



## Large plaques on a baby boy

This newborn's prenatal lab work and delivery were normal. So, too, were his Apgar scores and neurologic exam. So why was he covered in brown and black plaques?

A 25-YEAR-OLD G2P1 MOTHER gave birth to a boy at 40 and 6/7 weeks by vaginal delivery. Labor was induced because of oligohydramnios complicated by chorioamnionitis. The mother was treated with vancomycin and gentamicin. Prenatal lab work and delivery were otherwise unremarkable.

The delivering physician (CG) noted that the neonate had numerous brown, red, and black plaques distributed over his abdomen, lower back, groin, and thighs (FIGURE). Some

plaques were hypertrichotic and other areas, apart from the plaques, were thinly desquamated. Apgar scores were 8 and 9 and the remainder of the exam, including the neurologic exam, was normal. The Dermatology Service (JK) was consulted.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

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**FIGURE**

**Newborn with multiple plaques and areas of thin desquamation**



IMAGE COURTESY OF CARRIE GRIFFIN, DO

➤ **By age 10, up to 8% of patients with giant congenital nevi will develop melanoma within the nevi.**

**Diagnosis:  
Giant congenital nevus**

Congenital melanocytic nevi (CMN) are pigmented lesions that are present at birth and created by the abnormal migration of neural crest cells during embryogenesis.<sup>1</sup> Nevi are categorized by size as small (<1.5 cm), medium (1.5-20 cm), large (>20 cm), and giant (>40 cm).<sup>2</sup> Congenital nevi tend to start out flat, with uniform pigmentation, but can become more variegated in texture and color as normal growth and development continue. Giant congenital nevi are likely to thicken, darken, and enlarge as the patient grows. Some nevi may develop very coarse or dark hair.

CMN can cover any part of the body and occur independent of skin color and other ethnic factors.<sup>3</sup> Giant congenital nevi are rare, with an incidence of approximately one in 50,000 live births and with males and females equally affected.<sup>3,4</sup> The condition is diagnosed at birth, based on the appearance of the lesions.

■ **The differential diagnosis** for CMN includes café au lait macules, blue-gray spots (aka Mongolian spots), nevus of Ota, nevus spilus, and vascular malformations (TABLE).<sup>5</sup> CMN may present in almost any location and may be brown, black, pink, or purple in color. Café au lait macules, blue-gray spots, nevus of Ota, nevus spilus, and vascular malformations have individual location and color characteristics that set them apart clinically.

**Monitor patients  
for melanoma, CNS complications**

Patients with CMN are at increased risk of neurocutaneous melanosis (NCM) and cutaneous melanoma.

■ **Neurocutaneous melanosis**, a complication of giant congenital nevi, is a melanocyte proliferation in the central nervous system (CNS). Between 6% and 11% of patients with giant congenital nevi develop symptomatic NCM in childhood. Thus, any CNS symptoms should be fully evaluated.<sup>4,6</sup> NCM can result in seizures, cranial nerve palsy, hydrocephalus, and leptomeningeal melanoma.

Besides giant congenital nevi, risk factors for NCM include male sex, large numbers of satellite nevi, and the presence of nevi over the posterior midline or head and neck.<sup>7</sup> The prog-

nosis is poor for patients who develop neurologic symptoms. NCM is associated with other malignancies, including rhabdomyosarcoma, liposarcoma, and malignant peripheral nerve sheath tumors.<sup>4</sup>

Magnetic resonance imaging (MRI) is helpful to exclude NCM. Ideally, an MRI should be ordered before 4 months of age, at which time myelination begins to make the identification of melanin deposits in the CNS more challenging.<sup>7</sup> Not all patients with imaging findings that are consistent with NCM will develop symptoms.<sup>8</sup>

■ **Melanoma.** By age 10, up to 8% of patients with giant congenital nevi will develop melanoma within the nevi; most of these cases occur during the first 2 years of life.<sup>7,9</sup> Patients with NCM are at even greater risk: their rate of malignant melanoma is between 40% and 60%.<sup>6</sup> As a result, patients should be monitored closely for any signs of the disease. Total body photography, serial clinical photos, and patient self-exam are helpful to detect changes and de novo lesions. New lesions or ulcerations superimposed on existing nevi may indicate malignancy.<sup>7</sup> Sun protection is critical to reduce the risk of melanogenesis.

