



CASE REPORT

Giant congenital melanocytic nevus of the back: a case report

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ABSTRACT

BACKGROUND

Giant congenital melanocytic nevus (GCMN) is a rare disease with an extremely low incidence, that is present from or develops at birth and typically affects the dermis but may also affect other skin layers. Its incidence is estimated at <1 in 20,000 newborns. Despite its rarity, this lesion is important because it may be associated with severe complications such as malignant melanoma. A thorough follow-up is crucial since the probability of malignancy can vary depending on the clinical course. As such, careful observation is necessary to support possible management plans.

CASE DESCRIPTION

In this case report, we present a three-day-old newborn male with abrasions on a black patch on his back. He presented with fever, jaundice, and black patches on more than 50% of his trunk down to the sacral area. The black raised patches resembling nodules had wounds on the lower back near the gluteus. A histopathology examination of specimens taken from 3 nodules on the back revealed hypocellular tissues with lymphocytes, histiocytes, neutrophils, fat droplets, and mature fat cells interspersed with some erythrocytes. The lesion was, therefore, diagnosed as a giant congenital melanocytic nevus (GCMN). Parents were counseled regarding the possible future course and were asked to come for regular follow-ups.

CONCLUSIONS

In this instance, we document a rare occurrence of GCMN that warrants recognition and appropriate treatment. To accumulate evidence for improving disease prognosis and outcomes, children with congenital melanocytic nevus should be included in a systemic follow-up study from birth.

Keywords: Giant congenital melanocytic nevus, black patch, melanoma transformation, neuroblastoma, infant

INTRODUCTION

Giant congenital melanocytic nevus (GCMN) is usually defined as a melanocytic lesion that is present at birth and may reach a diameter of ≥ 20 cm in adulthood. Its incidence is estimated as $< 1:20,000$ newborns, of which about 6% develop melanoma at the site of the nevus. A rare tumor in children, malignant melanoma makes up 0.9% of all pediatric cancers.⁽¹⁾ There is a 0–3.8% chance that GCMN will develop into malignant melanoma. Patients with GCMN should have regular evaluations for neurological abnormalities and psychosocial impact in addition to malignant change.⁽²⁾

This skin lesion develops due to neural crest-derived melanocyte precursors overgrowing or migrating irregularly. They are also called garment nevi, stocking nevi, and coat sleeve nevi. The lesions are flat to elevated nevi and range from brown to black in color.⁽³⁾ One of the criteria used to classify congenital melanocyte nevus (CMN) is the maximum diameter that the nevus is expected to reach as an adult. A CMN is categorized as giant-1 if its maximal diameter is between 40 and 60 cm, giant-2 if it is higher than 60 cm, large-1 if its maximal diameter is between 20 and 30 cm, and large-2 if it is between 30 and 40 cm. Additional morphologic features of the CMN are also included in this classification method, such as the number of smaller satellite nevi, surface rugosity, hypertrichosis, anatomic placement, and degree of color heterogeneity.⁽⁴⁾ Giant congenital melanocytic nevi are birthmarks with a diameter of more than 20 cm. The risk of malignancy rises with the number of satellite lesions near the large nevus.⁽³⁾

Despite its rarity, GCMN is significant because of its link to serious side effects such as malignant melanoma and central nervous system (CNS) involvement, as well as a significant psychosocial impact on the patient and his family because of its unattractive look.⁽⁵⁾ There has been controversy about the incidence of melanoma in cases of CMN and thus the clinical management of CMN, which is partly due to the difficulties of histological diagnosis and partly due to publishing bias towards cases of malignancy. Giant congenital melanocytic nevus is the main risk factor for the development of melanoma in childhood.⁽¹⁾ To date, no absolute guidelines to treat GCMN have been established, and therefore, the subject remains one of the most controversial

issues in dermatologic surgery and dermatologic oncology.

The development of a malignant neoplasm on the lesion is the only unequivocal indication for surgery in cases of large congenital melanocytic nevi. Other treatment options include nonsurgical management. Congenital melanocytic nevi are managed differently depending on several variables, including the lesion's location, size, impact on cosmesis, patient age, and likelihood of malignant development. Many doctors are treating small and medium-sized congenital melanocytic nevi with baseline photography and routine follow-up, with the frequency of follow-up depending on the clinician's experience and practice, until evidence is produced on which to build clear treatment guidelines.⁽⁶⁾

To address these issues, this case report presents a thorough description of a three-day-old infant who was diagnosed with GCMN, with a focus on GCMN as the major diagnosis. The report aims to provide significant insights into this uncommon disease by examining the clinical and pathological aspects of the case.

CASE REPORT

A three-day-old newborn male presented with abrasions on a black patch on his back with an abrasion that was mildly bleeding. He had been delivered spontaneously with a birth weight of 3000 grams. At the time of the physician's first visit, the patient presented with a fever and jaundice. Vital signs showed a fever of 38°C , a pulse of 140 times per minute, and a respiratory rate of 40 times per minute. General examination was unremarkable despite the presence of black patches on more than 50% of his trunk down to the sacral area (Figure 1).



