CASE REPORTS

TOPICAL FLUOCINOLONE ACETONIDE ACETATE OINTMENT IN AUTOSOMAL DOMINANT CONGENITAL HYPOTRICHOSIS

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Hypotrichosis present at birth occurring as an isolated defect in three members of an Afghan family and transmitted as an autosomal dominant trait, responded to topical applications of fluocinolone acetonide acetate ointment. The patients were a 9-year-old boy, his 7-year-old sister and their 30-year-old mother who since birth, had sparse, light-coloured and thin hairs on their scalp which would not grow longer than I cm in length. Microscopic examination revealed the hairs to be thin and fragile, but of a uniform thickness. Some of the hairs showed secondary trichorrhexis. Following topical applications of 0.1% fluocinolone ointment, the hairs became coarser and increased in length from an average of 0.7 cm before treatment to 6.2 and 7.1 cm in the two children after 10 months. The mother who started the treatment later also showed improvement.

Key Words: Hypotrichosis, Hereditary, Congenital, Autosomal dominant, Treatment, Corticosteroids

Introduction

Hereditary hypotrichosis is usually seen in association with other abnormalities of the skin, nails, teeth and bones.[1-5] These include anhidrosis, onychodystrophy, keratosis pilaris, sparse conical teeth, mental deficiency, dwarfism etc. Rarely, it occurs as an isolated defect[2,3] when it is called hereditary hypotrichosis simplex (HHS), and still less commonly, it may be limited to the scalp.[3] Only a few families having HHS have been described in the past, the mode of inheritance in these families being autosomal dominant in most of the cases,[3-5] and autosomal recessive in a few.[1,5]

The treatment of this disease is far from satisfactory. Recently, we treated three members of a family afflicted with autosomal dominant hereditary hypotrichosis simplex with topical fluocinolone acetonide acetate ointment which led to a significant increase in the length and thickness of the hair, and a cosiderable cosmetic improvement.

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Case Reports

In July 1993, a 9-year-old Afghan boy and his 7-year-old sister were brought to us for consultation. Both of them had sparse, short and light-coloured hairs (Fig.1a) on their scalp which would not grow beyond a length of 1 cm or so. This abnormality was present in their 30-year-old mother as well, but the eldest brother 11 years in age and the father were normal. A detailed family history revealed that several other members of the mother's family had also been afflicted (Fig.2) while the family members on the paternal side were normal. There was no history of consanguinity. The mode of transmission seemed to be autosomal dominant.

The abnormality in each of these cases was manifest at birth and remained unchanged ever since in spite of treatment with some indigenous local applications for 3-4 years and a one-month spell of systemic corticosteroids. At birth the child would be noticed to have very fine, light-coloured short hairs sparsely growing on the scalp, while the eyebrows and eyelashes would be normal.

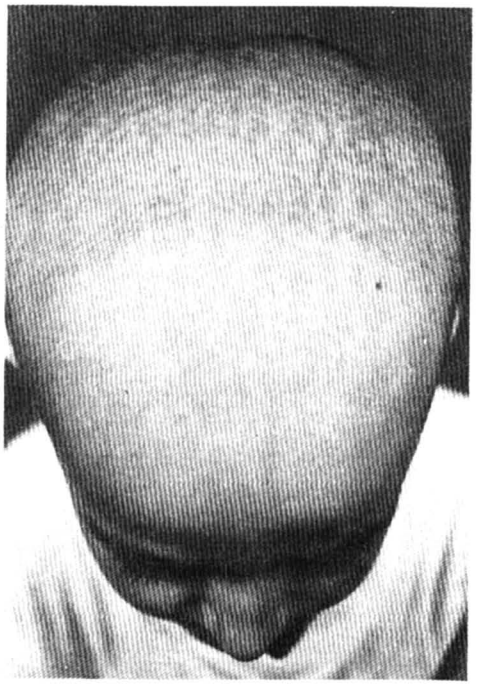

Fig. 1a. Scalp hair before starting the treatment.

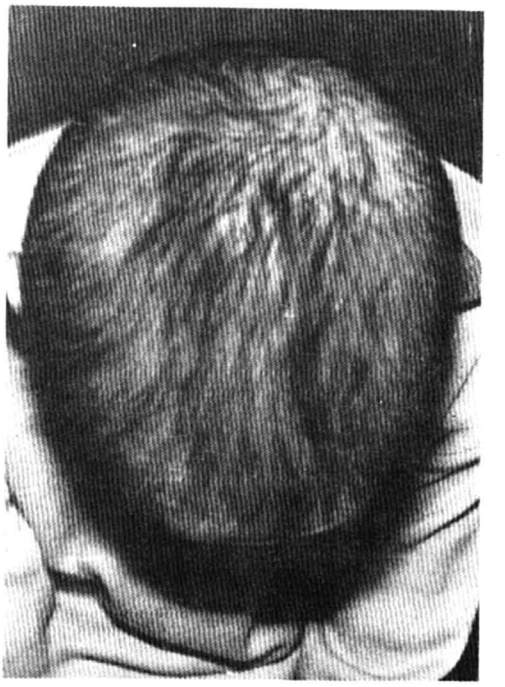

Fig. 1b. The same patient (Fig. 1a) 4 months after starting the treatment.

