CICATRICIAL ALOPECIA

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Introduction

In layman's terms alopecia = baldness. Though it is not a life-threatening disease, it can have a huge psychological impact on both males and females.

For centuries scalp hair has been used to differentiate females from males (though not so much these days). Various cultures are defined by their hair styles; the Rastas, Hare Krishnas, the Sikhs and Punks. Though a bald scalp is used as a trade mark for Skinheads, majority of patients are very distressed by alopecia. Alopecia can be divided into non-scarring and scarring. The non-scarring types are more common and if detected early, are usually reversible. Scarring (cicatricial) alopecia is more often permanent and little can be done to reverse it. In cicatricial alopecia there is permanent destruction of the pilosebaceous units and they are replaced by fibrous tissue. There is loss of the hair ostia on the skin surface and with time the scalp may appear smooth and shiny.

Cicatricial alopecia is divided into either primary cicatricial alopecia (PCA) or secondary cicatricial alopecia (SCA). In primary cicatricial alopecia, the hair follicle is the target of inflammatory destruction, with little effect on other dermal components. In secondary cicatricial alopecia, the hair follicle is an "innocent bystander" and is destroyed indirectly.

Primary cicatricial alopecia

Table1: Common causes of Primary cicatricial alopecia

Lymphocytic
Lichen planopilaris Frontal fibrosing alopecia Graham- Little syndrome
Pseudopelade of Brocq
Central centrifugal cicatricial alopecia
Discoid lupus erythematosus
Alopecia mucinosa
Keratosis follicilitis spinulosa decalvans
Neutrophilic
Folliculitis decalvans Tufted folliculitis
Dissecting cellulitis/folliculitis
Mixed
Acne keloidalis nuchae
Erosive pustular dermatosis
Non-specific (end stage)

1. Lichen planopilaris

Lichen planopilaris is a disorder of largely unknown cause, of which there are three main subtypes: Lichen planopilaris, Frontal fibrosing alopecia and Graham-Little syndrome.

Lichen planopilaris occurs at any age, although it is most common in females above the age of 30years. The scalp is usually the only site affected, though skin elsewhere i.e. mouth and genitalia, may be affected. Recent scalp lesions may present as papules, perifollicular scales (hyperkeratosis) and erythema around affected follicles. Affected hairs can easily be pulled with minimal traction (Pull test). The hair that is extracted may show a long glassy ensheathed root tip, the sign of a growing hair, or anagen hair. As the disease progresses there is scarring and marked absence of follicular ostia in areas of alopecia. Usually the disease progresses slowly with few bald patches that are well camouflaged by the remaining hairs. In some instances, the disease can be more aggressive and results in extensive hair loss within a few months.

2. Frontal fibrosing alopecia

Frontal fibrosing alopecia is considered as a variant of Lichen planopilaris based on clinical and histopathologic findings. It occurs predominantly in postmenopausal women (no connection to hormonal status), though few cases have been reported in premenopausal women and rarely in men.

This condition presents as a marked recession of the frontal and temporal hairline. There is loss of follicular ostia and the scalp appears pale and shiny in contrast to the forehead. The hairs at the margin of the recession may show perifollicular erythema and scaling. The hairline can move back, anything from few millimetres to several centimetres.

3. Graham-Little syndrome

First described in 1915, by Graham-Little, it is still being argued whether this syndrome is an entity on its own or still part of Lichen planopilaris. Graham-Little syndrome occurs in adults, usually females between 30 to 70 years. It is marked by patchy scarring hair loss of the scalp and loss of hairs in the axillae and pubic region, without clinically evident scarring.

There is also rapid development of Keratosis pilaris. The cicatricial alopecia may precede or occur simultaneously with the Keratosis pilaris.

The ostia of the affected follicles in the scalp are filled by horny plugs. The follicles are progressively lost and eventually an atrophic epidermis covers a sclerotic dermis. The axillae and pubic region do not show atrophy of the skin.

4. Pseudopelade of Brocq

This condition is named after the French dermatologist who was the first to study this condition in the late 1800s. ‘Pelade’ is the word used for alopecia areata in French. Pseusopelade was coined due to its similarity in clinical presentation to alopecia areata.

Pseudopelade of Brocq is an idiopathic, chronic, slowly progressive, patchy cicatricial alopecia that occurs without any evidence of inflammation. It presents as small patches of alopecia in middle-aged adults. The affected scalp surface may resemble “footsteps in the snow”. The scarred areas are usually devoid of any hairs and the skin supple and white. Braun-Falco et al have put forward certain diagnostic criteria for this condition; however it is still not always easy to diagnose.

