¹⁴



Figure 4: Discoid lupus erythematosus. Note the hyperpigmented border around the lesions.

Although discoid lupus erythematosus is a clinical diagnosis, biopsy is often necessary. Histological features are quite diagnostic. Laboratory tests such as antinuclear antibody (ANA) are also helpful. ANA is positive in approximately 35% of cases.¹⁴ Topical corticosteroids and chloroquine are useful in the treatment of discoid lupus erythematosus, but the associated scarring alopecia is often permanent.

Other causes of secondary cicatricial alopecia are thermal burns on the scalp, radiation dermatitis, scleroderma, sarcoidosis, bacterial and fungal infections of the scalp, and neoplastic infiltrations on the scalp.

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