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Case Report

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Association of Vitiligo with Congenital Melanocytic Nevus, A Rare entity.

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Abstract

Development of depigmented patches of vitiligo within or around congenital melanocytic nevus (CMN) is a very rare phenomenon. We hereby report a case of 16-year-old female who presented to us with giant CMN since birth and vitiligo lesions within and around the nevus and over lips, developed 1 year back and are gradually progressing.

Keywords: Vitiligo, Congenital Melanocytic Nevus, Becker's Nevus

Introduction

Congenital Melanocytic Nevi (CMN) occur in 0.6-1.6% of newborn.¹ They are divided into small (<1.5 cm), medium (1.5-10 cm), large (11-20 cm) and giant CMN (>20 cm).

Vitiligo is an acquired disease, usually progressive, a depigmentary disorder of multifactorial etiology affecting 0.5-2% of world population.² To explain the etiology of vitiligo, various theories like autoimmune, free radical, composite and neural hypothesis was proposed. Vitiligo may be associated with other autoimmune diseases particularly Graves's disease, Hashimoto's thyroiditis, and pernicious anemia and

endocrinopathies such as Addison's disease and diabetes mellitus which is proved by the presence of melanocytic antibodies and lymphocytic infiltrates in the serum of vitiligo patients, which is a widely accepted hypothesis.³

Giant CNM is a rare condition with an incidence of one in twenty thousand births,^{4, 5, 6} but vitiligo developing within giant CMN is a much rarer presentation.

We describe a patient with CMN, who presented with depigmented patches within the nevus, followed by the development of similar patches in other areas than the primary lesion.

Case Report

A 16-year-old female patient presented with a giant melanocytic nevus over back (Fig.1), right lower limb (Fig.2) with coarse hair on its surface since birth. On examination, lesions were in the form of multiple, discrete CMN of varied sizes ranging from 1x2cm to those covering half of the back. One year back she developed large depigmented patches within and

around the giant nevus and also over lips. There was no evidence of other associated autoimmune disorders or melanoma. A detailed clinical examination and laboratory investigations including computed tomography of the brain revealed no abnormalities. HPE of the lesions showed loss of melanin and melanocytes with nevoid cells and perivascular lymphocytic infiltrate in the dermis.(Fig.3)



Fig 1: Depigmented patch of vitiligo over congenital melanocytic nevus



Fig 2: Nevus hypertrichosis over Right lower limb

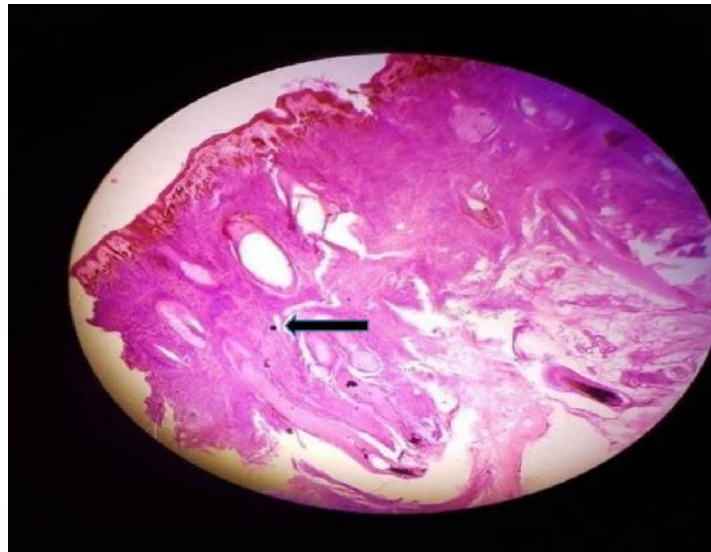


Fig 3: Nevus with vitiligo focus and nevoid nests in dermis

Discussion

The association of the CMN and the vitiligo is a rare co-existence. The evolution of the Nevus with depigmented patches varies widely. It may remain unaltered throughout life or undergo malignant degeneration or sometimes even shows spontaneous regression.

The incidence of CMN is 0.6-1.6% in newborns.¹ they are being classified on the basis of their size. There is 1% to 42% risk of developing malignancy in the large and giant CMN lesions with the risk being negligible in small and medium-sized CMN. Though, after puberty chances may increase with small and medium-sized nevi too.⁷

A depigmentation can develop around or inside the nevus. Itin and Lautinschlag⁸ reported a case of a 6-year-old boy who developed a white colored halo around the nevus when he was 3 years old with vitiligo lesions occurring 1 year later.

Recently Dainich *et al.*⁹ reported a case where CMN disappeared after the parallel onset of the vitiligo at the age of 19 years leading to almost disappearance of all nevus cells from a lesion of CMN. In contrast, our patient showed the development of vitiliginous patch over nevus suggesting that T-cells not just attacked melanocytes of epidermis but also nevoid cells.

A giant CMN *per se* carries a considerable risk of malignant transformation.¹⁰

A common immunological mechanism against similar molecular targets is suggested by the presence of cytotoxic CD8 positive T-cells in both the lesions.¹¹ It is most difficult to predict the evolution of CMN as it could regress³ or develop a halo nevus, vitiligo lesions or in contrast, could remain stable. Hence, CMN must be followed for life, particularly in the case of giant nevi, where the risk of melanoma is very high and also because of the additional reason of association between vitiligo and melanoma.

Conclusion

In view of the multiple small and medium melanocytic nevi over the trunk and limbs in addition to giant melanocytic nevus over back, our patient requires close observation for the development of melanoma at a later stage.

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