

Alopecia areata: Clinical presentation, diagnosis, and unusual cases

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ABSTRACT: Alopecia areata (AA) is a nonscarring hair loss disorder with a 2% lifetime risk. Most patients are below 30 years old. Clinical types include patchy AA, AA reticularis, diffuse AA, AA ophiasis, AA sialopho, and perinevoid AA. Besides scalp and body hair, the eyebrows, eyelashes, and nails can be affected. The disorder may be circumscribed, total (scalp hair loss), and universal (loss of all hairs). Atopy, autoimmune thyroid disease, and vitiligo are more commonly associated. The course of the disease is unpredictable. However, early, long-lasting, and severe cases have a less favorable prognosis. The clinical diagnosis is made by the aspect of hairless patches with a normal skin and preserved follicular ostia. Exclamations mark hairs and a positive pull test signal activity. Dermoscopy may reveal yellow dots. White hairs may be spared; initial regrowth may also be nonpigmented. The differential diagnosis includes trichotillomania, scarring alopecia, and other nonscarring hair loss disorders such as tinea capitis and syphilis.

KEYWORDS: alopecia areata clinical presentation, alopecia areata classification, alopecia areata diagnosis

Epidemiology

Alopecia areata (AA) is not uncommon. The prevalence has been reported at 0.1%, the lifetime risk at about 2%. The disorder affects children, men, and women of all hair colors (1). Most patients are younger, although the disorder is uncommon below the age of 3. The highest prevalence is seen between the second and fourth decades of life. Up to 66% of patients are below 30 years old, while only 20% are older than 40.

Clinical presentation

The typical patient with AA presents with one or a few nonscarred focal hairless patches of round or

oval, sharply demarcated shape (FIG. 1). The location and size of the patches is absolutely random.

Sometimes, the patient can tell when the hair fell out and may show bundles of collected hair to the doctor. But in most cases, the exact disease duration is unclear.

The skin of the affected patches is usually normal and smooth, rarely a slightly pinkish coloration can be observed. A soft, cushion-like infiltration may rarely be felt.

Usually, the patches are symptomless. But occasionally, patients describe some tingling, itching, or dysesthesia, at times preceding the hair loss.

Different clinical types of AA can be observed (Table 1).

Patchy AA is the most common form, occurring in up to 75% of patients.

Alopecia reticularis represents multiple active, stable, or regrowing patches (FIG. 2), which may merge to form a mosaic pattern.

A subset of patients (10–20%) have complete alopecia on the scalp, i.e., *alopecia totalis* (FIG. 3).

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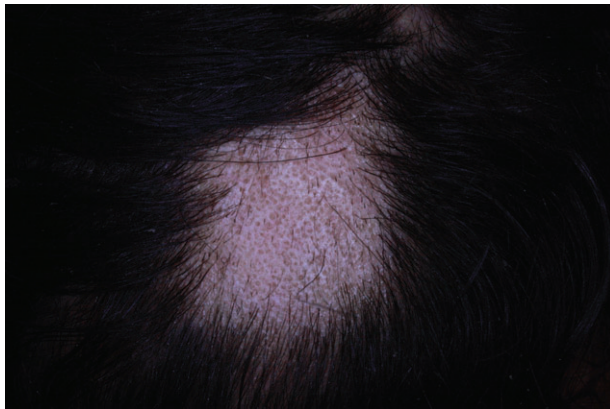


FIG. 1. Patchy AA.

Table 1. Clinical classification of alopecia areata

AA classification by pattern	Patchy type Reticular type Ophiasis type Sisiapho type Diffuse type Perinivoid type ±AA of the beard ±AA of eyelashes ±AA of eyebrows ±AA of body hair ±AA of the nails
AA classification by extent	AA circumscripta AA totalis AA universalis

When all scalp and body hairs are lost, the AA type is called *alopecia universalis*. This also includes loss of eyebrows, eyelashes, nose, and ear hairs.

A special band-like pattern of AA, which winds along the occipital hairline extending toward the temples has been called *ophiasis* (Greek for snake) type (FIG. 4). It usually has a more difficult prognosis and is often treatment recalcitrant.

Even more unusual is a band-like pattern on the frontal hairline (FIG. 5) which should not be confused with frontal fibrosing alopecia.

The opposite of ophiasis type, where hairs are lost centrally and spared at the margins of the scalp, is called *sisiapho* (FIG. 6) (2). It may mimic androgenetic alopecia.

