Clinical profile of alopecia areata in pediatric population
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INTRODUCTION
Alopecia areata (AA) is a common, chronic, inflammatory disease that causes non-scarring hair loss. The term “alopecia areata” was first used by Sauvages, in 1763 (cited by Hebra and Kaposi). The various synonyms for this condition are alopecia circumscriptum, pelade, and area celsi.

Epidemiology
It is a common disease with the lifetime risk of developing this condition reported to be 1.7%. Pediatric AA constitutes approximately 20% of all AA cases. It is rarely reported in infancy. Etiology and Pathogenesis The exact etiology is not known. Many factors have been described in its pathogenesis including genetic, family history, the atopic state, non-specific immune and organ-specific autoimmune reaction, possible emotional stress, infectious agents, and neurological factors. At present, AA is considered to be an autoimmune disease with a definite evidence for the role of T lymphocytes. AA has been found associated with Hashimoto’s thyroiditis, Addison’s disease, and pernicious anemia. There is also evidence of association with other autoimmune disorders such as vitiligo, lichen planus, morphea, Sjogren’s syndrome, myasthenia gravis, and diabetes mellitus.

KEY WORDS: Autoimmune disease, clinical profile, pediatric alopecia areata

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of family history of AA have been reported to be between 10% and 51.6%.[9]

Several studies have shown the association between AA and atopic disease. The incidence of atopy in patients with AA has been reported to be between 11% and 38.2%.[10]

Clinical Features
AA can occur on virtually any hair-bearing area, but it affects the scalp in approximately 90% of cases seen in dermatology clinics.[11]

Initial lesion is characteristically a circumscribed, hairless, and smooth patch with the involved skin appearing normal or slightly reddened. Several such hairless areas may arise and the individual lesion may spread to involve larger area as the disease progresses.

The margins show hairs that are short, tapered proximally with distal widening and are easily breakable, called as “exclamation mark” hairs. The presence of exclamatory hairs at the border and the hair pull test with six or more hairs from the periphery suggests that the patch may be active and progressive.

AA can be classified depending on extent and pattern of hair loss [Table 1].[12]

The most common clinical variant is the patchy AA, in which hair loss occurs in single to multiple patches. Involvement of the entire scalp is termed as alopecia totalis when hair loss over the scalp is incomplete but impending, it is termed as alopecia subtotalis. Involvement of all the body hairs is called alopecia universalis.[12] The reticular form represents multiple, active, stable, or regrowing patches which merge to form a mosaic or reticulate pattern. Ophiasis (snake-like) is a band-like AA along the posterior occipital and temporal margins. Sisaipho, also called as ophiasis inversus, presents with alopecia involving the frontal, temporal, and parietal scalp but spares hair along with the scalp periphery, mimicking androgenetic alopecia.[13]

Sometimes, AA may occur in a linear pattern called as linear AA. A rare form with patches around a nevus is called perinevoid AA.[13] Acute diffuse AA is a newer variant characterized by female preponderance, generalized thinning, rapid progression, tissue eosinophilia, extensive involvement, brief clinical course, and favorable prognosis.[14]

Chelidze and Lipner in a 2018 review observed that nail changes are seen in patients with AA with an average prevalence of 30%.[15]

Various clinical changes that have been seen in the nails are enumerated in Table 2.[15]

The diagnosis is clinical in most cases and investigations are rarely required. Fungal infections causing hair loss can be ruled out by potassium hydroxide (KOH) mount and culture. Syphilis and lupus erythematosus can be ruled out by serology. In certain cases, biopsy may help in ruling out cicatricial alopecia. Histopathology reveals peribulbar and intrabulbar lymphocytic inflammatory infiltrate around anagen follicles, resembling “swarm of bees” with more number of telogen hairs, and decreased anagen to telogen ratio (to 1:1 from a normal 7:1).[16]

Treatment
There is a wide variety of treatment options for AA, but none has shown to alter the course of disease and the response with extensive involvement is low. Leaving AA (which is localized and non-progressive) untreated is a legitimate option. Spontaneous remission occurs in up to 80% of patients with limited patchy hair loss of short duration within 1 year.[17]

Localized disease can be treated with topical agents. Depending on the extent of involvement, oral agents can be resorted to and may also aid topical therapy in severe and rapidly progressive disease.

