

Becker's Melanosis in Association with Ipsilateral Linear Verrucous Epidermal Nevus and Woolly Hair Nevus-A Coincidence or New Type of Epidermal Naevus Syndrome?

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Introduction

Becker melanosis is an acquired melanosis and hypertrichosis usually in a unilateral distribution, first described in 1948 by S. William Becker. It is characterized by peripubertal development, male preponderance, hypertrichosis, occasional development of acneform lesions within the patch, and rare association with accessory scrotum in the genital region. Androgens may play a significant role in its pathogenesis [1,2].

Woolly hair is characteristically unruly and frizzy and with resemblance to negroid hair and could present as autosomal dominant (hereditary), autosomal recessive (familial) woolly hair, symmetrical circumscribed allotrichia and as a woolly hair nevus (WHN) [3,4].

WHN is an uncommon non-genetically determined nevus condition first described by Dr. Fred Wise (1927) characterized by circumscribed patch of abnormal tightly coiled and abnormal in color hair upon unremarkable scalp. Both sexes are equally affected. Hairs are often thinner and lighter than the unaffected hair, demonstrate prominent axial twisting, narrower oval cross section and form narrow in diameter curls. Follicles may be curved. Electron microscopy shows marked flattening of the hair shafts, absence of cuticle and presence of trichorrhexis nodosa. Condition develops either when the scalp hair begins to grow, later in childhood or adolescence. There is a report on initial presentation as neonatal alopecia [5].

Epidermal nevi are congenital hamartomas of embryonal ectodermal origin classified on the basis of their main component as sebaceous, apocrine, eccrine, follicular, or keratinocytic. Approximately one third of individuals with epidermal nevi have associated skin conditions or involvement of other organ systems; hence, this condition is considered to be an epidermal nevus syndrome. An association between epidermal nevus and WHN has been described in the literature [5,6]. There is no report of association of Becker melanosis with WHN and epidermal nevus.

Case

We report the case of an 11-year-old Caucasian female patient in good general condition who at birth was reported to have alopecic patches on the occipitoparietal area of the scalp and three confluating hyperpigmented papillomatous slightly hyperkeratotic linear lesions on the ipsilateral side of her neck. Several months later finer tightly curled and slightly lighter than in the rest of the scalp hairs grew over the alopecic patches. At the age of 10 light brown sharply demarcated large areas of hyperpigmentation developed on her back, chest and arms. On her chest we observed 40/30 cm large, well circumscribed slightly hyperpigmented macule on the right side with sharp demarcation on the median line. On her back- several hyperpigmented macules varying in size from 20/30 cm to 5/10 cm with phylloid shape, sharply demarcated on the median line, were present (Figure 1a and 1b). In the last year, fine hairs appeared on the hyperpigmentations on the arms and new hyperpigmentation appeared on her face. On clinical examination, we noticed 3/5 cm circumscribed patch of woolly textured curly light in

color hair on the left occipitoparietal region of the scalp separated with areas of normal hair. On the ipsilateral side of her neck, three linear, sharply demarcated hyperpigmented papillomatous plaques with slightly raised surface on the left side of the median line, following the Blaschko lines, were present (Figure 2). Symmetrically, on the outer side of her arms, hyperpigmented bands covered with fine sparse hair were present. The rest of the skin, scalp and nails were normal. There were no teeth abnormalities. Family history was negative. The patient was diagnosed clinically with woolly hair nevus of the left parietal and

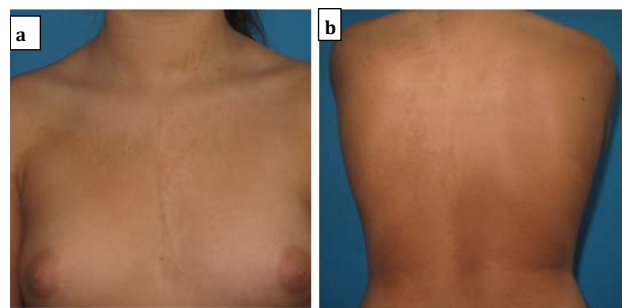


Figure 1a and 1b: several hyperpigmented macules varying in size from 20/30cm to 5/10 cm with phylloid shape, sharply demarcated on the median line on chest and neck.



Figure 2: Sharply demarcated hyperpigmented papillomatous plaques with slightly raised surface on the left side of the median line.

