

CASE REPORTS

Becker Nevus Syndrome

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Abstract. Becker nevus is a hyperpigmented hamartoma with an irregular outline and often hairy. It is normally found on the shoulders and chest, although it can appear in other areas. Becker nevus is sometimes associated with other muscular, skeletal, or cutaneous abnormalities such as ipsilateral breast hypoplasia or scoliosis. This characteristic phenotype of Becker nevus associated with unilateral breast hypoplasia or other abnormalities is referred to as Becker nevus syndrome. Although the lesions usually become apparent during adolescence, they are present from birth and represent part of the spectrum of so-called epidermal nevus syndromes. We present 4 cases of Becker nevus syndrome in which Becker nevus was associated with ipsilateral breast hypoplasia and, less consistently, other abnormalities.

Key words: Becker nevus, Becker nevus syndrome, breast hypoplasia, epidermal nevus.

SÍNDROME DEL NEVO DE BECKER

Resumen. El nevo de Becker es un hamartoma hiperpigmentado, de bordes geográficos y a menudo con hipertrichosis en superficie. Sus localizaciones habituales suelen ser los hombros o el tórax, aunque puede aparecer en cualquier otra ubicación. En ocasiones el nevo de Becker se asocia a diferentes anomalías musculares, esqueléticas o cutáneas como la hipoplasia ipsilateral de la mama o la escoliosis. A este fenotipo característico de nevo de Becker asociado a hipoplasia unilateral de la mama u otras anomalías se le denomina síndrome del nevo de Becker. Aunque las lesiones suelen manifestarse en la adolescencia, éstas están presentes desde el nacimiento y constituyen una parte del espectro de los llamados síndromes de nevo epidérmico.

Presentamos 4 casos de síndrome del nevo de Becker, en el que el nevo de Becker se asoció a hipoplasia mamaria ipsilateral y, menos consistentemente, a otras anomalías.

Palabras clave: nevo de Becker, síndrome del nevo de Becker, hipoplasia mamaria, nevo epidérmico.

Introduction

Becker nevus, also called Becker melanosis or pigmented hairy epidermal nevus, is a benign cutaneous hamartoma that develops as a light or dark brown macule with well-defined but irregular borders and can present hypertrichosis.¹ Although the most common sites are the scapular region or the chest, Becker nevus can appear on any part of the body.² Histologically it is characterized by moderate acanthosis with elongation of the rete ridges and a variable degree of hyperkeratosis. The basal

layer of the epidermis is hyperpigmented, although there is no increase in the number of melanocytes. Melanophages are present in the dermis and there is an increase in the number of arrector pili muscles, making differentiation from smooth muscle hamartoma sometimes impossible.^{3,4}

Becker nevus is androgen dependent and, in consequence, although it is congenital, it is not uncommon for it to become more pronounced during adolescence, particularly in males.⁴⁻⁶ It is occasionally associated with other abnormalities, such as hypoplasia of the ipsilateral breast and the presence of muscle, skeletal, or cutaneous abnormalities. The presence of any of these abnormalities associated with Becker nevus defines a characteristic phenotype called Becker nevus syndrome.⁷ These abnormalities usually but not invariably appear on the same side of the body.^{4,7}

We present 4 patients with Becker nevus and associated abnormalities within the spectrum of what is usually found in Becker nevus syndrome.

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Case Descriptions

Case 1

A 13-year-old boy presented a lesion that appeared spontaneously at 3 years of age; the lesion was situated over the right scapular region and extended onto the right shoulder and arm. It had a hairy surface and was associated with hypoplasia of the ipsilateral breast. The patient also presented clinical and radiological signs of scoliosis (Figure 1).



Figure 1. Case 1: extensive Becker nevus and scoliosis.

Case 2

A 12-year-old boy presented a brown lesion on the anterior aspect of the chest that had been present since 6 months of age. The lesion had a hairy surface and biopsy confirmed the diagnosis of Becker nevus. Ipsilateral breast hypoplasia was detected on examination (Figure 2). Clinical and radiological examination did not reveal scoliosis. The patient also had 3 café-au-lait spots on the back.



Figure 2. Case 2: Becker nevus and ipsilateral breast hypoplasia.

