

Case Report

Alopecia Universalis in a Girl with Down's Syndrome

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Abstract

Observation: Alopecia areata, an autoimmune disease of hair follicles is often associated with Down's syndrome. Alopecia universalis is complete loss of scalp and body hair and response to treatment is poor. We report a case of alopecia universalis in a girl with Down's syndrome showing excellent response to oral corticosteroid.

Introduction

Alopecia areata (AA) is an autoimmune disorder of potentially great cosmetic concern accounting for at least 2% of patients attending dermatology clinic [1]. Its association with other autoimmune diseases like thyroid disorders, vitiligo and pernicious anemia is well established. There is up to 8.8% increased frequency of AA in patients with Down's syndrome.

A patient of AA usually presents with patchy loss of hair of scalp, beard or eyebrows. Alopecia universalis with all the body hairs lost from body is rare.

Treatment of severe AA possesses a great therapeutic challenge. Systemic corticosteroids are one of the most commonly used therapies [1].

Here we report a case of Down's syndrome with alopecia universalis treated successfully with oral prednisolone.

Case Report

A nine-year-old female child diagnosed as a case of Down's syndrome was referred to the dermatology outpatient of a tertiary hospital as she had very little hair over her entire body. Her scalp was



Figure 1. Alopecia universalis in a girl with Down syndrome



Figure 2. Down child with alopecia universalis demonstrating clinodactyly

entirely bald and she had very little hair over eyebrows, eyelashes, and the limbs. Whole body of the child was almost barren and she presented a smooth and glossy look (**Figure 1**). She did not have any other cutaneous finding. The patient suffered from frequent episodes of cough and cold and her intelligence was below normal. Her face had a broad forehead with low set ears, depressed nasal bridge and slanted palpebral fissures and she had clinodactyly (**Figure 2**). Her nails were normal; she was 92 cm in height with a body weight of 22 kg. Routine laboratory investigations including thyroid profile were normal. A sample of her heparinised whole blood was sent for chromosomal analysis using PCR based DNA analysis and the report was 47, t21, XX. There were no chromosomal rearrangements. Though her birth history was normal her mother gave a history of stillbirth on two occasions. The child has three sisters all of whom are normal. She was diagnosed as a case of Down's syndrome with alopecia universalis and was put on oral prednisolone, 20 mg on alternate days as a single dose after breakfast. Regular follow up was possible with X-ray chest and blood sugar at the end of 1 month and again after 2 months of treatment.

After 2 months with this regime patient showed remarkable increase of hair on scalp and eyebrows. Occipital region did not show hair growth (**Figure 3**). Oral prednisolone was stopped at the end of this period. She was kept on regular follow up and at the end of 4 months there was no significant



Figure 3. Occipital area of child not showing hair growth even after treatment

hair fall even though she was off corticosteroids (**Figure 4**).

Discussion

The pathogenesis of AA is still unknown. Among the factors under investigation, genetic constitutions as well as non specific immune and organ specific auto-immune reactions have been the main areas of concern. Probably, a T-cell mediated auto-immune mechanism occurring in genetically predisposed individuals cause patchy and at times total loss of body hair. Persons with



Figure 4. Case of alopecia universalis in Down syndrome girl, 4 months after start of treatment

Down's syndrome are predisposed to immunologic deficiency in T-cell function and are vulnerable to develop vitiligo and alopecia areata. There is up to 8.6% increased frequency of AA with *Down's* syndrome suggesting involvement of a gene located on chromosome 21 in determining susceptibility to AA [2]. Furthermore a polymorphism in interleukin-1 (IL-1) receptor antagonist gene may be associated with the severity of AA [3]. In a study by *duVivier* and *Munroe*, sixty cases of AA were found among 1000 cases of *Down's* syndrome while only one case was seen among 1000 subnormal control [2]. Similarly, *Tan* et al. in their study of 219 Asians with AA in Singapore found 1.4% with *Down* syndrome [4].

