

## Case Report

# Large facial congenital melanocytic nevus: a case report

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**Received:** 25 July 2013

**Accepted:** 4 August 2013

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### ABSTRACT

Congenital nevi are hyperpigmented macular lesions that are derivatives of the melanoblasts. Crude incidence estimates suggest that approximately 1 in 20 000 are born with a large CMN and 1 in 500 000 are born with a very large (giant) CMN. They occur in less than 1% of the neonates in any site of the body. The giant congenital nevus is greater than 20 cm in size, pigmented and often hairy. We report a classical case of large congenital melanocytic nevus present over face. The case report is being presented because of its rarity.

**Keywords:** Congenital melanocytic nevus (CMN), Giant congenital melanocytic nevus (GCNM), Malignant melanoma

### INTRODUCTION

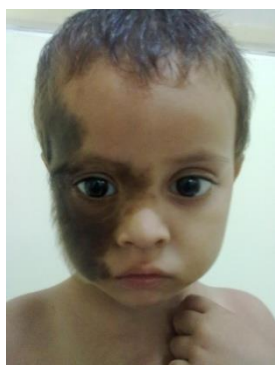
The Congenital Melanocytic Nevus (CMN) usually presents as a circumscribed, light brown to black patch or plaque, covering a surface area of any size involving any site of the body. The distribution of nevus over face is devastating and may be associated co-morbidities.<sup>1</sup> They are formed by overgrowth of the melanocytes. They are caused by malformations of the neuroectoderm that are comprised of melanocytes and occasionally neural elements, following dysregulated growth and arrest of melanocytes during migration from the neural crest to the skin.<sup>2</sup> Most CMN grow in proportion to the growth of the child except during early infancy when some CMN can grow rapidly. As the child has grown, CMN may get darker, lighter, lose pigmentation, become more heterogeneous and may develop a nodular surface or, rarely, regress.

CMN are classified as 'large' if the greatest diameter of the lesion, in adulthood, is 20 cm or more; 'medium' if its greatest diameter measures between 1.5 and 19.9 cm;

and 'small' if it measures less than 1.5 cm. Assuming proportionate expansion of body surface area, a 9 to 12 cm CMN on the head or a 6 to 7 cm CMN on the body of an infant will measure approximately 20 cm in adulthood.<sup>3</sup> The very large CMN, with diameters greater than 50 cm, are also known as 'giant' or 'garment nevi.' A giant congenital melanocytic nevus (GCMN), giant hairy nevus or nevocellular nevus represents a special group of melanocytic lesions that generally covers large areas of the body and have a potential risk for developing malignant melanoma.<sup>4</sup> The nevus location, nevus size, cosmetic issues regarding the nevus or resultant surgical scars, risk of anesthesia, risk of surgery, psychological implications, risk of melanoma and risk of NCM all need to be taken into account in this decision making process.<sup>3</sup> Surgical removal of CMN, especially large or clinically atypical CMN, may lower the risk for developing melanoma; however, this has not been confirmed in any controlled study. The present case is a classical case of large facial congenital melanocytic nevus and we report the case because of its rarity.

## CASE REPORT

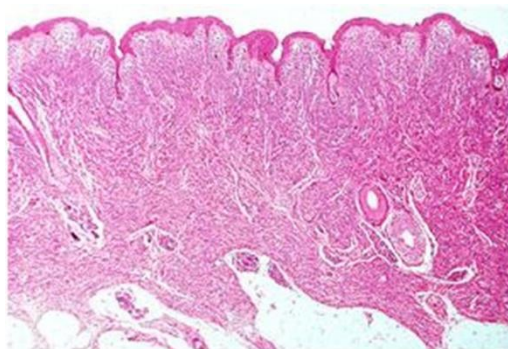
A two and half year old male child presented to us with chief complaint of large black colour skin patch over facial region since birth. He was a product of non-consanguineous marriage and was third in birth order. His elder brother who is 12 year old is a diagnosed case of Down's syndrome and elder sister is healthy and alive. His birth history and antenatal history was uneventful. Developmental mile stones were also normal according to age. There was no similar history in any of the relatives of both the parents. On examination child was of average built, weighing 13.2 kg and having a height of 90.2 cm, head circumference was 48.2 cm. Weight, height and head circumference were lies on 50<sup>th</sup> centile according to WHO growth curves. Vital parameters were stable. On examination of face there was a large pigmented patch of size 12 – 15 centimetre present over right side, covering nearly half of the face (Figure 1). 0.5 to 1.0 cm size hairs were present all over the lesion (Figure 2). Rest physical and systemic examination was normal. There were no other associated congenital anomalies. Fundus examination, X ray spine, CT head and ultrasound abdomen were all normal. The biopsy of the patient was taken and the histopathological findings were consistent with congenital melanocytic nevus. There were nest & cords of nevus cells filling the dermis and extending into the subcutaneous tissue (Figure 3). The dermis consisted entirely of heavily pigmented nevus cells containing melanin. No junctional activity or evidence of malignant transformation was seen.