Case 3

The patient was a 12-year-old boy with a past medical history of hydronephrosis secondary to bilateral ureteral stenosis who consulted for an asymptomatic brown patch situated on the right side of the chest that had appeared spontaneously over the previous 2 or 3 years. Biopsy was compatible with a diagnosis of Becker nevus. Examination revealed thinning of the right pectoralis muscle over which the nevus was situated. The patient did not present scoliosis or other skeletal abnormalities.

Case 4

The patient was a 12-year-old boy who consulted for an asymptomatic brown patch on the right side of the chest

that had appeared when he was 8 years of age. On examination, a nonhairy brown patch with irregular borders was observed. The pectoralis muscle over which the lesion was situated was thinner than the contralateral muscle. The patient had clinical signs of moderate scoliosis.

The findings of these cases are summarized in Table 1.

Table 1. Case Descriptions

Case	Sex	Age at Appearance	Localization	Hypertrichosis	Ipsilateral Breast Hypoplasia	Other Findings	Family History
1	Male	3 years	Scapular, shoulder, right arm	Yes	Yes	Scoliosis	No
2	Male	6 months	Left hemithorax	Yes	Yes	Café-au-lait spots	No
3	Male	9-10 years	Right hemithorax	No	Yes	Bilateral ureteral stenosis	No
4	Male	8 years	Right hemithorax	No	Yes	Scoliosis	No

Table 2. Clinical Spectrum of Becker Nevus Syndrome⁴

Becker nevus
Ipsilateral breast hypoplasia
Ipsilateral hypoplasia of the shoulder
Ipsilateral hypoplasia of the arm
Vertebral defects/scoliosis/spina bifida occulta
Other musculoskeletal defects:
Cervical rib/fused ribs
<i>Pectus excavatum</i>
<i>Pectus carinatum</i>
Bilateral tibial torsion
Scapular asymmetry
Hypoplasia of the sternocleidomastoid muscle
Ipsilateral dental hypoplasia and facial asymmetry
Umbilical hernia
Other skin defects:
Hypoplasia of extramammary subcutaneous tissue
Hypoplasia of the contralateral labium minus
Accessory scrotum
Sparse hair in the ipsilateral axilla
Skin hypoplasia over the temporal bone
Supernumerary nipples

Discussion

The majority of publications prior to the definitive description of Becker nevus syndrome reported the association of Becker nevus with scoliosis or unilateral breast hypoplasia. Subsequently, other associated abnormalities were added to the clinical spectrum of what became known as Becker nevus syndrome (Table 2).⁴ Although scoliosis falls within this clinical spectrum, it is not sufficient in itself to establish the definitive diagnosis.⁷ Becker nevus syndrome is included within the group of epidermal nevus syndromes, together with nevus sebaceus syndrome, nevus comedonicus syndrome, phakomatosis pigmentokeratocica, Proteus syndrome, and CHILD (congenital hemidysplasia with ichthyosiform erythroderma and limb defects) syndrome.⁸ Epidermal nevi are hamartomatous lesions derived from epidermal components; they arise from mutations in pluripotent cells during the early stages of embryogenesis.^{8,9}

Although the majority of published cases of Becker nevus syndrome are sporadic, familial grouping can be observed very rarely,¹⁰ probably due to a paradominant inheritance phenomenon.⁷ The genetic basis of Becker nevus syndrome has not been established, but it is assumed to be due to a postzygotic autosomal lethal mutation that survives in mosaic form; in fact, Becker nevus usually presents a pattern of mosaicism, either as a patch with serrated borders or, less frequently, as a segmental or “flag-like” lesion.¹¹ The association of Becker nevus with the other abnormalities

is probably due to mutations occurring earlier than those responsible for isolated Becker nevus.

Becker nevus syndrome affects males and females in equal proportions, whereas it has been reported that the male to female ratio in Becker nevus is 2 to 1; this could be related to the dependence of the lesion on androgens. The discrepancy might also be explained by the more clinically pronounced breast hypoplasia in females than in males.^{4,12} Our 4 cases were sporadic and all of them occurred in males. No further abnormalities other than those described were subsequently detected in any of the 4 cases after a follow-up of between 1 and 12 years. Finally, given that the appearance of Becker nevus-associated abnormalities in this syndrome can be relatively subtle, it is likely that many cases have passed undetected, and that the syndrome is more common than may be thought from the limited number of cases published.

Conflicts of Interest

The authors declare no conflicts of interest.

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