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An Unusual Case of Korean Brother and Sister with Woolly Hair

Seung Il Choi, M.D., Sung Yul Lee, M.D.

Department of Dermatology, Soonchunhyang University Cheoan Hospital, Soonchunhyang University College of Medicine, Cheonan, Korea

Dear Editor:

Congenital woolly hair is a rare disorder in Asians and Caucasians, resulting from structural defects in the hair shafts. It is characterized by tightly-coiled, fine hair involving the entire, or part of, the scalp of an individual of non-African ethnicity[1]. Most patients exhibit only woolly hair, without any associated disorders. However, Naxos and Carvajal syndromes are conditions that dermatologists should treat with caution because they can be lethal at an early age, due to a predisposition towards cardiac arrythmias[2].

A four year old Korean girl and her younger brother

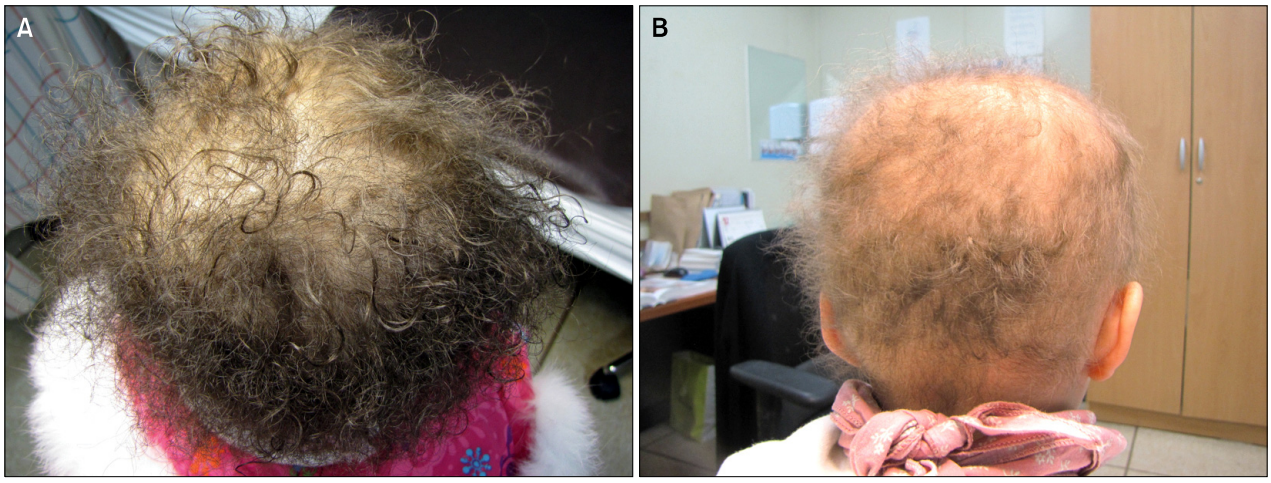

Fig. 1. Very fine, thin and lightly colored curled hairs of siblings were noticed over the whole scalp. Younger brother (B) has more lightly brownish colored hair than his sister (A).

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Corresponding author: Sung Yul Lee, M.D., Department of Dermatology, Soonchunhyang University Hospital, 31 Suncheonhyang 6-gil, Dongnam-gu, Cheonan 330-721, Korea. Tel: 82-41-570-2272, Fax: 82-41-570-2783, E-mail: dermsung@schmc.ac.kr

