

A Rare Cause of Breast Asymmetry: Becker's Nevus Syndrome with Two Cases

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Abstract

Observation: Becker Nevus, characterized by hyperpigmented macules and patches which can go along with hypertrichosis. Becker nevus is usually unilaterally located on the chest wall, back, shoulder, and, upper arm. Becker nevus occasionally co-exists with abnormalities of muscular, skeletal, skin, soft tissue and breast hypoplasia. We reported two girls with early diagnosis with Becker nevus syndrome.

Introduction

Becker Nevus, characterized by irregularly bordered, unilaterally localized, hyperpigmented macules and patches which can go along with hypertrichosis, is a kind of hamartomatous lesion that predominantly affects males. Although Becker nevus is a congenital disorder, it is more conspicuous during puberty owing to its androgen dependency. Becker nevus is usually located on the chest wall, back, shoulder, and, upper arm. Becker nevus occasionally co-exists with abnormalities of muscular, skeletal, skin, soft tissue and breast hypoplasia. When Becker nevus is presented with one of the abnormalities, it is called Becker nevus syndrome. In contrast with Becker nevus syndrome observed in adults, clinical findings in children, together with comorbidities and features of lesions, are different.

We have reported two girls with early diagnosis with Becker nevus syndrome, one of

whom has a lesion on her face in which is poorly documented place, and the other has a little lesion whose diagnosis is proven pathologically. Becker nevus syndrome must be considered in differential diagnosis in the case of combination of one or more Becker nevi with ipsilateral breast hypoplasia. We have aimed to discuss these cases accompanying them with the literature.

Case Reports

Case 1: The 10 year-old-girl patient was presented to our department with a history of increased number of hairs on the right side of her face by birth. She stated that the spot on her face has existed since 3 years old, and there has been increase in the number of hairs recently. Her past medical history was unremarkable. There was no family history with a similar lesion. In the physical examination, it was clear to see the mild facial asymmetry, the brown colored patch and hypertrichosis on the right side of her face. Her body



Figure 1. Brown colored, hypertrichosis over the patch on the right side of the face and right breast hypoplasia

length was 127 cm (3rd-10thp), her weight was 24 kg (3rd-10thp), and her bone age was 9. In addition, she had right breast Tanner Stage 1 and left breast Tanner Stage 3 (**Figure 1**). In her laboratory examination, routine blood and urine analyses of her were normal. Endocrine tests performed together with the previous tests, adrenocorticotrophic hormone (ACTH): 28 pg/ml, dehydroepiandrosterone sulfate (DHEA-S); 30 µg/ml, testosterone; 6 ng/dl, estradiol; 34 pg/ml, follicle-stimulating hormone (FSH); 0,55 mIU/ml, luteinizing hormone (LH); 0,35 mIU/ml, prolactin; 9 ng/ml levels were normal regarding her age and pubertal development.

During her pelvic ultrasound, her uterine and ovarian dimensions were normal for her age and pubertal status. In breast ultrasound, there was not any breast tissue on the right. A biopsy wasn't conducted owing to localization of the present lesion. This condition was named Becker nevus syndrome on account of the current lesion and ipsilateral breast hypoplasia. There was not a skeletal deformity in the assessment in terms of additional comorbidities.

Case 2: The 11 year-old-girl was referred to have no breast development. At anamnesis, she expressed that her left breast did not grow although her



Figure 2. An irregular outlined brown colored patches on the left scapular region

right breast had grown for 2 years. Her past medical history was unremarkable. There was no family history of a similar lesion. In her physical examination, her body height was 132 cm (3rd-10thp), weight was 26 kg (3rd-10thp), and her bone age was 10. There was an irregular-bordered brown patch which has approximately 2 cm in diameter on her left scapular region. She had left breast Tanner Stage 1 and right breast Tanner Stage 4 (**Figure 2**). In her laboratory examination, routine blood and urine analyses of her were normal. Endocrine tests, conducted together with these tests, (ACTH: 18pg/ml, DHEA-S: 50µg/ml, testosterone: 6ng/dl, estradiol: 41pg/ml, FSH: 2,3mIU/ml, LH: 1,2 mIU/ml, prolactin:11ng/dl) were normal regarding her age and pubertal development. During her pelvic ultrasound, her uterine and ovarian dimensions were normal for her age and pubertal status. In breast ultrasound, there was not any breast tissue on the left. The biopsy of the pigmented lesion on the left scapula showed acanthosis and hyperpigmentation of the basal layer. These histopathological features supported the diagnosis.

Both patients were diagnosed with Becker nevus syndrome owing to their lesions and ipsilateral

breast hypoplasia. There was not a skeletal deformity in the assessment in terms of additional comorbidities.

