Case Report

Becker's nevus with neurofibromatosis type 1

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Abstract

Neurofibromatosis type 1 is an autosomal dominant disorder which primarily affects the growth and development of neural cell tissues. It presents as multiple tumor-like growths over the skin that arises from the nerves and is associated with other abnormalities like pigmentation over the skin and bone deformities. Becker's nevus or hairy pigmented epidermal nevus is a benign cutaneous hamartoma which is characterized by hyperpigmented macule with hypertrichosis. It is rarely associated with neurofibromatosis. We report a 22-years-old male with coexistent Becker's nevus and type 1 neurofibromatosis.

Key Words

Becker's nevus, Becker's nevus syndrome, neurofibromatosis

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Introduction

Neurofibromatosis (NF) is a multisystem genetic disorder that commonly is associated with Cutaneous, neurologic, and orthopedic manifestations. Neurofibromatosis type 1 (NF-1) is inherited in an autosomal dominant fashion.[1] Approximately 50% of the cases are inherited and the rest are due to sporadic mutations.[2] Neurofibromatosis type 1 is one of the most common single-gene disorders affecting neurological function in humans. Mutation in NF-1 i.e., neurofibromin gene leads to Neurofibromatosis-1.[3] This gene is located at chromosome 17q11.2.[3] This neurofibromin gene affects primarily the development and growth of neural cell tissues and the regulation of melanogenesis. NF-1 is thus characterized by multiple tumors originating from the neural cell tissues and produces pigmentary skin changes, vascular and skeletal dysplasias. It is a well-known fact that neurofibromatosis is associated with various pigmented skin lesions like cafe au lait macules, axillary freckling, smooth muscle hamartomas, malignant melanoma and various types of nevi. Becker's nevus is a benign cutaneous hamartoma with epidermal and/or dermal elements. It is characterized by a large, hyperpigmented macule with irregular borders and hypertrichosis, usually located over the upper trunk. Smooth muscle hamartoma is seen associated with Beckers nevus. Unilateral breast hypoplasia is seen associated with becker nevus.[4] Other associations include iplsilateral limb shortening, ipsilateral foot enlargement, pectus carinatum, unilateral or ipsilateral pectoralis major aplasia, spina bifida, scoliosis, congenital adrenal hyperplasia,[5] localized lipoatophy,[6] polythelia[7] and accessory scrotum.[8] However, the association of beckers nevus with neurofibromatosis is a rare entity. Thus we report a case, a 22-year-old male, who presented to us with Becker's nevus with coexistent type 1 Neurofibromatosis.

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Case Report

A 22-year-old male presented to us with multiple, painless, firm tumor-like nodules all over the body since 4 years of age which is gradually increasing in number. He also had a pedunculated mass hanging from the left upper arm [Figure 1]. There were also multiple cafe-au-lait macules all over the body present since birth and increasing in both number and size [Figure 1]. There was no history of neurological or musculoskeletal complaints. Systemic examination was normal. There were no bony defects. He also had an asymptomatic large hyperpigmented lesion in the right shoulder region. The lesion was present since birth and was progressively increasing in size. On examination the hyperpigmented lesion measured about 20 × 12 cm in size with irregular borders and a tuft of hair over it [Figure 2]. He revealed that he was born out of a consanguineous. The other two siblings had no history of any similar complaints. Histological examination of the hyperpigmented lesion revealed Schwann cells, fibroblasts, endothelial cells, perineural fibroblasts and axons arranged in a haphazard manner in a fibrous and myxoid

stroma. [Figure 3]. The patient was diagnosed to have a Becker's nevus with a coexisting neurofibromatosis type 1 based on the clinical and histopathological findings. The benign nature of Becker's nevus was explained to the patient. He was counselled about the complications of neurofibromatosis and the need for regular follow-up.

