

# Becker nevus syndrome. Report of three cases

Paula C. Luna,<sup>1</sup> Mabel J. Nocito,<sup>2</sup> Nadia G. Cañadas,<sup>2</sup> María Laura Castellanos Posse,<sup>2</sup> Carolina Marchesi,<sup>3</sup> María Marcela Lustia,<sup>4</sup> Miguel A. Mazzini,<sup>5</sup> Graciela Carabajal<sup>6</sup>

## Abstract

Becker's nevus is a relatively frequent pigmentary disorder. Becker's nevus syndrome is the association of this characteristic nevus with other bone, muscle, or skin defects, generally ipsilateral to the lesion. This rare syndrome belongs to the epidermal nevus syndrome group. We describe three new cases of this syndrome: one male patient with ipsilateral deltoid hypertrophy, and two female patients, one with scoliosis and marked subcutaneous tissue atrophy underlying the nevus, and another with breast hypoplasia ipsilateral to the nevus, and scoliosis (*Dermatol Argent* 2008;14(5):379-382).

**Key words:** *Becker's nevus syndrome, Becker's nevus, epidermal nevus syndrome.*

## Introduction

Becker's nevus is a lesion characterized by the presence of a hyperpigmented macule with sharply demarcated and irregular margins and small peripheral islets of affected skin; in some cases, the lesion may also show hypertrichosis or an embossed surface due to an underlying smooth muscle hamartoma. Although relatively frequent, it may often go overlooked or is misdiagnosed.<sup>1</sup>

Happle and Koopman<sup>2</sup> denominated Becker's nevus syndrome (BNS) the association of Becker's nevus with breast hypoplasia or other bone, skin, or muscle alterations ipsilateral to the nevus.

We describe three new cases of this rare association (**Table 1**).

## Case series

### Case 1

An 18-year-old male patient, without relevant history, who consulted for acne. Physical examination showed, a hyperpigmented 15 × 8 cm diameter macule with sharply demarcated, irregular margins with the characteristic peripheral archipelago-like image, slight hypertrichosis, in the right pectoral area. The lesion was asymptomatic. The patient referred that the alteration began at puberty. An asymmetry appeared between both pectoral areas was also detected during the physical examination (**Figure 1**).

A biopsy from the lesion was obtained; it showed melanotic hyperpigmentation of the basal cell layer, papillomatosis, and elongation of the rete ridges and leading to the diagnosis of Becker's nevus.

To assess the origin of the asymmetry, a breast ultrasonography was obtained, resulting in: right pectoral hypertrophy (30.5

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1. Chief Residents.
2. Resident.
3. SAD-Fellowship.
4. Staff Physician.
5. Head of Department.
6. Pathologist Dermatology.

Complejo Médico Policial "Churrucá-Visca". Ciudad Autónoma de Buenos Aires, Argentina.

## Correspondence

Paula C. Luna: Arenales 2557 1º A - Autonomous City of Buenos Aires, Argentine Republic  
paulacarlinaluna@gmail.com

mm against 20 mm of left pectoral). There was no differences between breast glands, skin, or subcutaneous tissue.

### Case 2

A 39-year-old female patient without relevant history, who consulted for the presence of a 30-year-evolution hyperpigmented lesion. The lesion involved a large part of the left half of the body, from the submammary area to the knee. Characteristically, it appeared with sharply demarcated margins and peripheral involved skin islets. There were no hypertrichosis, but marked underlying tissue atrophy.

Three biopsies of the skin lesion were obtained, in order to rule out morphea. All skin samples showed the diagnosis of Becker's nevus (**Figure 2**).

The complementary studies performed showed scoliosis (**Figure 3**) in dorsolumbar spine. Comparison X-ray of both legs did not show asymmetries.

### Case 3

A 57-year-old patient, without relevant history, consulted for the presence of a long-term lesion.

The physical examination evidenced a hyperpigmented lesion with irregular margins, without hypertrichosis, beginning on the left scapular area and extended to the anterior aspect, involving the ipsilateral breast. Noteworthy was a marked mammary asymmetry, with hypoplasia ipsilateral to the pigmented lesion (**Figure 4**).

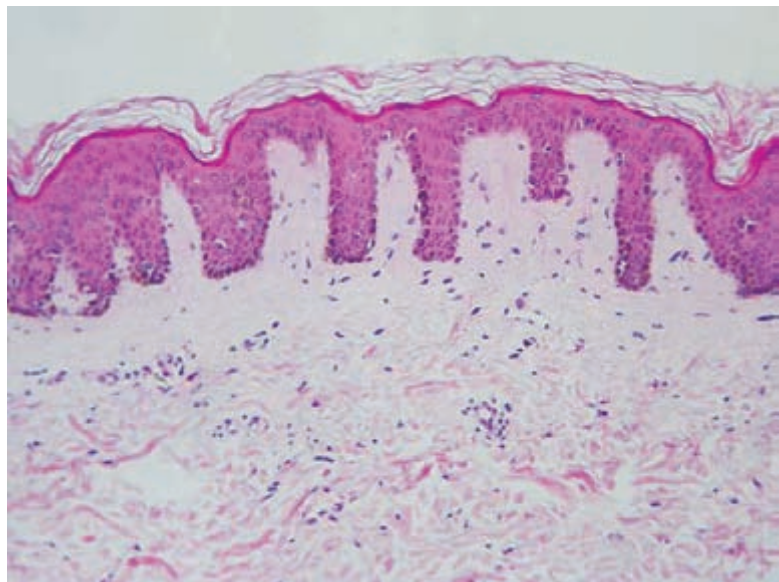
Biopsy findings were compatible with Becker's nevus. The breast ultrasonography reported left mammary hypoplasia, and the spinal X-ray, showed scoliosis.

