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Congenital melanocytic nevus - important facts and current opinions regarding management and treatment necessity

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Abbrevations: NCM Neurocutaneous Melanosis; CNS: Central Nervous System; MRI: Magnetic Resonance Imaging

Editorial

Congenital melanocytic nevi are present at birth or develop during the first year of life. They derived from neural crest melanoblasts and present in 1-2% of newborn infants. They begin as pale brown to tan macules, which become darker and more elevated during adolescence. Usually the nevi are growing proportionately with the child and are asymptomatic for life. As the child grows older, the lesions develop coarse terminal hairs and may become more verrucous in appearance, hence may possess a cosmetic concern depending on their location.

The classic and most useful classification of congenital nevi is based on the size of the nevus, and divided into 3 groups according to size:

i. Small: Less than 1.5 cm

ii. Medium: 1.5-19.9 cm

iii. Large or giant: Greater than 20 cm

It is important to measure every individual nevus and to use this classification method since the natural history, prognosis, risk for malignant transformation and management is based on size.

In general, management of congenital melanocytic nevus could be divided into two main approaches: first is surgical remove and second is conservative with monitoring and education.

Small and medium benign- appearing congenital nevi can be safely monitored clinically. Their risk of malignant transformation is too low to warrant removal, thus patients should be instructed about signs of malignant transformation and regular periodic skin examinations should be suffice. I would recommend referral to dermatologist for baseline photo and documenting of the lesion and for dermoscopic evaluation. Patients and their parents should be familiar with the "ABCDE" mnemonic for the signs of melanoma:

i. Asymmetry

ii. Borders (irregular)

iii. Color (variegated)

iv. Diameter change (other than expected proportional growth with the child)

v. Evolving over time

Although the size of the lesion is the most important factor in regard to management, other factors may require a removal rather than conservative approach. Such factors are:

i. Other risk factors for melanoma (fair skin, freckling, light hair, personal/family history of melanoma, Immune suppression etc.)

ii. Location that may be difficult to follow up (underwear area, scalp)

iii. Atypical appearance

iv. Cosmetic concern

Large or Giant congenital nevi (>20cm) occurs in 1:500,000 newborns and always present at birth. The overall lifetime risk of developing melanoma is 4.5-10% and it is usually occur before age of 5 years. In this case as well, I would recommend referral to dermatologist and the plastic surgeon to consider complete removal which may be the best way to prophylactically decrease the risk for malignant transformation (although to date: no controlled studies to support this theory). Complete removal of giant congenital nevus often requires multiple surgeries with tissue expansion, skin grafting, and/or artificial skin replacement. It is eminent to mention that even with complete removal of the nevus; melanoma may still arise from remnants of the lesion or from extra-cutaneous sites. Partial thickness removal techniques such as dermabrasion, curettage and laser, are not recommended due to lack of data regarding efficiency, and due to the lack of penetrance to deeper tissue levels. This in turn may result in high recurrence rates and may evade clinical detection until the melanoma reaches a more advanced stage, since it may be more likely to occur deeper in the tissue.

Aside from melanoma, other complication of large or giant congenital nevus could be neurocutaneous melanosis and other malignancies (rhabdomyosarcoma, liposarcoma, malignant peripheral sheath tumor). Neurocutaneous melanosis (NCM) is the presence of congenital melanocytic nevi and melanotic neoplasms of the central nervous system. It is results from an error in the morphogenesis of the neuroectoderm and associated with large or giant congenital nevus that usually located on head, neck and back. This may result in increased intracranial pressure due to hydrocephalus or a mass lesion and malignant melanoma of the CNS. Symptoms usually present

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during the first 3 years of life and prognosis of symptomatic NCM is poor, >90% die (70% before 10 years of age). An MRI to screen for neurocutaneous melanosis should be considered in neonates with large posterior axial lesions or multiple satellite nevi. Asymptomatic neurocutaneous melanosis can be monitored with repeat scans. Symptomatic neurocutaneous melanosis carries such a poor prognosis that surgical removal of the large congenital melanocytic nevus is typically not pursued.

To conclude, children with small and medium congenital nevi are usually monitored without the need for any intervention, not like in the case of large or giant congenital nevus that require surgical management. It is recommended to refer those patients with congenital nevi to a dermatologist for base line evaluation and proper documentation and education.

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Conflicts of Interest

There is no conflict of interest.

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