WOOLLY HAIR A RARE PRESENTATION

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ABSTRACT: Woolly hair is a rare congenital abnormality of scalp hair. It is marked by extreme kinkiness of hair in non-Negroid races. Woolly hair are either seen at birth or appear in first months of life. Here we are discussing a 5-year-old non-atopic boy born of a consanguineous marriage with woolly hair over the scalp evident since 3 years of age. There were no delayed milestones, physical or mental retardation or photosensitivity. Examination revealed short, tightly coiled, thin, dry, poorly pigmented, brittle hair over the scalp. The palms and soles were not involved and the nails, teeth and genitalia were normal. His systemic examination including a cardiac assessment was normal. There was no ocular or skeletal involvement. No relevant family history was evident.
KEYWORD: Kinky hair, Naxos disease, Desmoplakin, Carvajal disease, Plakoglobin.

INTRODUCTION: The phenomenon of Woolly hair was first observed and described in Europeon family by Gossage in 1907[1]. Woolly hair is a congenital structural abnormality of scalp hair occurring in Caucasians. It is either sporadic or genetic. Woolly hair is extremely curly, the average diameter of curls goes upto 0.5 cm, lie closely together and usually make the hair difficult to comb, In addition the hair may be more fragile than usual, which is probably due to simultaneous occurrence of trichorhexis nodosa [4]. The hair growth is usually normal but the anagen phase may be truncated with the result that the hair does not grow to be long. The hair shaft may exhibit an elliptical cross section, an axial rotation and a kink. The hair texture resembles sheep's hair whereas the hair color most often shows no unusual characteristics [2].Mohr and Hoffmann compared this of the hair shaft to structure of curly hair in black people [3]. The curled hair of black people lies typically separately while the curls of woolly hair tend to merge [2, 3]] There are mainly three types of woolly hair;

1) The Autosomal dominant woolly hair, also called hereditary woolly hair
2) Autosomal recessive familial woolly hair.
3) Localised woolly hair in form of woolly hair nevus [5]

The various forms of woolly hair may occur in association with other anomalies such as ocular (cataract), Palmopantar hyperkeratosis and heart anomalies.

CASE REPORT: A five year old male child presented with progressive curling of scalp hair since three years of age. He was a full term child of a consanguineous marriage with no known or distant African ancestary. Child had a normal milestones. None of the other family members had similar hair type.

On examination, the hair all over the scalp were found to be short, tightly coiled (curl diameter being ~ .5 cm), thin, dry, brittle, brown and sparse. Underlying scalp was normal. The palms and soles, nails, teeth and genitalia were normal. Nutritional status was normal. There was no

ocular, auditory or skeletal involvement.. There was no associated congenital abnormality. Systemic examination was normal. Echocardiography did not reveal any evidence of cardiomyopathy.



Fig. 1 & 2: Short and tightly curved hair, covering entire scalp

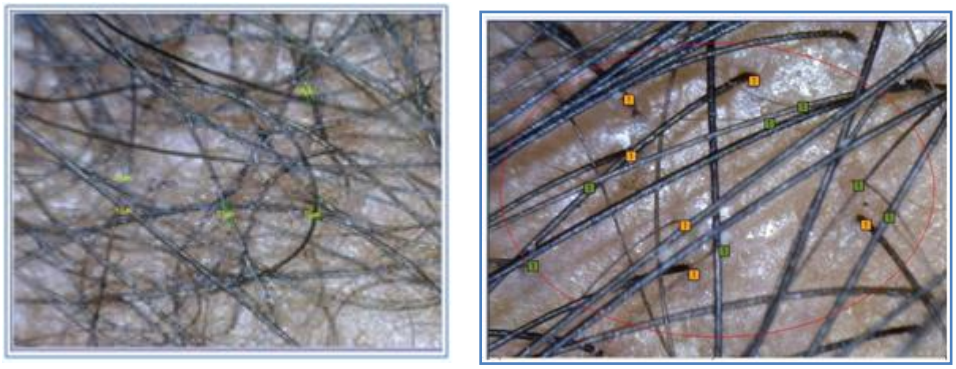


Fig. 3: No evidence of dental involvement



Fig. 4 & 5: No evidence of keratoderma

Trichoscan and microscopic study was compatible with the diagnosis of woolly hair (Figure 3). Trico scan examination revealed the presence of tight spirals and a clear diameter reduction as compared with normal hair.On hair microscopy Trichorrhexis nodosa was observed.



Tricho scan: Patient's woolly hair compared with normal hair at similar age



MICROSCOPY: TRICHORRHEXIS NODOSA

DISCUSSION: The term woolly hair refers to an abnormal variant of hair that is fine, with tight coiled curls, and often hypopigmented. In 1907, Gossage [1]was the first to describe a case of woolly hair in a European family, comparing this hair anomaly with the characteristic curly hair of black people. Woolly hair is extremely curly, with the average diameter of hair recorded up to a maximum of 0.5 cm. It is different from the curly hair of black people, in that the curled hair of black people lies separately while the curls of woolly hair usually merge. [2,3]

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Woolly hair is extremely curly, with the average diameter of hair recorded up to a maximum of 0.5 cm. It is different from the curly hair of black people, in that the curled hair of black people lies separately while the curls of woolly hair usually merge.[2,3] Woolly hair can appear as a part of systemic disease (Woolly hair syndrome) or without systemic findings (non-syndromic woolly hair). The latter can be inherited as either an autosomal dominant or recessive disorder.

Autosomal dominant woolly hair has been reported in six generations of a German family. A dominant form of woolly hair has also been reported in an English family with associated ichthyosis and deafness. [6] Additional features in a Dutch family with woolly hair included dental caries, acral keratoderma and facial abnormalities. [7] A case of woolly hair with keratosis follicularis spinulosa

decalvans, teeth abnormalities, scarring alopecia and ophthalmological abnormalities has also been reported. The dental abnormalities included agenesis, inclusions and malformed teeth.[8] Another case of woolly hair associated with pachyonychia congenita, nail changes in the form of yellowish brown hypertrophy and wedge-shaped subungual hyperkeratosis of all 20 nails with natal teeth has been reported.[9].

Two characteristic associations of woolly hair are Naxos disease and Carvajal disease. Naxos disease is characterised by woolly hair, palmoplantar keratoderma and dilated cardiomyopathy with right ventricular dysplasia.[10] It is an autosomal-recessive disorder occurring due to mutation in the plakoglobin gene.[11] Carvajal disease is similar clinically to Naxos disease, except for left ventricular involvement and presentation at a younger age, and it is due to mutation in the desmoplakin gene. [12]

Acquired woolly hair occurs most commonly in the context of patterned hair loss. It may also be caused by drugs like valproate and retinoids.[13] Our patient did not have any palmoplanter, dental, skeletal or cardiac involvement, and there was no history of any drug intake prior to onset of lesions.

MANAGEMENT AND TREATMENT: Treatment for woolly hair is not currently available,although in some patients the hair may become darker and less curly with time.[14]Physical and chemical traumatic measures should be avoided.

In syndromic occurrence palmoplantar keratosis a symptomatic treatment involving the mechanical abrasion of the hyperkeratosis and moisturizing measures are effective.

In the case of heart anomalies intensive internal and cardiological care is necessary. The pharmaceutical treatment of arrhythmias and anticoagulants as a prophylactic measures for embolic thrombosis are recommended. In cases of disturbed stimulus conduction it may be necessary to consider implanting pacemaker. The final option for treating therapy-resistant dilated cardiomyopathy(NYHA IV) is a heart transplant.

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