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Case Report

GIANT CONGENITAL MELANOCYTIC NEVUS: A CASE REPORT

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ABSTRACT

Congenital melanocytic nevi are present in approximately 1% of infants and are formed by overgrowth of melanocytes. These are usually present at birth and are also known as bathing trunk, coat sleeve or stocking nevi. These are commonly found over the trunk and thigh areas. Giant congenital naevi are of sizes greater than 20 cm and are very rare. They have association with leptomeningeal melanocytosis and predisposition for development of malignant melanoma. The overall incidence of multiple melanoma arising in a giant congenital nevus is estimated to be approximately 5-10 % and half of all melanoma that arise in giant congenital nevus do so by 5 year of age. So, an early removal is recommended. This paper highlights a rare case of giant nevus.

Keywords: Giant Congenital Naevi, Leptomeningeal Melanocytosis, Malignant Melanoma

INTRODUCTION

Congenital melanocytic nevus is defined as melanocytic nevus present at birth or presenting within first few months of life (Schaffer and Bolognia, 2000). Giant Congenital Melanocytic Nevi (GCMN) of sizes which are larger than 20 cm diameter are rare and occur in 1 per 5 lakh newborns. GCMN occur most commonly on posterior trunk but may also appear on the head or the extremities (Kimberly and Goodman, 2007). GCMN have irregular margins, dark brown to black in colour, verrucous surfaces and satellite lesions which are present beyond the periphery of main lesion (Mackie, 1993). Asymptomatic leptomeningeal melanosis on MRI scan is seen in approximately 30% individuals with GCMN (Arons, 2002). Here, we present a case of GCMN.

CASES

An 8 months old male child who was born of non consanguineous marriage by a normal vaginal delivery at term to a second gravida, with an uneventful antenatal history ,presented with an extensive pigmented patch over his body since birth [Figure 1,2]. There was no family history of any similar lesion.



 Figure 1 and 2: Shows extensive pigmented patch over the body of patient

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The physical examination revealed an extensive pigmented patch size 20×25 cm over the back, thighs and the legs. Multiple pigmented satellite lesions of 4-5 cm size were also present over the body and the extremities. Tufts of coarse and lusterless hair were scattered all over the lesion at the back. His neurological examination was normal.

DISCUSSION

Congenital melanocytic nevi are pigmented cutaneous lesions which are formed by a combination of epidermally and dermally derived nevi cells (Rhodes, 1999). These nevi have been categorized by size: small(<1.5 cm), medium (1.5-19.9 cm) and giant(>20 cm)(Rhodes AR, 1986). They have equal prevalence in males and females. GCMN begin as flat, brown or brownish black patches but with increasing age may become elevated and develop a mottled appearance and nodular surface (Ruiz-Maldonado et al., 1992). They appear darker at birth than a few weeks later and are found most commonly on the trunk, followed by the limbs and the head. Giant nevi are almost always confined to skin but rarely, invade underlying fascia and muscle (Madaree et al., 1997). GCMN are thought to be caused by spontaneous mutation but in some families, the frequent appearance of these lesions suggests that they may be inherited. The culture of melanocytes from such lesion show chromosomal rearrangements which involved the chromosomal regions 1p, 12p and 19p. Researchers postulate that the a body protein called HGF /SF (hepatocyte growth factor/scatter factor) is responsible for encouraging the neuroectodermal cells to develop, migrate and scatter (Habit, 2004). Early and complete surgical excision is the recommended treatment for GCMN as they have increased lifetime risk of malignant melanoma and other neural crest malignancies (Schaffer and Bolognia, 2000). Giant nevi on the scalp, in a posterior axial location and those with satellite lesions are at greater risk of malignant transformations. Abnormalities reported in association with GCMN are limb hypoplasia, ear deformities and angiomas (Frieden et al., 1994). The present case had no such associated abnormaities.

More importantly, congenital melanocytic nevi may be associated with leptomeningeal melanocytosis so radiographic imaging which include MRI is warranted to evaluate the CNS. It is also important to remember that in a child with GCMN, a normal MRI brain study does not rule out the possibility of future occurrence of CNS melanosis (Frieden *et al.*, 1994). Thus serial MRIs are recommended in every child with GCMN who is at risk of developing CNS melanosis. It is impractical to prophylactically excise all the non- giant congenital nevi and so, yearly examination for first three years is recommended. The surgical treatment of GCMN is addressed at the age of 6 months. The procedures which are used in the surgical treatment include serial excision and reconstruction with skin grafting, tissue expansion, local rotation flaps and free tissue transfer. Due to the depth of some lesions, especially if the leptomeninges are involved, excisions may not eliminate the risk for developing melanoma (Mark *et al.*, 1973).

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