5. Central centrifugal cicatricial alopecia

This condition, seen mainly in patients of African descent, is commonly blamed on the chronic use of physically and chemically traumatic hair-care products. Like Pseudopelade of Brocq, follicular inflammation is minimal. However few patients complain of intense pruritus, tenderness or even a tingling sensation in the affected areas.

It usually starts in the midline, affecting the central area of the scalp (vertex) and gradually moves in a symmetric centrifugal fashion over many years. The affected area becomes shiny and soft to touch with time.

6. Discoid lupus erythematosus (DLE)

DLE is part of Lupus erythematosus, which is an autoimmune disease. DLE affects the skin and the lesions on the scalp are the same as elsewhere in the body. In approximately 20% of males and 50% of females, the scalp is involved. It may be focal or widespread, (Fig 1). Patients typically complain of patchy hair loss associated with pruritus, burning and stinging. It often starts as a small bare patch that enlarges, showing erythema and follicular plugging in the centre.

The scarring alopecia that results is due to inflammation of the infundibular region of the hair follicle that contains stem cells. Histology shows hyperkeratosis with follicular plugging, a perivascular and periadnexal lymphoid infiltrate and focal basal vacuolar degeneration.

There is a 5% to 10% chance of developing Systemic lupus erythematosus, which is seen more commonly in patients with widespread cutaneous disease. Unlike other cicatricial alopecia, hair loss in DLE is potentially reversible if aggressive treatment is started early. High potency topical corticosteroids, intradermal steroids and chloroquine can be tried.

7. Folliculitis decalvans and tufted folliculitis

These are chronic scarring folliculitis of the scalp in which Staphylococcus aureus is commonly cultured. In the vast majority of people who develop folliculitis of the scalp, the disease is transient and does not result in scarring. However some people are more unlucky, in that even after a course of antibiotics, the disease tends to come back in the same area and result in permanent destruction of the hair. An abnormal host response to Staphylococcus aureus is postulated, which may be the result of a defect in cell-mediated immunity.

Patients typically present with a complaint of “pimples” and hair loss of the scalp, which may be very painful and pruritic. Perifollicular erythema, pustules, crusting and oozing are usually evident.

Management involves repeated cultures and sensitivity studies of pustules and the administration of appropriate antibiotics. Long term treatment with Cloxacillin, is the initial drug of choice. For those who do not respond, Rifampicin and Clindamycin (or Doxycycline) are given as a combination, for a period of 10 weeks. This can result in sustained disease-free periods.

8. Acne keloidalis nuchae

This entity is common in patients of African ancestry. It can resemble a scar or keloid, but is not considered a bona fide keloid. Patients complain of the presence of pimples affecting the nape of the neck. They are usually multiple brown to red, firm papules and nodules that present individually or as crops. In some they may be pruritic and painful. The mechanism is not clearly understood and may include a combination of ingrowing hair, friction from collars, chronic folliculitis, and seborrhoeic dermatitis. Treatment includes:

A) Avoid shaving posterior scalp
B) Class I or II corticosteroid gel combined with tretinoin gel every night
C) Oral and topical antibiotics (for anti-inflammatory effects or to treat secondary infection)
D) Intralesional steroids to try and shrink the papules and nodules (sometimes it tends to be easier to inject after cryotherapy with liquid nitrogen)
E) Extensive surgical removal and healing by secondary intention may be required in severe cases

8. End-stage or Burnt-out cicatricial alopecia
This entity does not indicate a specific disease but rather the end-point common to several entities. If possible, a repeat biopsy from the active sites of the disease is recommended.

Secondary cicatricial alopecia

Causes of secondary cicatricial alopecia are numerous. The most common ones are listed in table 2. A few of them will be discussed below.

Table 2. Common causes of secondary cicatricial alopecia

Traumatic
Radiation dermatitis
Mechanical trauma
Burns
Traction alopecia
Sclerosing disorders
Morphea
Scleroderma
Lichen sclerosus et atrophicus
Granulomatous
Sarcoidosis
Infectious
Bacterial : Folliculitis, Carbuncle/Furuncle, Syphillis, TB
Fungal : Kerion, Favus, Tinea capitis(rarely scarring)
Viral : Shingles, Varicella
Neoplastic
Cylindroma and other adnexal tumours
Basal cell carcinoma
Squamous cell carcinoma
Alopecia neoplastica

1. Cicatricial alopecia secondary to radiation therapy

High dose radiation therapy used for treating intracranial malignancies can permanently destroy hair follicles, resulting in cicatricial alopecia. It is believed that because most anagen follicles are approximately 4 mm deep in the skin, if the dose of radiation, superficial to a depth of 5mm is kept under 16GY, which is the approximate lethal dose for hair follicles, the incidence of radiation induced cicatricial alopecia could be markedly reduced.

