DOI:10.24041/ejmr2021.19

GIANT CONGENITAL MELANOCYTIC NEVI – A RARE CASE REPORT

Govind Kumar, Kshitij Saxena, Aliza Shoeb Zaidi, Reyan Abdul Jamil, Garima Choudhary

Department of Dermatology

Era's Lucknow Medical College & Hospital, Era University, Sarfarazganj Lucknow, U.P., India-226003

ABSTRACT

Giant congenital melanocytic nevi (GCMN) are the melanocytic lesions that arise due to the abnormal migration of melanoblasts during the embryogenesis, affecting approximately one in 500,000 live births. There is gain-of-function mutation in the NRAS gene is usually associated with GCMN, causing abnormal proliferation of embryonic melanoblasts that are usually present since birth with change in their morphological characteristics with time, and increasing in their size reaching upto a diameter ≥ 20 cm in adulthood. These are characterized by dark brown to black in color with irregular margins and having

verrucous surfaces, with or without satellite lesions which are present beyond the periphery of the central lesion. Depending on their distribution these can be classified as bathing trunk, coat-sleeve, or stocking nevi. We hereby report a case of a sixteen year old female presenting with GCMN. The development of a malignant neoplasm on same lesion is only absolute indication for surgery in GCMN.

KEYWORDS: Giant, Melanoblast, Neoplasm, NRAS gene.

INTRODUCTION

Congenital melanocytic nevi (CMN) are melanocytic nevi which are benign, pigmented present at birth, caused by a somatic mutation in utero.1 Congenital melanocytic nevi (CMN) are defined as brown to black benign nevomelanocytic proliferations present at birth and most commonly located on the back and the thigh areas (2). Giant Congenital Melanocytic Nevi (GCMN) is rare which is larger than 20 cm diameter in size and they occur in 1 per 500,000 newborns (3). GCMN are dark brown to black in color with irregular margins and vertucous surfaces, with satellite lesions are present beyond the periphery of the main lesion (3). They may be scattered all over the skin surface. These lesions are differently called as bathing trunk, coat sleeve or stocking nevi, according to their regional distribution (3). The giant nevi located on the scalp and the neck may be associated with leptomeningeal melanocytosis and neurologic disorders like neurofibromatosis and epilepsy (3).

CASE REPORT

A Sixteen years old female patient presented to the Dermatology OPD at Era's Lucknow Medical College and Hospital with the chief complain of large pigmented patches over her body. It was smaller in size at the time of birth which then, gradually increased in size covering the neck, back, chest and both the arms with age. It was not associated with itching and any other complaints.The Received on : 18-04-2021 Accepted on : 04-06-2021

Address for correspondence

Dr. Kshitij Saxena Department of Dermatology Era's Lucknow Medical College & Hospital, Era University, Lucknow-226003. Email: drkshitijSaxena@live.com Contact no: +91-9208015391

parents of the patient were not consanguineous married. None of the close family members had similar skin lesions.

ON EXAMINATION

Multiple, well to ill defined, discrete to coalescing, hyperpigmented, verrucous, plaques, of various sizes largest being >20cm in diameter and smallest being 2mm in diameter, present over the neck, chest, back and b/l arms. Multiple, well defined, discrete, hyperpigmented plaques of size 0.5 mm to 2 mm in diameter present over the neck and back. There was no variation in the density of hair over the lesion.



Fig. 1. GCMN present over upper back with satellite lesions at periphery Fig. 2. GCMN present over chest extending to neck and bilateral upper limb



Fig. 3. Histologic specimen shows epidermis with increased melanin in badal layer and dermis with extension of naevus cells around nerves, vessels and adnexae

INVESTIGATIONS

A MRI scan of brain and spine was advised which were within normal limits. Her neurological examination was normal she had no other congenital anomaly.

ON HISTOPATHOLOGY

The histopathological examination shows stratified squamous epithelium with basal cell proliferation. There was an increased melanin pigmentation and dermis was composed of band like infiltrates of nevus cells with vesicular nuclei, inconspicuous nucleoli, and moderate amount of pigmentation.

TREATMENT

Patient had given two sessions of ND-Yag laser, then she was lost to follow up due to COVID lockdown.

