Linear Congenital Becker Nevus

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Becker nevus is an acquired disorder that usually manifests in late childhood or adolescence as a hyperpigmented hypertrichotic patch usually located on the upper trunk or proximal upper extremities. Only a few cases of congenital and familial Becker nevus have been described. Although the lesions may have various shapes, they consistently have a geographic or blocklike configuration in an irregular fashion; a linear pattern has rarely been reported. We describe a case of linear congenial Becker nevus following Blaschko line that appeared at birth on the right shoulder, with hypertrichosis developing 4 years later.

Cutis. 2005;75:122-124.

In 1949, Becker nevus was first described as a "concurrent melanosis and hypertrichosis in the distribution of nevus unius lateralis."[1] Becker nevus typically appears on the upper half of the trunk or on the proximal upper extremities as a circumscribed hyperpigmented patch with an irregular, gradually enlarging outline with associated hypertrichosis that develops several years later.[2] Although the lesion usually appears in late childhood or adolescence, limited cases of congenital[3-7] and familial[7-11] Becker nevus have been described. In most instances, the lesions have a geographic or blocklike configuration in an irregular fashion.[2,12] To the best of our knowledge, linear congenital Becker nevus has rarely been reported. We describe this uncommon pattern of Becker nevus, with special reference to Blaschko line.

Case Report

An 8-year-old boy presented with 2 asymptomatic, hyperpigmented, linear patches on his shoulder that had been present since birth. The area had increased in size, and the surface had slowly become verrucous. At 3 years of age, the boy was seen in our department, and the lesion was clinically diagnosed as linear verrucous nevus. One year later, hypertrichosis became noticeable in the area of hyperpigmentation; thereafter, the intensity of the pigmentation and the verrucosity began to decrease.

Results of a physical examination revealed 2 linear, slightly brownish, verrucous patches with hypertrichosis. The main lesion measured approximately 0.9×11.0 cm, and the satellite lesion measured 0.5×1.0 cm on the right shoulder (Figure). Results of laboratory investigations, including complete blood count, blood biochemistry tests, and urinalysis, were normal. Histologic examination results showed mild hyperkeratosis, acanthosis, elongation and clubbing of rete ridges, and increased pigmentation of the basal cell layer; however, there were no nevus cells and no increase in smooth muscle bundles in the dermis. These findings were consistent with Becker nevus.

Comment

Blaschko lines define the distribution pattern of many congenital and acquired skin diseases.[12,13] The question of whether Becker nevus follows Blaschko lines remains controversial. Jackson[13] reviewed the literature in 1976 and listed Becker nevus as one of the nevoid skin diseases that follows Blaschko lines. Bolognia et al,[12] however, insisted that among the lesions listed by Jackson, Becker nevus and linear scleroderma do not follow Blaschko lines. Bolognia and collegues[12] stated that Becker nevus has a consistent blocklike configuration and that they were unable to find a case with a linear scleroderma for which distribution was along the Blaschko line. However, there are increasing reports of linear scleroderma following Blaschko lines.[14,15] Some dermatoses that usually present with a block configuration have sometimes been described with a linear configuration. These include segmental neurofibromatosis and segmental tuberous sclerosis arising from somatic mutation, where the abnormal phenotypic expression occurs only in the affected segment of the body.[16,17]

Accepted for publication May 5, 2004.

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The authors report no conflict of interest.

