

A Case of Woolly Hair

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Woolly hair is a rare disorder in Korea. We recently examined a seven-year-old girl with familial woolly hair showing abnormal, tightly curled, fine, light brown hairs on the entire scalp since birth. Her hairs tend to be short and easily break. In transmission and scanning electron microscopic studies there were decreased melanin pigment in her hair cortex and disruption of the outer hair cuticle, and damaged cuticle with cuticular splintering.

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Woolly hair is one of the hair shaft coiling and twisting diseases. It is normal in most blacks but is usually abnormal in individuals of Non-Negroid origin.¹ Hairs of woolly hair are very curly, turning in slow spirals, changing direction of curvature along the shaft as with wool fibers.² No cases of woolly hair have been reported in the Korean literature. We, herein, report a case of woolly hair with electron microscopic findings and a patient's pedigree.

REPORT OF A CASE

A 7-year-old Korean girl had abnormal, tightly curled, fine, light brown hairs on the entire scalp with diffuse hair loss. Her hairs were short, curly and hypopigmented and led to an impression of thinning. At best, her uncut hairs were not longer than 10cm (Fig. 1,2). Her parents had noticed her unruly hairs that would not form naturally into locks from the first year of her life. There in no family history of any ectodermal defect.

She had suffered no major physical or emotional trauma and had no history of remarkable drug ad-

ministration or systemic illness. On family history, other members of the patient's family had not noticed woolly hair (Fig. 3), but her father gave the appearance of curly hair and had hair loss. Laboratory examinations including complete blood cell count, urinalysis, and VDRL were within normal limits or negative. A biopsy specimen was obtained from the scalp. Histologic finding of the hair follicle showed wound follicle and no other definite abnormality (Fig. 4). Transmission electron microscopic examination of the patient's hair shaft revealed a decrease of melanin pigment in the hair cortex and disruption of the outer hair cuticle (Fig. 5), compared with normal hair shaft (Fig. 6). Scanning electron microscopy showed damaged cuticles with cuticular splintering (Fig. 7).

DISCUSSION

Human hair can be divided into 5 groups, such as straight, wavy, curly, woolly, and peppercorn in form.² Among them, the subject of woolly hair has been confused in the literature by use of many other descriptive terms including kinky, frizzy, crimped, and steely.

Clinically woolly hair, the acquired progressive kinking of hair, and whisker hair have a similar curling and coiling hair appearance, and resemble

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Fig.1. Abnormal, short tightly curled hairs on the entire scalp with diffuse hair loss.



Fig.2. Fine and whitish brown colored wool-like hairs.

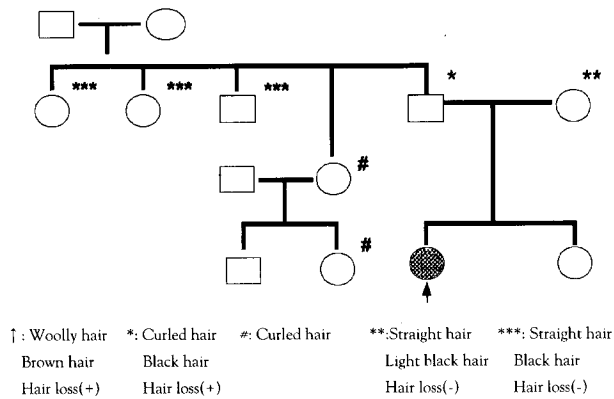


Fig.3. Pedigree of the patient.



Fig.4. Hair follicle of the scalp showed coiled curvatures in light microscopic examination.

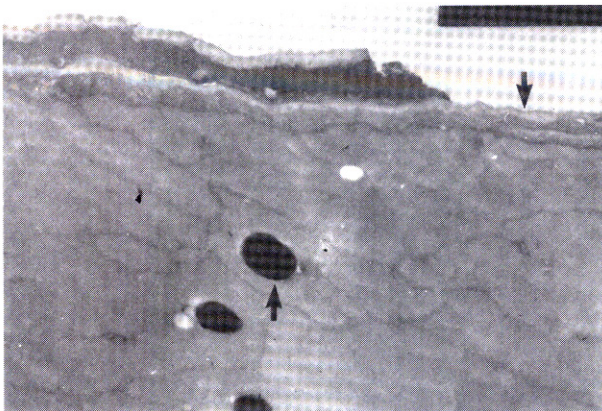


Fig.5. Transmission electron microscopic examination of the patient's anagen hair shaft revealed a decrease of melanin pigment(↑) in the hair cortex and disruption(↓) of the outer hair cuticle, compared with normal hair($\times 10,000$).

