

## CASE REPORTS

# Woolly Hair Nevus Associated With an Ipsilateral Linear Epidermal Nevus

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**Abstract.** We report a 4-year-old boy with two areas of woolly hair in the right parietotemporal region and a linear epidermal nevus in the areas of woolly hair as well as in the ipsilateral hemiface and chin. Evaluation by scanning electron microscopy showed woolly hair with oval transverse section and longitudinal groove. A complete examination ruled out associated anomalies.

**Key words:** woolly hair, woolly hair nevus, lineal epidermis nevus.

## NEVUS DE PELO LANOSO ASOCIADO A NEVUS EPIDÉRMICO LINEAL IPSILATERAL

**Resumen.** Presentamos un paciente varón de 4 años con dos áreas de pelo lanoso en zona parietotemporal derecha y un nevus epidérmico lineal en las zonas de pelo lanoso, así como en hemicara y mentón ipsilateral. La imagen de microscopía electrónica de barrido mostraba el pelo lanoso con sección transversal oval y acanaladura longitudinal.

El estudio sistémico para descartar anomalías asociadas fue normal.

**Palabras clave:** pelo lanoso, nevus de pelo lanoso, nevus epidérmico lineal.

## Introduction

Woolly hair is the normal structure for the hair of black individuals. However, in white individuals with no black ancestry it is a hereditary anomaly of the hair.

The woolly hair nevus is an uncommon nonhereditary disorder. It is characterized by the presence on the scalp of one or more clearly circumscribed regions of extremely kinky, smooth, and shiny hair, of a woolly texture, habitually of a lighter color than the normal hair, wrongfully giving the appearance hypotrichosis.<sup>1</sup>

In all the cases cited in the literature, the onset of this hair dysplasia is during the first 2 years of life, and in more than half of all cases it is associated with an epidermal nevus which may or may not involve the area of affected hair.<sup>2</sup>

We present a case of woolly hair nevus associated with an epidermal nevus with a slightly raised surface on palpation located in the same cephalic region, as well as on the ipsilateral side of the face and chin.

## Case description

A 4-year-old male patient of white European descent, with no hair abnormalities, ectodermal dysplasias, or history of consanguinity amongst his family members, was brought by his parents to the clinic because of very fine and kinky hair on his scalp since birth (Figure 1).

Physical examination confirmed the patient was a blond, phototype II child with blue eyes, normal teeth and nails, and no other cutaneous findings of interest.

Inspection of the scalp revealed two different populations of hair: the head was generally covered with light chestnut colored hair that was smooth, thick, and shiny, while in the right parietotemporal region we saw two clearly circumscribed areas of fine, extremely kinky, blond, and woolly-textured hair in a blaschkoid distribution, separated by normal hair. There was no real alopecic plaque or hypotrichosis. When the pull test was applied no hair was plucked.

In the same region of the head, and on the right side the face and chin, we found a slightly hyperkeratotic lesion of a light brown color and linear distribution, compatible with a linear epidermal nevus (Figure 2).

An ophthalmologic study was carried out to rule out strabismus, exudative retinitis, cataracts, and other associated anomalies, and the findings were normal. Electrocardiography, chest radiography, and echocardiography were performed to

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Electron microscope images courtesy of Dr Juan Ferrando Barberá.



**Figure 1.** Two clearly circumscribed areas of fine, kinky, blond hair could be seen in a blaschkoid distribution on the right parietotemporal zone of the scalp, separated by normal hair.

rule out cardiac disorders; in all cases the findings were also normal. Neurological examination ruled out retarded mental development and hemiparesis, and the electroencephalogram did not present any relevant findings. The skeleton was studied through a series of radiographs to rule out kyphosis, scoliosis, cysts, or delayed bone growth, and these showed no abnormalities.

Hair samples from the areas where the hair appeared finer and curlier than the normal hair were examined under an optical microscope.

The scanning electron microscope revealed an elliptical transverse section (Figure 3), a longitudinal groove running the length of the shaft of the hair (Figure 4), and a reduction in the number of cuticle cells.