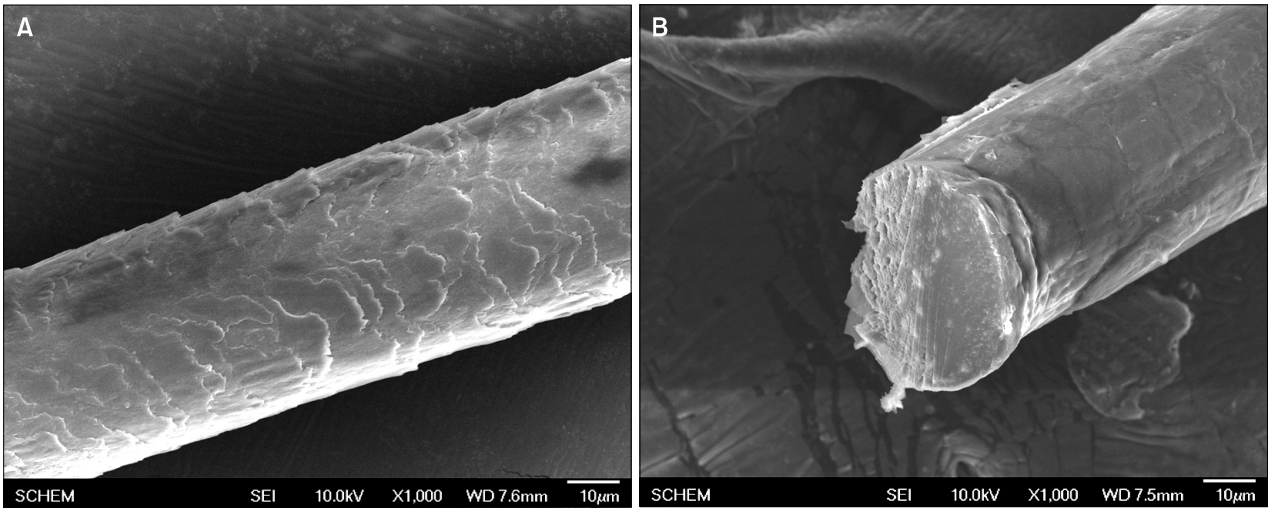

Fig. 2. (A) Hair shaft weathering by mild cuticular damage on scanning electron microscopy (×1,000). (B) Cuticular damage seen by transverse section (×1,000).

presented with woolly hair. Upon closer investigation, tightly curled, easily breakable hairs were evident over the whole scalp. The hairs were very fine, thin and lightly colored (Fig. 1). There were no abnormal conditions associated with their skin, nails, eyelashes, eyebrows, or skeletal and teeth development. They exhibited no mental retardation or cardiomyopathy, and their palms and soles exhibited no hyperkeratosis. The hair of the family, except for the siblings and father side, who had had woolly hair since birth, were normal. Under light microscopy, their hairs exhibited a knotted formation, caliber variation and axis rotation. On scanning electron microscopy, mild cuticular damage was evident as hair shaft weathering (Fig. 2), but hair shaft splintering was not evident, though it has been reported in previous cases.

Based on family history, clinical findings and microscopic examinations, we diagnosed the siblings with hereditary congenital woolly hair that is inherited in an autosomal recessive fashion. From the first report of woolly hair by Hutchinson in 1974, Cairns et al. have further classified woolly hair into three variants: woolly hair nevus, autosomal dominant hereditary woolly hair and autosomal recessive familial woolly hair[3]. One of the three variants, woolly hair nevus, is readily distinguished from the others by a partial involvement of the scalp and a well-circumscribed border that separates the nevus from the otherwise unaffected hair[3]. Hereditary woolly hair presents as various degrees of tight curling in all hairs throughout the scalp. On the other hand, familial woolly hair is characterized by abnormal, tightly-curled, fine, white or blond hair that tends to be short[3].

Mutations in the P2RY5 gene, which is expressed in the inner root sheath, encodes recently have been shown responsible for autosomal recessive woolly hair (ARWH). A mutation in P2RY5 may lead to defects in transmitting signals from oleoyl-L-α-lysophosphatidic acid. Therefore, it may affect proliferation and differentiation of inner root sheath cells[2,4]. Furthermore, it has recently been found that both LPAR6 and lipase H (LIPH) mutations cause ARWH/hypotrichosis. The mechanism of this mutation is based on the regulation of hair follicle development via LPA6 activation by a LIPH-catalyzed lipid mediator[4,5]. It is difficult to distinguish congenital woolly hair and congenital hypotrichosis, and predict woolly hair will later develop hypotrichosis, because there is no clear genotype-phenotype correlation and clinical variation can occur even within members of the same family[4].

In conclusion, this report details the first report of siblings with woolly hair (brother and sister) in Korea.

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