

A mini review: Giant congenital melanocytic nevus

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Abstract

Congenital melanocytic nevi with a diameter of at least 10 cm or covering 1% body surface area on the face/head/neck or 2% on rest of the body at any age, and by an extension those least 6 cm on the trunk and 9 cm on the head in a neonate are known as giant congenital melanocytic nevus (GCMN). Although, a rare entity they may pose possible risk of malignancy and neurocutaneous melanosis, apart from cosmetic concerns. The psychosocial distress posed by them has also been studied extensively by many authors. On histopathological analysis epithelioid lymphocytoid and neuroid nevus cells can be visualized. Owing to such myriad presentations the management option for GCMN has to be tailored to every patient. There is a common consensus that prophylactic removal of a GCMN has little to no advantage. The partial debulking of a GCMN can be done by mechanical or chemical exfoliation, curette, shave excision or laser therapy. However, in view of potential risk of malignancy total or subtotal deep excision followed by plastic reconstruction must be considered, especially in cases involving the head and neck area.

Keywords: Giant Congenital Melanocytic Nevus • Congenital Melanocytic Nevus • Melanoma • Skin Neoplasia • Neurocutaneous Syndrome • Nevus

Description

The noncancerous multiplication of melanocytic nevus cells is known as melanocytic nevus. These nevus cells can be found in the epidermis dermis or deeper tissue in a nest like arrangement. Melanocytic nevus can be divided into two groups, namely congenital and acquired [1]. While most authors consider congenital melanocytic nevi (CMN) to be those that are present since birth, some authors have extended this limit to those developing until two years of age. The reason for inclusion of the nevi developing until two years of age lies in the fact that it might take the nevus cells up to two years to develop the amount of pigmentation or cell mass necessary to be visible on the surface of skin [2].

Various definitions have been put forth for the giant congenital melanocytic nevus (GCMN). CMN with a diameter of at least 10 cm or covering 1% body surface area on the face/head/neck or 2% on rest of the body. In view of the expected rate of growth in a neonate, another definition has been proposed defining a CMN measuring at least 6 cm on the trunk and 9 cm on the head as GCMN in a neonate [2-8].

While most CMN are benign in nature and pose only cosmetic concerns, GCMN has a tendency to turn malignant, especially into malignant melanoma. Additionally, those involving the head and neck can invade the central nervous system. Owing to the uncertain prognosis of these nevi there is still no definite treatment modality and management options are still ambiguous at best.

Etiopathogenesis

CMN arise from neuroectodermal cells, resulting in unchecked proliferation of melanoblastic cells. They arise during the 5th to 24th week of intrauterine development. Therefore giant and medium CMN would be formed when proliferation starts, during migration of melanoblasts from the neural crest to the skin [9-11]. 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 in case of a small CMN the dermo-epidermal melanoblasts start proliferating right before or shortly after birth while in case of acquired nevus this proliferation

occurs later after birth, therefore, both types of nevi have similar clinical and histopathological appearance [12-14].

In case of a GCMN there is overexpression of either hepatocyte growth factor/scatter factor or the c-met and c-kit proto oncogenes, leading to exponentially aberrant melanocyte differentiation, proliferation and migration.

Clinical Features

Typically, a GCMN in a newborn consists of a mildly hyperpigmented macule which over the course of time tends to develop into a hyperpigmented plaque. The most commonly involved site is trunk, followed by limbs and the head and neck, with predominant involvement of multiple sites on body or satellite lesions. GCMN can also involve oral mucosa [15-17].

While most of these changes are benign some premalignant/malignant or severe transformations have also been reported including neurocutaneous melanosis and development of malignant melanoma. Central nervous system involvement can make an individual prone to spinal dysraphism, astrocytoma, cystic malformation in the arachnoid matter, schwannomas, liposarcomas, to name a few. Various disorders have been found to have a strong association with GCMN including hypertrichotic transformation of the cranium, asymmetrical limb dystrophy, multiple lipomas, kyphoscoliosis, dermal melanosis, etc.

A few special variants of the GCMN include the bathing trunk, garment type, coat sleeve nevus and development of halo phenomenon around the nevus.

Histopathological Findings

There exists a marked overlap between the histopathological picture of a congenital and acquired nevus with presence of hyperkeratosis, epidermal hyperplasia, hyperkeratosis, elongated rete ridges, up regulation of melanocyte number with nest like arrangement of these cells in the epidermis. However, there exist some subtle differences between the two with a deeper penetration of the nevus cells upto subcutis in GCMN with periappendageal involvement, unlike acquired nevi which tend to stay limited up to the upper half of the dermis. In case of GCMN the nevus

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cells are arranged in an Indian file like arrangement between the collagen. Three variants of the dermal nevus cells have been identified, namely, type-A (epitheloid) nevus cells which mature into Type-B (lymphocytoid) nevus cells which in turn mature into Type-C (neuroid) dermal cells during progressive downward migration. However, these type-C cells are often found to be found in the adipocyte or neural metaplasia [18].

Natural History

Over the course of time the lesions tend to develop hypertrichosis with pruritus in some cases owing to the loss of appendages at involved site leading to reduced sweating and increased dryness or superficial erosive or ulcerative changes. Secondary changes include papular, nodular, verrucous and cerebriform transformation. Lesions run an unpredictable course with darkening or fading in lesions, along with spontaneous regression in few cases [19].

The prime concern in a GCMN apart from cosmetic unacceptability is the development of malignant melanoma. However, while most melanomas arise from epidermis, the ones arising from GCMN originate from dermal or subcutaneous nevus cells. Patients with satellite lesions, or those with lesions involving the paravertebral sites, central back, head and neck are more prone to such transformations. These malignant cells can spread to the central or peripheral nervous system or gastrointestinal tract. According to a review by Marghoob et al. upto 70% cases of melanoma from GCMN arise during childhood period [20].

Dermatoscopic Analysis

Dermatoscopy can be a quick handy tool as a non-invasive option for diagnosis of CMN. On dermatoscopy multifocal globules, dots and central hyperpigmentation can be seen present disperse throughout along with hypertrichosis, perifollicular dyspigmentation and pseudomilia [21].

Treatment

Owing to such myriad presentations the management option for GCMN has to be tailored to every patient. There is a common consensus that prophylactic removal of a GCMN has little to no advantage. Although, a group of authors suggest that prophylactic excision could be considered in case of rough, irregularly textured, non-homogenous raised nevi. However, in case of a garment like pattern of nevus complete removal isn't feasibly due to extensive size. Therefore, many authors advocate reduction in melanocyte bulk by superficial removal.

The partial debulking of a GCMN can be done by mechanical or chemical exfoliation, curette, shave excision or laser therapy. However, in view of potential risk of malignancy total or subtotal deep excision followed by plastic reconstruction must be considered, especially in cases involving the head and neck area. It is also advisable for patients suffering from GCMN to get periodic clinical examinations throughout their life in order to facilitate early detection and timely management of potential malignancies [22-27].

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