Discussion

This form of nevus was first described in 1949 by *Samuel Becker*. Though Becker nevus is common, it was reported that it occurred rarely in children. The reason why it appeared rarely is that it may have been overlooked, misdiagnosed or not well-known [1, 2, 3, 4, 5, 6, 7, 8]. Two cases of the close ages, who were diagnosed with Becker nevus syndrome, were discussed clinically and histologically in the light of literature.

Becker nevus usually appears in the first or second decade. Other related lesions are frequently hypoplasia. During puberty, development of breast, nipple and areola of both genders depends on estrogen [9]. In Becker nevus syndrome, even though the formation of the breast hypoplasia problems is considered to have resulted from the increased androgen receptor sensitivity's counterbalance of the estrogen effect in its pathogenesis, this hypothesis can not explain the existing mechanism for musculoskeletal abnormalities [1, 2, 3, 4, 5]. According to a large-scale study of 118 children, androgenic stimulation is not suggested to have played a pathogenic role in girls with Becker nevus contrary to the conditions in adults, because most of the patients were under 6 years old [4].

Panizzon et al. described 3 types of BN with regard to clinical findings which are melanocytic, hypertrichotic and mix types. Particularly, melanocytic type is characterized by hyperpigmentation or vellus, and its diagnosis is not generally easy [11]. When our cases were evaluated in terms of this, they were hypertrichotic and melanocytic respectively. The first case was not histologically diagnosed due to the lesion region, while there were clinically diagnosable cases in the medical literature [1, 2, 3, 4].

In the event of unusual lesion region like the face, there may be difficulties with diagnosis in the case of lacking of hypertrichosis and unremarkable hypermelanosis. Becker nevus is usually single and a sharply outlined hypochromic brownish hairy patches located on the shoulder, anterior trunk or scapular

region. This type of nevus has a geometric configuration and tendency to be permanent and asymmetric. It often grows slowly and irregularly. A face has been reported to be the rare place where Becker nevus is seen in the literature [4, 5, 7]. Our first case has a hairy lesion on her face.

This nevus predominantly affects males (2-5:1) and occurs during pre-pubertal period. Reports of this condition in women (1,5:1) are much more common due to the easy diagnosis of breast hypoplasia. It has been reported that the nevus has been seen higher in adults, whereas it has not been found much in children [2, 3, 4]. Our both cases of the two girls support these recent researches.

In 1997, *Koopman and Happle* reviewed 23 cases in which Becker nevus was associated with breast hypoplasia and other muscle, skin or bone changes ipsilateral to the nevus, which they called Becker nevus syndrome [2]. This syndrome is associated with Becker nevus with breast hypoplasia and other anomalies. The most frequent ones are lumbar spinal bifida, thoracic scoliosis and pectus carinatum. The most rarely seen ones are anomalies along with Becker nevus, including connective tissue nevus, aplasia of pectorals major muscle, ipsilateral limb shortening, localized lipoatrophy, spina bifida, scoliosis, pectus carinatum, congenital adrenal hyperplasia, and an accessory scrotum [2, 3, 4, 5, 7, 9]. Both two cases are evaluated by orthopedists through X ray in terms of orthopedic problems with which they may be accompanied.

Although Becker nevus usually appears in pubertal period, a few congenital cases have been reported be found in the other periods. Becker nevus, whose etiopathogenesis is not known, is accepted as a hamartoma of ectomesodermal tissues, and this is seen sporadically. Besides, autosomal dominant inheritance with incomplete penetrance has been documented [4, 5, 9]. Family cases of patients' siblings, uncles, and cousins endorse the transmission of an autosomal dominant trait [1, 2, 13]. When we had carried out a pedigree analysis, there were no cases with similar eruption or problem of breast development. Hence, we thought that our cases are sporadic.

Acquired Becker nevus is easy to be diagnosed due to its clinical manifestation. In addition to this, a Becker nevus with congenital, hairless, hyperpigmented may be difficult to be sorted out from a giant cafe au late macule and some big congenital pale melanocytic nevus in the differential diagnosis. Thus, the diagnosis of some cases may be confirmed just by using dermatoscope and histopathology [10, 12]. A congenital Becker nevus can occasionally be clinically indistinguishable from a congenital melanocytic nevus. A Becker nevus does not have nevo cellular nevus cells in its histologic examination. Another required differential diagnosis situation is McCune Albright Syndrome which has pigmented macule by birth [10, 11, 12]. In our first case, a biopsy was not conducted due to the lesion region; our second case was histologically diagnosed with Becker nevus by biopsy.

As a consequence, our cases are presented to highlight the fact that Becker nevus syndrome must be considered as a differential diagnosis in patients with breast asymmetry who have breast hypoplasia and ipsilateral patches like our cases, this report emphasizes that patients may have mild clinical signs in the admission.

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