

Case Report



Address for correspondence:

Dr. Laura Miguel Gomez,
Department of Dermatology,
Ramon y Cajal Hospital,
Madrid, Spain. Carretera de
Colmenar Viejo km 9,1, 28034
Madrid, Spain.
E-mail: lmg_0007@hotmail.com

A Rare Presentation and Histopathologic Findings of Woolly Hair Nevus

Laura Miguel Gomez, Lorea Bagazgotia, Luis Requena¹

Department of Dermatology, Ramon y Cajal Hospital, ¹Department of Pathology, Fundación Jimenez Diaz Hospital, Madrid, Spain, Europe

ABSTRACT

Woolly hair nevus is a rare disease whose diagnosis is challenging. We present a case of this condition presenting in a 27-year-old healthy male. We describe a histology pattern consisting in the presence of several terminal hair follicles ending in the same dilated follicular infundibulum, a perifollicular lymphocytic infiltrate and an excessive amount of normal apocrine glands in deep reticular dermis, some findings non-previously reported. Clinicopathological correlation is very important for making a correct diagnosis.

Key words: Hair disorders, histopathologic pattern, woolly hair nevus

INTRODUCTION

Woolly hair nevus (WHN) is characterized by an area with fine, curly and often hypopigmented hair, on a circumscribed area of the scalp.^[1] Hutchinson *et al.*^[2] classified three variants of woolly hair: Hereditary woolly hair (autosomal dominant), familial woolly hair (autosomal recessive) and a WHN. Woolly hair syndrome (WHS) affects to the whole scalp and it may be associated with other cutaneous anomalies and extracutaneous anomalies, such as Noonan syndrome or carvajal disease.^[3-5] In contrast to the WHS, WHN is not a hereditary condition.^[6] Recently, it has been found in two cases of WHN, somatic HRAS c. 34G > A, p.G12S mutation in affected in hair. Interestingly, this mutation has also been observed in epidermal nevus.^[7] Also, a HRAS c. 37G > C, p.Gly13Arg mutation has been described in nevus of Jadassohn and Schimmelpenning syndrome.^[8]

CASE REPORT

We report the case of a 27-year-old healthy male who presented with an area of curlier and thinner hair than the rest of his scalp localized on the right parietal area. He referred that just at that location, his hair had been different since childhood, curlier than the rest. Physical examination revealed a plaque with shorter and curlier hair on an area of 10 cm in diameter on

the right parietal area of the scalp. A decreased density of hair was observed. The skin on the patch was apparently healthy. There were not exclamation mark hairs or miniaturization of hair follicles. A hair pull test was negative [Figure 1]. A skin biopsy showed a mild perifollicular lymphocytic infiltrate, telangiectatic capillaries in the superficial dermis and several terminal hair follicles emerged in the same dilated follicular infundibulum. Interestingly, an excessive amount of normal apocrine glands were found in the deep reticular dermis. Counting of hair follicles was normal [Figure 2a-c]. Based on clinical appearance and histopathologic studies, a diagnosis of WHN was established.

DISCUSSION

WHN usually appears during the 1st years of life and it remains stable throughout adulthood. It may be associated

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Figure 1: (a) A plaque with shorter and curlier hair on an area of 10 cm in diameter on the right parietal area of scalp. Skin on the patch was apparently healthy. (b) An area of curlier hair than the rest of his scalp localized on the right parietal area since childhood

with pigmented or epidermal nevus, usually on the neck, arm or even coexisting with WHN. Histopathologic findings of WHN are scarcely described in the literature, probably because most cases are diagnosed clinically. A wavy appearance of hair follicles with perifollicular infiltration of inflammatory cells has been described,^[9] but normal hairs and cutaneous appendages may also be observed.^[10] In our case, we found a different histopathological pattern, with several involved terminal hair follicles ending in the same infundibulum, a perifollicular lymphocytic infiltrate and an excessive amount of normal apocrine glands, findings similar to those of nevus of Jadassohn. It may be possible that WHN is the clinical manifestation of a wide range of different histopathologic hamartomatous lesions involving the hair follicles, some of them clearly evident, with thinner and curved hair follicles and accompanying epidermal nevus in the overlying epidermis, and subtle histopathologic changes in other cases, as in our case.

CONCLUSION

We described a case of WHN showing some nonpreviously reported histopathologic findings. These findings described indicate that clinicopathologic correlation is necessary to establish a diagnosis of WHN with confidence.

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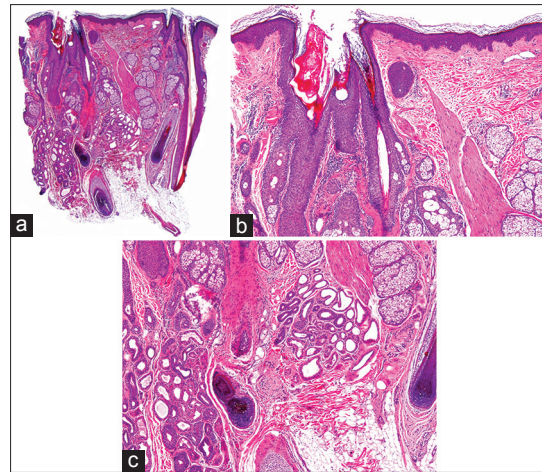


Figure 2: (a) Several involved terminal hair follicles ending in the same infundibulum, a perifollicular lymphocytic infiltrate and an excessive amount of normal apocrine glands (H and E, $\times 4$). (b) Several involved terminal hair follicles ending in the same infundibulum and a perifollicular lymphocytic infiltrate (H and E, $\times 20$). (c) An excessive amount of normal apocrine glands (H and E, $\times 20$)

Conflicts of interest

There are no conflicts of interest.

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