

Case report: Becker's nevus in Neurofibromatosis Type 1

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Abstract

Becker's nevus, also called Becker's melanosis or pigmented hairy epidermal nevus, is a benign cutaneous hamartoma characterized by hyperpigmented macule usually with hypertrichosis. It is rarely associated with neurofibromatosis. We report a 2 ½ yrs old male child with coexistent congenital Becker's nevus and type 1 neurofibromatosis.

Keywords: Becker's nevus, neurofibromatosis, melanosis

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Introduction

Becker's nevus is a cutaneous hamartoma characterized by a large, hyperpigmented macule with irregular borders and hypertrichotic elements usually located over the upper trunk. There are a variety of conditions reported to be associated with becker's nevus (smooth muscle hamartomas, malignant melanoma, acneiform eruptions, etc.) and one of the rarest being neurofibromatosis [1,2]. We report a 2 ½ yr old male child who presented to us with Becker's nevus with a coexistent type 1 neurofibromatosis.

Case report

A 2 ½ yr old male child born of a non-consanguinous marriage presented to us with a large hyperpigmented lesion in the right lumbar region. The lesion was present since birth and is progressively increasing in size. On examination the hyperpigmented lesion measured about 6.5x4 cms in size with irregular borders and a tuft of hair over it (Fig. 1). There were also multiple café-au-lait macules all over the body.

On enquiry, the parents revealed that the café-au-lait macules are present since birth and increasing in both number and size. There was no history of neurological or musculoskeletal complaints. Systemic examination was normal. There were no chest asymmetry or limb anomalies. Spine appeared normal. Slit lamp examination revealed lisch nodules at 4'o clock and 10'o clock positions. No cafe-au-lait macules were noted in the older sibling or parents.

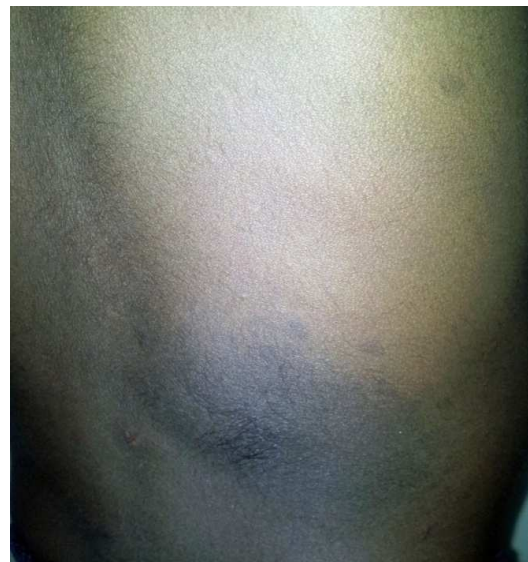


Figure 1. Becker's nevus - Hyperpigmented hairy patch with irregular borders over the right lumbar region.

Histological examination of the hyperpigmented lesion revealed elongated rete ridges with altered pigmentation in the basal layer (fig2). There were focal areas of dense melanin pigmentation in the basal layer while few areas showed hypopigmentation (absence of melanin). The patient was diagnosed to have a becker's nevus with a coexisting neurofibromatosis type 1 based on the clinical and histological findings. The benign nature of becker's nevus was explained to the parents. The parents were counseled about the complications of neurofibromatosis and the need for regular follow up. He is on regular follow up with periodic ophthalmologic examination, neurologic assessment, blood pressure monitoring and scoliosis evaluation.

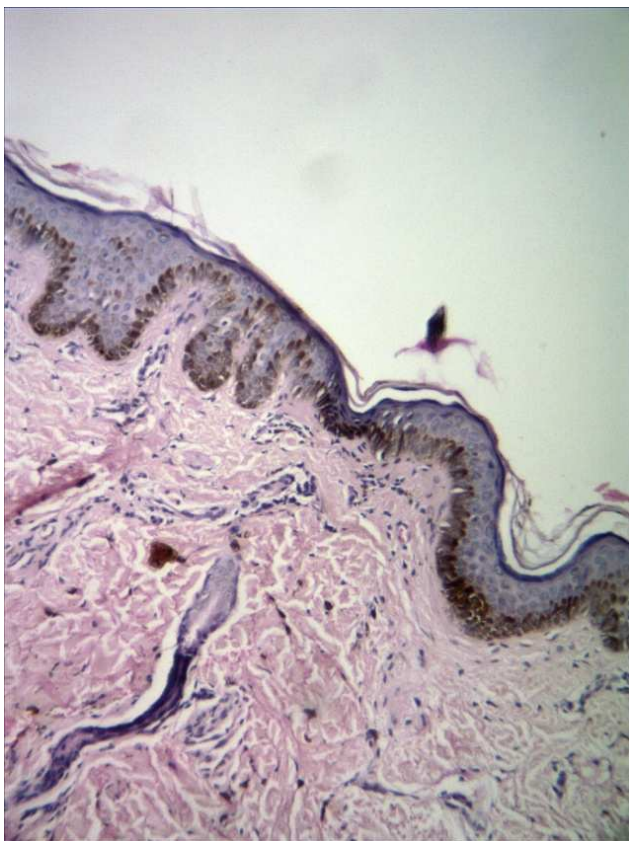


Figure 2. Histopathology of the hyperpigmented patch showing elongated rete ridges with hyperpigmentation of the basal layer.

Discussion

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 [1]. The prevalence of Becker's nevus is around 0.5 % and males are reported to be more commonly affected than females [2]. Androgen dependency may explain the increased prevalence in males, peripubertal development and hypertrichosis [3]. 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 [4]. Becker nevus syndrome affects males and females in equal proportions, whereas it has been reported that the male to female ratio in Becker's nevus is 2 to 1. A variety of cutaneous associations including intradermal nevi, malignant melanoma, leiomyoma, lymphangioma, acneiform eruptions have been associated with Becker's nevus [5].

Therapeutic intervention for Becker's nevus is primarily for cosmetic reasons. Laser therapy has been used with variable success for the treatment of both hypertrichosis and hyperpigmentation in Becker's nevus [6].

Neurofibromatosis has been rarely reported to be associated with Becker's nevus. In a study on 614 patients with neurofibromatosis, 6 of them underwent biopsy for concomitant hairy hyperpigmented spots, out of which 1 was diagnosed to have Becker's nevus [7]. Kim et al described a 30 yr old male with Becker's nevus overlying a neurofibroma in neurofibromatosis type 1 [8]. Both being 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 case of Becker's nevus with neurofibromatosis might be evidence supporting this suggestion.

Contributors: UP and RSR diagnosed the case. RSR and HG followed up the case. UP and IV conceptualized the article. UP and HG reviewed literature and prepared the manuscript. IV and RSR drafted the article and edited the final manuscript. UP will act as guarantor for the article.

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