Our case was predisposed to recurrent attacks of cough and cold but she was not atopic and did not have any family history of atopy. There seem to be limited data on association of alopecia universalis with *Down* syndrome. Our patient did not have any ocular complication and was also non-diabetic. She was euthyroid.

Skin changes in *Down's* syndrome are quite frequent and include hyperkeratotic plaques, single palmar crease, xerosis, syringoma, milia, elastosis perforans serpiginosa, idiopathic calcinosis cutis etc [5]. Though there is often a family history in AA (10-42%), no other family members were affected in our case.

The association of alopecia areata and *Down* syndrome may be explained by the chromosome 21 genes. The *Down* syndrome region of chromosome 21 has the MX1 gene that encodes interferon-induced p78 protein MxA. This protein is strongly expressed in lesional anagen hair bulbs from patients with alopecia areata. In a case-controlled study, the MX1 (+9959) polymorphism was significantly associated with alopecia areata, with an increased risk for early onset disease [6].

Sethuraman et al. in 2006, reported a case of *Down* syndrome who presented with progressive and patchy hair loss [7]. In contrast, our case presented with alopecia universalis since birth. Also, our case did not have any other cutaneous manifestations, which are usually reported in *Down* syndrome.

Treatment of severe alopecia areata is a great therapeutic challenge. Systemic corticosteroids are one of the most commonly used treatment modality in extensive alopecia areata. Pulse corticosteroid therapy was introduced by *Burton* and *Shuster* in 1975 [8]. However, experience with oral prednisolone pulse therapy in extensive AA is still limited. Few cases have been tried with betamethasone oral mini pulse therapy, along with minoxidil 2% lotion with satisfactory results [7]. Our patient responded very well to 20 mg prednisolone on alternate days and showed a cosmetically acceptable regrowth of scalp hair and eyebrows after 2 months of therapy. She is maintaining good hair even on stoppage of oral steroid and showed no significant side effects of oral prednisolone.

We report this case as it is a rare presentation of alopecia universalis in *Down's* syndrome with no family history of AA and satisfactory response to treatment.

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References

1. Madani S, Shapero J. Alopecia areata updates. J Am Acad Dermatol 2000; 42: 549-566. PMID: 10727299
2. du Vivier A, Munro DD. Alopecia areata and Down syndrome. Br Med J 1975; 191-192. PMID: 122906
3. Tarlow JK, Clay FE, Cork MJ. Severity of alopecia areata is associated with a polymorphism in the interleukin-1 receptor antagonist gene. J Invest Dermatol 1994; 103: 387-390. PMID: 8077705
4. Tan E, Tay YK, Goh CL, Chin Glan Y. The pattern and profile of alopecia areata in Singapore- a study of 219 Asians. Int J Dermatol 2002; 41: 748-753. PMID: 12452996
5. Scherbenske JM, Benson PM, Rotchford JP, James WD. Cutaneous and ocular manifestations of Down syndrome. J Am Acad Dermatol 1990; 22: 933-938. PMID: 2159488
6. McDonagh AJ, Tazi-Ahnini R. Epidemiology and genetics of alopecia areata. Clin Exp Dermatol 2002; 27: 405-409. PMID: 12190641

7. Sethuraman G, Malhotra AK, Sharma VK. Alopecia universalis in Down syndrome: Response to therapy. *Indian J Dermatol Venereol Leprol* 2006; 72: 454-455. PMID: 17179625
8. Burton JL, Shuster S. Large doses of glucocorticoids in the treatment of alopecia areata. *Acta Derm Venereol* 1975; 53: 493-496. PMID: 55045
9. Leventhal LJ, Straka PC, Schumacher HR Jr. Jacoud arthropathy and acroosteolysis in KID syndrome. *J Rheumatol* 1989; 16: 1274-1277. PMID: 2810288
10. Coto-Puckett WL, Gilbert-Barness E, Steelman CK, et al. A spectrum of phenotypical expression of Neu-Laxova syndrome: three case reports and a review of the literature. *Fetal Pediatr Pathol* 2010; 29: 108-119. PMID: 20334486