Figure 1. Generalized black pigmentation on the upper back down to the gluteal region with nodules on the lower back

Physical examination showed multiple well-defined hyperpigmented macular lesions, 5 mm to 2 cm in diameter (areas of the scalp, face, anterior thorax, and both legs). The abdomen, posterior thorax, gluteus, and bilateral femoral area showed extensive hyperpigmented well-defined maculae and plaques, with hair on the left femoral area and soft in consistency.

Laboratory examination revealed hyperbilirubinemia with a total bilirubin concentration of 10.32 mg/dL and a direct bilirubin concentration of 0.73 mg/dL. Standard hematology examination results were regular. A histopathology examination was performed on June 24th, 2021, with the specimens taken from 3 nodules at the back revealing hypocellular tissues with lymphocytes, histiocytes, neutrophils, fat droplets, and mature fat cells interspersed with some erythrocytes between the tissues. The child was diagnosed with GCMN, and the parents were educated to observe the lesions for any growth, asymmetry, or bleeding. Written informed consent was obtained from the parents of the patient for publication of the case report and any accompanying images.

DISCUSSION

Three case reports that have been published offer new insights about giant congenital melanocytic nevus (GCMN). According to the first report, a 6-year-old boy had a tumor that covered approximately 20% of the surface area of his body and accounted for 28% of his weight.⁽⁵⁾ In the second case, a 12-year-old girl from Cameroon is described as having a brownish-black, nodular, hypertrophic skin lesion that has grown larger over time, measuring about 45 centimeters, from the time when she was just 2 days old.⁽⁷⁾ And the third case, a 6-month-old girl presented with hyperpigmented skin lesions over the back, gluteal regions, perineum, thighs, and scalp since birth.⁽⁸⁾ (Table 1)

These case reports add to our understanding of GCMN by illuminating the difficulties in managing the disorder and providing insightful clinical information. They stress how crucial it is to diagnose and treat GCMN as soon as possible because of its potential for serious complications and its psychological effects on patients and their families.^(5,7,8)

It is crucial to identify congenital skin abnormalities early in the newborn dermatologic examination, establish an accurate diagnosis, and

treat them. Although benign cases are frequent during this stage of life, clinical presentations can be much more dramatic, exaggerated, and stressful for parents.⁽⁹⁾ Many differential diagnoses must be considered when a newborn presents with hyperpigmented skin areas, such as Mongolian spots, epidermal hyperpigmentation, congenital melanocytic nevi, and dermal melanosis.⁽¹⁰⁾

Congenital melanocytic nevi are categorized clinically according to their size. Small nevi are typically defined as having a diameter of less than 1.5 cm, medium nevi as having a diameter of 1.5 to 19.9 cm, and large or giant congenital melanocytic nevi as having a diameter of 20 cm or more. Furthermore, a more accurate description of giant congenital melanocytic nevi compares the size of the lesion with the total body surface area; lesions that occupy 2% or more of the body surface area are defined as giant nevi. This is because the growth of these lesions is proportionate to overall growth. Massive nevi can manifest with multiple smaller satellite lesions and frequently have "bathing trunks" and "glove stocking" distributions.⁽¹¹⁾

Giant congenital melanocytic nevus frequently has a disfiguring appearance that is difficult to conceal with a standard dressing. As a result, GCMN puts a significant psychological strain on the patients and their parents. The incidence of GCMN is 1:20,000 as reported by Arad and Zuker,⁽¹²⁾ which is far more uncommon than that of 1:9,450 as found by Recio et al.⁽²⁰⁾ In Indonesia, only a few GCMN case reports have been identified.^(13,14) Several plausible explanations for the rarity of GCMN in Indonesia are the lack of awareness of this disease, the paucity of clinicians publishing the cases despite encountering them, or the fact that GCMN is just that rare.

It is crucial to diagnose CMN because the size of the nevus has been linked to an increased risk of melanoma in CMN patients.⁽¹⁵⁾ Compared to CMN and the typical acquired melanocytic nevi, people with a GCMN have a lifetime melanoma transformation risk as high as 5–10%.^(16,17) Because of this greater malignancy rate, clinicians should pay more attention to GCMN. According to studies, CMN may manifest in several ways genetically; for example, neuroblastoma RAS (N-RAS) is mutated in 97.4% of cases with GCMN.⁽¹⁸⁾

