**Giant Hairy Nevus - A Case Report**

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**Introduction:** Newborns can present with a host of varying birth marks. Nevi can represent a benign condition or be a harbinger of a more concerning underlying condition or genetic syndrome. Hyperpigmentation in the newborn is very common and has a wide spectrum of presentation from large areas of dark discoloration to spotty segmental patches to solitary light brown macules. Some nevi may have increased vascularity. These nevi are of special significance because of their association with leptomeningeal melanocytosis and their predisposition for development of malignant melanoma. Review of the literature reveals that congenital malignant melanoma is an exceptionally rare occurrence and has a generally poor prognosis. Congenital nevi have been stratified into 3 groups according to size. Congenital nevi, depending on size and location, may have a significant impact on the shape of the body. No racial predilection is recognized for congenital nevi. Congenital nevi occur in both sexes. Treatment consists of frequent examination to check for skin cancers and surgery to remove the nevus. Larger nevi may need to be removed in several stages. All birth marks should be evaluated by healthcare provider. We report a case of Giant Hairy nevus in a neonate.

**Keyword:** Congenital giant hairy nevus.

**Case report:** A male neonate with skin abnormality since birth was born to a 28 years old primigravida mother, at full term after Cesarean section. Patient’s birth weight was 2.75 kilogram, length 48 centimeter and head circumference 34 cm. Skin lesion covered posterior trunk, scalp, perineal region & left leg. Small nevi of 3-4 cm diameter were also present on the right upper and lower limb, abdomen and forehead above the left eye (See Fig.1 below). Clinically there was no neurological deficit and other systems were normal. The baby was on breast feeding and was discharged in stable condition.

**Discussion:** Congenital melanocytic nevi (CMN) occur in approximately 1% of infants but giant congenital nevi occur in 1 in 1000 to 1 in 20,000 newborns.
Garment nevi involve a very large body surface area; for example the entire back or an extremity and occurs in about 1 in 500000 newborns (1). Nevi result from a proliferation of benign melanocytes in the dermis, epidermis or both (2). Giant congenital pigmented nevi occur most commonly on the posterior trunk but may also appear on the head or extremities. They may be present at birth or develop within the first 2 postnatal years, they appear as confluent brown-to-black patches or plaques with possible satellite lesions. The surface may have a verrucous or rugated texture with pigment irregularity. They have an irregular margin and often long dark hair and may become increasingly lobulated and hairy with time (3) by convention, they are described as small, intermediate, large, or giant. Small CMN are 1.5 cm or less, intermediate are between 1.6 cm and 19.9 cm, and large are 20.0 cm or larger. Some authors describe giant CMN as those greater than 40 to 60 cms. In infants, large CMN correspond to nevi that measure 9.0 cm on the head or 6.0 cm on the body. Nevus cells within the leptomeninges and brain parenchyma may cause intracranial pressure, hydrocephalus, seizures, retardation, motor deficits and may result in melanoma (4). In the absence of neural melanosis early excision and repair aided by tissue expanders is useful tool for reconstruction or grafting and reduce the burden of nevus cells (5). There is currently much debate about the risk for melanoma in small and intermediate CMN. Many recent studies estimate the risk of development of melanoma in these lesions to be no greater than that for acquired nevi, fortunately melanoma remains an uncommon malignancy in prepubertal children, with an annual incidence of 0.7 cases per million children aged 0-9 years (6). Treatment is, therefore, dependent on other risk factors. When changes within the lesions occur, a skin biopsy and possible surgical excision may be warranted. Excision may also be undertaken due to esthetic concerns. Many authorities also recommend removal of lesions in areas that are hard to monitor clinically for malignant changes, such as the scalp. Patients who have large CMN are at risk for myriad problems that may arise within the nevi, including rhabdomyosarcoma, neuroblastoma, liposarcoma, primitive neuroectodermal tumors, and more benign findings such as subcutaneous fat atrophy, benign proliferative nodules, and hemangiomas (7).

The two associations that are most common are melanoma and neurocutaneous melanocytosis (NCM). The increased risk of melanoma in large CMN is from 2% to 18% however, the most recent studies and many authorities now believe that the correct percentage is between 5 to 8%. More than 50% of such melanomas occur before the age of 5 years, with most occurring before the prepubertal years. Unfortunately many of the melanomas that arise within these lesions are dermal and thus, difficult to detect therefore, if any worrisome changes occur, the clinician should biopsy the lesion to rule out malignancy. The risk of Cutaneous melanoma has been considered higher in large axial CMN (posterior midline lesions), but recent reviews have questioned this thinking and after multivariate analysis determined that large nevi in a truncal location, not axial, increase this risk, although confounding factors may affect this finding (8,9) The risk for NCM is higher in patients who have multiple (10) satellite lesions, although this particular numeric value is arbitrary and not based on established evidence (11). Patients who have large axial CMN and headache or other central nervous system symptoms should receive magnetic resonance imaging evaluation, but when and if asymptomatic patients should obtain such imaging is still under debate. Research on both adult patients and parents of the patients has demonstrated that satisfaction with medical encounters predicts important outcomes such as compliance with the medical regimen (12). On the basis of these data, algorithms for the extent of resection and subsequent reconstructive options for giant congenital nevi were developed. Larger nevi need to be removed in several stages. Many treatment options have been postulated for giant CMN,
including dermal abrasion, CO2 laser ablation, phenol chemical peels, curettage, excision and skin grafting, use of expanded full thickness skin grafts, serial excision, tissue expansion, and free tissue transfer (13). Research on both adult patients and parents of pediatric patients has demonstrated that satisfaction with medical encounters predicts important outcomes such as compliance with the medical regimen. Recent practice has been one of surgical excision after staged tissue expansion, and the final goals are reduction and control of symptoms, aesthetic improvement, psychosocial well-being, maintenance of function, and decrease in the risk of malignant degeneration (14). All birthmarks should be evaluated by healthcare provider. A biopsy of suspicious areas may be obtained for examination to determine if the cells have become cancerous. An MRI of the brain might be performed if the lesion is over the spine to prevent melanoma in all people, but particularly those with large congenital pigmented naevi, reducing ultra violet light exposure, reducing peak sunlight exposure during most intense sun periods. Use of hats and clothing to shield from intense sunlight (15,16).

References:
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Conflict of Interest: None
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