

A Newborn with Giant Scalp Nevus and Satellite Melanocytic Nevus on the Rest of the Body

Jorge Sales Marques*

Department of Pediatric and Neonatology, Centro Hospitalar Conde S. Januário, Macau, China

***Corresponding Author:** Jorge Sales Marques, Department of Pediatric and Neonatology, Centro Hospitalar Conde S. Januário, Macau, China.

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Abstract

Large and giant congenital nevi, particularly those on the head, neck and back, may be associated with a condition termed neurocutaneous melanosis (NCM). Patients with NCM have a proliferation of nevus cells in the central nervous system (leptomeningeal melanocytosis) and are predisposed to seizures, malignant melanoma of the CNS, and neurologic symptoms related to increased intracranial pressure or spinal cord compression.

A 4-day life twin 1 girl was transferred from the maternity because of multiple skin nevus lesions over the body with the larger one covering more than 50% of the scalp region. Brain MRI is normal in our case but this cannot rule out NCM. Management of extensive CNN should be individualized on a case-by-case basis. The prognosis for symptomatic NCM is poor, with > 90% of patients dying of the disease and around 70% of those dying before 10 years of age.

Keywords: Neonatal Neurocutaneous Melanosis; Brain MRI

Background

Large and giant congenital nevi, particularly those on the head, neck and back, may be associated with a condition termed neurocutaneous melanosis (NCM).

NCM, as well as melanoma risk, appears to be more common in patients with greater numbers of satellite nevi.

Male and female, the same as all races, are equally affected. The incidence of large/giant NCM is 0.005% of the population. The etiology is likely multifactorial. Animal models suggest that aberrant expression of the hepatocyte growth factor/scatter factor (HGF/SF) may be involved. Rare familial cases have been reported.

Congenital melanocytic nevi are derived from neural crest-derived melanoblasts embryologically migrating to form the nevus after 10 weeks in utero but before the sixth uterine month. It is unclear what causes these melanoblasts to migrate differently from other melanoblasts when forming the skin.

Histologically, melanosis may be seen at the base of the brain, ventral surfaces of the pons, medulla, upper cervical, or lumbosacral spinal cord.

Patients with NCM have a proliferation of nevus cells in the central nervous system (leptomeningeal melanocytosis), and are predisposed to seizures, malignant melanoma of the CNS, and neurologic symptoms related to increased intracranial pressure or spinal cord compression. In symptomatic cases macrocephaly may be seen as a result of hydrocephalus with or without increased intracranial pressure, which may be due to obstructed CSF flow, excess absorption due to subarachnoid infiltration by pigmented cells, or associated Dandy-Walker syndrome [1-5].

Case Report

A 4-day life twin 1 girl was transferred from the maternity because of multiple skin nevus lesions over the body with the larger one covering more than 50% of the scalp region (Figure 1).



Figure 1: A giant nevus covering more than 50% of the scalp.

The baby born by cesarean section. Gestational age of 37 weeks. Rupture of membranes < 6 hours. Amniotic fluid is clear. Birth weight: 2.2 kg.

Apgar10/10/10. She is a twin 1. The other twin 2 is normal at birth with no skin lesions.

The maternal history is unremarkable. This is a G2P3 pregnancy. No history of intrauterine infection during this period.

On physical examination revealed a normal phenotype. A giant nevus on the scalp was observed. Some small nevus was found over the whole body but two bigger one was detected on the lower back (Figure 2). Heart and lungs auscultation, abdomen, muscle tone and reflex and anterior fontanelle are all normal.



Figure 2: Satellite melanocytic nevus on the rest of the body.

Blood test showed complete blood count and biochemistry within normal range. The abdominal ultrasound showed normal result. Heart ultrasound revealed a persistent ductus arteriosus. Brain ultrasound demonstrated echogenic intraparenchymal lesions. The brain MRI is normal at this stage.

Discussion

Leptomeningeal melanoma develops in 40 - 62% of symptomatic patients, often within the first 3 years of life, though delayed presentations after puberty are reported.

The symptoms are lethargy, irritability, headache, recurrent vomiting, seizures, increased head circumference, bulging anterior fontanelle, photophobia, papilledema, neck stiffness and occasionally nerve palsies, particularly of cranial nerves VI and VII [6].

Magnetic resonance imaging (MRI) to screen for NCI should be considered in neonates with large posterior axial lesions or multiple satellite nevi. Asymptomatic NCI can be monitored with repeat scans.

NCM reveals focal areas of high signal on T1-weighted images in one or multiple areas of the brain, including the lobes, cerebellar hemispheres, pons, medulla and anterior temporal horns, particularly the amygdala. T2 shortening may also occur.

A normal MRI does not fully rule out NCM. Cytologic examination of cerebrospinal fluid may be necessary [1-3].

The differential diagnosis includes melanoma, including prenatal metastases.

In one study, 45% of neurologically asymptomatic children with NCI had these radiologic findings.

However, a questionnaire-based study of 186 patients with large congenital nevi who were imaged revealed that only 4.8% of those with positive MRI findings for NCM were asymptomatic. Hence, the exact prevalence of asymptomatic NCM remains unclear.

Overall, the prognosis for symptomatic NCM is poor, with > 90% of patients dying of the disease, and around 70% of those dying before 10 years of age.

Management of extensive NCM should be individualized on a case-by-case basis.

There are many factors that must be considered in the decision-making process regarding surgical excision of such lesions.

Large congenital nevi are often treated with surgical excision. The primary reason for this recommendation is the potentially-decreased risk of malignant transformation, although controlled studies supporting this hypothesis are lacking.

Unfortunately, excision of large lesions does not usually result in complete removal of all nevus cells, and melanoma may still develop from this residual tissue.

Large/giant NCM (20 cm) are clinically difficult to manage since the risk of malignant transformation is present at birth, plus they are logistically more difficult to remove.

It is important to follow these lesions clinically with measurements and/or photographs. Complete surgical removal of the lesion is difficult and often requires multiple surgeries with tissue expansion, skin grafting, and/or artificial skin replacement. In addition, melanoma may develop with increased frequency at extracutaneous sites in these patients, and the significance of neurocutaneous melanosis (if present) must also be factored into this decision. Melanoma arises in ~5 - 15% in the reported literature. Other tumors developing in NCM: schwannoma, neuroid tumor, lipoma, rhabdomyosarcoma, neurofibroma and others.

Partial thickness removal techniques (i.e. dermabrasion, curettage, and laser therapy) have been advocated by some, but the impact of these procedures on malignant transformation or clinical surveillance of the lesion must be further defined. Nowadays are not recommended [5].

A multidisciplinary approach must be employed for families of children with large congenital nevi. This includes the primary care physician, dermatologist, plastic surgeon, and diagnostic radiologist. Emotional support should be provided, and the family should be given information on support groups.

Our index case, the newborn showed a giant nevus on the scalp, with satellite melanocytic nevus on the rest of the body. Although the brain MRI was normal at this moment, we cannot rule out NCM. We decide not to do the lumbar puncture for the cytologic examination of cerebrospinal fluid during this stage and follow up the skin lesions with short time observation in the outpatient and repeat brain MRI, if necessary.

Conclusion

In a case of giant nevus on the scalp with satellite melanocytic nevus on the rest of the body, neurocutaneous melanosis need to be rule out. Normal brain MRI does not exclude the diagnosis. The prognosis is poor, due of the huge extension of the skin lesions.

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