First-line agents and the mainstay of treatment are the corticosteroids which can be given topically, intralesionally, orally, and parenterally depending on the nature and extent of the disease. Localized disease (patchy AA) responds well to potent and mid-potent topical corticosteroids and intralesional injections of triamcinolone. Adverse effects include telangiectasia, folliculitis, and atrophy.[18] Oral and parenteral steroids can be resorted to if topical therapy fails to show response and as the first-line regimen in generalized disease as pulse therapy. Other topical agents which have shown definite response include minoxidil, topical irritants such as anthralin

Table 1: Classification of alopecia areata

<table>
<thead>
<tr>
<th>Based on extent</th>
<th>Patchy AA</th>
<th>Alopecia totalis, subtotalis</th>
<th>Alopecia universalis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Based on pattern</td>
<td>Reticular</td>
<td>Ophiasis</td>
<td>Sisaipho</td>
</tr>
<tr>
<td>Unusual variants</td>
<td>Perinevoid AA</td>
<td>Acute diffuse</td>
<td>Linear</td>
</tr>
</tbody>
</table>

AA: Alopecia areata

Table 2: Nail changes in alopecia areata

| Scotch plaid effect of pitting in nail plate | Red or mottled lunulae |
| Nail plate discoloration | Thinning and ridging of nail plate |
| Sandpaper-like roughening (trachyonychia) | Nail plate dystrophy |
| Splitting of nail plate | Onycholysis and onychomadesis |
and topical immunotherapy which involves contact sensitizers such as dinitrochlorobenzene and diphenylcyclopropenone. Photochemotherapy and topical prostaglandin analogs are other miscellaneous agents that have been tried to show improvement in cases of AA.\cite{19} Besides steroids, other oral agents that have shown response in some patients include immunomodulators such as levamisole, and sulfasalazine and azathioprine.\cite{20} Cyclosporine and methotrexate have been given with variable success rates, but adverse effect profile renders them as poor choice and should be tried only in recalcitrant and severe cases.\cite{20}

Patients with extensive scalp alopecia may benefit with the use of camouflage techniques such as hairpieces (wigs) and hair additions. Lost eyebrows can be disguised with semi-permanent tattooing. Counseling the patient regarding the spontaneity and unpredictable course of the disease is of utmost importance. It is crucial to evaluate the impact of the disease on the child’s physical and emotional well-being, including issues such as self-confidence, self-image, and acceptance by peers. Some patients might require the intervention of a psychologist and psychiatrist if such issues arise. Parental anxiety, frustration, guilt, and expectations must also be proactively managed to ensure an overall holistic management of the patient.\cite{21}

Aims and Objectives

Aims

This study aims to know the clinical profile of AA in pediatric population.

Objectives

The objectives of the study were as follows:
1. To know the incidence of various clinical types of AA in children
2. To assess the age and sex distribution of AA in children
3. To assess the association of AA with other autoimmune diseases and concomitant infections in children
4. To know the association of positive family history and nail changes in pediatric AA.

MATERIALS AND METHODS

Children <14 years, presenting with AA to the Dermatology Outpatient Department of Rohilkhand Medical College and Hospital, Bareilly, during the period of November 2016–October 2017 were included in the study after proper consent.

A detailed clinical history of each patient was taken. This included patient identification data, duration of disease, history, and family history. History of complaints suggestive of concurrent illnesses was noted.

Thorough clinical examination included local examination of the lesion, associated nail changes, and examination for any concurrent disease. Clinical test performed was hair pull test and clinical sign included the presence of “exclamation mark” hairs. Relevant investigations were done only when there was a doubt in clinical diagnosis. These included KOH mount, and fungal culture, hair microscopy, skin biopsy, and serology for syphilis. The clinical data were entered into Microsoft Excel spreadsheet to assess the clinical profile.

RESULTS

A total of 84 patients who presented to the Dermatology Outpatient Department of Rohilkhand Medical College and Hospital were enrolled to study the clinical profile after proper consent.

Age and Sex Distribution

The highest number of patients belonged to the age group of 6–10 years, constituting 51% of the total patients enrolled followed by 11–14 years constituting 40% and the rest belonged to 1–5 years of age, as shown in Table 3 and Figure 1.

There was a slight female preponderance (55%) with male-to-female ratio being 1:1.2 [Table 3].

The median age of onset was 10 years and the mean age of onset was 9.5 years.

Duration of Disease at Presentation and History of Lesions Corresponding to AA

Majority of the patients (46%) presented during the period of 1–3 months after the onset of the first lesion [Figure 2]. The earliest presentation was 15 days after the onset and the longest was 1 year.

For most patients (81%), this was the first episode of such symptoms [Figure 3].

| Table 3: Age and sex distribution of patients enrolled |
|-----------------|---|---|---|---|
| Age group (years) | 1–5 | 6–10 | 11–14 | Total |
| Number of females | 3 | 25 | 18 | 46 |
| Number of males | 5 | 17 | 16 | 38 |
| Total number of patients | 8 | 42 | 34 | 84 |

Figure 1: Graph depicting age and sex distribution
**Associated Diseases**

Majority of the patients (60%) had no comorbidities [Figure 4]. Atopy was found to be associated in 17.5% of patients and such patients gave a history suggestive of allergic rhinitis, asthma, and/or atopic dermatitis [Figure 4]. Among the autoimmune diseases that were found to be concomitant in three patients, one patient had concurrent vitiligo, another had systemic lupus erythematosus, and one patient showed lesions of hypertrophic lichen planus. Anemia was found to be associated with 6% of patients.

