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CASE REPORT

Giant Congenital Melanocytic Nevus in an Infant: Case Report

Iuli Ferro da Silva Krieck[1], Vanessa Borges Platt[1]

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Abstract

Objective: To describe a rare dermatological disease condition in the pediatric population, with an occurrence rate of approximately 1 in 20,000 to 1 in 500,000 births. **Case Description:** A 4-month-old infant, with no comorbidities and appropriate anthropometric measurements and neuropsychomotor development for age, presented with multiple circular, brownish nevi of varying diameters and distributions on the face, scalp, trunk, buttocks, upper and lower limbs, hands, and feet. The largest nevus measured over 20 cm in diameter and was characterized as the "bathing trunk" type. **Comments:** The diagnosis is clinical, with particular emphasis on using the "6B Rule" classification to measure the size of lesions in cases of giant congenital melanocytic nevi. There is a recommendation for performing central nervous system imaging at an appropriate age to rule out neurological involvement. In this specific case, the patient has not yet undergone such examinations, which underscores the need for continued follow-up and monitoring

[1] Universidade Federal de Santa Catarina, Hospital Universitário - Florianópolis – Santa Catarina - Brazil.

Correspondence to:
Vanessa Borges Platt
R. Profa. Maria Flora Pausewang - Trindade, Florianópolis - SC, Brazil, CEP: 88036-800. E-mail: vanessabplatt@gmail.com

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INTRODUCTION

Congenital giant melanocytic nevi (CGMN) are uncommon skin lesions, with an estimated occurrence of about 1 in every 20,000 to 1 in every 500,000 births[1,2]. They reach a diameter greater than 20 cm in adulthood and typically present as brown or black patches or plaques, which can be flat or elevated, with or without association with hypertrichosis and the presence of satellite lesions.[3]

The diagnosis is clinical, but additional exams can be useful for investigation, especially to assess the involvement of the Central Nervous System (CNS), given the possibility of more serious complications such as neurocutaneous melanosis and malignant melanoma[3]. Although it is considered a risk factor for melanoma, the incidence of malignancy is still controversial in literature, ranging from 5% to 10% over a lifetime[4].

The unesthetic appearance of the nevi can have psychological repercussions for the patient and their family[5]. Treatment includes monitoring the lesions, surgical removal when indicated, and multidisciplinary care for the patient, who should be regularly followed up.

Due to the low incidence of congenital giant melanocytic nevi, this study is essential to expand clinical and scientific knowledge about this rare and complex condition. These nevi, present at birth, can vary in size and pose risks, such as malignancy. Documenting specific clinical cases helps identify patterns in diagnosis, monitoring, and therapeutic interventions, providing more appropriate and effective management in newborns.

CASE REPORT

A male infant, 4 months old, with adequate prenatal care, born via vaginal delivery at term, without complications, at 39 weeks and 2 days of gestational age, considered appropriate for gestational age. He was hospitalized for 4 days in the general pediatric ward of a tertiary hospital due to acute viral bronchiolitis (AVB). The viral panel did not detect the presence of respiratory syncytial virus, influenza A and B, or SARS-CoV-2, and the condition showed good progression. He remained afebrile, exclusively breastfed. The vaccination schedule was up to date according to the National Immunization Program (PNI).

On physical examination, the patient appeared to be in good general condition with a normal clinical impression. The anthropometric data showed a weight of 6.5 kg (z-score of -0.82) and a height of 64 cm (z-score of 0.14), both appropriate for the age. Neuropsychomotor development was consistent with age. In addition to mild respiratory discomfort, multiple circular, brownish nevi of varying sizes and distributions were observed on the face, scalp, trunk, buttocks, upper and lower limbs, as well as on the hands and feet. The largest nevus measured over 20 cm in diameter. (Figure 1).

The nevus affected almost the entire left lower limb, sparing only the left ankle and foot, including the genital region, left buttock, and part of the right buttock, extending to the thoracolumbar transition area. The observed area was dark brown in color, with patches of heterogeneous pigmentation, irregular borders, and areas with a rough surface, as well as the presence of fine and long hair on the side of the left thigh (Figure 2).