each other.³

Woolly hair can be easily diagnosed among these groups through early onset of disease, diffuse scalp involvement, and tight curling in boiling water^{2,4,5,6}. Woolly hair, which does not form locks nor lie flat, consists of tightly coiled spirals with an average curl diameter of approximately 0.5 cm. The curly character of woolly hair is associated with the curvature of the follicle in the dermis⁷

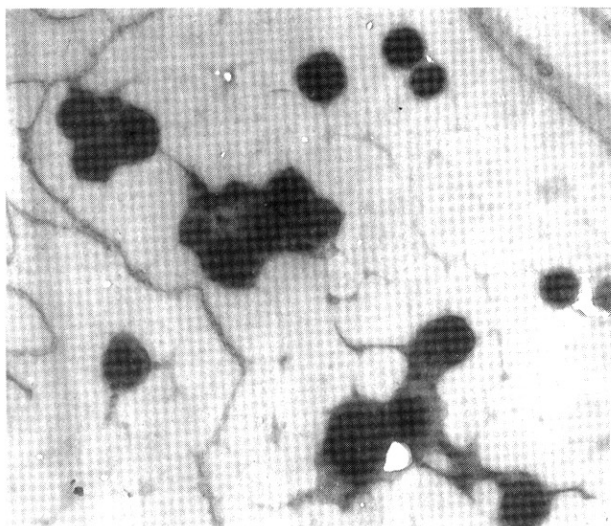


Fig.6. Transmission electron microscopy of normal hair shaft ($\times 10,000$).

and the crosssectioned surface is oval in shape compared to round in straight hair.⁸ The growth rate was probably normal, but the hair does not grow longer than a few centimeters, perhaps because of a shortened growth cycle.⁹

Woolly hair is classified into three distinct types.¹⁰ Diffuse or generalized scalp involvement is typically inherited as an autosomal dominant trait (hereditary woolly hair) or as an autosomal recessive trait (familial woolly hair). The third form of woolly hair appears well demarcated from the surrounding field of normal hair and is often associated with epidermal nevi on the ipsilateral side (woolly hair nevus).¹¹

Our patient had tightly curled, fine, light brown colored hairs on the entire scalp with diffuse hair loss. Her hair abnormality had developed since birth and the hairs failed to grow longer than 10 cm. When these hairs were placed in boiling water, we could not observe a tight curl of regular diameter, as seen in the woolly hair of a black patient.² On the transmission electron microscopic studies, we found decreased melanin pigment in the hair cortex, which is consistent with the patient's light brown color of the hairs. The fragility of patient's hair could be explained by scanning electron microscopic findings which showed damaged cuticle and cuticular splintering.

Her father had black curled hairs with hair loss and her mother had normal hairs except for a light black hair color. Although we could not

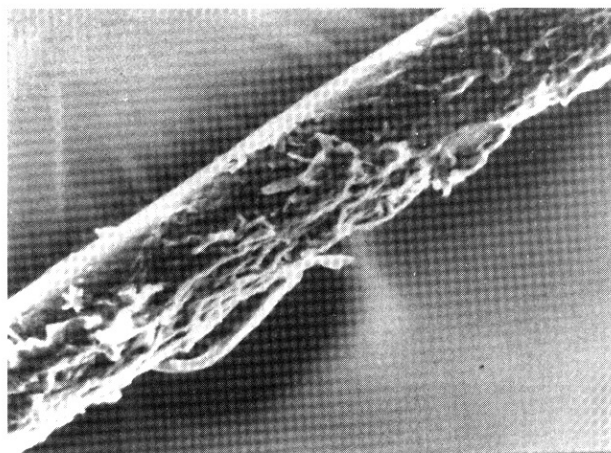


Fig.7. Scanning electron microscopy showed a damaged cuticle with cuticular splintering ($\times 200$).

confirm the pattern of inheritance, we think our case can be classified as familial woolly hair among the three types of woolly hair, because there was no other woolly hair in her family, sporadic occurrence and distinctive clinical and electron microscopic features.¹⁰

In summary, our case revealed the early onset of disease, easily fragile hairs due to cuticular damages, poor hair growth, and change of hair color to light brown due to decrease of melanin pigment in the hair cortex. Because there is no effective treatment for woolly hair up to the present time, we have observed the patient without giving any specific treatment.

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