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Case Report

Becker's Nevus in a Patient of Type 1 Neurofibromatosis

Authors
Anuj Sharma[1]*, Renu Rattan[2]
[1]Consultant Dermatologist, Dept of Dermatology, Regional Hospital Bilaspur, Himachal Pradesh-174001
[2]Consultant Dermatologist, Dept of Dermatology, Deen Dyal Upadhyay Hospital, Shimla
Email: *renurattan@gmail.com*
*Corresponding Author
Anuj Sharma MD
Consultant Dermatologist, Dept of Dermatology, Regional Hospital Bilaspur, Himachal Pradesh-174001
Email: *anujtandian@gmail.com, Phone no-+919418051677*

Abstract
Neurofibromatosis is an autosomal dominant disorder presenting with multiple tumor like growths over the skin and various abnormalities of pigmentation, and bony deformities. Becker's nevus is a benign cutaneous hamartoma characterized by hyperpigmented macule with hypertrichosis. The co-occurence of Neurofibromatosis and Becker's nevus in a patient is a rare entity. We hereby report a case of Neurofibromatosis Type 1 with Becker's nevus in a 21-year old male. The patient was evaluated for systemic manifestations, counselled and was advised regular follow up.
Keywords: *Neurofibromatosis, Becker's nevus, association.*

Introduction

Neurofibromatosis Type 1(NF-1), is a multisystem genetic disorder inherited in an autosomal dominant fashion, and is associated with various cutaneous, neurological and orthopaedic manifestations.[1] It is associated with various skin lesions like cafe au lait macules (CALMs), axillary freckling, smooth muscle hamartomas, malignant melanoma and various types of nevi.[2] Becker's nevus is a benign cutaneous hamartoma, characterized by a large, hyperpigmented macule with irregular borders and hypertrichosis, usually located over the upper trunk. There are a variety of conditions reported to be associated with becker's nevus like smooth muscle hamartoma,[3] unilateral breast hypoplasia,[4] various skeletal abnormalities, congenital adrenal hyperplasia,[5] localized lipoatophy,[6] polythelia,[7] and accessory scrotum.[8] However, the association of Beckers nevus with neurofibromatosis is rare, with only a few case reports mentioning the co-occurrence of these two conditions.[2,9-11] We hereby report a case of a 21-year-old male, who presented to us with Becker's nevus with coexistent type 1 Neurofibromatosis.

Case Report

A 21-year-old male patient, not born out of consanguineous, presented to our out-patient

department with multiple, painless, firm, tumor-like nodules all over the body (more over face and extremities) since early childhood which were gradually increasing in number and size. There were also multiple CALMs, variable in size (many being > 15mm in size) all over the body present since birth and increasing in both number and size [Figure 1]. He also had an asymptomatic large hyperpigmented lesion measuring 14 × 8 cm in size with irregular borders and a tuft of hair over it in the left infra-scapular region [Figure 2] which was present since birth and was progressively increasing in size. There was no history of neurological or musculoskeletal complaints. Systemic examination was normal. Similar complaints and features were present in the younger brother. 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 Neurofibromatosis-1 as per the criteria along with co-existent Becker's nevus on the basis of 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.

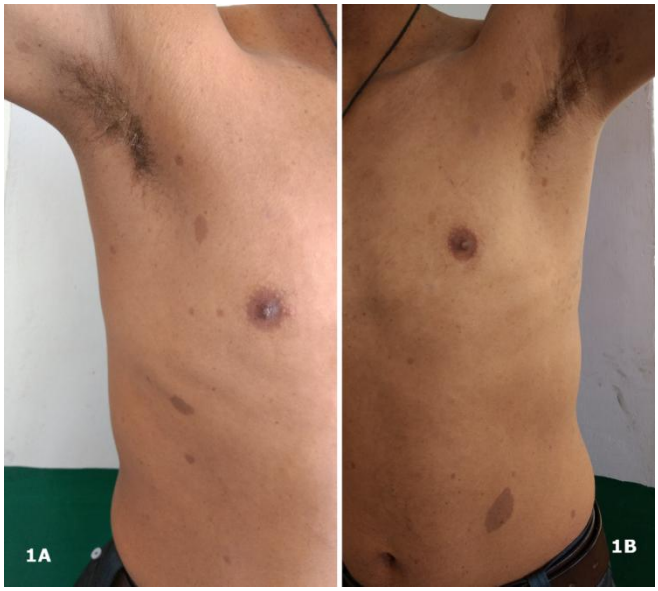

Figure 1: 1A and 1B showing multiple CALMs over the chest and trunk area.

