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Case Report

Becker's nevus syndrome

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Introduction

Becker's nevus (BN) was first described by Becker in 1949.¹ BN typically presents as hyperpigmented patch with irregular borders that gradually enlarges for few years and then remains stable. Hypertrichosis within lesion is common but not universal. Happle in 1997, described Becker's nevus syndrome (BNS) or hairy epidermal nevus syndrome, where BN was associated with multiple cutaneous and musculoskeletal abnormalities, mostly ipsilateral.²

In Armed Forces, a number of candidates for recruitment are referred for medical fitness regarding BN. The associated abnormalities, which may cause physical disablement if not looked for specifically, may be missed.

Case report

30 years old male patient presented with history of gradual loss of girth of right leg for about 2 years and gradually progressive difficulty in squatting for about 8 months. He gave history of fasciculations in affected muscles. There was no history of injury to right knee or lower back. He did not complain of pain, paraesthesia or numbness of affected limb or any symptoms suggestive of leprosy. Individual was diagnosed as a case of motor neuron disease (anterior horn disease) nearly 12 months after the onset of symptoms and was under follow up.

General examination was within normal limits. Examination of musculoskeletal system revealed wasting of right leg muscles (Fig. 1A) with reduced tone. Girth over most bulky part of right leg muscles was reduced by 6.5 cm as compared to left leg. There was restriction of dorsiflexion of right ankle joint beyond initial range, which was because of stiff right tendo Achilles, as it could not be stretched fully as compared to the left. This caused inability to squat fully (Fig. 1B). Power of dorsiflexors in the range of mobility; and planter flexors was IV/V. Ankle jerk was absent on the affected side. There was no sensory loss. There was scoliosis of thoracolumbar spine, slight drooping of right shoulder, and wasting of right pectoral muscles (Fig. 1C).

Dermatological examination revealed 11 cm × 4 cm faintly hyperpigmented macule with irregular margins, over left interscapular region. The patch did not have coarse hair or acneiform lesions. Supernumerary nipple was present in left inframammary region (Fig. 1C). There was no hypesthetic or hypopigmented patch or peripheral nerve thickening.

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Fig. 1 – Clinical features. (A) Hypoplasia right leg with reduced girth, (B) Incomplete squatting due to restricted dorsiflexion right foot, (C) Wasting right pectoral muscles, supernumerary nipple in left inframammary region.

Radiograph of dorsolumbar spine showed scoliosis with convexity towards right of lumbar and towards left of dorsal spine and spina bifida at the level of S1 (Fig. 2A, B). Skin biopsy from hyperpigmented macule showed increased melanocytes in basal cell layer, mild pigment incontinence and mild perivascular lymphonuclear infiltrate. Rete ridges though tended to be regular, were not elongated. Smooth muscle fibres were unremarkable. Nerve conduction study was normal. EMG showed no evidence of spontaneous activity in muscles of right leg. Hormonal assay for androgens was within normal limits.

A case of non archetype Becker's nevus, with contralateral features of hypoplasia of right leg with shortening of tendoachillis, hypoplasia of right pectoral muscles and ipsilateral accessory nipple on left side, scoliosis of dorsolumbar spine and spina bifida occulta, case was thus having multiple features of BNS.

Discussion

BNS is a genetic hamartomatus disorder having BN with associated cutaneous and musculoskeletal abnormalities

(Table 1).^{3–6} The prevalence of BN in men is estimated to be 0.25%–4.2% in various countries. The male to female ratio for BN is 2:1 to 6:1.^{2–4} BNS is however, more frequently reported in females (1.5:1). The variations can be explained by absence of hypertrichosis and easily noticeable breast hypoplasia in females.^{3,4,7} Autosomal dominant inheritance with incomplete penetrance and variable expressivity was previously assumed, however, sporadic occurrence of BN is better explained by paradominant mode of inheritance, wherein mutation in early stage of embryogenesis results in mosaic population of hemizygous or homozygous cells in otherwise heterozygous individual.^{2,3} BN is considered androgen dependent based on onset at adolescence and associated features of adrenal hyperplasia, accessory scrotum, and hypertrichosis and improvement of breast hypoplasia with spironolactone.^{4,8,9}

Our patient had multiple abnormalities as mentioned earlier. Unusual features being, contrary to mostly ipsilateral involvement, hypoplasia of pectoral and leg muscles was on contralateral side, whereas supernumerary nipple was on ipsilateral side. Restriction of dorsiflexion of ankle was due to shortening of tendoachillis, which has not been reported in

Fig. 2 – Radiograph of dorsolumbar spine. (A) Scoliosis, (B) Spina bifida.

Table 1 – Clinical spectrum of Becker's nevus syndrome.^{3–6}

Common features
Becker's nevus
Ipsilateral breast hypoplasia
Ipsilateral hypoplasia of pectoralis major
Ipsilateral limb reduction or asymmetry
Vertebral defects/scoliosis/spina bifida occulta
Other musculoskeletal defects
Cervical rib/fused ribs
Pectus excavatum
Pectus carinatum
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

literature. This has probably not been noticed because full squatting position as adopted by Indians is rarely adopted by people in Western countries, from where the most cases are reported. Clinically non archytype BN with incomplete histopathological features, in presence of other features of BNS; hyperpigmented macule can be considered as forme fruste of BN.¹⁰

Candidates found to have BN during recruitment medical examination should therefore be examined thoroughly for the associated musculoskeletal features. Conversely with any of

the musculoskeletal abnormalities described, deliberate search for BN be made to make right diagnosis.

Conflicts of interest

All authors have none to declare.

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