

Giant Congenital Melanocytic Nevus in a Pediatric Patient: Case Report

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Summary: The giant congenital melanocytic nevus is a rare entity with an extremely low incidence that appears at the time of birth and generally involves the dermis but may also affect other skin layers. According to its clinical evolution, the probability of malignancy may vary, so proper follow-up is essential for potential management. There is no consensus in the literature about the greater benefit of surgical versus nonsurgical management. In this case report, we present the surgical management of a school-aged patient using dermal substitutes and skin grafts, subjectively obtaining an improvement in his quality of life. (*Plast Reconstr Surg Glob Open* 2021;9:e3940; doi: [10.1097/GOX.0000000000003940](https://doi.org/10.1097/GOX.0000000000003940); Published online 17 November 2021.)

Although the etiology is unclear, it is believed that melanocytic nevi can appear due to a pathological accumulation of melanocytes of neuroectodermal origin in an ectopic location,¹ which occurs around the fifth and 25th weeks of gestation.^{2,4} They can be classified into nevi that appear at birth, called congenital melanocytic nevi (CMN), or ones that appear after birth, called acquired melanocytic nevi.^{1,2,7} Within the CMN, there is a size subclassification where small, medium, and large/giant nevi are found.

Its importance arises from the potential risk of malignancy, the inherent psychosocial impact of the lesion, and its aesthetic effect. Adequate management of the lesion, whether surgical or nonsurgical, is definitive for a successful outcome. We describe a case of giant congenital melanocytic nevus, where satisfactory progress was achieved after total resection and the use of dermal substitutes and partial thickness skin grafts.

CLINICAL CASE

A 6-year-old male child presented to the outpatient setting with 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 28% of his body weight, located on the back and abdomen, and satellite lesions in the extremities, face, and scalp. Marked splenomegaly with increased abdominal perimeter was found on the physical examination (Fig. 1). Two skin biopsies were performed; the first biopsy was obtained from the largest lesion and reported intradermal melanocytic nevus with congenital traits and lateral resection margins compromised by the lesion. The second biopsy was taken from a nodular hyperpigmented lesion on the left knee and reported a compound melanocytic nevus.

The histological pattern was characterized by predominant melanocytic cell presence and architecture, hyperpigmented and circumscribed mainly to the dense, reticular superficial dermis. Visceral and cerebral involvement was ruled out with magnetic resonance imaging. Given these findings, a multidisciplinary consensus discussed the surgical management to decrease the potential for malignancy and functional and cosmetic improvement. The patient underwent an initial surgery that removed 90% of the lesion which was found in the posterior trunk, and a second surgery 8 days later that removed the last 10%, achieving full removal. The reason why the procedure was performed this way was exclusively for the patient's comfort and preferences for the postoperative period. The procedure was performed on the right side of the abdomen, from caudal to cephalic and medial to lateral direction, using cautery coagulation for bleeding control, after administration of Toledo solution under the nevus to achieve vasoconstriction. The exposed area was covered with sheets of dermal matrix (Integra), fixed with mechanical staples (Fig. 2), and a vacuum-assisted closure system at continuous 150 mm Hg was placed. The partial skin graft procedures began fifteen days after the

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Fig. 1. A 6-year-old male child with a tumoral mass covering approximately 20% of the total body surface and 28% of his body weight.

total resection, taking the grafts from the lower limbs and gluteal areas. Twelve procedures were needed in total to graft enough tissue to cover the full extent of the exposed area. Between each procedure, it was necessary to allow the epithelialization of the donor area.

The patient was hospitalized in the pediatric ward for medical and rehabilitation support. He was discharged 3



Fig. 2. The exposed area was covered with sheets of dermal matrix (Integra) and fixed with mechanical staples.

months after the resection, with satisfactory postoperative progress, with the integration of more than 90% of the skin grafts and adequate epithelization of donor areas. After resection of the gigantic congenital melanocytic nevi (GCMN), the spontaneous resolution of organ enlargement was observed. The patient has been followed for 9 years (Fig. 3), and despite the growth of satellite lesions, he has not required new surgical interventions nor has he presented malignancy. Additionally, although it is assessed subjectively, the patient's quality of life has circumstantially improved.

DISCUSSION

GCMN are lesions that are present at birth and that will reach a size of more than 20 cm in adulthood,² lesions that affect a significant portion of a specific area (the face, hand, etc.), or nevi that involve more than 1% of the total body surface area on any anatomical location.¹ Other authors define GCMN as bigger than 100 cm or lesions that cannot be resected with only one procedure.⁴ CMN form when there is abnormal proliferation and migration of melanoblasts, which migrate from the neural crest to the skin, mucosae, leptomeninges, mesentery, eyes, and ears.^{1,2,4} This can be caused by genetic abnormalities such



Fig. 3. Eight years after surgery.

as NRAS (Q61) and BRAF (V600) mutations, in addition to the involvement of hepatocyte growth factor/scatter factor (HGF-SF).^{1-3,7,8,9}