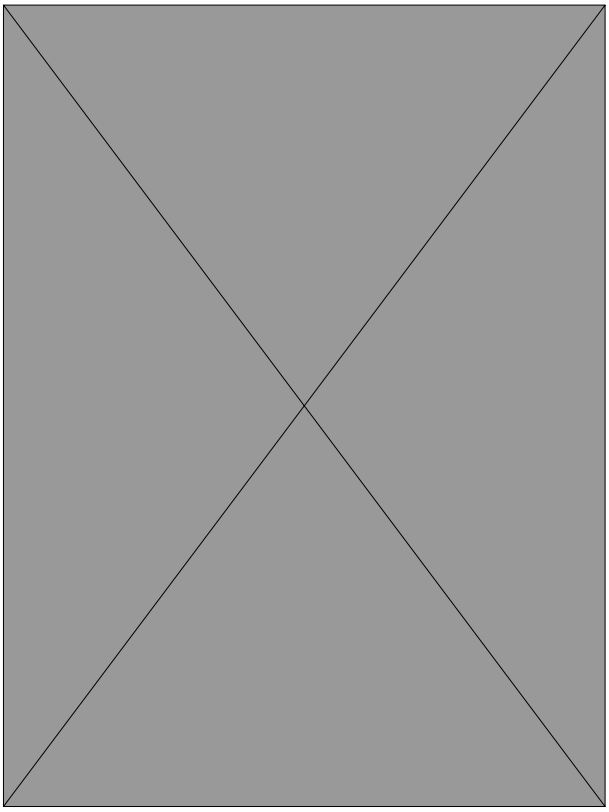

Figure 1. Giant congenital melanocytic nevus in the shape of "swimming trunks"

The patient had a family history of small diameter nevi (father and older siblings), but there were no other reports of giant congenital nevi in the family.

After hospital discharge, the patient was referred for evaluation by pediatric specialists (dermatologist, neurologist, and ophthalmologist), where no other abnormalities were identified in the specific exams. The management plan recommended was expectant care, with outpatient follow-up, and an MRI of the neuro-axis should be performed after 6 months of age.

The research was approved by the Institutional Ethics Committee under Opinion No. 7.310.440 dated 19/12/24 (CAAE: 84996324.8.0000.0121), with adherence to Brazilian ethical guidelines for research involving human subjects and the use and sharing of images.

DISCUSSION

Giant congenital melanocytic nevi (GCMN) result from a somatic mutation of melanocytic cells during the intrauterine period, triggering uncontrolled growth of melanocytes. They

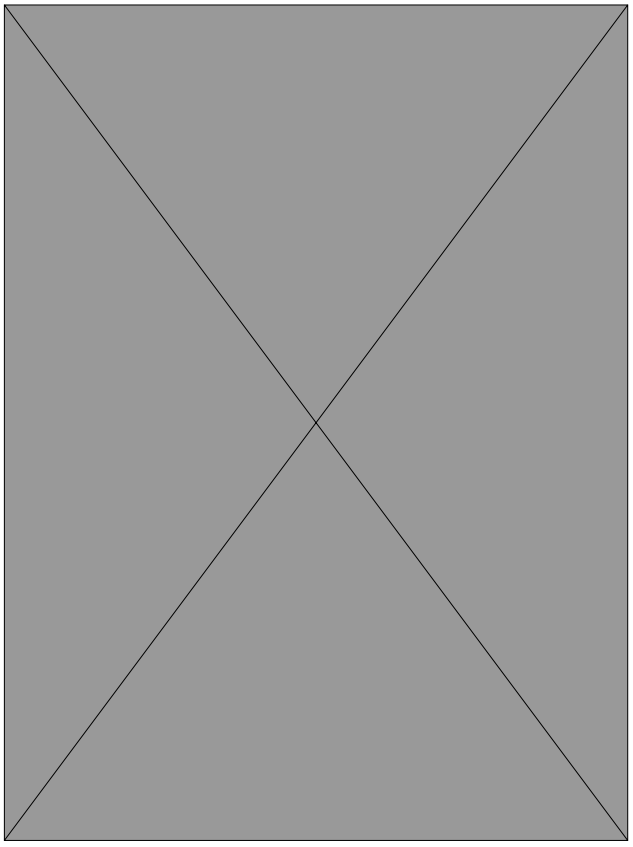

Figure 2. Giant nevi and associated satellite lesions.