DISCUSSION

The congenital melanocytic nevi are pigmented cutaneous lesions which arise by an epidermally and dermally derived nevus cells, which occur in about 1% of the newborns. They are classified as small (<1.5 cm), medium (1.5-19.9cm) and large or giant nevi (>20cm)1 according to their sizes.1

The incidence of various CMN as small nevi is 1 in 100 births, that of the medium nevi is 6 in 1000 births, and that of GCMN which are larger than 20 cm in diameter is 1 per 20,000 to 500,000 newborns1. The reported female to male ratio ranges between 1.17:1 to 1.46:11. They are believed to be caused by spontaneous

mutations or during the fetal development, but because of the frequent appearance of CMN in some families, suggests that they may be genetically inherited4.

Classically, GCMN is an asymptomatic disease but some patient may complain of pruritus and xerosis1. The melanocytes culture from such lesions showed chromosomal rearrangements which involved the chromosomal regions 1p, 12p and 19p. CMN are identified by their projected adult size, number of smaller satellite lesions, anatomical localization, and degree of rugosity, hypertrichosis, nodularity, and color heterogeneity (4). A nevus with a projected adult size greater than 40 cm is classified as GCMN, as in our case.

Further, GCMN are classified into six categories according to their anatomical location as bolero, back, bathing-trunk, breast/belly, body extremity, and body. GCMN has a significantly greater risk of transforming into malignant melanomas, as compared to smaller-sized nevi (5 to 10%)6. Neuro cutaneous melanosis (NCM) is another complication associated with GCMN in which there is abnormal melanosis of the central nervous system and frequently asymptomatic at birth (2).

Most common histological findings are hyperkeratosis and hyperplasia, elongation of epidermal ridges, and increased number of melanocytes (5). Management of GCMN is symptomatic and palliative. The various surgical treatment include serial excision and reconstruction with skin grafting, tissue expansion, local rotation flaps and free tissue transfer. In case of leptomeninges, excisions may not eliminate the risk for developing melanoma due to their depth (5). Varoius non excisional procedure for treatment include dermabrasion, laser ablation, curettage, and chemical peel (2). The carbon dioxide laser, the Er: YAG and the Q switched ruby laser have all been recently used for resurfacing and for selectively treating the deep pigmentations (2).

CONCLUSION

Giant congenital melanocytic nevus is a very rare condition. Its early and correct diagnosis is essential due to the risk of development of Malignant melanoma. 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.

REFERENCES

1. Endomba F.T, Mbega C.R, Tochie J.N et al. Giant congenital melanocytic nevus in a Cameroonian child: a case report. 2018. Med Case Reports 12, 175.

- Salim S, Ilham EM, Asmae S, Hassam B Giant Congenital Melanocytic Nevus: About a Case. 2018 Clin Med Img Lib 4:110. doi.org/10.23937/2474-3682/1510110.
- 3. Kinsler A. Veronica and Sebire J Neil. Congenital naevi and other developmental Abnormalities affecting the skin. In Rook's textbook of dermatology. 9th ed. Publisher: Wiley-Blackwell; 2016. p. 75.9.
- 4. Das SK, M Amarendra, Subudhi Monalisa. Giant congenital melanocytic nevi: a case report. Clin

Diagn *Res*. 2013;7(1):154-155. doi:10.7860/JCDR/2012/4832.2693.

- 5. Viana ACL, Gontijo B, Bittencourt FV. Giant congenital melanocytic nevus. An Bras Dermatol. 2013;88(6):863-78.
- 6. Bhagwat PV, Tophakhane R S, Shashikumar B M, Noronha TM, Naidu V. Giant congenital melanocytic nevus (bathing trunk nevus) associated with lipoma and neurofibroma: Report of two cases. Indian J Dermatol Venereol Leprol 2009; 75:495-498.

How to cite this article : Kumar G., Saxena K., Zaidi A. S., Jamil R. A., Choudhary G. Giant Congenital Melanocytic Nevi – A Rare Case Report. Era J. Med. Res. 2021; 8(1): 93-95

licencing Information

Attribution-ShareAlike 2.0 Generic (CC BY-SA 2.0)

Derived from the licencing format of creative commons & creative commonsmay be contacted at https://creativecommons.org/ for further details.