**Should patients pursue surgery?  
It's debatable**

Options for patients with large and giant CMN include early curettage (prior to 2 weeks of life), local excision (often with tissue expansion), dermabrasion, and laser therapy.<sup>2</sup> There is considerable debate about surgery. Advocates of surgery cite psychosocial relief as a major treatment benefit and speculate about prevention of melanoma. Opponents worry that excessive surgical intervention may cause melanogenesis in a scar or deep in an area of treatment. And, while smaller congenital nevi are easier to surgically remove, they have a low associated risk of developing melanoma and are typically monitored clinically.

**Children with congenital nevi  
will need support**

Several nonprofit organizations offer resources for children with congenital nevi and their

TABLE

## Differential diagnosis of congenital melanocytic nevi<sup>5</sup>

Diagnosis	Presentation
Café au lait macules	Often present as well-demarcated, brown, flat patches on the trunk. Relatively common and can be associated with syndromes such as neurofibromatosis.
Blue-gray spots (aka Mongolian spots)	Evenly pigmented blue to gray patches. Often on the sacrum in darker skin types. Tend to resolve with time and remain flat.
Nevus of Ota	Speckled pigmentation on the periorbital skin. Involves pigmentation of the sclera in darker skin types.
Nevus spilus	Also known as a speckled lentiginous nevus, this is a localized patch of various shades of brown to black macules and thin papules. It is often circumscribed to a patch with a lighter background than the rest of the skin.
Vascular malformations	Usually red to violet in color, these vascular malformations may be more deeply pigmented in some patients, resembling melanocytic proliferations.

families. Nevus Outreach ([www.nevus.org](http://www.nevus.org)) is an organization devoted to improving awareness and providing support for people with CMN and NCM. The group maintains a registry of patients with large nevi in an effort to help researchers improve treatment and identify a cure.

For children with congenital nevi and other skin conditions, the American Academy of Dermatology offers its “Camp Discovery” at locations across the country (<https://www.aad.org/public/kids/camp-discovery>). Camp Discovery provides full scholarships and includes transportation to each of the individual camps for attendees.

■ Our patient underwent an MRI on his fifth day of life. The results were normal and he hadn’t developed any neurologic symptoms at 4 months of age. The child sees his family physician for routine well-child visits and a dermatologist annually. The dermatologist is carefully monitoring the nevi, which continue to grow.

JFP

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## References

1. Sarnat HB, Flores-Sarnat L. Embryology of the neural crest: its inductive role in the neurocutaneous syndromes. *J Child Neurol.* 2005;20:637-643.
2. Gosain AK, Santoro TD, Larson DL, et al. Giant congenital nevi: a 20-year experience and an algorithm for their management. *Plast Reconstr Surg.* 2001;108:622-636.
3. National Organization for Rare Disorders. Giant congenital melanocytic nevus. National Organization for Rare Disorders Web site. Available at: <http://rarediseases.org/rare-diseases/giant-congenital-melanocytic-nevus>. Accessed April 29, 2016.
4. Vourc’h-Jourdain M, Martin L, Barbarot S, aRED. Large congenital melanocytic nevi: therapeutic management and melanoma risk: a systematic review. *J Am Acad Dermatol.* 2013;68:493-498.e1-e14.
5. Jackson SM, Nesbitt LT. *Differential Diagnosis for the Dermatologist.* 2nd ed. Berlin: Springer; 2012.
6. Jain P, Kannan L, Kumar A, et al. Symptomatic neurocutaneous melanosis in a child. *JAMA Neurol.* 2013;70:516.
7. Kinsler VA, Chong WK, Aylett SE, et al. Complications of congenital melanocytic naevi in children: analysis of 16 years’ experience and clinical practice. *Br J Dermatol.* 2008;159:907-914.
8. Agero AL, Benvenuto-Andrade C, Dusza SW, et al. Asymptomatic neurocutaneous melanocytosis in patients with large congenital melanocytic nevi: a study of cases from an Internet-based registry. *J Am Acad Dermatol.* 2005;53:959-965.
9. Zayour M, Lazova R. Congenital melanocytic nevi. *Clin Lab Med.* 2011;31:267-280.



For children with congenital nevi and other skin conditions, the American Academy of Dermatology offers summer camps across the country.



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