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occipital scalp, an associated ipsilateral epidermal nevus, and Becker's melanosis.

Light microscopic examination of clinically abnormal hair showed axial twisting of the hair shaft and presence of trichorrhexis nodosa (Figure 3). Histopathological examination of biopsy specimen obtained from the lesion on the neck revealed epidermal nevus (Figure 4a). Histopathological examination of a biopsy from hyperpigmented hypertrichotic macule on the left arm showed mild acanthosis with hyperkeratosis with elongation of the epidermal rete ridges with increased melanin in the basal layer. This histological picture is characteristic for Becker's melanosis (Figure 4b).

Ophthalmologic examination showed no structural abnormalities of the eyes. MRI of the head was made due to slight diplopy and showed no particularities. Diplopy was attributed to astigmatism of the left eye that will be corrected with lenses.

Electrocardiography and echocardiography were normal.

Radiographic examination of the teeth excluded abnormalities.

Discussion

Becker's melanosis (Becker's nevus, Becker's pigmented hairy nevus) is a common acquired cutaneous hamartoma characterized by epidermal hypermelanosis. It affects predominantly affecting young males. Congenital and familial cases have been reported. Usually it presents as single unilaretal hyperpigmented macule and hypertrichosis located on the chest, back or shoulder. Rarer manifestations are multiple Becker's nevi, on different from the above mentioned sites. Segmental Becker's melanosis is a rare clinical presentation, with only one published report in the literature.



Figure 3: Light microscopic examination of clinically abnormal hair showed axial twisting of the hair shaft and presence of trichorrhexis nodosa.

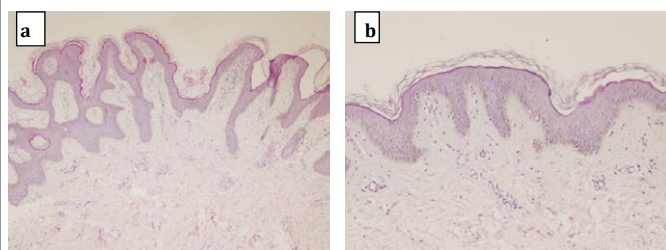


Figure 4: (a) Histopathological examination of biopsy specimen obtained from the lesion on the neck revealed epidermal nevus. (b) Mild acanthosis with hyperkeratosis with elongation of the epidermal rete ridges with increased melanin in the basal layer- histological picture is characteristic for Becker's melanosis.

Occasionally, Becker nevus may be associated with smooth muscle hamartoma. Rarely, hypoplasia of underlying structures, such as unilateral breast hypoplasia, has been reported.

Other associations seen with Becker nevus include unilateral or ipsilateral pectoralis major aplasia, ipsilateral limb shortening, ipsilateral foot enlargement, spina bifida, scoliosis, pectus carinatum, localized lipoatrophy, congenital adrenal hyperplasia, polythelia and accessory scrotum.

WHN may initially manifest as neonatal alopecia, which is also confirmed in this case.

WHN may be associated with extracutaneous symptoms (WHN syndrome). Association of WHN has been reported with ocular defects- persistent papillary membrane and retinal defects. (WHN may also be associated with digital abnormalities and auditory disturbance. In one case an association with precocious puberty has been reported. Epidermal nevus syndrome, has been ruled out in this case by appropriate examinations.

WHN has been reported in association with other cutaneous lesions. Sebaceous naevi and white sponge naevi have been reported in association with WHN.

WHN could be associated with epidermal or melanocytic nevi. Epidermal nevus may or may not involve the affected area of the scalp. Reports in the literature suggest that an association of WHN with an ipsilateral epidermal nevus occurs in around half of the cases.

Recently the association between woolly hair and epidermal nevus has been attributed to a somatic mutation present in both conditions.

Reports show the association of WHN with Mongolian spot and café- au- lait spot. To our knowledge no association of WHN with Becker melanosis has been reported so far.

Conclusion

There is no previous report about an association of Becker's melanosis with WHN. Our case could be considered an addition to different associations of WHN with pigmentary disorders. In this aspect this observation adds to the growing spectrum of epidermal lesions associated with WHN and raises the question about a previously undescribed epidermal naevus syndrome.

Our case of WHN debuted as neonatal alopecia. It confirms a previous observation that WHN arises on alopecic regions on the scalp present at birth. Furthermore, it is consistent with previous reports for association of WHN with ipsilateral epidermal nevi.

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