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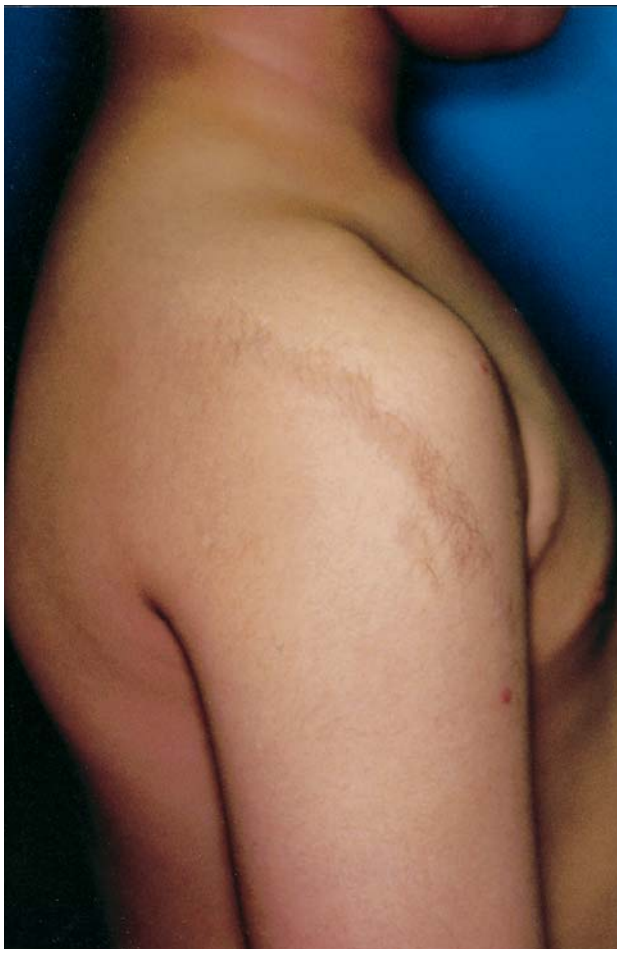

Two linear, brownish verrucous plaques with hypertrichosis on the right shoulder.

The mutant cell clones are left with a selective growth advantage and may expand beyond the linear Blaschko segment, showing a block or linear configuration.

The exact nature of Blaschko lines is unknown; however, it is generally believed that they represent the pattern of embryonic migration of skin cells and can often be produced by genetic mosaicism.[18,19] Becker nevus has been found to be associated with chromosomal mosaicism.[20] In addition to Blaschko line, mosaicism may produce cutaneous patterns such as checkerboard, phylloid, and patch without midline separation.[19] Recently, Khaitan et al[21] reported a case of multiple Becker nevus with 7 distinct lesions in checkerboard patterns characterized by alternating squares of hyperpigmentation with a sharp midline separation.

Several authors have reported cases of Becker nevus associated with other anomalies, such as unilateral hypoplasia of the breast or scoliosis.[2,22-24] Lucky et al[25] suggested that Becker nevus might be part of the spectrum of epidermal nevus because both have similar skeletal anomalies. Happle and Koopman[24] proposed the term *Becker nevus syndrome*, which defines a phenotype characterized by the presence of a particular type of organoid epithelial nevus showing hyperpigmentation; increased hairiness and hamartomatous augmentation of smooth muscle fibers; and other developmental defects such as ipsilateral hypoplasia of breast and skeletal anomalies, including scoliosis, spina bifida occulta, or ipsilateral hypoplasia of a limb. This remarkable regional correspondence between Becker nevus and associated anomalies is believed to indicate a common origin from an early postzygotic mutational event, giving rise to mosaicism.[23,24]

Chima et al[2] suggested that androgen stimulation may play a role in the pathogenesis of Becker nevus because of the condition's common occurrence in peripubertal boys and the occurrence of hypertrichosis. In one study, the androgen receptor level in the skin lesion present in Becker nevus was markedly increased compared with contralateral normal skin.[26] The gene encoding the androgen receptor is on the X chromosome and has been found to be subject to random inactivation (lyonization),[27,28] which has been proposed to be the mechanism behind the X-linked disorders occurring along Blaschko lines.[29]

In conclusion, our case provides the possible genetic basis of Becker nevus because it occurred at birth as a linear lesion following the Blaschko line. However, considering the fact that most cases of Becker nevus occur during late childhood or adolescence, it is highly likely that the tendency for development of Becker nevus is predetermined during embryogenesis with the formation of a clone of vulnerable cells. Appropriate triggers such as hormonal change or sunlight exposure may result in the occurrence of a lesion of Becker nevus. Becker[1] originally noted a history of sunburn prior to Becker nevus in his 2 cases. Others also have described sun exposure before the appearance of the lesion.[20,30,31] Hynes and Shenefelt[32] reported a similar case of unilateral nevoid telangiectasia that may have resulted from a localized increase in estrogen levels caused by a chromosomal mosaicism that is unmasked at times of relative estrogen excess.

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