**Figure 1: Congenital melanocytic nevus.**



**Figure 2: Hairs present on the lesion.**



**Figure 3: Nests & cords of naevus cells in dermis and subcutaneous tissue.**

## DISCUSSION

The congenital melanocytic nevi are pigmented cutaneous lesions occurs in about 1-6 percent of the newborns. Their colour is due to the melanin pigment of nevomelanocytes. Nevomelanocytes are the derivatives of melanoblasts and they compose the cellular format of the neoplasm.<sup>5</sup> Colour of the nevi is typically dark brown to black and 95% of them have dark, coarse surface hair. Satellite lesions are often present beyond the periphery of the main lesion and may be scattered over the entire skin surface. Nearly at the age of 10 years the giant nevus becomes more elevated, verrucous, and hyperkeratotic and the surface hair thicker.<sup>6</sup> Giant hairy nevi on the scalp and neck may be associated with leptomeningeal melanocytosis and neurologic disorders like neurofibromatosis, epilepsy or focal neurologic abnormalities, whereas lesions over the vertebral column may be associated with spina bifida or meningocele.

The incidence estimates suggests that approximately 1 in 100 infants are born with a small CMN; 1 in 1000 are born with a medium-sized CMN; 1 in 20 000 are born with a large CMN and 1 in 500 000 are born with a very large (giant) CMN.<sup>3</sup> Prevalence is same for males and females. Autosomal dominant inheritance with incomplete penetrance or multifactorial determination occurs in families with small CMN.<sup>7</sup> GCMN are thought to be caused by spontaneous mutations or other events during fetal development, but in some families, the frequent appearance of these lesions suggest that they may be genetically inherited. The genetic basis of these lesions is not known.<sup>6</sup> They may be associated with other birth defects. The differential diagnosis for CMN may include café-au-lait macule, Becker's nevus, dysplastic nevus, melanoma, nevus of Ota/Ito, Mongolian spot and speckled lentiginous nevus.

Behavioral and emotional problems are reported to occur in as many as 30% of patients with large CMN.<sup>3</sup> The psychological burden on patients and parents may stem from the cosmetic appearance of the CMN, the anxiety associated with the knowledge that complications such as melanoma can develop, the discomfort associated with

the often multiple staged surgical treatments rendered and the cosmetic appearance of resultant scars.<sup>3</sup> In addition, some CMN and/or nevi excision scars may develop tenderness, pruritus and/or skin fragility thus adding to the patient's discomfort.

Risk for potential development of malignant transformation within the nevus has been ranges from 0.05% to 10.7% depending on the size of the CMN.<sup>7</sup> Greater risk is associated with larger nevi.<sup>8</sup>

The impact of large CMN is greater because of the considerable cosmetic disfiguration which is very distressing to the parents along with its higher malignant potential. Radiographic imaging, including MRI, is warranted to evaluate melanocytic depositions in the CNS. The baseline MRI should be obtained when the patient is aged 4-6 months. Serial MRIs are frequently required in patients with meningeal melanocytosis.<sup>6</sup>

The management and treatment of patients with large and Giant CMN remains controversial. No absolute guidelines can be recommended. Treatment interventions may include full-thickness excisions, partial-thickness excisions, dermabrasions, curettage, laser treatment and chemical peels. Improving the cosmetic appearance frequently requires the use of a combination of different treatment interventions.<sup>3</sup> In terms of preventing the development of melanoma (prophylactic removal), any of the above-mentioned procedures will reduce the overall number of melanocytes which theoretically should lower the risk of melanoma. However, with the exception of full-thickness surgical excision, these procedures do not adequately address the risk for developing melanoma.<sup>3</sup>

Surgical excision of CMN depends on the size and site of the lesion. The larger lesions that require surgical excision are usually followed by tissue expanders, tissue

grafts, and tissue flaps in order to repair the large defects. Surgical curettage is relatively an alternate simpler procedure when performed during the neonatal period yields acceptable results.<sup>5</sup>

Our case had large CMN over face and his parents want removal of the lesion because of cosmetic reason as well as of malignant potential. We have referred this case to plastic surgery unit for further management.

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DOI: 10.5455/2320-6012.ijrms20131148

**Cite this article as:** Sharma D, Sharma CM. Large facial congenital melanocytic nevus: a case report. *Int J Res Med Sci* 2013;1:576-8.