

Case Report

Giant congenital melanocytic nevi (garment variety): a case report

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ABSTRACT

Giant congenital melanocytic nevi (GCMN) are large brown-to-black skin lesions caused due to genetic mutations which lead to defective proliferation, differentiation and migration of melanoblasts which are precursor cells of melanocytes. There is a mutation in the NRAS gene causing abnormal proliferation of embryonic melanoblasts. Congenital melanocytic nevus is primarily a clinical diagnosis. The malignant melanoma and neurocutaneous melanosis are the two major complications associated with GCMN. The risk of transformation of GCMN to malignant melanoma varies between 0 and 3.8%. About 1% of live births presents with a CMN. The incidence of GCMN is estimated at less than 1: 20,000 newborns. The variety 'garment-like' of GCMN is even scarcer, 1: 5,00,000. GCMN has got major psychosocial impact on the patient and his family due to its unsightly appearance. Treatment includes surgical and non-surgical procedures, psychological intervention and clinical follow-up, with special attention to changes in color, texture on the surface of the lesion. We presented a case of 1-day-old female neonate born with GCMN in our hospital.

Keywords: Giant congenital melanocytic nevi, Neurocutaneous melanosis, Malignant melanoma, Nevus, pigmented, Skin neoplasia

INTRODUCTION

The melanoacytic nevi are benign proliferations of melanocytic cells arranged in nests in the epidermis, dermis or in other tissue.¹⁻⁴ These are pigmented lesions present at birth or in the first year of life without a known etiology. They are large brown to-black lesions caused due to genetic mutations which lead to defective proliferation, differentiation and/or migration of melanoblasts, the precursor cells of melanocyte.¹⁻⁴ GCMN with a diameter greater than 20 cm have an estimated incidence of between 1/20,000 and 1/500,000 births.

The risk of transformation of GCMN to malignant melanoma varies between 0 and 3.8%.⁵ Besides malignant transformation, patients with GCMN need to be periodically assessed for neurological abnormalities and psychological impairment. GCMN are usually managed surgically.⁶ However, new chemotherapy-based therapies are being developed. GCMN may be associated with

severe complications such as malignant melanoma and central nervous system (CNS) involvement, along with major psychological impact on the patient and his family, due to its unsightly appearance.^{1,2,8}

'Garment-like' GCMN is rare 1: 5,00,000. The male to female ratio is 1.17:1 to 1.46.^{1,2} The indication for surgical intervention in GCMN is the emergence of a malignant neoplasm on the lesion.^{1,2,4}

It has been suggested by Kopf et al to classify the lesions by surface diameter: small lesion <1.5 cm, intermediate lesions between 1.5 to 20.0 cm and giant lesions >20.0 cm. In 75% of the cases CMNS are accompanied by small, multiple lesions called 'satellites'.

Their number must be recorded because a number >29 is connected to leptomeningeal involvement. We presented a rare case of 1-day-old female newborn with GCMN, discussing its etiology and latest in management.

CASE REPORT

This patient was a 1-day-old female neonate, presented to our pediatric department with diffuse and large black pigmented patch over lower thoracic, abdomen, pelvic, groin, upper thighs, buttocks, and the lumbosacral region of his body. The mother of the patient was a multigravida, and gave birth to her child via normal vaginal delivery at term without any complications. The parents of the child did not have a consanguineous marriage. None of the close family members had similar skin lesions.

Examination revealed an extensive pigmented nevus was observed encompassing the abdomen, pelvis, groin, upper thighs, buttocks, and the lumbosacral region like a garment. Within the pigmented nevus there were several irregularly shaped macules, papules, and plaques of various colors with areas of breaking in. On palpation the surface of the nevus was irregular. Hypertrichosis was absent (Figure 1 to 4).

No other congenital anomaly was observed. Neurological examination was unremarkable, A clinical diagnosis of GCMN was made. Routine blood and urine examination did not revealed any abnormalities. Biopsy sample revealed findings consistent with GCMN without malignant changes.

Transfontanellar ultrasonography and X-ray of the spine were unremarkable. The Patient has been advised frequent follow-up visits and an MRI scan of the brain and spine was unremarkable.



Figure 1: One day old newborn with garment variety GCMN. Extensive pigmented nevus abdomen, pelvis, groin, upper thighs, buttocks, and the lumbosacral region at the back, within the pigmented nevus irregular nodules seen with areas of skin giving in.



Figure 2: One day old newborn with garment variety GCMN. Extensive pigmented nevus around abdomen, pelvis, groin, upper thighs, buttocks, and the lumbosacral region.



Figure 3: One day old newborn with huge garment variety GCMN- genitalia involved- irregular surface, no hypertrichosis.



Figure 4: One day old newborn with garment variety GCMN. Whole of the back, buttocks and upper thigh involved in this newborn GCMN, Within the pigmented nevus there are several irregular macules, papules, and plaques of various colors with areas of thickening and wrinkling.