Table 1. Previous reported cases

Author	Patient	Chief complaint	Accompanying symptoms	Physical examination	Additional examination	Diagnosis	Management	Outcome
Merchan-Cadavid et al. ⁽⁵⁾	6-year-old boy	Dark, raised lesions that had been present since birth.	None.	<ul style="list-style-type: none"> • Hyperpigmented, raised lesions that had been present since birth, and later developed into a tumoral mass covering approximately 20% of the total body surface and accounting for 28% of his body weight, located on the back and abdomen, with satellite lesions on the extremities, face, and scalp. • Marked splenomegaly with increased abdominal perimeter was also found. 	<ul style="list-style-type: none"> • First biopsy from the largest lesion showed intradermal melanocytic nevus with congenital traits. • Second biopsy from nodular hyperpigmented lesion on the left knee showed a compound melanocytic nevus. • Histology examination showed predominant melanocytic cell presence and architecture, hyperpigmented and circumscribed mainly to the dense, reticular superficial dermis. 	Giant congenital melanocytic nevi (GCMN).	Surgical management to decrease the potential for malignancy and to functional and cosmetic improvement	After 9 years, there was growth of satellite lesions that did not require new surgical interventions nor presented with malignancy.
Endomba et al. ⁽⁷⁾	12-year-old girl	Progressively extensive, pruritic, and painless pigmented skin lesion on the back, persistent since she was 2-days old.	None.	<ul style="list-style-type: none"> • A large, irregular, well-demarcated and unequally pigmented (bluish-brown to black) multinodular hypertrophic nevus occupying almost all her back. • Largest diameter was 45 cm. • Surface was rough and had several excoriation marks. 	None.	Giant congenital melanocytic nevus (GCMN).	Planned for surgical excision and referred to a psychologist.	Followed up clinically and psychologically on a weekly basis while waiting for surgery.

Abubakar et al. ⁽⁸⁾	6-month-old girl	Hyperpigmented skin lesions over the back, gluteal regions, perineum, thighs, and scalp since birth.	Increasing number of other smaller satellite lesions on the trunk, extremities, and skull, increasing in size as compared to initial ones in the preceding 4 weeks.	<ul style="list-style-type: none"> • Extensive black nevus on the back extending from T12 vertebra to the gluteal, right iliac, right femoral, and perineal regions. measuring 31 cm in its largest diameter. Multiple satellite nevi on the abdomen, back, scalp, and extremities, with diameters ranging from 0.5 to 2 cm. 	<ul style="list-style-type: none"> • Full blood count, and renal and liver function tests were normal. • EEG, CT of the brain, and skin biopsy could not be done. 	Giant congenital melanocytic nevi (GCMN).	Lost to follow up.
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Giant congenital melanocytic nevus can darken or lighten over time, become more heterogenous or homogenous in its pigmentation, exhibit a rise in hair growth, develop a more uneven surface, or, less frequently, spontaneously regress. Lesion thickness typically increases with time. Nodules in the nevus typically correlate with the growth of neuroid tissue components.⁽¹⁹⁾ Hence, clinicians must educate the parents on what to watch out for as these developing lesions become malignant.⁽⁶⁾

Giant congenital melanocytic nevus is treated symptomatically and palliatively. The use of tissue expanders followed by resection, serial resection, or excision followed by skin grafts or substitutes are surgical methods for GCMN.⁽²⁰⁾ Surgery may be indicated based on the nevi's size and location, risk of cancer or neurocutaneous melanocytosis, potential psychological effects, and aesthetic considerations.⁽²¹⁾ Neurocutaneous melanosis is an uncommon condition that includes both CNS and leptomeningeal melanosis. The size, quantity, and location of CMNs directly correlate with the prevalence of NCM. A higher incidence of NCM is associated with nevi that originate in the posterior axis (head, neck, and trunk), presence of satellites, male gender, and neurological complications. Therefore, the presence of predisposing factors warrants further examination and management.⁽²²⁾ Dermabrasion, laser ablation, curettage, and chemical peeling are non-surgical treatments for GCMN.⁽²³⁾ One case report combined the use of Q-switched Nd:YAG (1064 nm), fractional CO₂ (10,600 nm), and diode (810 nm) lasers to treat GCMN.⁽²⁴⁾ However, these options may not be available everywhere in Indonesia.

CONCLUSIONS

While GCMN is a rare disease, it can potentially be malignant and a source of distress for the parents. Clinicians need to recognize this disease early, educate the parents empathically, and ensure that the proper treatment modalities are employed. It requires a correct diagnosis and proper management due to its risk of developing malignant melanoma. Regardless of what type of management is decided upon, be it surgical or observational, it must be remembered that most GCMN patients can have a healthy and productive life.

CONFLICT OF INTEREST

Author has no conflicts of interest to disclose.

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We thank the parents and family of the child who were willing to share this experience with all parents who may encounter similar situations and conditions. They have been very generous to share the challenges they faced and would like to use this opportunity to support other families.

Ethical Approval

The methods used in this study that involved human participants were done in line with the Helsinki Declaration of 1964 and any later revisions or equivalent ethical standards, as well as the institutional and/or national research committees. For the writing of this case report, ethical approval was not needed.

Informed Consent

Informed consent to be included in the study was obtained from the patient.

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Data Availability Statement

Not applicable.

Declaration Of Use Of AI in Scientific Writing

Nothing to declare.

Author's Contributions

The idea and design of the study, the gathering and processing of data, the analysis and interpretation of findings, and the writing of the paper all attest to the author's sole responsibility.

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