

Giant Congenital Melanocytic Nevi: Short Review

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Introduction

Giant congenital melanocytic nevi (GCMN) are variants of melanocytic nevi. They can affect any part of the skin and their size is greater than 20cm in diameter [1]. The incidence of GCMN varies between 1:20,000 to 1: 50,000 [2].

GCMN have irregular borders and they are darker in color than other congenital nevi, covered with moderate hair growth in more than 90% of lesions and usually associated with satellite lesions [3]. Patients with GCMN have higher risk of developing melanoma [4,5].

Characteristics

GCMN are characterized by presence at birth like other congenital nevi, larger size >20 cm, hypertrichosis, rough surface and deep blackish color. They are difficult to excise with favorable cosmetic appearance and have higher risk to develop melanoma in addition to bad psychological impact on the patient and the family [6-8].

Classification

Among several authors who classified congenital melanocytic nevi (CMN) Kopf et al classified them based on their largest diameter in adulthood, as follows: small (<1.5 cm), medium (1.5-19.9 cm), giant (≥ 20 cm) [9].

Ruiz-Maldonado [10] modified the classification by Kopf et al., where he considered CMN with size between 1.5 cm and 10 cm as medium and those between 11 cm and 20 cm as large and those >20 cm as giant. GCMN were further subdivided into: G1 (21-30 cm), G2 (31-40 cm) and G3 (>40 cm). Regardless of different classifications and because of the rarity of GCMN it is difficult to estimate the real incidence of melanoma in those patients.

Clinical Features

GCMN present at birth as raised brown to black in color usually hairy with well-defined borders. As the child grow up the nevi gets darker and develop more hair and become deeper in color. The surface of GCMN can be rough or verrucous [11] Usually asymptomatic but with time, patients may complain of mild pruritus [6,12] GCMN most frequently affect the trunk followed by the limbs and head [13-16]. Sometimes GCMN may affect more than one site of the skin which is known as bathing trunk [16,17]. The majority of cases of GCMN are associated with satellite lesions [18,19].

Histopathologic features

Histologically, GCMN of various sizes are separated from acquired melanocytic nevi by deeper location in the dermis and subcutis and

periappendageal involvement, while the changes in the epidermis are like those of acquired melanocytic nevi [20-22].

Management

The treatment of GCMN is a daunting challenge and can be surgical or non-surgical. There is no ideal treatment that reduces the risk of melanoma in patients with GCMN. Because of large size of GCMN and the deep location in the dermis the surgical excision will result in unfavorable scarring and will not remove all nevus cells.

The aims of all treatment modalities that have been tried were either to reduce the risk of melanoma or for cosmetic purposes. Unfortunately, none of these aims were fully achieved as 50% of melanoma that developed in patients with GCMN arises in other sites [18,23] and unsightly scar formation is inevitable.

Surgical excision of GCMN on extremities using tissue expanders, flaps or grafts will have the risk of contractures and other complications [14]. Procedures like shave excision, skin curettage, lasers and chemical peels can be used for improving the cosmetic appearance of GCMN but they only remove the superficial parts of GCMN [8, 24-27].

The best approach is to deal with each case of GCMN according to the clinical presentation taking into consideration the size, the location, the age of the patients, the risk of melanoma and the surgical complications. Teamwork is necessary for GCMN and should involve dermatologist, plastic surgeon and psychologist [6,28,29]. The role of the medical team is crucial for regular follow up both before surgical intervention and after any procedure [6,28]. The parents and patient if he is an adult should be informed and trained how to detect changes in the nevus like color, shape, bleeding or any symptom in order to seek medical advice [6].

Conclusion

GCMN rare melanocytic disorder which have a size of ≥ 20 cm in diameter. They have bad psychological impact on patients and their families. None of the treatment modalities will achieve desirable goals in reducing the risk of melanoma or improving the cosmetic appearance of the nevus.

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