Alopecia Areata: An Update

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ABSTRACT

Alopecia areata (AA) is a very common autoimmune disease that leads to unpredictable, relapsing patchy hair loss. Its chronic pathophysiology is still not fully understood. Hair follicles are not destroyed permanently due to which the potential for regrowth of hair is retained for many years, and is possibly lifelong. Clinical presentation varies from small alopecia patches most commonly on the scalp to full body involvement. Characteristic “swarm of bees” appearance on histopathology is confirmatory in acute cases. A variety of therapeutic options are available, but search for new modalities continues as there is a high relapse rate and a number of side effects associated with the available treatment options.

Keywords: Alopecia areata, Hair loss, Nonscarring alopecia.


INTRODUCTION

Most of the dermatological conditions are underestimated as a simple cosmetic concern. Their effect on patient’s quality of life and self-esteem is not taken into consideration. Alopecia areata is one such disease. Though it is a very common autoimmune disease leading to unpredictable, relapsing hair loss, its pathophysiology is still poorly understood. Owing to incomplete knowledge about how the disease specifically affects only pigmented hair follicles, no curative therapy has been established.

EPIDEMIOLOGY

Alopecia areata is one of the most common causes of hair loss due to inflammation, with a worldwide prevalence of 0.1 to 0.2%. It affects children as well as adults, and hair of all colors. There is generally no sex predilection, but men are more frequently affected than women. The disorder is more severe in males and in those with an onset in early childhood. There is an increase in the overall risk of other autoimmune disorders (16%) in those affected. For example, it is accompanied by lupus erythematosus in 0.6%, vitiligo in 4%, and autoimmune thyroid disease in 28% of patients.

ETIOPATHOGENESIS

In the normal hair growth cycle, hair follicles are continuously being recycled through various phases. During the anagen phase (I through IV), which lasts for around 1 to 8 years for scalp hair, a pigmented hair shaft is generated. This is followed by catagen phase that lasts several weeks, during which there is a rapid, apoptosis-driven organ involution. Melanogenesis is switched off and the hair shaft is transformed into a “club hair.” The hair follicle then enters telogen phase, which is a quiescent phase that lasts for several months. This completes the hair cycle and then hair returns to anagen phase. In patients with AA, the hair cycle is shortened and distorted. There is a characteristic inflammatory infiltrate that attacks only (or at least primarily) the pigment producing hair follicles, predominantly those in the anagen phase. The mixed inflammatory cell infiltrate contains T cells, mast cells, natural killer cells, and dendritic cells.

THEORY OF IMMUNE PRIVILEGE

The hair follicle has a crucial immunologic feature of relative immune privilege that prevents an autoimmune attack on intrafollicularly expressed autoantigens. This relative immune privilege is established mainly by suppression of the surface molecules required for presenting autoantigens to preexisting autoreactive CD8+ T cells. When various costimulatory circumstances occur during anagen (e.g., trauma, infection, stress), the clinical phenotype of AA results.

Some people are genetically predisposed to abnormalities in the microenvironment of the follicle, allowing follicular autoantigens to be presented to preexisting autoreactive CD8+ T cells. When various costimulatory circumstances occur during anagen (e.g., trauma, infection, stress), the clinical phenotype of AA results.

Stress is an important factor in the etiology, as it may precipitate the condition, and the condition may precipitate stress. The most frequent stressful events include the beginning of school or preschool education,
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CLINICAL FEATURES

Alopecia areata manifests as well-circumscribed patches of hair loss on normal-appearing skin, most commonly on the scalp and in the region of the beard. The condition is asymptomatic with a sudden onset. Patient usually acknowledges the alopecia patch during hair grooming. Spontaneous recovery is possible but is not the rule. In severe cases, the condition can involve the full scalp or even full body.

Various clinical forms of AA include

- Patchy/focal AA—hair loss occurs in patches on the scalp or on other parts of the body (e.g., face, abdomen, extremities)
- AA totalis—the loss of all hair on the scalp (including eyebrows and eyelashes)
- AA universalis—the loss of all or almost all body hair
- Alopecia maligna—a generalized long-term loss of hair, resistant to treatment
- Ophiasis or AA marginata—snake-shaped hair loss around the circumference of the head in the temporal, occipital, and frontal areas
- Sisaipho—hair loss beginning from the frontal area and extending backward
- Ophiasis inversus—the inverse pattern of hair loss, which expands from the central to the marginal area of the head
- Diffuse/reticular AA—diffuse or reticular hair loss where no separate bald patches can be distinguished
- Nail involvement is seen most commonly as regular pits on the nail plate

Dermoscopic signs

- Exclamation-mark hairs—hair shaft is thinner at the root, resembling an exclamation mark
- Cadaver hairs—dystrophic hair seen as black dots
- Coudability sign—a kink is seen in the hair shaft near the root
- Growth of/sparing of white hair in alopecia patches

Figs 1A to F: Various clinical presentations of AA: (A) Patchy/focal AA; (B) AA totalis; (C) ophiasis/AA marginata and sisaipho; (D) diffuse/reticular AA; (E) AA universalis; and (F) regular shallow pits in a patient with AA
Association with atopy and other autoimmune diseases further helps in establishing the diagnosis (Table 1).

### HISTOPATHOLOGY

Diagnosis of AA is usually straightforward and does not require a histopathological examination. However, in confusing cases, such as diffuse AA, a skin biopsy is usually diagnostic. In the acute phase, histological examination reveals a characteristic “swarm of bees pattern” of dense, perifollicular lymphocytic infiltrates around anagen hair follicles (Fig. 3). However, in patients with chronic disease, this pattern may be absent.18,19

### TREATMENT

Although diagnosing AA is usually easy, treating it is not. Curative therapy does not exist, and there is a paucity of well-conducted, long-term, controlled trials evaluating the efficacy of therapy and its effect on the quality of life.20 Poor prognostic factors include bald patches persisting for more than 1 year, onset or aggravation of hair loss before puberty, positive family history of AA, ophiasis pattern of involvement, associated nail changes, atopy, autoimmune disorders, and Down syndrome.15

Given the often unsatisfactory results, some clinicians rely on the high rate of spontaneous remission and will recommend a wig if remission does not occur.20 Spontaneous remission occurs in 80% of patients with AA within 1 year, and not all patients require intense therapy. Therefore, watchful observation is also a therapeutic option.21 Limited, but often helpful therapeutic options do exist for both acute and chronic, relapsing AA.

Intralesional corticosteroids (ILSs) are a time-tested modality of treatment for AA.22 Hair regrowth is promoted by their immunosuppressive effect.23 The time from injection to visible hair growth is 2 to 4 weeks and subsequent growth occurs at a constant linear rate.24 Therapy should be stopped if there is no hair growth by 6 months, as such individuals may lack adequate corticosteroid receptors in their scalp tissue. Disadvantages of using ILS include transient atrophy at the injection site25 and pigmentary changes (hypopigmentation > hyperpigmentation). Systemic side effects have also been described. The ILS near the orbit can lead to amaurosis.
Topical and oral corticosteroids are also effective. A high-potency corticosteroid can be used under occlusion for topical therapy. Systemically, betamethasone and prednisolone are commonly used as mini pulse therapies.

Other modalities found effective in the treatment of AA include topical minoxidil, oral and topical psoralsen and ultraviolet A (PUVA), turban PUVA, topical diphenylcyclopropenone as contact-sensitizing immunotherapy, methotrexate, azathioprine, platelet-rich plasma, sulfasalazine, narrow band ultraviolet B, and bexarotene gel. Biologics are ineffective and may cause aggravation during therapy.

REFERENCES


