CASE REPORT

Becker's nevus syndrome with bilateral skin involvement


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A R T I C L E   I N F O

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A B S T R A C T

Becker's nevus syndrome is a rare disorder characterized by Becker's nevus associated with developmental anomalies, such as hypoplasia of the ipsilateral breast or other cutaneous, muscular, or skeletal defects. We present a rare case of Becker's nevus syndrome with bilateral skin involvement but the associated anomalies, hypoplasia of the whole right upper limb and ipsilateral breast, remained localized to the more severe body side only.

Introduction

Becker's nevus, also known as Becker's melanosis or pigmented hairy epidermal nevus, was first reported by Becker in 1949. It is characterized by a light or dark brown patch with a sharply outlined but irregular border and always shows hypertrichosis after puberty in male patients. Becker's nevus may sometimes be associated with developmental anomalies, including hypoplasia of the ipsilateral breast, other cutaneous or muscular abnormalities, and skeletal defects. This phenotype is now described as Becker's nevus syndrome. Here, we describe a patient with Becker's nevus syndrome with bilateral skin involvement as follows.

Case Report

An 18-year-old man was admitted to our hospital presenting with brownish hairy macula on the right scapular, lateral back region (Figure 1A), prothorax, upper limb, thigh (Figure 1B), and rump (Figure 1C) together with the posterior left upper arm (Figure 1D). The lesion started when he was 3 years old, increasing proportionally in size with the growth of the child. Hypoplasia of the whole right upper limb and ipsilateral breast were also observed (Figure 1B). There was no family history of similar disorders. Physical examination revealed neither impaired sensation of skin lesions nor enlarged cubital nerve. Laboratory examination showed no abnormality in routine tests. Magnetic resonance imaging (MRI) of the right upper limb showed congenital dislocation of the capitulum radius. A skin biopsy specimen obtained from the patient's forearm revealed slight acanthosis, hyperpigmentation of the basal layer without melanocytes increasing in number, and a superficial and sparse perivascular lymphotohistiocytic infiltrate in the dermis (Figure 2A and B). Based on the clinical and histopathologic characters, a diagnosis of Becker's nevus syndrome was formed. The patient was being followed without any special treatment.

Discussion

Becker's nevus syndrome is included within the group of epidermal nevus syndromes, together with nevus sebaceous syndrome, nevus comedonicus syndrome, phakomatosis pigmentokeratotica, Proteus syndrome, and CHILD (congenital hemidysplasia with ichthyosiform erythroderma and limb defects) syndrome. Becker's nevus often presents as a unilateral, asymptomatic, and irregular tan-brown patch. The associated anomalies tend to be a regional correspondence to the nevus and are mostly ipsilateral. Various associated developmental abnormalities or structural defects have been described along with Becker's nevus, including ipsilateral breast hypoplasia, supernumerary nipples, short limb or other forms of limb asymmetry, scoliosis, hemi-vertebrae, cleft vertebralae, spina bifida occulta, pectus excavatum, and patchy hypoplasia, etc.

The etiology of Becker's nevus remains uncertain. There are two main hypotheses concerning this disorder. First, the majority of cases are always sporadic; familial grouping is very rare. Its genetic basis is assumed to be due to a postzygotic autosomal lethal mutation that survives in a mosaic pattern. Second, the hypothesis of
a hormone-dependent disorder based on the increase in the number of androgen receptors in the affected areas is also postulated, which would explain the appearance of lesions in puberty and alterations such as hypertrichosis and acneiform eruptions restricted to the affected regions.\(^5\)

The therapeutic indication for the nevus lesion is essentially cosmetic, and to date the most effective treatment has not been well defined. Two recent individual case reports describe the response to treatment with fractionate laser, without presenting side effects. However, this therapeutic modality still needs to be re-evaluated in the clinic and more research is needed to confirm it.\(^5\)

Hoon et al presented a case of Becker's nevus with ipsilateral breast hypoplasia: an improvement occurred following 4-week treatment with spironolactone.\(^6\)

In summary, we report a rare case of Becker's nevus syndrome with bilateral skin involvement, but the associated anomalies, hypoplasia of the whole right upper limb and ipsilateral breast, remained localized to the more severe body side only, which emphasizes the polymorphic spectrum of involvement of this condition.

Figure 1 (A,B) Brownish macula in the right upper limb and hypoplasia of the whole right upper limb and ipsilateral breast. (C) Brownish macula in the right rump and (D) posterior left upper arm.

Figure 2 (A) Histology compatible with Becker's nevus (hematoxylin–eosin stain, original magnification 100×). (B) Hyperpigmentation of the basal layer (hematoxylin–eosin stain, original magnification 400×).
References