Case Report

Giant congenital melanocytic nevus

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Abstract

Congenital melanocytic nevus is rare benign neoplasm which generally seen in one in 20,000 children. They vary in size from <1 cm to covering almost the entire body. Giant congenital melanocytic nevus (GCMN), variant of congenital melanocytic nevus, involving much of the body surface area are less common, around one in 200,000–500,000. It is characterized by its extensive size and is defined as melanocytic nevus measuring more than 20 cm in its greatest dimension. Malignant changes of giant nevus can occur at any age but most often occur in infants or toddlers. We are here reporting a case of GCMN in a 3-day-old female child.

Key words: Bathing trunk nevus, congenital nevus, garment nevus, melanocytic nevus

Introduction

Congenital melanocytic nevi are defined as benign nevomelanocytic proliferations present at birth. Congenital melanocytic nevi occur in approximately 1–2% of newborns with a female predominance and are usually classified according to their size. Giant congenital melanocytic nevus (GCMN) are even rare (1 in 200,000–500,000) and they are most simply defined as melanocytic nevi that are >20 cm in largest dimension. They are usually deeply pigmented, covered with a moderate growth of hair and often there are many scattered satellite lesions associated with them. Malignant melanoma (MM) and malignant neuroectodermal tumors might develop in these lesions.

Congenital nevi can exhibit distinctive histologic features on biopsy that can help in differentiating them from common acquired nevi. Here we are presenting a rare case of GCMN in a 3-day-old female child.

Case Report

Three days old female child with an uneventful antenatal history, presented with an extensive pigmented patch over chest since birth measuring 20.5 cm in length [Figure 1]. Similar lesions were also present on thighs and lumbosacral region.

On physical examination lesions were hyperpigmented, rugosed, plaque-like with moderate amount of hair growth. There was no family history of any similar lesion. There were no other associated congenital anomalies. The biopsy was taken for histopathological examination.

The histopathological examination revealed stratified squamous epithelium with basal cell proliferation along with increased melanin pigmentation and dermis composed of band like infiltrates of nevus cells with vesicular nuclei, inconspicuous nucleoli, and moderate amount of pigmentation [Figure 2]. These cells were also infiltrating skin adnexal structures and had angiocentric distribution [Figure 3]. No evidence of a malignant transformation was seen.
Immunohistochemically S-100 was showing diffuse and intense positivity in nevus cells [Figure 2].

**DISCUSSION**

Congenital melanocytic nevi are neurocristopathy (a disorder of the development of the embryonic neural crest), with unknown etiology. Majority of congenital melanocytic nevi, develop during first trimester of pregnancy. Vast majority are sporadic, but familial cases are also reported.\(^2\) Congenital melanocytic nevi vary greatly in size, macroscopic appearance, and histology. There is a practical need to subdivide congenital nevi according to size, since size differences have a direct bearing on cosmetic, therapeutic options, and probably on the chance of malignant transformation.\(^3\) They are classified according to their sizes as small (<1.5 cm), medium (1.5–19.9 cm) and large or giant (≥20 cm) congenital melanocytic nevus (giant pigmented nevus, giant cerebriform nevus, garment nevus, bathing trunk nevus, etc). The incidence of the small naevi is one in 100 births, that of the medium naevi is 6 in 1000 births, and that of GCMN, which are larger than 20 cm in diameter is 1/500,000 newborns.\(^4\)

The best parameters distinguishing a congenital nevus from an acquired nevus are its presence at birth and its larger size. The histologic appearance of giant congenital nevi differs from that of acquired nevi in terms of their greater size and depth, and in the involvement of skin appendages. The extension of nevus cells into the upper reticular dermis, within skin appendages, and angiocentric distribution is quite characteristic of congenital nevi.\(^5\)

The culture of melanocytes from such lesions showed chromosomal rearrangements which involved the chromosomal regions 1p, 12p, and 19p. Researchers think that a body protein which is called hepatocyte growth factor/scatter factor seems to be responsible for encouraging the neuroectodermal cells to develop, migrate, and scatter. It seems that either too much or a wrong type of this protein in some cells, develop extra pigment, and abnormal skin cells which are called nevus cells. These cells scatter around and so, we have naevi scattered all over the body.\(^6\)

While the association between GCMN and MM has been established beyond any doubt, the exact magnitude of the risk is still unknown. The lifetime risk of melanoma for patients with giant congenital nevi has been reported to range from 5% to 40%.\(^6\)

Management decisions of congenital melanocytic nevi, regardless of their size, should take into consideration the perceived risk of melanoma, the patient’s age, the cosmetic outcome, the surgical complexity, and the risk of anesthesia.\(^6,7\) The risk of developing melanoma in giant congenital nevi appears to be greatest in the first decade of life.\(^6,8\) Intervention, if recommended, must start early in life. The options for management of giant congenital nevi
include staged excision with grafting, dermabrasion, curettage, Q-switched ruby laser, and simply a close observation.\[^9\]

All patients with GCMN should be screened periodically for MM and neurocutaneous melanocytosis. Visual inspection can be facilitated by obtaining baseline images to use for comparison, which may help in the detection of subtle focal changes indicative of early Cutaneous Melanoma.\[^9\]

**CONCLUSION**

Giant congenital melanocytic nevus is a very rare condition. Its early and correct diagnosis is essential due to the risk of development of MM. Other congenital anomalies should be sought out early after birth. Regular follow-up and proper specialized management is required. Regardless of what type of management is decided upon, be it surgical or observation, it must be remembered that most GCMN patients lead to healthy and productive life.

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**Conflicts of Interest**
There are no conflicts of interest.

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