Concurrent infections were seen in 12.5% of patients. The focus of infection in such patients was present in the form of tonsillitis, otitis, dental caries, or lower respiratory tract infections.

**Family History**

A total of 9 patients (11%) gave a positive history suggestive of AA in the family [Figure 5].

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**Figure 2:** Graph depicting the duration of disease at presentation

**Figure 3:** Pie chart depicting previous episodes of lesions corresponding to alopecia areata

**Figure 4:** Graph depicting number of patients with associated comorbidities

**Figure 5:** Pie chart depicting the prevalence of family history in patients of alopecia areata

**Figure 6:** (a and b) Photographs of patients presenting with multiple lesions of patchy alopecia areata

**Figure 7:** Clinical variants of alopecia areata seen
Clinical Types and Number of Lesions

Majority of the patients (60%) had multiple patches of AA at presentation [Figure 6]. Single patch of AA was seen in 29 patients (34.5%) at the time of presentation [Figures 7 and 8].

Among the rare variants, ophiasis was seen in two patients [Figure 9]. AA subtotalis [Figure 10] and AA universalis [Figure 11] were seen in one patient each [Figure 7].

Clinical Signs and Tests

“Exclamation mark” hairs were seen in 17 patients (21%) and hair pull test was positive in 18 patients (22%) [Figures 12 and 13].
patients enrolled followed by 11–14 years constituting 41% and the lowest (10%) belonged to 1–5 years of age. These results are comparable to the study conducted by Netha and Merugu, in 2016, which showed that majority (60%) of the pediatric patients belonged to 6–10 years of age followed by 11–15 years (30%) and least (10%) in 0–5 years of age.\cite{23}
Sex Distribution
There was a slight female preponderance (55%) with male-to-female ratio being 1:1.2.

This is comparable to the study conducted by Vishwanath and Gowda, in 2014, that found the male-to-female ratio to be 1:1.17 in the pediatric population visiting the clinic for AA. [24]

Disease Duration and History
Majority of the patients (46%) presented during the period of 1–3 months after the onset of the first lesion. The earliest presentation was 15 days after the onset and the longest was 1 year. Majority (81%) of the patients had no history of lesions corresponding to AA. The patients presenting with a disease duration up to 6 months comprised 87% of the patients. In a clinical study conducted by Panda et al., in 2014, 72% of patients presented with a disease duration of up to 6 months. [25]

Diseases Associated with AA
Majority of the patients (60%) in our study had no comorbidities. In literature, atopy has been frequently associated with atopy. In our study, atopy was found to be associated in 17.5% of patients and such patients gave a history suggestive of allergic rhinitis, asthma, and/or atopic dermatitis. In a study conducted by Huang et al., in 2013, a high association (38%) of atopy in AA patients was observed. [8]

AA is now very well recognized to be an autoimmune disease. In our study, the autoimmune diseases were found to be comitant in three patients – one patient had concurrent vitiligo, another had systemic lupus erythematosus, and one patient showed lesions of hypertrophic lichen planus. Anemia was found to be associated with 6% of patients. This is comparable to the study by Thomas and Kadyan which observed the association of AA with other autoimmune diseases. [26] The study also observed anemia in 11% of the patients. Concurrent infections were seen in 12.5% of patients. The focus of infection in such patients was present in the form of tonsillitis, otitis, dental caries, or lower respiratory tract infections.

Family History
In our study, 11% of patients had a positive family history of AA. The same incidence of positive family history was observed by Xiao et al., in China. [27]

Clinical Types of AA
Most patients presented with patchy AA and among them most had multiple lesions. Ophiasis was seen in patients. This is comparable to the study conducted by Nageswaramma et al., in 2017, which found 60% of the pediatric patients presenting with AA of the patchy type. [22]

Clinical Signs and Tests
Among the clinical signs, “Exclamation mark” hairs were seen in 17 patients (21%) and hair pull test was positive in 18 patients (22%) in our study. The presence of “exclamation hairs” in AA has shown a variable incidence ranging from 12% to 71% as reported in a review by Waśkiel et al., in 2018. [28]

Associated Nail Changes
Nail changes were observed in 14% of our patients and the most common abnormality was superficial pitting of the nail plate which was seen in 7 patients (9%). Punctate leukonychia was seen in 4 patients (5%). However, majority of the cases (86%) did not show any associated nail abnormalities. Tan et al. conducted a study evaluating the clinical profile of AA in Asians and found association with nail abnormalities to be 10.5%. [29] Besides pitting, their study also observed other nail changes associated with AA such as trachyonychia and longitudinal ridging. Goh et al., in 2006, studied the clinical profile of AA patients and reported 22% association with nail changes. [30]

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