Woolly hair nevus: case report and review of literature

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Abstract

Woolly hair nevus consists of a patch of curly and hypopigmented hair that is restricted to an area of the scalp. It is usually benign but it can be associated with other systemic findings. Trichoscopy and dermoscopy may be useful when analyzing this entity. The authors describe a case of woolly hair nevus in a 5-year-old boy and present a review of the literature of woolly hair nevus, including classification, histopathology, associated systemic findings, and the recent described genetic mutations.

Keywords: woolly hair, trichology

Introduction

Woolly hair is a rare condition. It is characterized by coiled, curly, and slightly hypopigmented hair with an oval form at transversal section. The affected hair has a smaller diameter but without increased fragility [1, 2]. It is not a form of hypotrichosis [3]. The term, wooly hair nevus (WHN), describes a case of woolly hair restricted to a limited area of the scalp [3, 5].

Several associations have been described such as ophthalmologic, auditory, renal, cutaneous, dental, and skeletal manifestations. There is an ipsilateral verrucous epidermal nevus in half of patients with WHN. Recent genetic findings suggest that WHN represents a mosaic RASopathy.

Case Synopsis

We describe a previously healthy 5-year-old boy who presented to our department with abnormal hair findings since the first years of life. Examination revealed a solitary patch of light and coiled hair, localized at the right parietal region (**Figure 1**). The texture of the hair was different from the rest. The remaining hair was straight dark brown. Trichoscopy of the patch showed hypopigmented, light, and coiled hair (**Figure 2**). There were no relevant findings by dermoscopy of subjacent scalp. The child did not show any other cutaneous stigmas, or cardiac or ophthalmologic alterations. His past medical history was unremarkable. His parents denied similar clinical observations in other family members. A diagnosis of woolly hair nevus was made. Owing to the typical clinical picture no scalp biopsy was performed.

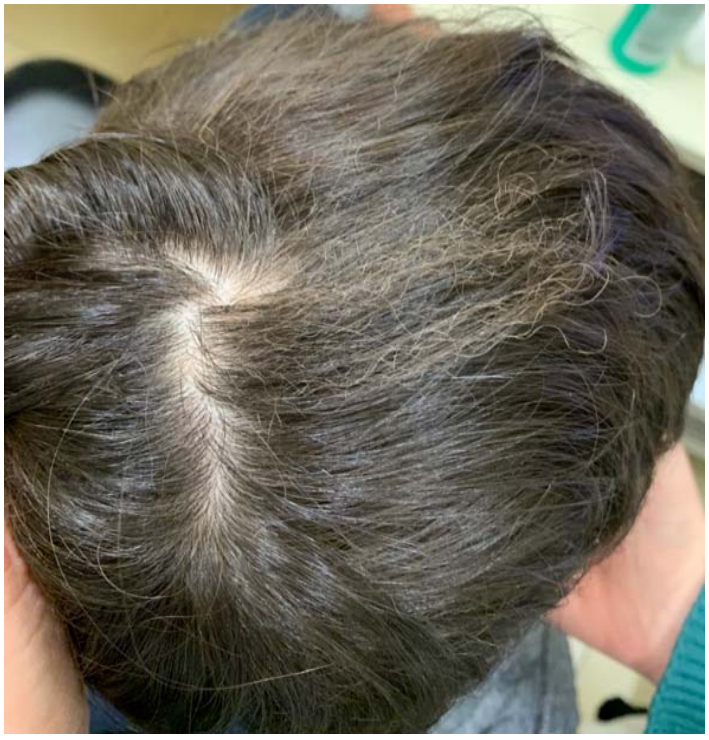

Figure 1. *Patch of hypopigmented and curly hair on the right parietal area.*

Case Discussion

Hutchinson et al. classified woolly hair as hereditary wooly hair (autosomal dominant), familial woolly hair (autosomal recessive), and woolly hair nevus [4]. The first two are generalized and genetically transmitted forms whereas the third type is a localized non-hereditary form. Wooly hair nevus describes a case of woolly hair restricted to a limited area of the scalp [3, 5].

Post subdivided the WHN into type 1 (no cutaneous involvement), type 2 (associated linear verrucous epidermal nevus), and type 3 (acquired in young adult patients in which scalp has short, dark, twisted hair), which has been termed acquired progressive kinking of scalp hair [6].

The onset of WHN is usually noted within the first two years of life, although some cases may be detected in adolescence [7, 8]. Most cases reported have a solitary patch, but up to four patches have been described in the same individual [8]. When presenting in a neonate, it may manifest as a patch of alopecia instead of a coiled and curly hair patch. It is not true alopecia but rather an area of sparse thin hair, which reflects the slower growth rate of the hair in comparison with normal hair [9]. Some authors consider the growth rate of the affected hair to be normal, but the hair may be short owing to the shortened growth cycle [1].

