

A Case of Woolly Hair

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Woolly hair is the presence of Negroid hair on the scalp of non-Negroid people. We report a rare case of woolly hair on a 14-year-old Korean girl who presented with tightly-curled, fine and light brown hair which she had had since birth. Light microscopy disclosed flattening of the hair shafts with partial twists at irregular intervals. On scanning electron microscopy, the patient's hair revealed weathered cuticles, and twisted and flattened hair shafts. (*Ann Dermatol (Seoul)* 19(2) 72~74, 2007)

Key Words: Woolly hair

INTRODUCTION

The term "woolly hair" refers to an abnormal variant of fine, tightly-curled hair that often exhibits decreased pigmentation. After the initial report of woolly hair by Gossage¹ in 1907, Hutchinson et al.² in 1974 further classified woolly hair into three variants: autosomal dominant hereditary woolly hair, autosomal recessive familial woolly hair and woolly hair nevus. Since then woolly hair has also been observed in association with several genetic conditions, most notably Noonan syndrome and cardio-faciocutaneous syndrome³.

To our knowledge, there have only been two cases of woolly hair in the Korean dermatologic literature^{4,5}. We herein report a case of woolly hair in a 14-year-old Korean girl.

CASE REPORT

A 14-year-old Korean girl presented with hair loss associated with the presence of fine, short and kinky

hairs on the scalp which she had had since birth. She had easily-broken, slow-growing hair that seldom required cutting. On physical examination, her scalp hair was noted to be light brown, evenly-pigmented, coarse-textured, and extremely curly (Fig. 1). Her scalp, eyebrows, eyelashes, body hair, nails, and skin were normal in appearance. Two-dimensional color echocardiography and electrocardiography were performed, but abnormal findings were not observed. Her medical history was also



Fig. 1. Scalp hairs of the 14-year-old patient, the abnormal hairs were light brown, evenly-pigmented, tightly-curled with diffuse hair loss.

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unremarkable. No family members had any hair or skin disorder, including alopecia. Histopathologic examination of the scalp revealed the presence of intermediate-sized hair follicles without other abnormalities. Light microscopic examination of clipped hairs from her crown and parietal areas disclosed flattening of the shafts with partial twists at irregular intervals. Her hair' index of ovality was 1.5. Scanning electron microscopic examination revealed flattening and irregular twisting of the hair shafts. Weathering of the cuticles was also observed (Fig. 2). These clinical, light and electron microscopic findings were consistent with a diagnosis of woolly hair.

DISCUSSION

Woolly hair is characterized by fair, fine hair. It is found most frequently in Negroes, but is unusual in people of non-Negroid origin. Woolly hair is present at birth and is usually most severe during childhood, when it is often impossible to brush the hair. Adult life brings variable amelioration of the condition.

The growth rate is probably normal, but the hair may not grow longer than a few centimeters, perhaps because the growth cycle is shortened. In the familial group, the hairs on the arms, legs, pubic and axillary regions and eyebrows may be short and pale. There are no associated cutaneous or systemic diseases. Hutchinson et al.² classified woolly hair into three groups: (1) hereditary woolly hair, which is typically inherited as an autosomal dominant trait; (2) familial or sporadic woolly hair, probably inherited as an autosomal recessive trait; and (3) woolly hair nevus, a nonhereditary, partial scalp involvement by woolly hair of a markedly reduced diameter. The former two

types are characterized by generalized scalp involvement. A fourth group was described by Ormerod et al.⁶, who reported a family with short, fine, kinky hair interspersed with normal hair throughout their scalp. We think our case can be classified as sporadic woolly hair, because there were no other woolly hair cases in her family, she had a sporadic occurrence and there were distinctive clinical findings.

Woolly hair has been reported in association with palmoplantar keratoderma and cardiac abnormalities. The triad of woolly hair, palmoplantar keratoderma and right ventricular cardiomyopathy defines Naxos disease, which in at least some cases may be caused by mutations in the desmosomal protein, plakoglobin⁷. Mutation in another desmosomal protein, desmoplakin, has been implicated in the pathogenesis of woolly hair with palmoplantar keratoderma and dilated cardiomyopathy, also called Carvajal syndrome⁸. Both Naxos disease and Carvajal syndrome are inherited in an autosomal recessive fashion, whereas most other cardiomyopathies are inherited through autosomal dominant transmission⁹. Of these conditions, electrocardiographic changes, arrhythmias, heart failure, and even sudden cardiac death have been reported. Therefore the presence of woolly hair with palmoplantar keratoderma should trigger further evaluation for cardiac disease. Skin fragility/woolly hair syndrome¹⁰, usually classified among the epidermolysis bullosa disorder, is an autosomal recessive disorder that, like Carvajal syndrome, exhibits woolly hair, palmoplantar keratoderma and mutation in the desmoplakin gene. Cardiac findings, however, are not characteristic of skin fragility/woolly hair syndrome.

Several other conditions such as curly hair-ankyloblepharon-nail syndrome and tricho-dento-osseous syndrome have been associated with very curly hair that clinically may be difficult to distinguish from true woolly hair.

On light microscopy, the shaft of woolly hair is oval and its diameter is significantly less than in normal hair. Hutchinson et al.² reported that the shaft of woolly hair was smaller than that of normal hair. Wright et al.¹¹ found that the ovality of woolly hair was larger than that of straight hair; the woolly hair' index of ovality was 1.55, and that of normal hair was 1.3. In our case, the woolly hair' index (1.5) of ovality was similar to that of Wright, et al.¹¹.

Inconsistent scanning electron microscopic results have been reported¹²: while Wright et al.¹¹ found

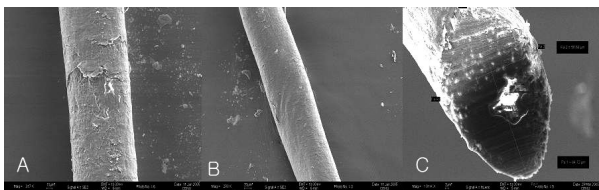


Fig. 2. On scanning electron microscopy the following conditions were observed: (A) weathering of the cuticles ($\times 317$), (B) twisting of the hair shaft ($\times 200$), (C) marked flattening of the hair shaft ($\times 1010$).

no scanning electron microscopic abnormalities, Crosti and Menni¹³ reported absences of the cuticle, trichorrhexis nodosa and occasional nodes. Lopez et al.¹⁴ showed oval and triangular hair shaft sections and longitudinal grooves without abnormalities of the cuticular structure. Goldin et al.¹⁵ showed twisting of the hair shaft and abnormal cuticular formation. The investigators also reported the presence of triangular cross sections and canaliform dystrophy similar to those reported in spunglass hair.

Our patient had a marked flattening of the hair shaft with twisting and cuticular structure abnormalities. Distinguishing woolly hair from curly hair may require the evaluation of hair shafts by either light or electron microscopy. The study of woolly hair with light microscopy revealed ovoid cross sections, 180-degree longitudinal twisting, trichorrhexis nodosa and pili annulati. Normal curly hair in the scalp has been described as oval in shape, without descriptions of twisting or trichorrhexis nodosa. Shallow longitudinal grooves can be seen, and are typical findings in the normal curly hairs of the pubic and axillary region¹⁶.

No specific treatment was administered because there is no effective treatment for woolly hair. In some patients, however, the woolly hair may become darker and less curly with time¹⁷⁻¹⁹. Our patient, however, had shown no significant changes since birth.

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