FAMILIAL WOOLLY HAIR

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A 22-year-old woman and her 12-year-old brother presented with tightly coiled, thin, brittle, dry, lustreless, sparse hairs all over the scalp with history of consanguinity in parents.

Key words: Familial woolly hair, Autosomal recessive, Hair

Familial woolly hair is one of the five variants of the clinical condition collectively called woolly hair syndrome. The other four being hereditary woolly hair, symmetric circumscribed allotrichia, acquired progressive kinking of hair and woolly hair naevus. Familial woolly hair is characterised by the presence of tightly coiled hair over a part of or entire scalp in non negroid races. There is marked reduction in the diameter of hair shaft. It could be poorly pigmented and of increased fragility.

Case Report

A 22-year-old Muslim woman and her 12year-old brother presented with loss of hair from different parts of the scalp. All the hairs over the scalp were found tightly coiled, thin, dry, brittle, poorly pigmented, lustreless and sparse (Fig. 1). These changes were present since infancy and scalp did not show any other abnormality. There was no history of photosensitivity or any major illness. Margins of patches of alopecia were irregular but well defined. Eyebrows, axillary and pubic hairs were very sparse in both the patients. Nails, teeth and genitals were normal. There were no skeletal or ocular abnormalities. The woman gave history



Fig. 1. Profile view of sister and brother showing woolly hair of normal menstrual cycle and the boy had low IQ. The parents and the other three siblings were unaffected. There was history of consanguinity in parents.

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Address correspondence to: Dr. Samir V Shah T/3 Dharnidhar Flat Lad Society Road Vastrapur, Ahmedabad-380015 Routine laboratory tests on blood and urine were normal.

Low power microscopic examination showed neither any beads nor any twisting in the course of the shaft.

Discussion

There could be many conditions with abnormalities of hair shaft. Most of them present to us only because of cosmetic problems like excessive fragility, loss of hair kinking or difficulty in combing. Woolly hair is one of them. Familial woolly hair is probably inherited as an auto-

somal recessive trait.² In our case there was history of consanguinity in the parents but parents and the rest of the siblings were unaffected. This

is reported because of the rarity of the condition in non negroid races.

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