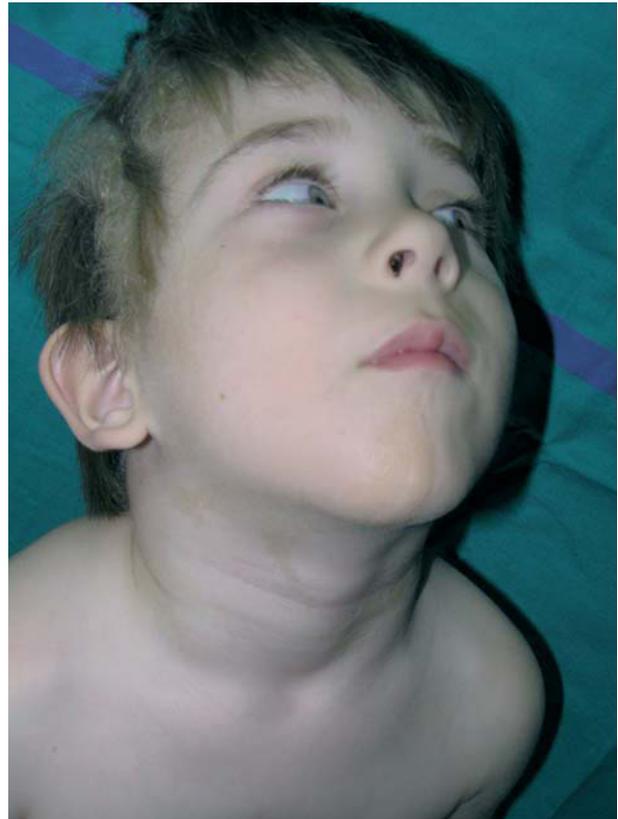
## Discussion

Gossage, in 1907, was the first to describe woolly hair in a European family<sup>3</sup> comparing this anomaly of the hair shaft with the curly hair characteristic of people of black ancestry. However, the clumping, which can be seen in this woolly hair, is not characteristic of that racial group.

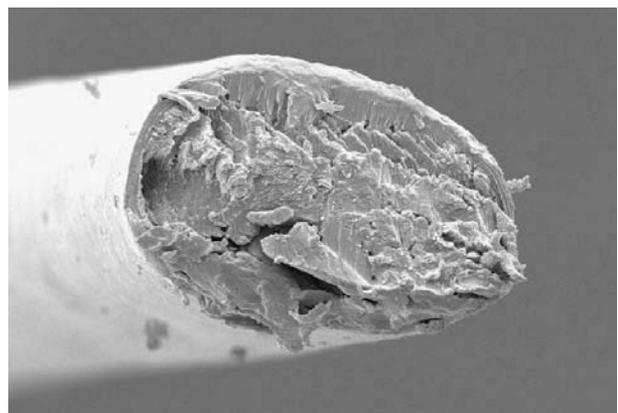
Woolly hair is extremely kinky, woolly textured, and often of a lighter color than normal hair. It tends to be difficult to comb, but is not fragile except when associated with trichorrhexis nodosa.<sup>4</sup>

Hutchison et al<sup>5</sup> classified woolly hair into three groups:

1. A hereditary form, of autosomal dominant inheritance in most cases (hereditary woolly hair),<sup>5-8</sup> characterized by the presence from the first weeks of life of an extremely kinky, occasionally hypopigmented, dark hair.<sup>6</sup>

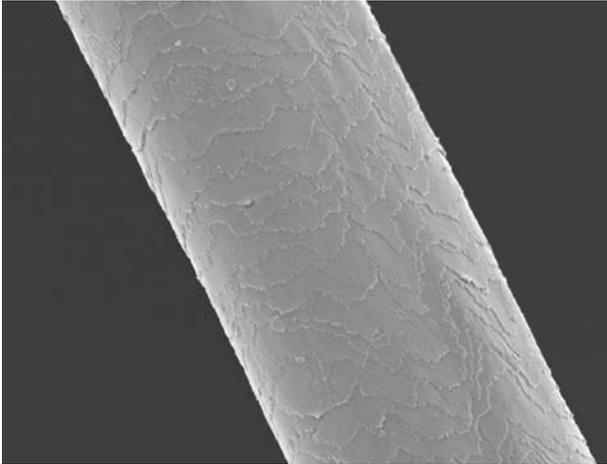


**Figure 2.** In the cephalic region bearing the woolly hair, and on the right side of the face and chin, a slightly hyperkeratotic, light brown, linear lesion could be seen, compatible with linear epidermal nevus.



