

Generalized wooly hair with new associations: A case report

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ABSTRACT

Wooly hair is a structural anomaly of the hair shaft that affects scalp hair and manifests itself as tightly coiled hair. Wooly hair may be localized or generalized, but the generalized form is rare and may be limited to the scalp or complicated by cutaneous and extracutaneous features. A four-year-old female complaining of short, tightly-coiled scalp hair present since birth. An examination of the skin revealed the scalp hair to be fine, coarse, and lightly-pigmented. The cutaneous surface was dry, with multiple hypopigmented macules on the upper trunk and a larger, ash-leaf-shaped, located on the anterior trunk. Also, a large hyperpigmented patch, a congenital melanocytic nevus, was noted on the posterior trunk. An examination of the hair under a light microscope revealed various hair shaft abnormalities. To our knowledge, generalized wooly hair in association with an ash-leaf macule and a congenital melanocytic nevus has not been reported before.

Key words: Wool hair; Ash-leaf macule; Unruly hair; Melanocytic nevus

INTRODUCTION

Wooly hair is a disorder that results from a structural defect in the hair shaft, without increasing its fragility [1,2], showing as tightly-coiled, unruly hair in non-black races [3]. Wooly hair is generally a cosmetic problem but, in some, may threaten life [4]. It manifests itself either as the localized non-hereditary variant, a wooly hair nevus, or as the generalized hereditary form. Generalized wooly hair may be an isolated anomaly, affecting only the scalp hair, or associated with cutaneous and extracutaneous manifestations [5]. Herein, we present a case of generalized wooly hair in a four-year-old girl associated with skin anomalies but without systemic involvement.

CASE REPORT

A four-year-old girl brought by her father showed short, thin, light-colored, easily-fractured, tightly-coiled, and unruly hair that had not increased in length since birth. The child was born to a sanguineous marriage

and was the seventh and the only affected among eight siblings: five daughters and three sons. There was no family history of the same condition, atopy, or other genetic disorders. On examination, the patient looked well, apart from the affected scalp hair. The hair was short, fine, light-colored, coarse in texture, and tightly-coiled into short locks. (Figs. 1a and 1b). The eyebrows and eyelashes were normal. The skin surface was dry, with multiple pityriasis alba lesions on the face associated with widespread keratosis pilaris. Multiple hypopigmented spots were present, mainly on the upper trunk and in a confetti-like distribution (Fig. 2). An ash-leaf-like hypopigmented macule was present on the anterior trunk (Fig. 3) and another macule, but hyperpigmented, on the posterior upper trunk (Fig. 2). There was no palmoplantar keratoderma. The nails, teeth, and eyes were not affected. Light microscopy revealed various hair calibers: fine hair, hair twisted on its long axis, fractured hair, hair with knots, and trichorrhexis nodosa (Figs. 4a – 4d). Echo studies and hematological and biochemical investigations were within normal limits. An examination of the hair with

How to cite this article: Mancy A. Generalized wooly hair with new associations: A case report. Our Dermatol Online. 2021;12(3):304-306.

Submission: 08.12.2020; **Acceptance:** 28.02.2021

DOI: 10.7241/ourd.20213.18



Figure 1: Wooly hair on the scalp in (a) a 1.6-year-old patient and (b) a 4-year-old patient.



Figure 2: Hypopigmented spots on the posterior trunk in association with a hyperpigmented patch.

10% potassium hydroxide revealed no fungal elements. Growth milestones were normal for the patient's age and no abnormality was detected on systemic examination. Taking into consideration these findings, the case was mostly an autosomal recessive variant of generalized wooly hair with cutaneous anomalies, but without systemic manifestations.

DISCUSSION

Wooly hair is a hair shaft disorder either limited to a part of the scalp in the form of a wooly hair nevus or generalized as in familial wooly hair and hereditary wooly hair [6]. Wooly hair may be only a cosmetic concern if confined to the scalp or associated with fatal complications, such as cardiomyopathy [4]. Generalized wooly hair may be the only abnormality present or may be associated with injury to the skin, teeth, eyes, or heart [7]. Hair growth in generalized wooly hair of the scalp is normal or slower than in a healthy individual but the anagen phase is shorter, leading to shorter hair



Figure 3: The hypopigmented ash-leaf-like patch on the anterior trunk and keratosis pilaris.



Figure 4: (a) Hair microscopy showing (a) a fractured hair, (b) different hair calibers (fine, thin, and twisted), (c) a fine and broken hair shaft, and (d) a hair with a knot formation.

that does not grow to be more than several centimeters long [7,8]. Also, the presence of trichorrhexis nodosa leads to the breakage of hair, effectively preventing hair growth [3]. Despite this, the length and condition of hair may improve with age [9] (as in the patient shown in Fig. 1). Keratosis pilaris is a disease that results from a keratinization abnormality in which the hair follicle is affected by a keratinous plug [10]. Widespread keratosis pilaris, affecting most of the skin surface, as in the patient shown in Fig. 1, was reported by Budhwar et al. to be a cutaneous anomaly manifested by generalized wooly hair [8]. Vasudevan B et al. described a four-year-old patient with widespread keratosis pilaris in addition to generalized wooly hair [11].

Xerosis and pityriasis alba are non-specific and minor criteria for atopic dermatitis [10]. Nonetheless, our

case was not atopic and there was no family history of atopy, so these may be considered a non-specific association with generalized wooly hair. Multiple hypopigmented spots were reported by Pavone et al. in a 29-month-old female, mainly between the shoulders [12]. In our case, the hypopigmented spots were more widespread, distributed as if confetti, and present on the shoulders and the upper and lower trunk. An ash-leaf-like hypopigmented macule was present on the anterior trunk. The macule was a white, oval lesion, which is a feature of tuberous sclerosis also seen in familial progressive hyperpigmentation and hypopigmentation [13,14]. This has not been previously reported in association with wooly hair. A hyperpigmented patch is a brownish patch with a well-defined border, generally a congenital melanocytic nevus (CMN) of medium size. A CMN is a nevus of melanocytic origin, its color varies from light to dark brown, may be of any size, and any site of the skin may be affected [15].

Although the examination of wooly hair with a light microscope usually shows non-specific features, different anomalies may be observed, such as varying calibers of hair and twisted hair on its long axis [11,16]. Trichorrhexis nodosa, hair with tapered ends, and knot formations may also be seen [4,16]. In our case, most of these hair shaft anomalies were present. Two important syndromes are associated with generalized wooly hair: Naxos syndrome and Carvajal syndrome. However, these diseases are associated, in addition to wooly hair, with palmoplantar keratoderma and cardiomyopathy [4].

CONCLUSION

Although, in such cases, genetic studies are important to reach an accurate diagnosis, they are not available at our location. Taking into account the history, clinical presentation, and investigations performed, our patient is mostly a case of generalized wooly hair of the scalp in association with cutaneous anomalies of ectodermal origin, but without systemic involvement. Some of these anomalies have been mentioned in the literature, such as keratosis pilaris and hypopigmented spots. However, to our knowledge, we are the first to report an association with an ash-leaf macule and a congenital melanocytic nevus.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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Source of Support: Nil, **Conflict of Interest:** None declared.