Becker's Nevus Syndrome

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Becker's nevus (BN) is a unilateral hyperpigmented, often hairy, cutaneous hamartoma, with geographic borders. It is usually localized on the shoulder, anterior chest, scapula, or upper arm, but there have been reports of BN in other areas (eg, lower extremities, face). In most cases, the lesion appears in late childhood or adolescence, and both congenital and familial cases have been described.

The term *Becker's nevus* (BN) describes a frequently observed hamartomatous lesion characterized by circumscribed hyperpigmentation, often associated with an irregularly bordered hypertrichosis, mainly located on the upper part of the trunk or on a shoulder. Other sites have been described in unusual places, such as the face or lower limbs, mainly in a unilateral disposition.[1] Its onset is relatively belated, during the first or second decade of life, although congenital cases of BN have been reported.[2] In 1983, Glinick et al[3] highlighted the hamartomatous nature of a whole group of manifestations—either acquired or congenital—deriving from various cellular lines (keratinocytes, melanocytes, sebaceous glands and body hair, fibroblasts, and nonstriated muscle and nerves). In 1981, Chapel et al[4] coined the term *organoid skin hamartomas* owing to increased reports of associated osseous abnormalities. Happle and Koopman[5] have recently suggested the term *Becker's nevus syndrome*, thus defining a phenotype characterized by a particular type of epithelial organoid nevus associated with homolateral mammary hypoplasia, topographically correlated skeleton abnormalities, or both.

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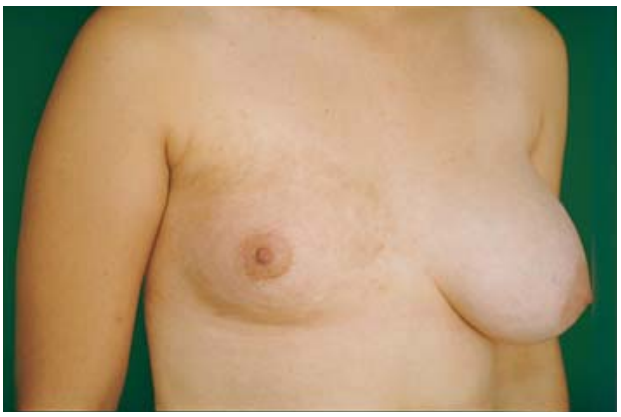

Becker's nevus and ipsilateral breast with hypoplasia.

Case Report

A 16-year-old girl presented for evaluation. The findings from the physical examination revealed an irregularly margined hyperpigmented patch on the right side of the mammary and axillary region that had areas that variedly overlapped each other, without hypertrichosis. Moreover, an ipsilateral breast and areola hypoplasia was observed that caused the right breast to be much smaller than the left (Figure). Specifically, the first patches appeared when our patient was 4 years of age, whereas the breast hypoplasia only became noticeable at about 13 to 14 years of age. Comparative ultrasound of the mammary region confirmed the clinical result of evident gland hypotrophy. Histology of the skin lesions confirmed the diagnosis of BN. The following hormone examinations were all within normal values: dehydroepiandosterone, cortisolemia, adrenocorticotropic hormone, sex hormone–binding globulin, androstenedione, prolactin, free testosterone, dihydrotestosterone, 3-α androstenediol, follicule-stimulating hormone, luteinizing hormone, and thyroid hormones. In addition, radiography of the entire spine and upper limbs did not show any alterations. Surgical correction of the hypoplasia was performed several months later with an additive

ipsilateral mastoplasty on the right side, which produced a satisfactory aesthetic result.

Comment

We thought it would be interesting to report this case, not only for its rare clinical manifestations but also to reassess the concept of epidermal nevus. In 1949, Samuel Becker was the first to report a melanosis associated with hypertrichosis with a nevus unius lateris–type distribution, later called *Becker's nevus*.[6] The diagnosis is mainly clinical, but it can be confirmed by histology because of the bulge of acanthosis, papillomatosis without nevus theca, and a slight increase of melanocytes in the basal layer. There have been a series of disorders associated with this type of neviform that are specifically characterized by their loco-regional correspondence with the nevus itself.[7-9] These connections did not receive attention until 1995, when Happle defined the term *pigmented hair epidermal nevus syndrome*.[10] Later, Happle and Koopman,[5] reexamined 23 case histories published over 30 years and coined the term *Becker's nevus syndrome*, correlating the skin disorders with those of the organ. This definition acknowledges the independence of this clinical entity from the more general definition of *epidermal nevus syndrome*. In Happle's opinion, the definition of epidermal nevus syndrome is inaccurate and cannot be referred to any specific clinical entity.[11] At least 5 different forms of nevus syndromes can be distinguished according to the following genetic, histopathologic, and clinical criteria[10]: Schimmelpenning, nevus comedonicus, BN, Proteus, and CHILD syndrome. However, according to Happle, frequent and benign manifestations of the cutaneous form of BN demand the research of other abnormalities, especially at a skeletal level, because these abnormalities are often observed in the epidermal nevus syndromes.

The regional association of a mammary hypoplasia with a nevus is typical of this syndrome. In fact, 11 cases have been reported in the literature[2,12-15] in which the onset of BN in the mammary area during prepubertal age was followed by breast hypoplasia. For this reason, the androgen skin receptors were examined, and the results revealed high levels of these receptors in the affected skin.[14] This explains the mammary hypoplasia of the specific cases, as well as the frequent hypotrichosis, the possible presence of acneform lesions, and the thick derma to which the nevus is associated. Hence, we could hypothesize that BN is an androgen-dependent nevus that becomes evident after puberty. The distribution of this lesion does not follow the typical pattern of Blaschko lines but that of pigment disorders, for example, the systematized lentigo nevus[10,16] (checkerboard pattern). In Happle's opinion, the BN's congenital and familial cases[1,14] confirm the hereditary model as the predominant one. Therefore, during embryogenesis, heterozygote healthy individuals would undergo the somatic mutation of a cell clone (which has lost heterozygosity) where the hamartomatous area presumably originates. This hypothesis of paradominant heredity also supports the necessary loco-regional correspondence between BN and associated disorders that may commonly originate from a primitive postzygotic mutation responsible for the mosaic resemblance.

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