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Case Report

Giant Congenital Melanocytic Nevus

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ABSTRACT

Context: Giant congenital melanocytic nevus is a very rare type of congenital melanocytic nevus causing a considerable cosmetic problem for children and their families. The giant congenital nevus is defined as large as patients palm if it occurs on the head and neck or more than 30% of body surface or greater than 20 cms in size. Although asymptomatic, its management is difficult and challenging with the available therapeutic modalities and aesthetic improvement remains a goal to achieve till date. Here we report a unique case of giant bathing trunk nevus in 3 year old female child.

Keywords: Giant congenital melanocytic nevus, therapeutic modalities, congenital melanocytic nevus.

INTRODUCTION

Congenital melanocytic nevi (CMN) are hyper pigmented macular lesions which arise due to normal benign proliferation of melanocytes. Congenital nevi are found in 1 to 3% of new born infants [1] and are more common in black or Asians. [2] CMN is classified as small, intermediate and giant depending on the size of lesion as in less than 1.5 cm, 1.5 cm to 20 cm and more than 20 cm respectively. [3] Giant congenital melanocytic nevi (GCMN) are large macular lesions present since birth and develop coarse terminal hair, become rugose or warty or can develop hamartomatous nodules subsequently over a period of time. It is also variously known as Bathing trunk, cape, garment, vest, coat sleeve nevi depending on the local distribution. [4] GCMN is commonly situated on the back and thighs but can occur at any site on the body. The smaller satellite melanocytic nevi are seen elsewhere in the body. The life time risk of malignant changes in GCMN is about 2% to 15%.

CASE REPORT

A three year old female child presented with a bathing trunk nevus involving most of the trunk, buttocks, external genitalia and upper one third of both legs with gross enlargement of external genitalia and few scattered nodules over the lesion since birth. There were terminal hairs over the lesion. There was three intermediate congenital melanocytic nevi over the face, chest and below left knee joint and multiple small congenital nevi widely distributed over the body. (Figure 1, 2, 3) No history suggestive of ulceration and bleeding from the lesions. There was no history of seizures, focal neurological deficit, musculoskeletal or any associated systemic complaints. The child achieved her mile stones in time according to her age. The child was born out of non consanguineous marriage by a full term normal vaginal delivery with uneventful antenatal history. The general physical and systemic examination was normal. No abnormality was detected in the routine blood investigations and radiological

examination of spine. The biopsy of lesion was not done.

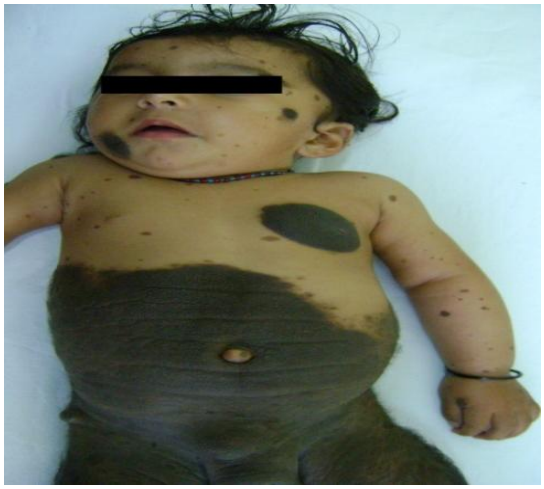

Figure 1: Bathing trunk nevus with satellite lesions, Intermediate congenital melanocytic nevi over right cheek and chest

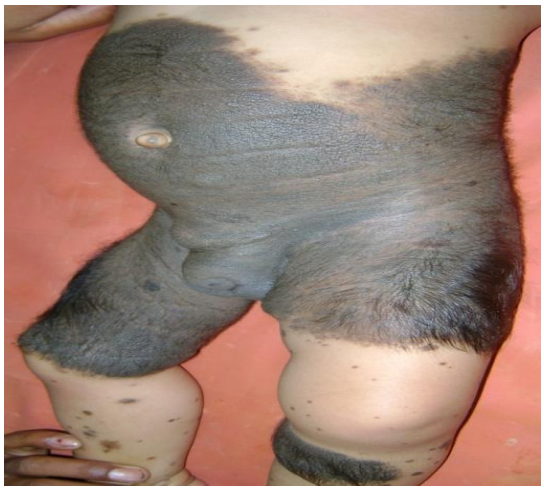

Figure 2: Bathing trunk nevus involving abdomen and thighs, intermediate congenital melanocytic nevus below left knee joint, gross enlargement of external genitalia and Tufts of terminal hair

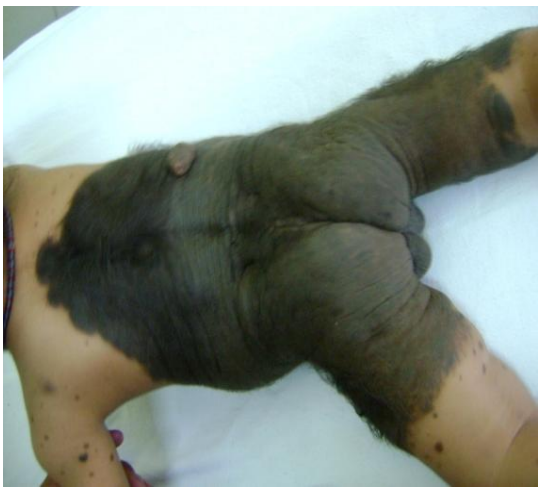

Figure 3: Bathing trunk nevus covering back, buttocks, upper one third legs with scattered nodules.

DISCUSSION

The congenital nevi are developmental abnormalities of skin, due to genetic mosaicism which reflects the migration paths of individual clones of genetically identical cells. The Masson's theory hypothesizes that nevus cells in the upper dermis develop from epidermal melanocytes and those present in the mid and lower dermis are derived from Schwann cells. The skin and nerves develop from neuroectodermal between the 8th and 24th weeks of gestation. Melanoblasts migrates laterally from neural crest first to the dermis and then to the basal lamina of the epidermis between 8th to 10 weeks of fetal life. [5]

A bathing trunk nevus may be associated with other congenital anomalies such as spina bifida, neurofibromatosis and lipomas. [6] Malignant melanoma [7] and rapidly growing ulcerative tumor - nodular proliferative neurocristic hamartoma may develop at birth or thereafter. The classic histopathological features of CMN is the presence of nevus cell in reticular dermis, extension of nevus cell into the collagen bundles in Indian file appearance, increased concentration of nevus cell around blood vessels, nerves and adnexal tissues. The GCMN shows involvement of whole dermis with patterns of a compound or an intradermal nevus, a neural nevus, a blue nevus, an epithelioid cell nevus in variable proportion. [4]

The cosmetic and psychological impact of GCMN is quite obvious for the patient and families with risk of malignant melanoma poses a great challenge to find out the best modalities of treatment. The treatment options includes many surgical procedures, dermabrasion, curettage, Q switched ruby laser, carbon dioxide laser and erbium: YAG laser.

The management is individualized and no absolute guidelines can be recommended. The currently available methods did not sufficiently address the need of patient and physician and search for better modalities of treatment is on. The main focus is on

education and awareness about the benign nature, psychotherapy, appropriate follow up and timely intervention in case of suspicion of malignant changes in the lesions.

CONCLUSION

As the management of GCMN is difficult as is true with all other developmental disorder, so, early intervention is better as size of nevus increases proportionate to the body growth. Parents counseling to allay fears and normal psychological development of child are to be encouraged. The biopsy from suspicious lesions and radiological investigation to rule out neurocutaneous melanosis and spinal dysraphism is recommended for subsequent management.

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