Diffuse AA extends over the whole scalp but usually does not affect all hairs (FIG. 7). A highly positive pull test, the additional presence of patches and dermoscopic signs of AA reveal the diagnosis (FIG. 8). In unclear cases, a biopsy will be



FIG. 2. AA reticularis.

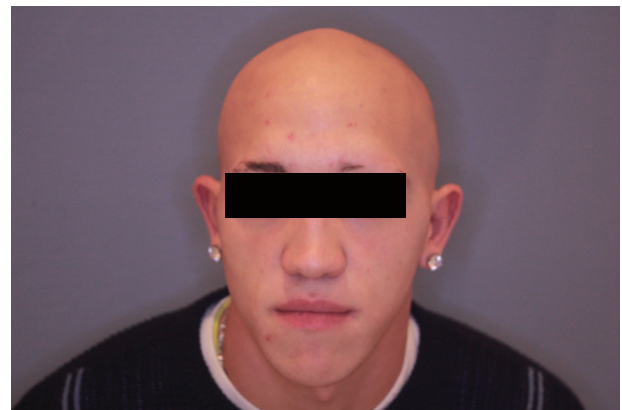


FIG. 3. AA totalis with partial eyebrow involvement.

necessary. Some patients have eyebrow or eyelash alopecia or patches on the beard or body with or without scalp alopecia.

Recently, a variant of acute diffuse and total alopecia has been described. It is characterized by rapid progression and extensive hair loss, but has a favorable prognosis (3,4).

An unusual and rare form with patches around a nevus is called *perinevoid alopecia areata* (5).

Rarely, inflamed areas such as psoriatic lesions are spared from AA.



FIG. 4. AA ophiasis type.



FIG. 5. AA ophiasis on the frontal scalp, patch confirms the diagnosis.

Nail changes can be seen in a portion of patients (10–66%), rarely even without alopecia. Small shallow pits (>30%) up to trachyonychia (sandpaper nails; >10%) are typical (FIG. 9), rarely other changes. A red-spotted lunula and periungual erythema have been postulated as a sign of acute nail involvement.

AA is associated with atopy in 10–22%, which is twice the general prevalence. AA is also associated with (autoimmune) thyroid disease in 8–28% (6), and with vitiligo in 1–4% (FIG. 10). AA is more frequent in patients with Down's syndrome and sickle cell anemia.

Course and prognosis

The course of the disease is practically unpredictable. In most cases, AA takes a chronic but mild course with episodic patches.



FIG. 6. AA sisiapho type.



FIG. 7. AA diffuse type.

In up to 50% of patients with patchy AA, spontaneous regrowth occurs within 12 months, in 66% within 5 years. However, the risk of recurrence is 85%.

Severity of AA at the time of the first consultation is an important prognostic factor (7). In alopecia totalis and universalis, the chance of full recovery is less than 10% (8).

Children with severe and long-lasting AA have the most difficult prognosis.

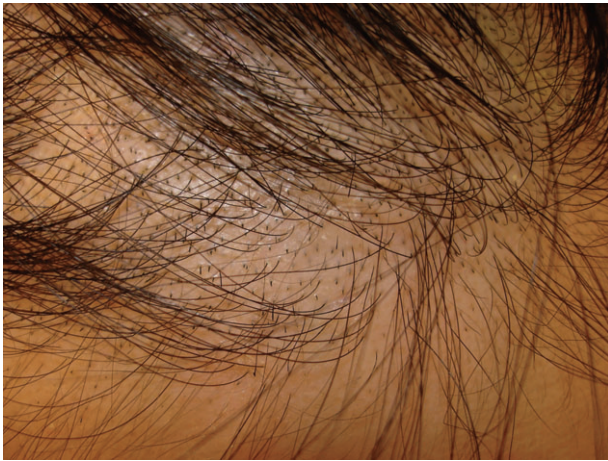


FIG. 8. AA diffuse type with exclamation mark hairs.



FIG. 10. AA sisiapho type with vitiligo.



FIG. 9. AA of the nails.



FIG. 11. AA with white hairs spared.

In patients with AA before puberty, the risk of AA totalis is 50%, in older patients about 25% (8,9).

A positive family history, duration of more than 12 months without regrowth, more than 50% scalp alopecia, nail involvement, atopy, and associated changes are negative prognostic factors.

According to prognosis, AA has been categorized into four types by Ikeda (10).

Based on this, Rook identified three types (11). Type I is associated with atopy, starts in early life, and has a negative prognosis. Type II has no associations, usually starts after the age of 20 and has a favorable prognosis. Type III is associated with autoimmune endocrinopathy, starts later in life, and has an intermediate prognosis. Another study identified onset before 16, female gender, atopy, and autoimmunity as negative prognostic factors (12).