Different histopathology patterns have been reported. Fernandes et al. found usual density of hair follicles, mostly terminal and anagen, with no significant inflammatory infiltrate. Some hair shafts were slightly oval-shaped in the sections [5]. Gomez et al. described several involved terminal hair follicles ending in the same infundibulum, a perifollicular lymphocytic infiltrate, and an excessive amount of normal apocrine glands [10]. Veraitch et al. described a diffuse miniaturization of the hair follicles with variation in size among hair follicles when compared with the biopsy from unaffected scalp. Comparison of the terminal-to-vellus (T:V) ratio between affected and unaffected scalp showed T:V=1:1 in affected scalp and T:V=7:1 in the control scalp. Affected scalp had a reduced anagen-to-telogen ratio of A:T=4.2:1, whereas unaffected scalp had an anagen-to-telogen ratio of A:T=8:1. The mean diameter of woolly hair shafts (63±19.6µm) was significantly smaller than normal hair shafts (88±3.2µm), [11].

Trichoscopy usually shows a hair shaft irregularity with a "snake crawl appearance" [12]. Dermoscopy is useful to search for cutaneous findings in the scalp.Light microscopic examination of the hair usually reveals no structural abnormalities [13]. Sometimes it can show irregularities of the cuticle, varying diameter of a single hair shaft, twisting of hair, and trichorrhexis nodosa-like features [8, 12]. Electron microscopy reveals twisting of the hair shaft at irregular distances with a longitudinal groove reminiscent of pili canaliculi. In some cases a cuticle may exhibit a reduced number of layers. The cross-section is usually elliptical and thinner than normal normal hair [14, 15].

There have been several associations between woolly hair nevus and ophthalmologic (heterochromia iridis, persistent papillary membrane), [16, 17], auditory (auditory disturbance), [18], renal, cutaneous (pigmentary demarcation lines, linear epidermal nevus, white sponge nevus), [2, 13, 16, 19], dental (diastema, decayed lower canine teeth), [5,13], and skeletal (growth impairment, brachyphalangy), [18] manifestations. In half the cases, patients with WHN have an ipsilateral verrucous epidermal nevus. The areas most commonly affected by this condition are the face, the neck, and the ipsilateral upper limb [15].

Figure 2. *Trichoscopy: detail of the hypopigmented and curly hair, with no relevant findings on the scalp.*

Generalized woolly hair may be associated with systemic diseases such as Naxos and Carvajal diseases, Noonan syndrome, or cardiofaciocutaneous syndrome [20].

Genetic studies have recently given some clues about WHN. Woolly hair associated with epidermal nevi may be caused by an *HRAS* p.G12S mosaic mutation. Nishihara et al. identified the *HRAS* c.34G>A (pG12S) mutation in the hair roots of the woolly hair but not of the straight hair in a child who also presented with an epidermal nevus. The same mutation was also present in the epidermis of the epidermal nevus [21]. Levinsohn et al. also reported the findings of *HRAS* c.34G>A, p.G12S mutation in affected tissue from two patients with mosaic woolly hair and epidermal nevi. The authors suggested that WHN represents a mosaic RASopathy and in contrast to the strong activation of RAS mutations found in nevus sebaceous (which drive hair follicle progenitors towards sebocyte differentiation), the weaker activation of the mutation in WHN permits an intermediate phenotype with abnormal curly hair without sebaceous hyperplasia [22]. Nishiara et al. suggests that epidermal nevus patients who have the mosaic *HRAS* mutation both in epidermal nevi and also in other tissues may be at risk of developing neoplasms similar to those found in Costello syndrome. The authors conclude that if the *HRAS* p.G12S mutation is identified, periodical follow-up is necessary to screen for carcinogenesis from early childhood [21]. Kuentz et al. reported a postzygotic *BRAF* mutation in a patient with a subtype of phacomatosis pigmentokeratotica (PPK) who had agminated melanocytic nevi, epidermal nevus, woolly hair nevus in a mosaic pattern, focal cortical dysplasia (FCD), and cerebral cavernous malformation. The authors found a somatic heterozygous *BRAF* c.1803A>T (p.Lys601Asn) mutation in her epidermal and woolly hair nevi [23].

There is no effective treatment. Improvement has been reported with topical minoxidil 5% in an 11-year-old boy, but the hair relapsed on suspension of the therapy [11]. The woolly hair may become darker and less curly with the years in some patients [7]. However, a progressive course has been reported in the number of patches of one patient [8].

Conclusion

Woolly hair nevus is a rare non-hereditary condition in which the patient presents with a patch of curly, hypopigmented hair in the first years of life. It is usually a benign condition, although it may be associated with other systemic findings. The genetic diagnosis may be important in those cases. It is therefore recommended to perform a detailed examination of a patient who presents with wooly hair nevus.

Potential conflicts of interest

The authors declare no conflicts of interests.

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