

Giant congenital melanocytic nevus: a case report at Osmania General Hospital

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Abstract: Giant congenital melanocytic nevus is usually defined as a melanocytic lesion present at birth that will reach a diameter ≥ 20 cm in adulthood. The giant congenital nevus is greater than 20 cm in size, pigmented and often hairy. Since approximately 50% of the melanomas develop by the age of two, and 80% by the age of seven, early removal is recommended. However, their large size poses a great treatment challenge. The objective of this paper is to present a unique case of giant nevi along with a review of the literature. Its incidence is estimated in $<1:20,000$ newborns. Despite its rarity, this lesion is important because it may associate with severe complications such as malignant melanoma, Between 4% and 6% of these lesions will develop into a malignant melanoma also affect the central nervous system (neurocutaneous melanosis), and have major psychosocial impact on the patient and his family due to its unsightly appearance. Giant congenital melanocytic nevus generally presents as a black or brown lesion, with flat or mammilated surface, well-demarcated borders and hypertrichosis. Congenital melanocytic nevus is primarily a clinical diagnosis. However, congenital nevi are histologically distinguished from acquired nevi mainly by their larger size, the spread of the nevus cells to the deep layers of the skin and by their more varied architecture and morphology. Although giant congenital melanocytic nevus is recognized as a risk factor for the development of melanoma, the precise magnitude of this risk is still controversial. The estimated lifetime risk of developing melanoma varies from 5 to 10%. On account of these uncertainties and the size of the lesions, the management of giant congenital melanocytic nevus needs individualization. Treatment may include surgical and non-surgical procedures, psychological intervention and/or clinical follow-up, with special attention to changes in color, texture or on the surface of the lesion. The only absolute indication for surgery in giant congenital melanocytic nevus is the development of a malignant neoplasm on the lesion.

Keywords: Giant Congenital melanocytic nevus, staged excision, skin grafting

I. Introduction

Nevus, a Latin word Knee-vus meaning “birthmark,” or “mole,” is a general term for a congenital mark on the skin. Congenital melanocytic naevi are brown or black moles which are present at birth or which develop in the first year of life [1]. They are formed by overgrowth of the melanocytes. A giant congenital melanocytic nevus (GCMN), giant hairy nevus or nevocellular nevus represents a special group of melanocytic lesions that generally covers large areas of the body and have a potential risk for developing malignant melanoma [2,3]. The lesion is variously called bathing trunk, cape, vest, coat-sleeve or stocking naevus depending on regional distribution [4]. Melanocytic nevi are benign proliferations of melanocytic cells (also known as nevus cells) arranged in nests in the epidermis, dermis or in other tissue.

History

The interest in studying GCMN has increased over time mostly because of the hypothesis that there would be an increased risk of melanoma among these patients. Reports of individuals (usually children) with GCMN who later developed melanoma from late nineteenth and early twentieth century already raised the suspicion that these two conditions were linked.

Genetics

[5] Congenital melanocytic nevi (CMN) can be associated with neurological abnormalities and an increased risk of melanoma. Mutations in NRAS, BRAF, and Tp53, their role in the pathogenesis of multiple CMN and development of associated features is not been clear. A single post zygotic mutation in NRAS could be responsible for multiple CMN, as well as for melanocytic and nonmelanocytic central nervous system (CNS) lesions. Oncogenic missense mutations in codon 61 of NRAS are found in affected neurological and cutaneous

tissues. Loss of heterozygosity is associated with the onset of melanoma in implying a multistep progression to malignancy. A single postzygotic NRAS mutations are responsible for multiple CMN and associated neurological lesions

Pathogenesis

[6]CMN originates between the 5th and 24th weeks of gestation. It is believed that a morphological error occurs in the neuroectoderm during embryogenesis, leading to unregulated growth of melanoblasts, the precursor cells of melanocytes. Both CMN and acquired nevus arise from an accelerated proliferation of cells of melanocytic lineage sometime during development. Therefore giant and medium CMN would be formed when proliferation starts, during migration of melanoblasts from the neural crest to the skin. Nevi will be larger and deeper when this process starts during the embryonic or early fetal periods. The later the onset of cell proliferation, the smaller the melanocytic lesion will be. From a molecular standpoint, it is believed that the development of melanocytes is partially controlled by the proto-oncogenes c-met and c-kit. The hepatocyte growth factor (HGF / SF – hepatocyte growth factor or scatter 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 could be related to the occurrence of GCMN

Clinical features

GCMN usually presents as a brownish lesion with well-defined borders and hypertrichosis. 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, roughed, warty or cerebriform. Although GCMN can affect any region of the tegument, its most frequent location is the torso, followed by the limbs and head. Commonly, however, GCMN affects more than one body segment.

Some peculiar locations led to the term “GCMN in garment”. These nevi are described as “bathing trunk”, “stole” or “coat sleeve.” The presence of smaller pigmented lesions scattered over the skin surface, known as satellite lesions, is common in individuals with GCMN, occurring in up to 78% of the cases. Patients may complain of pruritus, local and intermittent stimuli of afferent sensory fibers, which would be caused by xerosis and hypohidrosis secondary to the functional impairment of adnexal structures such as sebaceous and eccrine glands. The presence of nevus cells may also lead to the disruption of the normal skin architecture, leading to cutaneous fragility that would be responsible for the occurrence of erosions and superficial ulcerations.

The impact of GCMN is greater because of the considerable cosmetic disfiguration which is very distressing to the parents along with its higher malignant potential

Management

In this major teaching hospital most of these patients go to dermatovenerology department first. There the faculty and the postgraduates of this dermatovenerology department counsel the patient and take the patient into confidence and explain to them the possible modalities of management which includes Phenol chemical peels, dermabrasion, curettage, Q switch Ruby laser or just simple close observation, Staged serial excision and reconstruction with skin grafting, tissue expansion, local rotation flaps, and free tissue transfer, etc.. The patients have immense faith in the primary contact physicians (Dermato venerologist) and follow their advice in toto. In the following cases also as per their advice this patient came to plastic surgery department for further management.

II. Case Report

A 35 year male born out of non-consanguineous marriage by normal vaginal delivery at term to a primigravida with uneventful antenatal history presented with an extensive pigmented patch over the body since birth. Physical examination revealed an extensive hairy pigmented patch with 18x22 cms hyper pigmented patch over the back extending either side of dorsal spine horizontally from one loin to other vertically extending below the level of lower border of scapula to sacrum lesion was black in color with convolutions within the lesion, Tufts of coarse and lusterless hair were scattered all over the lesion at the back. His neurological examination was normal. No other satellite lesions seen. There were no associated anomalies, History of febrile seizures at the age of 11 months, MRI brain and spine shows no evidence of deposits of Melanocytes in brain parenchyma. Fundus examination, X ray spine, and ultrasound abdomen were all normal. The rest of the examination was normal. There was a negative family history of similar lesions; Punch biopsy shows extent of lesion to dermis showing dermal nevus

Management was done single staged excision and reconstruction with skin grafting. Donor area of skin graft was left thigh. Post op follow up showed 100% skin graft take up. Patient is advised to come for follow up to both plastic surgery and dermatovenerology departments.



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