### Comment

Becker's nevus is a hamartomatous lesion that many authors consider an organoid nevus, and thus probably reflects cutaneous mosaicism;<sup>2</sup> characterized by the presence of two or more populations of genetically different cells derived from the same zygote.<sup>3</sup> It is described as an isolated, acquired lesion, hyperpigmented lesion in a color ranging from light tan to dark brown, measuring several centimeters in diameter, with sharply demarcated but irregular margins, and small peripheral affected skin "archipelagos" surrounding the lesion. Most lesions are described as hypertrichotic, and the most typical location is on the deltoid area. Different series refer a male dominance, with a male/female ratio from 2:1 to 6:1.<sup>4,5</sup> It is androgen-dependant and thus considered a "functional" nevus. This explains a more frequent



**Figure 1.** Becker's nevus and chest asymmetry with right side dominance.



**Figure 2.** Skin biopsy showing acanthosis, papillomatosis, basal layer melanotic hyperpigmentation, and flattening of the ridge base.

diagnosis in males, given the fact they develop more hyperpigmented lesions and more hypertrichosis than females, where it may go unnoticed.<sup>6</sup> There is no consensus regarding the time of appearance. In both genders, it is usually more evident during puberty, but so far it is not well established if it is a congenital lesion that is expressed by the increase of sexual hormones, or if it is a lesion acquired during adolescence.

TABLE 1.

	Patient 1	Patient 2	Patient 3
Gender	Male	Female	Female
Age	18	39	57
Appearance	Puberty	Puberty	Puberty
Nevus location	Right deltoid	Right flank, thigh, and leg	Left scapula and breast
Alteration type	Deltoid muscle hypertrophy	Subcutaneous cell tissue hypoplasia and scoliosis	Breast hypoplasia and scoliosis



Figure 3. Spine X-ray evidencing marked scoliosis.

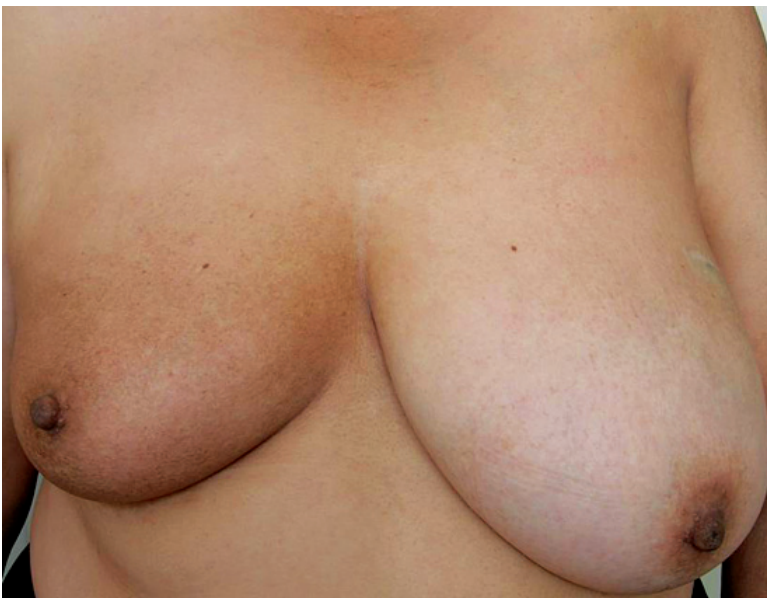


Figure 4. Hyperpigmented macula, irregular margins from scapular area to left breast, which shows atrophy.

As regards to its etiology, Happle suggested the term “paradominant” to explain its genetic base, because the occurrence of this nevus is almost exclusively sporadic, although in some opportunities it was described in different members of the same family. Generally, heterozygous patients are phenotypically normal, and the gene may go undetected for many generations. The lesion is expressed in carriers when a second somatic mutation takes place during embryogenesis and generates a mosaic plaque.<sup>7</sup>

Histopathologically, Becker’s nevus shows variable degrees of epidermal changes, slight acanthosis, elongated rete ridges with plain<sup>8</sup> quadrangular<sup>9</sup> baseline, melanotic basal hyperpigmentation, without increase of the amount of melanocytes or sheath formation,<sup>4</sup> and some melanophages in dermis. With relative frequency, there is a smooth muscle hamartoma under the lesion.<sup>5</sup> Some authors consider it a continuous spectrum from pigmented and hairy predominance to the arrector pili muscle dominance.