2. Burns

Burns remain a major problem in South Africa, due to the widespread use of paraffin stoves in poor communities. Burns to the scalp, can cause permanent damage to pilosebaceous units. The subsequent healing by fibrosis leads to cicatricial alopecia, (Fig 2).

3. Traction alopecia

In 1907, the first example of traction alopecia was reported in girls and women from Greenland who styled their hair in pony tails.

Traction alopecia is most commonly observed in females of African descent, who wear hairstyles that put continuous traction on hair follicles. This results in gradual thinning and loss of hair in the frontal and temporal areas. Unlike trichotillomania, which is a psychiatric disorder of compulsive hair pulling, traction alopecia is unintentionally induced by the hair styling processes such as braids, weaves, twists, locks and “corn rows”. In the initial stages the hair loss is reversible. However with prolonged traction, the alopecia can become permanent.

4. Sclerosing disorders

Morphea, Scleroderma and Lichen sclerosus et atrophicus can all cause alopecia. The eventual sclerosis and homogenization of the dermis leads to permanent damage of the adnexal structures. This leads to cicatricial alopecia.

5. Tuberculosis of the skin

Lupus vulgaris is one of the most common forms of tuberculosis of the skin. It usually presents as a solitary lesion on the face, however it may affect the scalp, causing fibrosis and scarring alopecia.

6. Tinea capitis

This is a disease caused by superficial fungal infection of the scalp, with a propensity for attacking hair shafts and follicles. It is a superficial dermatophytosis and is also known as ring worm of the scalp, (Fig 3). Tinea capitis is caused by fungi of species of genera Trichophyton and Microsporum. Some common organisms are Trichophyton tonsurans, Trichophyton violaceum and Microsporum canis. It is most common in childhood and uncommon thereafter, due to the fungistatic properties of the fatty acids in sebum.

Clinical presentation may vary from a scaly non-inflamed dermatosis resembling seborrhoeic dermatitis, to an inflammatory disease with scaly erythematous lesions and hair loss. The alopecia may progress to severely inflamed deep abscesses termed Kerion, with the potential of scarring and permanent alopecia.

Early treatment with oral Griseofulvin and a Selenium sulphide shampoo, to reduce viable spore shedding, is the treatment of choice in our department. Other effective oral treatments include: Itraconazole, Ketoconazole and Terbinafine. Due to their high costs they are not routinely used for treatment of tinea capitis.

Investigations

The first step in the management of a patient with suspected cicatricial alopecia is the selection of an optimal biopsy site. The chosen site must be hair-bearing, symptomatic and ideally at the site that yields anagen hair on the Pull test. One or two 4mm punch biopsies which include subcutaneous fat usually suffice. They are sent for haematoxylin and eosin staining and histologic examination.

Conclusion

The diagnosis of cicatricial alopecia can be very distressing to patients. In general the prognosis is poor, however in some instances, like Discoid lupus erythematosus and probably Folliculitis decalvans, the disease can be controlled if diagnosed and treated early.

Some centres offer surgical management, like scalp reduction surgery and hair transplantation, however it may not be suitable for all patients.

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LEGENDS TO FIGURES

1). FIGURE 1……CICATRICIAL ALOPECIA DUE TO DISCOID LUPUS ERYTHEMATOSUS
2). FIGURE 2……CICATRICIAL ALOPECIA DUE TO THERMAL BURNS
3). FIGURE 3……TINEA CAPITIS IN A YOUNG BOY

CPD QUESTIONS:

Please choose true or false for the statements below:

1. Cicatricial alopecia is always treatable.
2. Folliculitis decalvans is a predominantly neutrophilic type of primary cicatricial alopecia.
3. Surgery and healing by secondary intention may be considered in a patient with Acne kelodalis nuchae.
4. Traction alopecia is due to compulsive pulling of hair.
5. Tinea capitis is more common in adults.

