



Giant Congenital Melanocytic Nevus in Newborn: Case Report

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Abstract

Congenital melanocytic nevus is usually defined as a melanocytic lesion which presents within 2 years of birth. Giant congenital melanocytic nevus (GCMN) has incidence of approximately <1:20,000 newborns. Despite being rare, GCMN is significant because of its association with severe complications such as malignant melanoma and neurocutaneous melanosis. GCMN presents as a brown lesion, with flat or mammilated surface, well-demarcated borders and hypertrichosis. Congenital melanocytic nevus is primarily a clinical diagnosis. Although GCMN is recognized as a risk factor for the development of melanoma, the precise magnitude of this risk is still controversial. The estimated lifetime risk of developing melanoma varies from 5 to 10%. On account of these uncertainties and the size of the lesions, the management of giant congenital melanocytic nevus needs individualization. Treatment may include surgical and non-surgical procedures, psychological intervention and/or clinical follow-up, with special attention to changes in color, texture or on the surface of the lesion. Indication for surgery in GCMN is the development of a malignant neoplasm on the lesion.

Keywords: *Nevi; melanomas; Nevus, pigmented; Skin neoplasm.*

Introduction

Melanocytic nevi are benign proliferations of melanocytic cells arranged in nests in the epidermis, dermis or in other tissue.¹ Giant congenital melanocytic nevi (GCMN) are pigmented lesions seen at birth or shortly after birth.^{1,2}

About 1% of live births presents with a congenital melanocytic nevi.² The incidence of GCMN is estimated at less than 1:20,000 newborns.^{2,3} GCMN occur most commonly on the posterior trunk or with multiple satellite lesions but may also appear on the head or extremities².

Despite being rare, these melanocytic nevi are significant because of their association with leptomeningeal melanocytosis, major psychosocial impact on the patient and family, due to unsightly appearance and their predisposition for development of malignant melanoma.^{3,4}

Leptomeningeal involvement occurs most often when the nevus is located on the head or midline on the trunk and associated with multiple "satellite" melanocytic nevi (>20 lesions)^{2,5}. Nevus cells within the leptomeninges and brain parenchyma can cause raised intracranial pressure,

hydrocephalus, seizures, mental retardation, and motor deficits and may result in melanoma. Cytological examination of the cerebrospinal fluid for melanin-containing cells can be done to screen for malignancy.⁶ GMN located on the scalp may have tendency to gradually lighten and regress over time.⁷

Classification based on size by Kopf et al.⁸

Small size	<2.0 cm diameter	
Intermediate size	>2.0-20 cm	
Large size	>20 cm	>5% of BSA

Case Report

35 weeks 2 days female infant (TWIN 1) born to Primigravida by caesarian with weight of 2280gms. Uneventful postnatal transit with Apgar scores 7 and 8 at 1 and 5 minutes respectively, head circumference 33.4cm and length 47cm. Maternal serology labs negative with uneventful antenatal history.

Neonate was born with melanocytic nevi extending from thoracic vertebrae to lower trunk with flat black skin lesion with smooth velvety appearance (Fig 1 & 2) and multiple small satellite lesions present on the bilateral lower legs, right forearm, cheeks and on back. (Fig 3, 4, 5) All limbs and orifices were normal. Neonate had 2 natal teeth at time of birth (Fig 6).

No neurological abnormalities noted during the 5 days hospital course. Cerebrospinal fluid examination was done and no melanin containing cells were present. Routine blood tests were in normal range.

Ultrasound abdomen and Neurosonogram were normal. ECHO of neonate was normal. Skin biopsy was not done. Fundus of newborn was normal. Neonate was sent home and kept on follow up (1,3,6,12 months of age) and neurological examination was normal. Skin biopsy was done at 1 year of age and the histopathological findings were consistent with those of congenital melanocytic naevi. No evidence of a malignant transformation was seen. Second twin was normal with no skin lesions. There was no family history of any similar lesion.



Fig 1: Melanocytic nevi size 12cmx8cm



Fig 2: CMN



Fig 3



Fig 4



Fig 5



Fig 6

Discussion

GMN develop between the 5th and 24th week of gestation, due to a morphological error in the neuroectoderm which results in dysregulated growth of melanoblasts. Kissing nevus, or divided nevus of the eyelid, divided after 20th week of gestation, implying GMN's development before 20th week of gestation.^{2,3}

Neural crest derived melanoblasts may be linked to the c-met proto-oncogene, controlling expression of the tyrosine kinase receptor Met.¹⁰ Mostly affected individual with leptomeningeal involvement have associated pigmented nevi on the head, neck or dorsal spine area.¹¹

They may present at early age (before 2 years) with sign and symptoms of raised ICT, with second peak seen at puberty or during the adult life.¹²

The prognosis of symptomatic patients is poor, with majority of these patients die within 3 years of their presentation of neurological symptoms, either due to CNS malignant melanoma or from progressive growth of benign melanocytic cells.¹³ 85% patients with giant congenital melanocytic nevi involving the head and neck have noted to have neurological abnormalities, although neurocutaneous melanosis was not detected by CT or MRI. Patients with neurocutaneous involvement had a posterior axial location of their cutaneous lesions on the head, neck, back and buttock.¹³

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