

A Case of Woolly Hair

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We report a case of woolly hair. Woolly hair is found frequently in most blacks but is unusual in individuals of non-negroid origin. A 12-year-old female patient visited our clinic complaining of a hair abnormality. It had been tightly curled, fine, light brown, short and easily broken since birth. On scanning electron microscopy, many of the hairs showed damaged cuticles with cuticular splintering, and most hair shafts were round to oval on cross sectional examination. (Ann Dermatol 11(3) 161~164, 1999).

Key Words : Woolly hair

Hair shaft coiling and twisting diseases include woolly hair, pili torti, corkscrew hair, Menkes' disease, acquired progressive kinking of the hair, whisker hair, trichonodosis and circle hairs. Some authors classify unruly hair into congenital or acquired woolly hair, pili torti and spun glass hair.

Woolly hair is defined as a condition with an onset at birth or soon afterwards. Maximal severity is usually during childhood, when the average curl diameter is approximately 0.5cm. It is also very difficult to brush or comb the hair.^{1,2} The hair shafts show a variable number of 180° axial twists. To our knowledge, only one case of woolly hair has been reported in the Korean literature.³ We report the second case of woolly hair on scanning electron microscopic findings.

CASE REPORT

A 12-year-old Korean female had had unusual, tightly curled, fine, light brown hairs on the entire scalp since birth. Her hairs were short, easily broken and led to an impression of thinning, but had not grown beyond a length of 12cm (Fig. 1). The eyelashes, eyebrows, fingernails, toenails and teeth

were normal. The pubic and axillary hair had not grown because she was a child. Her growth and development were within normal limits. She had no history of significant drug administration. There was no family or personal history of any abnormalities of the teeth, nails and hairs. On laboratory examination, a complete blood count, urinalysis, a liver function test, and VDRL were within normal limits or negative. Under the light microscopic examination of the hair shaft, there was no specific diagnostic information suggestive of other hair shaft diseases, for example, intussusception of the hair shaft, twisting of the hair shaft on its own axis, alternating constrictions and fusiform enlargements, and split ends, etc.. A skin biopsy specimen was obtained from the vertex area of the scalp and sectioning was done parallel to the skin surface. A hematoxylin-eosin stain showed most hair follicles were round to oval and a few follicles were coiled, but not triangular or reniform (Fig. 2). On scanning electron microscopy, many of the hairs showed damaged cuticles with cuticular splintering, and most hair shafts were round to oval on cross sectional examination (Fig. 3, 4).

DISCUSSION

The diagnosis and classification of hair shaft abnormalities are keyed to easy recognition of the abnormalities under the light microscope. In order to simplify recognition of hair shaft abnormalities by

Received June 13, 1998.

Accepted for publication November 2, 1998.

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Fig. 1. Fine, light brown colored, short and tightly curled hairs on the entire scalp.

Fig. 2. Most hair follicles were round to oval and a few follicles were coiled on cross sectional examination (H&E, $\times 10$).

Fig. 3. On scanning electron microscopy, damaged cuticle with cuticular splintering was seen (SEM, $\times 300$).

light microscopy, Whiting⁴ subdivided them into four structural categories: fractures, irregularities, coiling and twisting, and extraneous matter. Hair shaft coiling and twisting diseases include woolly hair, acquired progressive kinking of the hair, whisker hair, pili torti, corkscrew hair, Menkes' disease, circle hairs and trichonodosis.

Woolly hair is very curly hair that is characteristically unruly and will not form naturally into locks.⁵ Woolly hair is found frequently in most blacks⁵ but is unusual in individuals of non-negroid origin.⁶ Three subgroups of woolly hair have been identified:² *Hereditary dominant woolly hair* is

Fig. 4. Round to oval shaped hair shaft on cross sectional examination (SEM, $\times 600$).

seen at birth or in the first few months of life. Woolly hair affects the scalp hair diffusely, but does not involve hair on other sites. The growth rate is normal, but some patients are unable to grow long hair, probably because of its increased fragility. Associated abnormalities are usually absent, particularly hair color change. *Familial or sporadic recessive woolly hair* appears at birth and is tightly curled and fine, with a white-blond color. The

growth rate is probably normal, but the hair may not grow longer than a few centimeters, perhaps because the growth cycle is shortened. A cuticular weathering can be increased.⁷ *Woolly hair nevus* has a partial scalp involvement, which has a markedly reduced diameter, and appears at or shortly after birth. Some cases are associated with a linear nevus, either verrucous or pigmented, usually on the neck or arm.⁸

Many hair shaft coiling and twisting diseases are included in the differential diagnosis of woolly hair. Acquired progressive kinking of the hair is an entity distinct from woolly hair in its onset at or after puberty, its predominant involvement of the front, temporal and vertex regions of the scalp as well as the supra-auricular and postauricular margins, and a tendency for affected hairs to resemble pubic hair both in texture and color.⁹ Whisker hair is short, curly, dark hair that grows around the ears in young men, aged 18 to 25 years, and is generally regarded as a precursor of androgenetic alopecia.⁹ Some authors have theorized that the acquired progressive kinking of the hair and whisker hair are variants of the same disorder.¹⁰ Our case is different from acquired progressive kinking of the hair and whisker hair because its onset and clinical findings. Pili torti is characterized by twisting of the hair shaft on its own axis. In the classic type, not associated with other disorders, it appears in early childhood when normal scalp hairs are replaced by brittle, spangled hairs, especially in the occipital and temporal regions.¹¹ The eyebrows and eyelashes may also be involved. It usually follows a dominant inheritance pattern, even if recessive and sporadic cases have been reported. On light microscopic examination, the hair shaft twisting is on its own axis. Circle or spiral hairs occur in middle-aged men on the back, abdomen and thighs, as small, dark circles, and represent an unusual form of ingrown hair, but is not associated with follicular abnormalities. The tightly coiled hair lies in a circular tract just below the stratum corneum and is easily extracted.¹² Trichonodosis or knotted hair can compromise a single or double knot of the hair shaft. The common causes of knots in hair are various cosmetic procedures and friction from pillows. Knotting is usually an incidental finding and mainly affects the scalp, but can occur in the pubis and other body hair. Reports of trichonodosis or knotted hair are uncommon, but Dawber¹³ stated that it was not rare

and knotting occurred mostly in short, curly hair of the Negroid and Caucasoid type.

Our patient had had tightly curled, fine, light brown colored hairs on the entire scalp since birth. Her hairs do not grow beyond a length of 12cm. A hair abnormality, except on the scalp, could not be noticed. The fragility of her hairs could be explained by the damaged cuticles with cuticular splintering on scanning electron microscopy. Under the light microscopic examination of the hair shaft, there was no evidence for presuming any specific diagnosis, such as trichorrhexis invaginata, pili torti, monilethrix, trichorrhexis nodosa etc.. On cross sectional examination of a skin biopsy and hairs on scanning electron microscopy, round to oval shaped hair follicles and hairs were seen respectively. These results were consistent with woolly hair, not uncombable hair syndrome. Although we could not clarify the pattern of inheritance, we think our case can be classified as familial or sporadic recessive woolly hair based upon sporadic occurrence, distinctive clinical findings and no family history.

Only one case of Woolly hair has been reported in the Korean literature. We report an additional example of this rare disorder in the orient. No specific treatment administered, because there is no effective treatment for woolly hair.

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