Histology shows intradermal or compound involvement, with melanocytes that can reach even the muscle, glands, vascular walls, nervous structures, and fasciae.⁵ There are many classifications available. Kopf et al proposed the most widely used, defining the CMN according to size and largest diameter. Small nevi are smaller than 1.5 cm, medium nevi are between 1.5 cm and 19.9 cm, and large or GCMN are larger than 20 cm.¹⁰ However, GCMN can also include those with a projected adult size larger than 20 cm, according to location and growth factors (projected size in the head is multiplied by a factor of 1.7; by 2.8 in the neck, by 2.8 in the trunk, and by 3.3 in the lower extremities).¹¹⁻¹³ Similarly, those lesions cover more than 1% of total body surface area on the face and neck, or 2% on any other anatomical location.¹⁴

The incidence of CMN in newborns has been reported to be between 0.2% and 2.1%, and it has been observed to affect women slightly more frequently (male/female ratios between 1:1.17 and 1:1.4).¹⁵ Its presence increases the risk of neurocutaneous melanosis and malignant melanoma,² which usually presents in one of two forms: the appearance of extracutaneous melanocytes in the central nervous system (CNS) or malignant degeneration.¹ Neurocutaneous melanosis arises in abnormal neuroectodermal development, which leads to unregulated proliferation of melanocytes in inadequate sites such as the CNS.¹ The presence of multiple satellite lesions and the involvement of the midline of the trunk or head increase the risk of CNS involvement. The gold standard for CNM diagnosis is magnetic resonance imaging of the brain and spinal cord, usually between the 4 and 6 months of life due to the cerebral myelination timeline.⁴ The development of malignancy with the CMN is the primary rationale for removal, but there is no precise information about the incidence of malignant transformation. Varying rates have been reported. Zaal et al¹⁶ performed a retrospective study on the national pathology database of the Netherlands and concluded that the incidence rate of malignant melanoma was higher than the expected rate of the general population. These patients have a risk 12 times higher than the general population, and women have an even higher risk of malignancy (14.1) versus men (6.4). Additionally, the study showed that GCMN have an even higher risk of malignant melanoma, with an incidence rate of 51.6.

A prospective study by Viana et al¹⁷ conducted between 1999 and 2011, including all patients with GCMN of the Giant Congenital Melanocytic Nevus Registry of the Minas de Gerais Federal University concluded that the lifetime risk of malignant melanoma in these individuals was around 5%. The most common location was the trunk (68.4%), followed by head and neck (17.5%), and then the extremities with 14.1%, which is why they suggest they tend to appear on axial locations. The GCMN were accompanied by satellite lesions in 84.2% of cases, which are themselves associated with an increased risk of malignant melanoma and cutaneous melanosis. Both Viana et al and Zaal et al^{16,17} recommend surgical excision as long as it is

feasible to decrease the number of nevomelanocytic cells, and thus reduce the risk of malignancy, under the assumption that the malignant transformation occurs inside the nevus. Krengel et al recommend prophylactic excisional surgery, considering that a systematic review of 14 studies with 49 cases of CMN melanoma, 67% of patients had malignant transformation within the nevus.¹⁵

Additionally, Krengel et al recommend prophylactic excisional surgery, considering that a systematic review of 14 studies with 49 cases of CMN melanoma, 67% of patients had malignant transformation within the nevus.¹⁵ Even though there is still debate about the reduction in malignant melanoma incidence with surgery, the aesthetic, and even functional improvements cannot be denied, and this alone may be the reason why the patient and family seek this type of treatment.

Different management strategies have been proposed, such as tissue expanders⁶ and partial and full-thickness skin grafts, but all of them have limitations and disadvantages. Schiessler et al¹⁸ proposed a study using Integra (Integra Artificial Skin) for surgical management of GCMN and found that it can be used successfully to cover all skin defects after complete nevus excision. Management consisted of two steps: resection of GCMN with Integra placement and then either partial or full-thickness skin grafts, depending on the case. The initial integration rates of the dermal matrix were 95%–100%. The risk of complications was relatively high (33%), but it reflected minor complications like loss of Integra or graft integration, or infections requiring partial or total removal of grafts. Complications were correlated to patient age and the size and anatomical location of the lesion, with the majority of cases presenting below the median age (3.8 years) and lesions above the median size (4.2% BSA). Results were measured according to a subjective scale where results with other methods were also considered, but we agree with the authors that such scoring systems are inadequate because they are subjective and have not been validated. Concerns regarding surgical excision can include the challenges that emerge to achieve a good aesthetic outcome, the need for multiple surgical procedures or the psychological impact, especially in facial locations.



Fig. 4. An adequate integration of the grafts without retraction was obtained due to the dermal matrix.

Early identification of possible complications is important for patients with a diagnosis of GCMN, by means of periodical follow-up visits, with skin evaluations and assessment of a multidisciplinary team if needed. Also, multiple factors (including age of initial presentation, neurological development, quality of life, emotional and behavioral concerns, pain, and stigmatization) must be evaluated and reevaluated in each consultation.²⁰

CONCLUSIONS

Giant congenital melanocytic nevi are uncommon entities that appear in the newborn and generally involve the superficial and deep layers of the skin, as well as other tissues. According to its evolution, the probability of malignancy increases or decreases, so proper monitoring is essential as a control method and potential management. The final result is striking, where an adequate integration of the grafts without retraction was obtained due to the dermal matrix that provides an adequate scaffolding for said grafts (Fig. 4). As demonstrated above, there was an evident improvement in terms of the patient's quality of life, mobility, and functionality, which is why it is considered a good alternative for this type of patient.

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