Case Report

BECKER'S NEVUS WITH IPSILATERAL BREAST HYPOPLASIA: A CASE REPORT AND REVIEW OF LITERATURE

Abbas Rasi MD[•], Alireza Taghizadeh MD, Babak Yaghmaii MD, Roya Setareh-Shenas MD

Specific cutaneous associations in patients with Becker's nevus have been reported. We present a patient with typical clinical and histopathological features clearly consistent with Becker's nevus associated with ipsilateral breast hypoplasia. The changes were distinct and could be separated from smooth muscle hamartoma. We include clinical and histological illustrations of our case.

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Introduction

In 1949, Samuel Becker was the first to report a melanosis associated with hypertrichosis with a nevus unius lateristype distribution, later called Becker's nevus.¹ This nevus is also called Becker's pigmented hairy melanosis. Although the lesion is called a nevus, there are no nevomelanocytic structures found. The diagnosis is mainly clinical, but it can be confirmed by histology.

Case Report

An 18-year-old girl presented to our dermatology section with a 15-year history of an irregular expanding patch of hypermelanosis with hypertrichosis over her chest wall on the left side associated with breast atrophy of the same side, but normal underlying pectoralis muscle, areola, and nipple of the hypoplastic breast. According to the patient's mother, the first pigmented patch appeared when she was 3 years old, whereas the breast hypoplasia only became noticeable at about 13 years of age.

Her medical history was not remarkable. No family members reported similar skin lesions or any other cutaneous or systemic disorders. Generally, the patient was in good health, and a review of systems revealed no abnormalities.

Physical examination revealed the presence of a solitary well-defined, nonpruritic, segmental, unilateral, uniform, and light-brown patch showing hyperpigmentation and hypertrichosis on her left chest and shoulder area with a mean surface area of 600 cm² (20 × 30 cm) (Figures 1 and 2). The patch was sharply, but irregularly, demarcated.



Figure 1. Hypermelanotic patch with breast mass hypoplasia are seen (left side). Note that nipple and areola are normal.

Authors' affiliation: Department of Dermatology, Iran University of Medical Sciences, Tehran, Iran.

[•]Corresponding author and reprints: Abbas Rasi MD, Department of Dermatology, Rasoul-e-Akram Hospital, Niayesh St., Sattarkhan Ave., Iran University of Medical Sciences, Tehran, Iran. Tel: +98-21-66516071-6, E-mail: dr_rasi2002@ yahoo.com. Accepted for publication: 16 January 2005



Figure 2. A hypermelanotic patch with well-defined and irregular border is seen.

There were no pustules, vesicles, perifollicular papular elevations, or indurations. The lesion did not show transient elevations on rubbing. Over the time, hypertrichosis gradually developed within it. There was no associated hypoplasia of underlying structures and asymmetry of the limbs.

The patient's blood pressure was 110/80 mmHg. Radiographic studies of the entire spine and upper limbs did not show any alterations. The results of routine blood tests and urine analysis were within normal limits. The following hormone examinations were all within normal values: free dihydrotestosterone, dehvdrotestosterone, epiandrosterone, prolactin, follicular stimulating hormone, luteinizing hormone, and thyroid hormones. A biopsy specimen of the left chest lesion showed: hyperkeratosis, pigmented increased basal laver pigmentation, regular elongation, and clubbing of rete ridges (elongated



Figure 3. A smooth muscle bundle independent of the hair follicle is seen (arrow).

rete ridges had flat tips). In the dermis, there was not any marked increase in smooth muscle fibers, but only one bundle of pilar smooth muscles was present, and no nevus cells were observed (Figure 3). Examination with trichrome stain demonstrated no increase in smooth muscle fibers.

On the basis of clinical and histopathologic findings, a diagnosis of Becker's nevus associated with ipsilateral breast hypoplasia was made.

Discussion

Becker's nevus_is a unilateral (rarely bilateral) solitary, macular light brown hyperpigmentation of varying size, with geographic borders. The border is irregular and sharply demarcated. Occasionally, lesions resembling acne vulgaris, including papules, pustules, and cystic nodules, may occur within the affected area.² Over time, hypertrichosis (which is a characteristic feature of the lesion), develops within the lesion in about 50% of cases.³ Hairs, which are confined to the hyperpigmented area, are darker and coarser than normal. The lesion varies in size and may cover the entire upper arm or shoulder. A typical lesion is about 125 cm², although lesions up to 500 cm² have been reported. These lesions are usually asymptomatic and are usually localized on the shoulder, anterior chest, or upper arms, but there have been reports in other areas (e.g., proximal upper extremities, lower extremities, face, forearm, wrist, neck, and upper and lower back).²

In a study of 19,302 men aged 17 to 26 years, a prevalence of 0.52% was observed.³ Reports of this condition in women are much rarer in the literature. The male-to-female ratio, approximated from case studies, has been reported to be anywhere from 4:1 to 6:1.^{4, 5} There is no racial predilection, but it is more common in young people with fair skin.⁶ Becker's nevus is usually acquired in adolescence (the majority of cases are first noticed sharply before, at, or after puberty), but a congenital onset has been recorded,⁷ as have familial cases in siblings, and in an uncle and a nephew.