Diagnostic procedures

The diagnosis can usually readily be made by inspection. The presence of circumscribed hairless patches or large alopecic areas with preserved follicular ostia is typical for AA. No laboratory tests are necessary, unless there is a hint for associated thyroid disease.

A detailed personal and family history for AA (extent, start, and course), atopy, thyroid disease, and other autoimmune disorders should be obtained.

Sometimes, long white hairs are preserved from alopecia leading to poliosis (FIG. 11) or sudden whitening of hair in extensive AA (canities subita).

A positive pull test with telogen or dystrophic anagen hairs at the margins of the lesion suggests activity (13). A trichogram usually is not necessary.

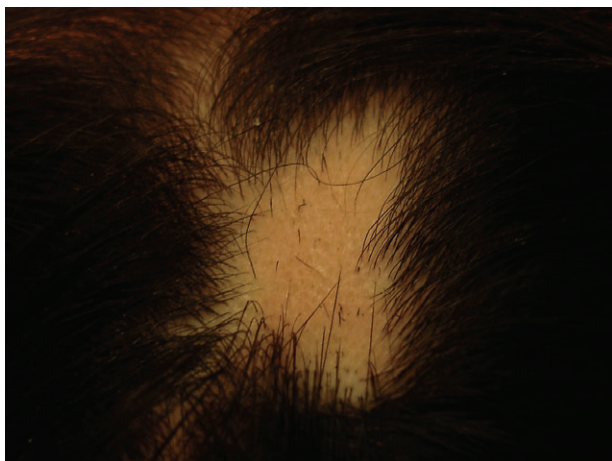


FIG. 12. AA patch with exclamation mark hairs.



FIG. 14. AA diffuse type with yellow dots.



FIG. 13. AA exclamation mark hairs with 20-fold magnification.

Exclamation mark hairs are another sign of acute disease. They represent short trichorrhexis-like broken hairs, which are tapered proximally (FIGS 12 and 13). Structural weakness has been observed (14). Comedo-like cadaver hairs (black dots) may also be present.

Dermoscopy or videodermoscopy is very helpful. It often reveals yellow dots, i.e., keratotic plugs in follicular ostia, although they are not specific for AA (15) (FIG. 14). In a larger study with Asian patients, black dots, yellow dots, and short vellus hairs correlated with the severity of disease. Black dots, tapering hairs, broken hairs, and short vellus hairs correlated with disease activity. For diagnosis, yellow dots and short vellus hairs were the most sensitive markers, and black dots, tapering hairs, and broken hairs were the most specific markers (16).



FIG. 15. AA patch with partial regrowth.

If history and examination are not conclusive, a biopsy will confirm the diagnosis. Histology shows a peribulbar lymphocytic infiltrate.

Short tapered hairs of equal length mostly in the center of the lesion are regrowing hairs (FIG. 15). They are often depigmented initially. These regrowing hairs can be demonstrated by holding a contrasting white (or black) paper card behind them.

A detailed guideline for the assessment of AA has been developed (17). It includes a SALT score, which can help to estimate the extent of disease to guide therapy and assess study results. The quality of life should also be evaluated in such studies.

Table 2. Differential diagnosis of alopecia areata

Congenital hypotrichosis
Ectodermal dysplasias
Loose anagen hair
Alopecia triangularis
Cicatricial alopecias (aplasia cutis, morphea, pseudopelade of Brocq, and others)
Tinea capitis
Trichotillomania
Friction alopecia in babies
Traction alopecia
Pressure alopecia
Toxic alopecia
Alopecia from tick bites
Syphilis

For follow-up, single patches can be measured or photographed to assess the disease course with or without therapy. To categorize the patient, the type of AA should be documented as well as disease activity (stable, active, resolving). Patients should be examined for associated signs such as nail changes and vitiligo.

Differential diagnosis

Many conditions can mimic AA. In children, tinea capitis has to be considered. It usually presents with signs of inflammation. Diffuse AA is sometimes hard to distinguish from telogen effluvium. The presence of dystrophic hairs and the dermoscopic signs are useful clues. The differential diagnosis is listed in Table 2.

The association of AA with androgenetic alopecia (FIG. 16) or trichotillomania (FIG. 17) is not uncommon.

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**FIG. 16.** AA in a patient with androgenetic alopecia.**FIG. 17.** Trichotillomania in a patient who initially had alopecia areata.

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