ANSWERS:

1. False
2. True
3. True
4. False
5. False

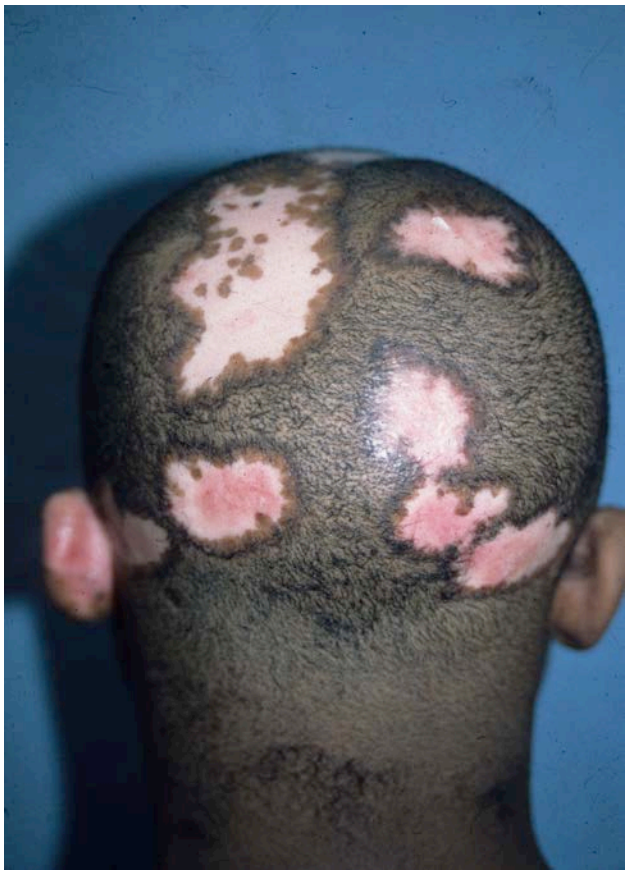

FIG 1

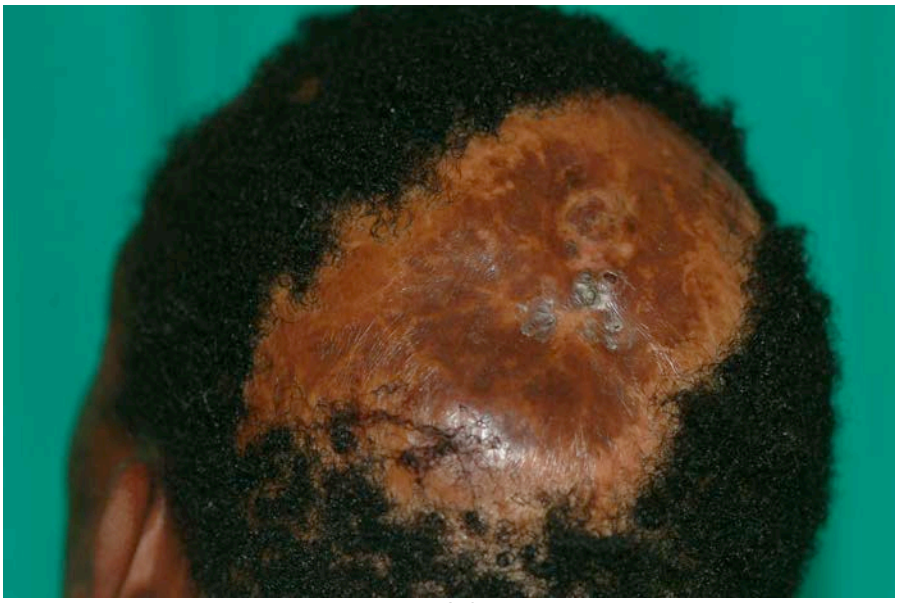

FIG 2

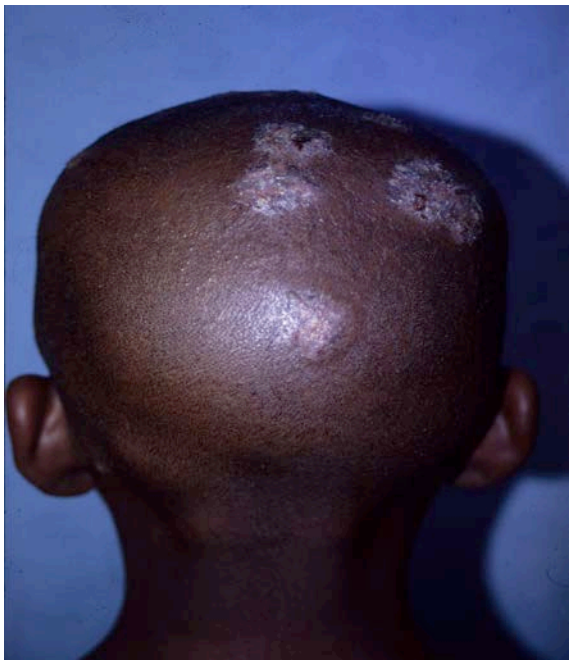

FIG 3