

## Case Report

# Giant congenital melanocytic nevus: a rare case report

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### ABSTRACT

Giant congenital melanocytic nevus is usually defined as a melanocytic lesion present at birth. Its incidence is estimated in <1 in 20,000 newborns. Despite its rarity, this lesion is important because it may be associated with severe complications such as malignant melanoma and may also cause neurological deficit including neurocutaneous melanocytosis. Giant congenital melanocytic nevus generally presents as a brown lesion with flat or mammillated surface, well-demarcated borders and hypertrichiosis. Congenital melanocytic nevus is primarily a clinical diagnosis. Recently, we encountered a 3 day old baby girl in whom giant congenital melanocytic nevus was noted at birth. She presented with large nevus over posterior trunk, head and extremities. Magnetic resonance imaging (MRI) brain was normal. Parents were counselled regarding possible future course and were asked to be in regular follow-up. Here, we report this rare case with giant congenital melanocytic nevus.

**Keywords:** Giant, Congenital melanocytic nevus, Hyperpigmentation

### INTRODUCTION

Congenital melanocytic nevus (CMNs) is defined as pigmented lesion that presents at birth or in the first year of life with unknown aetiology. Melanocytic nevi are benign proliferations of melanocytic cells arranged in nests in the epidermis, dermis or in other tissue.<sup>1,2</sup> CMN develops during the intrauterine life, the occurrence of these late congenital nevi might be explained by the insufficient initial production of melanin and/or by the small size of the nevus early on, hindering its detection.<sup>3-6</sup>

It has been suggested by Kopf et al to classify the lesions by surface diameter: small lesion <1.5 cm, intermediate lesions between 1.5 to 20.0 cm and giant lesions >20.0 cm. In 75% of the cases CMNs are accompanied by small, multiple lesions called “satellites”. Their number must be recorded because a number >20 is connected to leptomeningeal involvement.<sup>7</sup>

Despite its rarity, every patient with a giant CMN should be considered a suspect for development of neurodermal

melanosis regardless of the fact that he/she may or may not present neurological symptoms and magnetic resonance imaging (MRI) brain and the spinal marrow is advisable before the myelination of the brain.<sup>8-10</sup> Here we report a case of giant CMN in a 3 day old baby girl with review of literature.

### CASE REPORT

A 3 days old female baby brought in out-door patient department with complaint of blackish rough lesions all over body and some large swellings over nape of the neck. She was a product of non-consanguineous marriage of para 3 mother. Both parents were healthy and there was no significant family history. The neonate's weight at birth was 3000 grams, the head circumference was 34.5 cm and the body length was 54 cm. On physical examination, the neonate had a giant melanocytic nevus over back; of a brownish hue with geographical borders, flat surface and multiple satellite lesions with one neurofibromatous lesion over nape of the neck measuring 3×3.5 cm (Figure 1). Similar lesions were also present on arms, thighs and

lumbosacral region. The main lesion was of a developed constitution with a pronounced thickening and wrinkling; it lacked uniformity in colour and did not have definite borders. Multiple nodules with definite borders and dark, black colour were present on the posterior surface of the trunk (Figure 2). On systemic examination of other system were normal. There were no other associated congenital anomalies. Her MRI brain was normal. Parents were counselled regarding possible available treatment option regarding large area of disfigurement and future course and were asked to be in regular follow-up. Parents were also advised to seek opinion of dermatologist.



**Figure 1: Large melanocytic nevus covering almost whole body with large nodule over nape of the neck.**



**Figure 2: Multiple small nodules over chest.**

## DISCUSSION

CMN is defined as pigmented lesion that presents at birth or in the first year of life with unknown aetiology. Melanocytic nevi are benign proliferations of melanocytic cells arranged in nest in the epidermis, dermis or in other tissue.<sup>1,2</sup>

The giant CMN are evident at birth. In our case giant congenital melanocytic nevus also evident since birth, covering almost whole body. Neurocutaneous melanosis is a neurological and cutaneous disorder characterized by a non-normal concentration of melanocytes inside the central nervous system (CNS) and skin. Giant CMN was

first described by Rokitanski in 1861 in adolescent female with hydrocephalus and developmental retardation and after her death meningeal melanocytosis was found.<sup>8,9,11</sup> Neurocutaneous melanosis is a complication of large and giant CMN or of multiple small ones. Individuals with giant CMN and projected adult size (PAS) >40 with large and giant CMN on the back, on the neck or on the head are in increased danger for CMN even in absence of melanoma; inspite of the fact that many symptomatic patients survive with various neurological defects.<sup>11</sup>

The culture of melanocytes from such lesions showed chromosomal rearrangement which involved the chromosomal regions 1p, 12p and 19p. Researchers think that a body protein which is called 'hepatocytes growth factor/scatter factor' seems to be responsible for encouraging the neuroectodermal cells to develop, migrate, and scatter. It seems that either too much or a wrong type of this protein in some cells, develop extra pigment and abnormal skin cells which are called nevus cells. These cells scatter around and so we have nevi scattered all over the body.<sup>12</sup>

About half of the individuals with NCM will become symptomatic before the age of 5 years. Possible symptoms are hydrocephalus or other signs indicating increased ICP like headache, vomiting or seizures.<sup>11</sup> The cutaneous melanoma tends to penetrate deep in the skin or in the fat tissue of the large congenital melanocytic naevi (LCMN), rendering the early diagnosis difficult. MRI brain is advisable in multiple CMN or one CMN with more than 20 satellite nevi.<sup>8,9,13</sup> An equal prevalence exists in males and females. Also, every patient with newly appearing neurological symptoms like urination, tip-toe walking, epilepsy and sunset sign must undergo neurological evaluation and suitable imaging. Symptomatic neurocutaneous melanosis even without malignancy has an extremely poor prognosis.

It is important to diagnose the intracranial lesions at birth as this contributes to an earlier diagnosis of clinical manifestations. Knowledge of these lesions facilitates the differential diagnosis (d/d) of melanotic loci that are part of the disease or secondary metastasis of a malignant exchange of a giant CNM. In spite of the fact that the neurocutaneous melanosis is better imaged via MRI, the brain ultrasound is quite useful in early diagnosis in neonates. However, ultrasound does not visualize very small lesions in the cerebellum due to the artifacts of the bones.<sup>11</sup>

Surgical management is suggested to be accomplished in the first weeks of life to extract the cells early before migrating to deep dermis. Furthermore, following-up the patients in a long term period is essential part of the management. Treatment must be individualized. Psychological support is advised for all patients and their families, regardless of the therapeutic choice. Lifelong monitoring is necessary for patient with large and giant melanocytic nevi, regardless of the treatment adopted.

## CONCLUSION

GCMN is a rare condition that is defined as pigmented lesion present at birth or in the first year of life with unknown etiology. It requires a correct diagnosis and proper management due to its risk of developing MM. Other congenital anomalies should be sought out early after birth. Moreover, following-up the patients is an essential part of the management. Regardless of what type of management is decided upon be it surgical or observational, it must be remembered that most GCMN patients can have healthy and productive life.

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