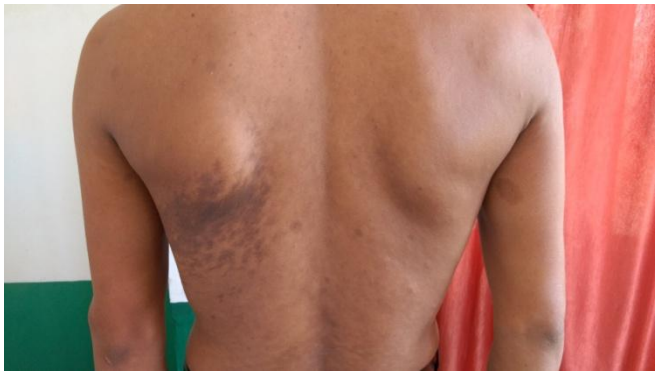

Figure 2: Becker's nevus-Hyperpigmented macule over the left infrascapular region with irregular borders and growth of hairs over it. CALMs can be seen in the vicinity.

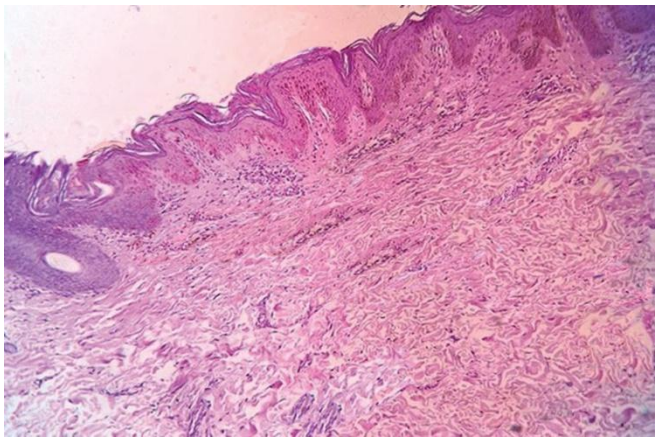

Figure 3: (Haematoxylin & eosin;100x) Histopathology of the hyperpigmented macule in figure 2 showing fibroblasts, endothelial cells, schwann cells, perineural fibroblasts and axons arranged haphazardly in a fibrous and myxoid stroma.

Discussion

Neurofibromatosis Type 1, also known as von Reckling-hausen disease, is a genetic disorder inherited in an autosomal dominant fashion. It is one of the most common single-gene disorders affecting neurological function in humans. The mutation occurs in NF-1 gene which is located at chromosome 17q11.2., encoding for the protein neurofibromin. 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. Various pigmented skin lesions like CALMs, axillary freckling, smooth muscle hamartomas, malignant melanoma and different types of naevi, have been mentioned in NF-1. The diagnostic criteria for NF-1 are listed in table1.[1]

Table-1. Diagnostic criteria for NF-1 (two or more required for diagnosis)

1.	Six or more cafe-au-lait macules over 5mm in greatest diameter in prepubertal individuals, and over 15mm in post pubertal individuals.
2.	Two or more neurofibromas of any type or one plexiform neurofibroma.
3.	Freckling in the axillary or inguinal regions.
4.	Optic glioma.
5.	Two or more Lisch nodules.
6.	A distinctive osseous lesion such as sphenoid dysplasia or thinning of long bone cortex with or without pseudoarthrosis.
7.	A first degree relative (parent, sibling or offspring) with NF-1 by the above criteria.

Becker's nevus, also called Becker's melanosis or pigmented hairy epidermal nevus, is a benign cutaneous hamartoma that develops as a light or dark brown macule with well-defined but irregular borders and can present with hypertrichosis.[12] The prevalence of Becker's nevus is around 0.5 % and males are reported to be more commonly affected than females.[3] Androgen dependency may explain the increased prevalence in males, peripubertal development and hypertrichosis.[13] 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.[14] A variety of cutaneous associations have been reported to be associated with Becker's naevus including intradermal nevi, malignant melanoma, leiomyoma, lymphangioma, acneiform eruptions,[15] smooth muscle hamartoma, [3] unilateral breast hypoplasia,[4] 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] The association of Beckers nevus with neurofibromatosis is a rare entity. Only a few cases have been reported in the past. Mahe et al in a study on 614 patients with neurofibromatosis did the biopsy of 6 patients for concomitant hairy hyperpigmented spots, out of which 1 was diagnosed to have Becker's nevus.[9] Kim et al described a 30 yr old male with Becker's nevus overlying a neurofibroma in neurofibromatosis type 1.[10] Uvaraj et al reported a case of NF-1 with Becker's nevus on the lumbar area of the back.[11] Kar et al reported a similar case of Becker's nevus on right upper arm in a patient of NF-1 and suggested that Becker's nevus association with neurofibromatosis can be a component of Becker's nevus syndrome.[2] Both of these conditions are hamartomatous proliferation of dermal and epidermal elements. Their association suggests that there could be common underlying pathology or Becker's nevus could be a component of neurofibromatosis. Our patient had no mammary hypoplasia, scoliosis or any other skin/muscular/skeletal alteration. This case of Becker's nevus with neurofibromatosis might be further evidence supporting the suggestion that Becker's nevus could be a component of neurofibromatosis.

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