

# Bathing trunk nevus with multiple satellite lesions and neuroid proliferations

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**Abstract** Giant congenital melanocytic nevus (GCMN) are rare melanocytic proliferations of the skin. These may be associated with benign neuroid proliferations, lipomas and abnormalities such as spina bifida. Major concern with GCMN is risk of neurocutaneous melanosis (NCM), melanoma, or other complications. We report a case of a 4-year-old girl with an extensive hyperpigmented plaque that covered her entire trunk involving almost 50% of her body surface area, highly suggestive of a giant congenital melanocytic nevus. Multiple neuroid proliferations were present.

**Key words**

Giant congenital melanocytic nevus, neuroid proliferations, satellite lesions.

## Introduction

Bathing trunk nevus is a term used for giant congenital melanocytic nevus (GCMN). These nevi are large, hyperpigmented patches with a diameter of more than 20 cm and usually present since birth.<sup>1</sup> The prevalence of single CMN is approximately 1-2% of new births.<sup>2</sup> The prevalence of giant CMN is 1 in 20000 to 1 in 50000 births.<sup>3</sup> Congenital melanocytic nevi are clonal proliferation of benign melanocytes during embryogenesis due to somatic gain-of-function mutations in NRAS.<sup>4</sup> As compared to CMN, giant CMN are deeper and extend into dermis up to subcutaneous tissue. These are irregular in margin, dark coloured and covered with coarse hair. Satellite lesions are frequently present.<sup>5</sup> Giant CMN can have involvement of leptomeninges and can present with seizures or neurological deficits. The size of lesions

increases proportionally to body size. Morphological changes take place over time as the lesions can become elevated, thicker, darker and can develop a mottled, cribriform or verrucous surface.<sup>6,7</sup> Superimposed nodules can develop in pre-existing patches. Changes such as rapid growth, colour change, and ulceration should raise a suspicion for melanoma development as there is a 5-10% of risk for malignant transformation in giant CMN before the age of 5 years.<sup>8</sup>

Here we are present a case of GCMN with multiple satellite lesions along with various tumour like proliferations resembling neurofibromas.

## Case report

A 4-year-old girl child born to non-consanguineous parents, presented with hyperpigmented lesions on her body covering almost 50% of her body surface area. The mother gave history of dark brown to black skin lesions of varying sizes on the patient's body since birth. Perinatal history was uneventful with

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**Figure 1** Giant bathing trunk melanocytic nevus over trunk, extending from lower chest till the knees.



**Figure 2** Multiple melanocytic nevi on face, lips, neck, chest, shoulders and arms and neck.



**Figure 3** Giant melanocytic nevus over back with a pebbly appearance and multiple protuberant skin proliferations.

a full-term vaginal delivery of the child. These lesions slowly progressed with increase in size and number till date. The biggest lesion was present on trunk from mid chest and extending up to knees. A few smaller brown to black patches were scattered over upper chest, face including lips and upper as well as lower limbs. There was associated increased hair growth over the lesions since birth. Lesions also developed a rough texture as the child grew and small soft swellings appeared over a few lesions mainly on the trunk. These swellings were not associated with any pain. There was no history of any vision abnormalities, seizures or neurological complaints and the child had a normal growth.

On examination, hyperpigmented patches and plaques with rough surface and uneven border were present covering approximately 50% of body surface area. Largest plaque extended from just below the nipples till the knees (**Figure 1**). Circular to oval hyperpigmented patches of sizes varying from approximately 0.5 to 2 cm in diameter were scattered on upper chest, face including lips, arms, hands, legs and feet (**Figure 2**). Hypertrichosis was present compared to surrounding unaffected skin. Few lesions had a pebbly appearance with well-

defined soft nodules arising from existing lesions. Diffuse neuroid proliferations of up to 4-5cm in size and firm texture were present on the truncal lesions (**Figure 3**). These were non tender and appeared later in life and were not present at birth. Eye examination including the fundus examination was normal. Head circumference and shape was normal for her age. MRI brain wasn't done as there was no history of seizures and neurological examination was normal. She was provided with psychological support and her family was counselled on warning signs of complications which would warrant an urgent consultation.

## Discussion

Giant CMN has been associated with neurofibromas, lipomas and spina bifida occulta.<sup>9,10</sup> These can be explained on the basis of defect in neural crest which is a common origin for melanoblasts, Schwann cells, ganglia, bone, fat, muscle and blood vessels.<sup>11</sup> Benign proliferations within CMN are common, primarily associated with large or multiple nevi. Clinically, benign proliferations can be divided as classic proliferative nodules and neuroid overgrowth. Classic proliferative nodules have a

well-defined edge, a round or oval outline and a smooth and sometimes shiny surface, and are soft or firm but not hard. Neuroid overgrowth areas have poorly defined edges, are usually round or ovoid/ fusiform, are several centimetres to >20cm in diameter, can be less pigmented than the surrounding CMN, and are soft or lipoma-like to the touch.<sup>12</sup> Diffuse neuroid proliferations can develop in existing plaques of CMN at any time during childhood and usually not present at birth. These usually become more active around puberty.

New recommendations for the categorization of cutaneous features of congenital melanocytic nevi have been given which include CMN size categories of small (<1.5cm); medium (M1: 1.5-10cm, M2: >10-20cm); large (L1: >20-30cm, L2: >30-40cm); and giant (G1: >40-60cm, G2: >60cm). Number of satellite nevi in the first year of life is categorized into none, 1 to 20, more than 20 to 50, and more than 50 satellites. Additional descriptors of CMN include anatomic localization, colour heterogeneity, surface rugosity and presence of hypertrichosis and presence of dermal or subcutaneous nodules.<sup>13</sup> On the basis of this classification our patient would be classified as giant CMN G2 (>60cm), CMN of trunk (localization), S3 (number of satellite nevi >50), with additional morphological features as C1 (moderate colour heterogeneity), R2 (marked surface rugosity), N1 (scattered dermal and subcutaneous nodules), H1 (notable hypertrichosis). These characteristics of GCMN can help predict risk for developing neurocutaneous melanosis (NCM), melanoma, or other complications. Therefore, it is important that all patients with GCMN having high risk phenotypic features should have a lifetime follow up for early detection and management of melanoma, NCM or other complications.

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