The hairs would grow to a maximal length of a cm or so and then each hair would fall off spontaneously to be replaced by a new hair which would follow the same course. The milestones in the children had been normal. There was no other structural or functional abnormality in any of the three patients except that the teeth in both the children were slightly yellowish in colour but normal in shape and size. The bones, the nails and the sweat gland function were normal, and all the three patients were physically active and intelligent. Follicular openings on the scalp especially in the occipital region were somewhat prominent, but there was no palmo-plantar keratoderma or any other cutaneous abnormality.

The routine laboratory investigations were normal. Ten hairs plucked randomly from different regions of the scalp from each of the patients revealed the following: It was easy to pluck the hairs but some hairs in each patient had apparently broken off because the root in at least 6 hairs was missing. The average length of the plucked hairs was 0.7 cm. The shaft was thin but uniform in thickness from one end to the other. There was no medulla. The hair roots seen in some of the hairs were normal. The distal end in some hairs were broken off with a brushlike ending suggestive of trichorrhexis nodosa. In other hairs the distal end was either tapering to a fine point or had been cut off neatly.

Both the children were advised to use a routine normal shampoo every day for their hair, avoid any hair oil, but massage 0.1% fluocinolone acetonide acetate ointment once a day all over the scalp and report after two months. On the next visit, the hair growth

seemed to have improved and there were no side effects attributable to the topical corticosteroid. The treatment was therefore continued (Fig.1b). After 8 months of

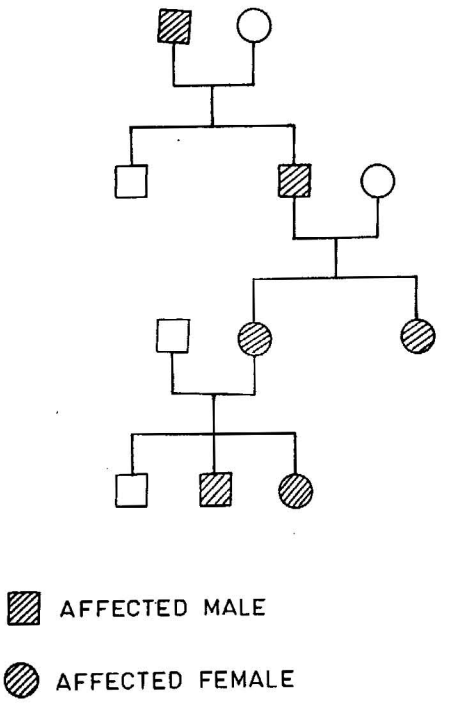

Fig. 2. The family tree showing affected members and autosomal dominant transmission.

treatment, the average length of the hair had increased to 5.8 cm and 2 months later to 6.2 cm in the girl and 7.1 cm in the boy. Seeing this response, their mother also felt encouraged to try the same treatment and started having improvement within 2 months. Microscopic examination of the hairs at this stage revealed the hairs to be thicker, though some of the hairs still had missing roots and brush-like ends.

Discussion

The literature has reference to six large families afflicted with autosomal dominant hereditary hypotrichosis simplex and a few case reports.[3,4] The manifestations however in different families have shown some differences. In the families reported by Petersen in 1915, and Toribio and Quinones in 1974,[3] the affected children had normal hair at birth, showed the first manifestations at the age of 4-5 years and the hair loss was limited to the scalp. In the family reported by Bentley-Phillips and Grace in 1974[4] the hair loss started by 7 years or so but it was not limited to the scalp. In contrast, the hypotrichosis in the families reported by Ledermann (1896), Marie Unna (1925) and Pajtas (1950) was present at birth as seen in our cases but not limited to the scalp.[3] Similarly, the family reported by Jeanselme and Rime (1924) also had generalised hypotrichosis. Our patients, therefore, seem to have a somewhat different pattern.

None of the previous reports have suggested any treatment. The patients are expected to accept their abnormality. A significant and progressive increase in the length of the hairs in both the children within a reasonable period after starting the treatment compared to the natural history of the disease in the mother and the other members of the family would exclude the possibility that the hair growth in the two children was a spontaneous recovery. The fact that the mother had shown no improvement till she also started using the same treatment further suggests that the response was indeed attributable to the topical corticosteroid. We had used the same treatment for an Indian girl also on a previous occasion who had a similar defect. In her case, the hairs had started growing to a length of 10 cm or so, though after attaining that length, the old hairs would fall off and this would be replaced by a regrowth of new hairs which would also grow back to the same length. In the process, the patient could have a reasonable hair style.

The maximal length to which a hair can grow depends upon the duration of the anagen phase in that hair follicle. Hair follicles on the eyebrows and eyelashes have a short anagen phase and thus grow smaller hairs, while hair follicles in the beard region and scalp have long anagen phases and grow very long hairs.[6] We believe that the main defect in our patients was that the scalp hairs had very brief anagen phases.[3] That is why the hairs could not grow beyond a cm or so in length. In addition, the hairs produced by these follicles were structurally weak and fragile and that is why the hairs would come off easily on traction and even break off during the routine hair care procedures. Fragile hairs are more likely to develop secondary trichorrhexis under mechanical/ chemical insults.[7]

The mechanisms which control the hair cycle, the duration of the anagen phase and thus the length of the hair are not well understood. But corticosteroids are known to produce hypertrichosis by converting fine and thin hairs into thicker and longer hairs. Corticosteroids were reported to be useful in a case of monilethrix[8] but have not been used extensively for this purpose, because there is a disproportionate fear about the side effects produced by corticosteroids. If it is realised that the side effects do not occur in all the individuals, one can easily monitor the patient by periodic checks, and most of the side effects are reversible on withdrawal of the treatment, this group of drugs can be usefully exploited to their maximum potential.

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