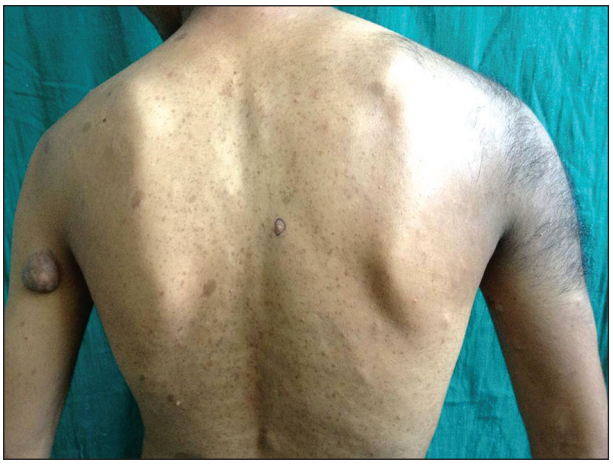

Figure 1: Multiple cafe au lait macules over back with pedunculated neurofibroma over left upper arm

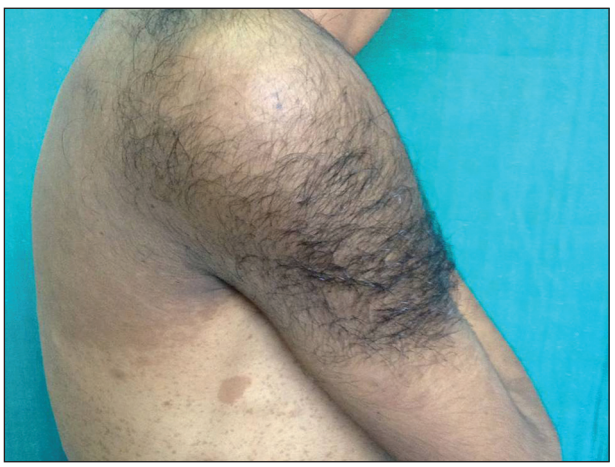

Figure 2: Hyperpigmented patch with tuft of hairs suggestive of Becker's nevus

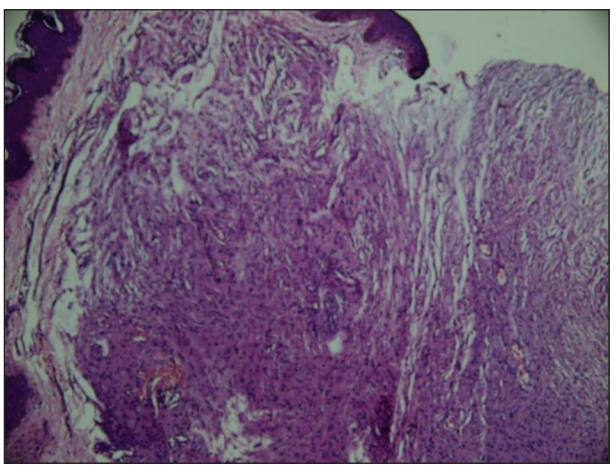

Figure 3: H & E-stained section showing Schwann cells, fibroblasts, endothelial cells, perineural fibroblasts and axons arranged in a haphazard manner in a fibrous and myxoid stroma

Discussion

Becker's nevus is also known as Becker's melanosis, Becker's pigmentary hamartoma and pigmented hairy epidermal nevus.[9] It is a skin disorder which affects predominantly males.[10] It is a benign cutaneous hamartoma that developed as a light or dark brown macule with well defined but irregular borders and can present with hypertrichosis.

Its etiopathogenesis is not clear yet. Lesion distribution reflects to a mosaic pattern. The majority of cases are sporadic but familial occurrence has also been reported which can be explained as a type of para-dominant inheritance.[11] Beckers nevus appears around puberty, the nevus contains increased number of terminal hairs and acne vulgaris and high male to female ratio points toward the possibility of androgenic stimulation as an underlying factor in its pathogenesis. A variety of cutaneous associations including intradermal nevi, malignant melanoma, leiomyoma, lymphangioma, acneiform eruptions have been associated with Becker's nevus but the association of neurofibromatosis type 1 is found to be very rare. Both the conditions are due to mutation of genes affecting both tumor suppression as well as melanogenesis. Becker's nevus can present as an isolated lesion or as Becker nevus syndrome. Becker nevus syndrome is the association of Becker's nevus with mammary hypoplasia, scoliosis or any other skin, muscular or skeletal alteration. Our patient had no mammary hypoplasia, scoliosis or any other skin/muscular/skeletal alteration. This case is presented to suggest that Beckers nevus association with neurofibromatosis can be a component of Beckers nevus syndrome.

There are very few case reports published so far showing the association of neurofibromatosis and Becker's nevus. Kim *et al.* reported a case of Becker's nevus with neurofibromatosis but in his case the patient developed CALM in adolescence while in our case he had these lesions since birth.[12] Uvaraj *et al* has also reported a case of similar association.[13]

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