occur sporadically, with few reports of familial predisposition, unlike what was observed in our patient[5]. The earlier the mutation occurs, the higher the probability of GCMN and the association with satellite nevi and involvement of the Central Nervous System (CNS).[6]

Classically, GCMN is an asymptomatic condition, but it can cause pruritus and xerosis in some cases[7]. Aesthetically, the hyperpigmented areas can affect any anatomical region, being most visible on the trunk (47%), followed by the extremities (30%) and the head (22%)[8] The lesions generally grow proportionally to the somatic growth of the child, as evidenced by longitudinal photographic records made by the parents (Figures 1 and 2).

The classification of the nevus is based on the evaluation of the maximum diameter projected for adulthood. It is classified as large-1 if it measures between 20-30 cm, large-2 between 30-40 cm, giant-1 between 40-60 cm, and giant-2 if the maximum diameter exceeds 60 cm[5]. The lesions can also be classified according to characteristics of color heterogeneity, presence or absence of hypertrichosis, dermal and/or subcutaneous nodularity, and the number of satellites nevi[9]. The patient's lesion was considered giant, as the projected diameter for adulthood was greater than 40 cm.

Some terms related to clothing are commonly used for the classification of lesions. The "6B Rule" pattern of giant nevi systematizes their distribution: "back" (involving only the back, excluding the shoulders or buttocks), "bolero" (upper back, including the neck), "bathing shorts" (genital and buttock regions) – as observed in the reported case, "chest/abdomen" (isolated to the abdomen or chest), "extremity of the body" (isolated to the extremities), and "body" (combination of the "bolero" and "bathing shorts" patterns).[10]

The diagnosis of this condition is clinical, but biopsy with histopathological evaluation may be indicated[5]. The results demonstrate intradermal involvement, with melanocytes of varied morphologies, which may reach muscles, glands, vessels, nerve structures, and fascia. It is recommended to perform an MRI of the brain and spinal cord between 4-6 months of age due to the brain myelination schedule[11]. In this case reported, no additional examination was performed at the time of hospitalization.

The complications of GCMN, present in about 1-5% of cases, include malignant melanoma and neurocutaneous melanosis[7]. Malignant melanoma is rare in pediatrics, representing 2% of melanomas in childhood, with the risk of development directly proportional to the size of the nevus.[11]

Neurocutaneous melanosis is another uncommon complication that worsens the prognosis of these patients. It consists of melanocytic proliferation, which can be benign or malignant, nodular or diffuse, within the leptomeninges and cerebral parenchyma[12]. It frequently occurs in individuals with lesions larger than 40 cm, located on the trunk, and often associated with satellite nevi.[13]

Therapeutic options for GCMN range from surgical excision to laser therapy[10]. The primary justification for surgical removal is the risk of malignancy, applicable to any region of the nevus that shows changes in growth, color, or homogeneity compared to the rest of the lesion[13]. Other criteria for surgical removal should be evaluated individually, considering the size, location of the lesion, and the aesthetic and functional impacts.[14]

The management of congenital melanocytic nevi, especially those presenting as large lesions and multiple satellite nevi, requires a meticulous, personalized, and integrated clinical approach. The indication for brain and spinal cord magnetic resonance imaging should be judicious, taking into account not only the morphological aspects of the lesion but also the presence of neurological symptoms suggestive of underlying involvement. Although the risk of neurocutaneous melanosis is low in isolated small and medium nevi, this risk significantly increases in patients with giant nevi and multiple lesions, justifying the adoption of imaging surveillance, as exemplified in the case under discussion.[15]

However, despite clear recommendations for imaging screening, the patient did not undergo magnetic resonance imaging screening, which underscores the importance of strategies to promote adherence to clinical follow-up and diagnostic protocols. Therapeutic decisions must carefully balance potential benefits and risks, as well as consider the psychosocial impact inherent to prolonged follow-up,

emphasizing the importance of continuous dermatological monitoring and multidisciplinary evaluation of the emotional aspects affecting both the patient and their family[15].

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