Becker nevus syndrome is the association of Becker’s nevus with mammary hypoplasia, scoliosis, or any other skin, muscular, or skeletal alteration.<sup>6</sup> This syndrome is included in the epidermal nevi syndrome group, together with: sebaceous nevus syndrome (Schimmelpenning), nevus comedonicus syndrome, Proteus syndrome, and CHILD (Congenital Hemidysplasia with Ichthyosiform Nevus and Limb Defects) syndrome.

Epidermal nevi are hamartomatous lesions derived from epidermal components generated by pluripotent cell mutations during embryonic stages.<sup>10</sup> In some opportunities, these nevi may be associated to different ocular neurologic, or skeletal alterations.<sup>11</sup> Each particular epidermal nevus syndrome has specific clinical features that are essential for diagnosis, as well as distinct genetic bases. Schimmelpenning’s syndrome associates an extensive sebaceous nevus, arranged following the lines of Blaschko, with brain, eyes, or skeletal alterations. Nevus comedonicus syndrome associates a nevus comedonicus lesion arranged following the skin cleavage lines with ipsilateral cataracts, skeletal defects, and electroencephalogram alterations.

Proteus syndrome associates partial hand and/or feet gigantism, macrocephaly, subcutaneous hamartomas, vascular lesions, and organoid-type epidermal nevi. CHILD syndrome associates an hemicorporal ichthyosiform inflammatory nevus with alterations of ipsilateral limbs and internal organs.

In contrast to the isolated Becker's nevus, BN syndrome is more frequently reported in females (1.5:1).<sup>5,6</sup> This difference could be explained by the fact that breast asymmetry is more notorious in females.<sup>6</sup>

Many lesions have been described in association with this nevus, such as ipsilateral mammary hypoplasia, limb shortening or asymmetry, scoliosis, and supernumerary nipples.

Usually, the simultaneous existence of these findings is explained by the androgenic stimulus of these tumors. Since estrogens are essential, for example in breast development, the androgenic dominance would locally inhibit female sexual hormones, thus preventing the normal mammary development. In fact, Jung et al.<sup>12</sup> used spironolactone in an adolescent patient with breast hypoplasia associated with Becker's nevus; evidence of improvement appeared after 4 weeks of treatment. Subcutaneous tissue and bone hypoplasias ipsilateral to the lesion are difficult to explain by this mechanism,<sup>6</sup> thus, the pathogenesis is yet unknown.

## Conclusions

Although Becker's nevus is a relatively frequent finding, the literature shows few BN syndrome reports.

A skin biopsy must be done to certify the diagnosis, because the presence of a plaque morphea with deep involvement may also be clinically expressed with associate asymmetries.

Screening for bone, muscle, and breast alterations is essential for an early (preventive or palliative) treatment of morphological defects.

So far, there are no cases of striated muscle hypertrophy associated with BN syndrome. We suggest an androgenic mechanism upon muscle receptors as possibly accounting for excessive muscular development.

## References

1. Luna P, Etcheverry M, Carabajal G, Miguez L, Mazzini MA. Liquen córneo hipertrófico sobre nevo de Becker. *Dermatol Argent* 2005; 11:222-224.
2. Happle R, Koopman RJ. Becker nevus syndrome. *Am J Med Genet* 1997; 357-361.
3. Barnhill R, Llewellyn K. Neoplasias melanocíticas benignas. In: *Dermatología Bologna*, Jorizzo, Rapini. Madrid: Elsevier Ed.; 2004. Vol 2 pp. 1759-1760.
4. Tymen R, Forestier JF, Boudtet B, Colomb D. Naevus Tardif de Becker, a propos d'une série de 100 observations. *Ann Dermatol Venereol* 1981; 108:41-46.
5. Danatri R, König A, Salhi A, Bittar M, Happle R. Becker's nevus syndrome revisited. *J Am Acad Dermatol* 2004; 51:965-969.
6. Weedon D. Alteraciones pigmentarias. In: *Weedon, Piel Patología*. Madrid: Marbán Libros; 2002. Vol 1, pp. 276-277.
7. Cabrera H, Garcia S. Nevos mixtos con nevos despigmentarios. In: *Cabrera H, Garcia S. Nevos*. Buenos Aires: Editorial Actualizaciones Médicas; 1998; 100-101.
8. Happle R. Epidermal Nevus syndromes. *Seminars in dermatology* 1995; 14:111-121.
9. Jung JH, Kim YC, Park HJ, Clinn YW. Becker's nevus with ipsilateral breast hypoplasia: improvement with spironolactone. *J Dermatol* 2003;30:154-156.
10. Happle R, Bittar M. Patrones del mosaicismo en la piel humana: comprendiendo aspectos actuales y futuros. *Dermatol Pediatr Lat* 2006; 4:171-181.
11. Vidaurri-de la Cruz H, Tamayo-Sánchez L, Durán-McKinster C, Orozco-Covarrubias ML, Ruiz-Maldonado RR. Epidermal Nevus Syndromes: Clinical Findings in 35 Patients. *Pediatric Dermatol* 2004; 21:432-439.
12. Happle R. Paradominant inheritance: a possible explanation for Becker's pigmentary hairy nevus. *Eur J Dermatol* 1992; 2:39-40.