The lesion is a developmental anomaly, but occasionally, lesions have been said to follow severe sunburn. Lesional tissue has been found to have an increased level of androgen receptors, suggesting that heightened local androgen sensitivity may result in the hypertrichosis.

Many abnormalities have been associated with

Becker's nevus such as:

- Smooth muscle hamartoma. This is the most frequent finding. In such cases, the area of Becker's nevus may exhibit slight perifollicular papular elevations or slight induration.⁸
- Unilateral breast hypoplasia. This is possibly as a result of enhanced androgen sensitivity.^{2,9}
- Hypoplasia of underlying structures and asymmetry of limbs.¹⁰

Other abnormalities include: various skeletal malformations, connective tissue nevus,⁹ aplasia of ipsilateral pectoralis major muscle, ipsilateral limb shortening, localized lipoatrophy, spina bifida, scoliosis pectus carinatum, congenital adrenal hyperplasia, and accessory scrotum.²

Unless associated with a hamartomatous process, histologic changes in Becker's nevus are minimal. The epidermal changes are variable, but usually there is hyperkeratosis, subtle acanthosis, papillomatosis,⁸ and basal laver hyperpigmentation. There are regular elongation of rete ridges, and elongated rete ridges tend to have flat, rather than pointed tips. Melanocyte proliferation is usually mild and not always obvious in routine sections; this increase is particularly evident when melanocytes are stained for dopa-oxidase activity in both involved and uninvolved skin nearby.⁸ The dermis is thickened and contains numerous, but often inconspicuous bundles of smooth muscle fibers, unrelated to hair follicles or blood vessels.⁸ Controversy exists about the relationship of these cases to smooth muscle hamartoma. Some authors believe that, in cases associated with smooth muscle hamartoma, irregularly arranged, thick bundles of smooth muscle are present in the dermis.⁸ There may also be an underlying increase in the number and size of hair follicles and sebaceous glands.¹¹ Some melanophages are seen in the dermis.¹ There should be no problem in making the diagnosis in a well-developed Becker's nevus. In early lesions, the age of onset and, generally, the site are perhaps the most helpful diagnostic features.

Clinically, congenital Becker's nevus may be confused with a congenital melanocytic nevus. Histologically, Becker's nevus does not have nevocellular nevus cells.

Albright's syndrome may be easily confused with Becker's nevus, but the pigmented macule in Albright's syndrome is present at birth.

Theoretically, Becker's nevus and smooth

muscle hamartoma are two quite separate entities. In fact, there are cases which could be considered as intermediate. As a matter of fact, a slight underlying smooth muscle hyperplasia can be seen in Becker's nevus; on the other hand, hypermelanosis of basal layer and hypertrichosis may be encountered in smooth muscle hamartoma. Both conditions can be considered polar forms of a spectrum of dermal smooth muscle hyperplasia.¹²,

¹³ Becker's nevus may enlarge slowly a year or two after presentation and then stabilizes and appears to persist indefinitely. The hyperpigmentation usually remains stable, although there have been reports of fading over many years.^{4, 14} Becker's nevus is usually too large to remove and is best left untouched. The hair may be shaved or permanently removed. The skin hyperpigmentation may respond to therapy with Q-switched ruby laser, although the results are unpredictable and recurrences are common.

Although Becker's nevus is most common in males, ipsilateral breast hypoplasia in Becker's nevus occurs infrequently. Cases associated with breast hypoplasia are more frequently reported in women. In fact, 13 cases have been reported in the literature in whom the onset of Becker's nevus in the mammary area during the prepubertal age was followed by breast hypoplasia.^{8, 9, 15 - 17} For this reason, the androgen skin receptors were examined, and the results revealed high levels of these receptors in the affected skin.⁹ This explains the mammary hypoplasia of the specific cases, as well as the frequent hypertrichosis, the possible presence of acneiform lesions, and the thick dermis with which the nevus is associated. We thought it would be interesting to report this case, not only because of the rare association of breast hypoplasia with Becker's nevus, but due to this case's early age of onset and the large size of the lesion.

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