**Figure 3.** An elliptical transverse section could be observed under the scanning electron microscope.

2. A familial form, of autosomal recessive transmission (familial woolly hair), with thin hair of a color lighter than that of unaffected family members, in some cases affecting body hair, and the lateral portion of the eyebrows. In Naxos disease this form of woolly hair has been



**Figure 4.** Longitudinal groove along the shaft of the hair.

associated with palmoplantar hyperkeratosis of the Vörner-Unna-Thost type, and cardiac disorders.<sup>9</sup>

3. A nonhereditary, localized form (woolly hair nevus).

In addition, Ormerod et al<sup>10</sup> described diffuse partial woolly hair in adolescents and young adults, characterized by the presence of woolly hair mixed in with normal hair, constituting from 20% to 30% of all the scalp hair. In these cases the woolly hair is short, fine, and hypopigmented.<sup>10</sup>

Wise, in 1927, was the first to describe the woolly haired nevus<sup>11</sup> in 2 girls of Syrian-Jewish origin, who presented a section of woolly hair, one of them having an associated linear epidermal nevus on the neck.

Post<sup>12</sup> divided the woolly hair nevus into three categories:

1. Type 1: with no associated scalp disorders or hairless skin.
2. Type 2: with associated linear verrucous epidermal nevus.
3. Type 3: acquired, in young adults with short, dark, kinky hair, which has been termed acquired progressive kinking of scalp hair.<sup>13</sup>

Type 2 woolly hair nevi tend to show a distinctive blaschkoid distribution, as can be seen in the present case.

Clinically, the woolly hair nevus is characterized by the presence from birth or the first months of life of one or several well circumscribed areas on the scalp of very curly, kinky, matted hair, generally of a smaller diameter and a lighter color than the remaining normal hair, although it can darken over the years.<sup>12,14</sup>

Woolly hair grows normally and is not fragile. When placed in boiling water it contracts into tight spirals with a diameter of less than 5 mm. Descriptions from electron microscope studies are characterized by flattened hairs with an elliptical cross-section, thinner than normal, with transverse diameters of between 0.044 mm and 0.086 mm, a longitudinal

groove reminiscent of pili canaliculi, and in some cases a cuticle structure with a reduced number of layers.<sup>15</sup>

Our case is reminiscent of those described previously, with elliptical sections and longitudinal grooves.

The electron microscope fundamentally provides a differential diagnosis from the so-called pili torti, which can, in turn, be associated with other hair dysplasias such as monilethrix or trichorrhexis nodosa, as well as Menkes syndrome (kinky hair).<sup>15</sup>

These nevi normally grow in proportion with the body, reaching a size which remains stable throughout adult life.

The scalp tends to be normal in color and consistency, although cases have been described with associated seborrheic dermatitis within the area of the nevus.<sup>16</sup> In one of their cases, Rodríguez Pichardo et al<sup>17</sup> observed a sebaceous nevus which was not biopsied.

In half the cases, patients with woolly hair nevus show an associated ipsilateral verrucous epidermal nevus,<sup>14</sup> as did our patient, thus falling into type 2 as described by Post. The areas most commonly affected by this condition are the face, the neck, and the ipsilateral upper limb.<sup>11,12,18,19</sup>

Peteiro et al<sup>2</sup> described the first case of an epidermal nevus underlying the woolly hair nevus, and subsequently new cases similar to ours were reported.<sup>2,20</sup>

Other extracutaneous anomalies have been noted which may be linked with the epidermal nevus, constituting epidermal nevus syndrome.<sup>21</sup> These anomalies include bone, neurological, and ophthalmological alterations, and, less often, cardiac and renal disorders, as well as other cutaneous disorders (sebaceous nevus, acanthosis nigricans, and hemangioma).

In our case—as would be expected given the limited area covered by the epidermal nevus—studies undertaken to rule out associated anomalies proved normal. The cause of the frequent association between woolly hair nevus and epidermal nevus is unknown.

Our patient presents the clinical and morphological features of woolly hair nevus.

Where an associated linear epidermal nevus is present there is an obligation to investigate and rule out the possibility of systemic anomalies associated with epidermal nevus syndrome, although this fundamentally occurs in cases of systematized epidermal nevus.

#### Conflicts of interest

The authors declare there are no conflicts of interest

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