

Alopecia universalis in Down syndrome: Response to therapy

Sir,

Patients with Down syndrome have increased prevalence of common dermatological conditions like folliculitis, atopy and vitiligo, besides having increased incidence of peculiar dermatoses like syringomas, milia-like calcinosis, acanthosis nigricans and elastosis perforans serpiginosa.^[1,2] The frequency of alopecia areata is also reported to be higher in Down syndrome than in the general population ranging up to 8.9%.^[3] We highlight alopecia universalis in an eight-year-old boy with Down syndrome and discuss the probable significance of this association.

An eight-year-old boy presented with gradual loss of hair all over the body that started at the age of four years. Initially, the hair loss was patchy involving the parietal area, but within two to three years there was progressive loss of hair all over the body. He did not have any other cutaneous and/or systemic complaints. He was the eighth sibling, born to non-consanguineous parents following an uneventful pregnancy, when the maternal age was 35 years. Cutaneous examination revealed alopecia universalis and scrotal tongue [Figure 1]. Dental evaluation showed retained teeth. Examination of the other systems did not reveal any abnormality. Karyotype confirmed the diagnosis of Down syndrome. He was treated with topical corticosteroids and minoxidil without any response. Hence he was started on betamethasone oral mini-pulse therapy (3 mg of betamethasone given on two consecutive days, weekly) along with minoxidil 2% lotion. There was significant regrowth of hair within three months of therapy, which was noticed all over the body including the scalp, eyebrows and eyelashes [Figure 2]. He is on regular follow-up.

The onset of alopecia areata in Down syndrome occurs



Figure 1: Alopecia universalis and scrotal tongue



Figure 2: Significant regrowth of hair after treatment

at an early age with a tendency towards extensive or severe disease. The response to standard treatment is also poor.^[1] Schepis *et al* reported six cases of severe alopecia areata in a series of 203 Down syndrome cases, which were refractory to the treatment.^[1] Alopecia areata was seen in 60 of 1000 cases of mongolism (6%), out of which 25 had alopecia totalis/universalis.^[4]

The figures of Down syndrome from the series of alopecia areata patients also show similar disease association. In a clinical study of 392 children of age less than 16 years, with alopecia areata, five (1.3%) had Down syndrome. Of these, four were males and all of them had extensive disease with > 50% area

involvement.^[5] In another report of 219 cases of alopecia areata, three (1.4%) had Down syndrome and in all of them, the onset was before the age of 15 years with extensive involvement.^[6] In a series of 212 cases of childhood alopecia areata from India, only one had Down syndrome.^[7]

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.^[8]

Our case had early onset extensive disease, but a good response was seen with a combination therapy of systemic steroids in the form of oral mini-pulse therapy along with topical minoxidil, which may be tried in such refractory cases.

REFERENCES

1. Schepis C, Barone C, Siragusa M, Pettinato R, Romano C. An updated survey on skin conditions in Down syndrome. *Dermatology* 2002;205:234-8.
2. Munoz-Perez MA, Camacho F. Acanthosis-nigricans: A new cutaneous sign in severe atopic dermatitis and Down syndrome. *J Eur Acad Dermatol Venereol* 2001;15:325-7.
3. Carter DM, Jegasothy BV. Alopecia areata and Down syndrome. *Arch Dermatol* 1976;112:1397-9.
4. Du Vivier A, Munro DD. Alopecia areata, autoimmunity and Down syndrome. *Br Med J* 1975;1:191-2.
5. Tan E, Tay YK, Giam YC. A clinical study of childhood alopecia areata in Singapore. *Pediatr Dermatol* 2002;19:298-301.
6. Tan E, Tay YK, Goh CL, Giam YC. The pattern and profile of alopecia areata in Singapore: A study of 219 Asians. *Int J Dermatol* 2002;41:748-53.
7. Sharma VK, Kumar B, Dawn G. A clinical study of childhood alopecia areata in Chandigarh, India. *Pediatr Dermatol* 1996;13:372-7.
8. McDonagh AJ, Tazi-Ahnini R. Epidemiology and genetics of alopecia areata. *Clin Exp Dermatol* 2002;27:405-9.

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