DISCUSSION

Congenital melanocytic nevus originates between the 5th and 24th weeks of intra uterine gestation. CMN and acquired nevus arise from an accelerated proliferation of cells of melanocytic lineage during fetal development.¹ The giant and medium CMN are formed as the proliferation starts, during migration of melanoblasts from the neural crest to the skin. Nevi are larger and deeper. It is believed that the development of melanocytes is partially controlled by the proto-oncogenes c-met and c-kit. The hepatocyte growth factor is a cytokine regulator of epithelial cells that express the tyrosine kinase receptor encoded by c-met. Over expression of this factor is associated with disorders of differentiation, proliferation and migration of melanocytes and may be the reason for the occurrence of GCMN.^{1,2}

In newborns it may have a lighter coloration and present few or no hair follicles, occurring as a macule or as an elevated lesion. The surface of the nevus may be papular, roughened, warty or cerebriform. The most frequent site of GCMN is the torso, followed by the limbs and head.⁹ GCMN may affect more than one body segment. For unusual sites the term 'GCMN in garment' has been coined, 'bathing trunk', 'stole' or 'coat sleeve' are other terms used for these nevi satellite lesions scattered over the skin surface are common in individuals with GCMN, in 78% of the cases. GCMN may also have important psychological consequences, especially when the lesions are very extensive and are located in visible areas such as the face. There may be impaired self-image along with emotional or behavioral problems.

One of the serious complications of giant congenital melanocytic nevus is neurocutaneous melanosis (NCM), a rare syndrome. It is characterized by the presence of benign or malignant melanocytic proliferations in the CNS associated with the occurrence of congenital melanocytic lesions. Melanocytic cells are found in large number in the leptomeninges of the brain and/or spinal cord.^{1,2,9,11}

Some babies may have diffuse lipomatosis, hypertrophy of cranial bones, atrophy of limbs, skeletal asymmetries involving hyper- or hypoplasia of soft tissues, scoliosis, urinary tract anomalies, capillary vascular malformations, cafe-au-lait spots, ectopic mongolian spots, fibroepithelial polyps and atopic dermatitis.^{1,2,6,10,13-15} Structural CNS malformations may be found, such as arachnoid cysts, choroid plexus papilloma, cerebellar astrocytoma, spinal dysraphism (associated with GCMN located in the lumbosacral region) and type I Arnold-Chiari syndrome (herniation of cerebellar tissue through the foramen magnum).

Dandy-Walker malformation may be associated with NCM or occur in the absence of CNS involvement by melanocytic cells. It is characterized by cystic enlargement of the fourth ventricle, aplasia or hypoplasia of the

cerebellar vermis and an increase of volume of the posterior fossa with or without hydrocephaly.^{1,2,9,10,14}

During neonatal period, various types of melanocytic tumors may arise on GCMN, and are distinct from melanomas but may mimic neoplasms. These proliferative lesions have varied sizes and can present with rapid growth and ulceration. During this period tumors with the most worrisome clinical features tend to have a benign behavior, and show stabilization or regression after a period of rapid growth. Nodules appearing on GCMN in newborns can be very difficult to classify histologically, but despite the similarity with melanoma-simulating lesions, they usually show some maturation and do not exhibit the same degree of atypia or the high mitotic rate observed in malignant melanocytic neoplasms. Findings such as inflammatory infiltrate, cellular necrosis and perineural invasion are also suggestive of malignancy.¹⁵

When melanoma arises in GCMN, the prognosis is bad. The approach should be individualized taking into consideration: age of the patient, size and location of the lesion, risk of melanoma, possibility of concomitant NCM; presence of changes suggestive of malignancy on the nevus; possible functional impairments resulting from invasive procedures and the psychological impact associated with CMN or the surgical scars, often unsightly. The treatment of GCMN includes surgical or non-surgical procedures, psychological and/or clinical interventions treatment in the form of prophylactic surgical excision is justified based on the assumption that melanoma arise on the nevic lesion.^{2,3,10}

The removal of the nevus does not guarantee protection against malignancy. It is difficult to measure the impact of total excision of GCMN or other therapeutic measures on the risk of developing melanoma. Probably the reduction of melanocytic cells reduces the incidence of malignancy. Medical treatment in the form of new chemotherapy-based therapies are being developed.

CONCLUSION

GCMN are large brown-to-black skin lesions caused due to genetic mutations which lead to defective proliferation, differentiation or migration of melanoblasts which are precursor cells of melanocytes. GCMN requires a correct diagnosis and proper management due to its risk of developing malignancy. The malignant melanoma and neurocutaneous melanosis are the two major complications associated with GCMN. GCMN has got major psychosocial impact on the patient and his family due to its unsightly appearance. Management remains challenging. The child will require a lifelong follow up and support. In spite of the disease most GCMN patients may have a useful, healthy and productive life.

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