



Congenital Melanocytic Nevus: An Unusual Presentation

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Abstract: The overgrowth of melanocytes produces congenital melanocytic nevus. They are commonly present since birth, and the common terms used to describe them are called 'bathing trunk,' 'coat-sleeve,' or 'stocking naevi,' and they are usually found over thighs and back. The giant congenital nevus of size larger than twenty centimeters is highly pigmented and usually hairy. It is more commonly found in females compared to males. It is well recognized that the lesion's displeasing nature might have psychological and social ramifications, negatively affecting the patient's self-esteem, particularly females. Although congenital melanocytic nevus is mostly clinically diagnosed, the histological investigation is required to confirm the diagnosis and rule out malignant changes. The likelihood of transforming the giant nevus to malignant melanoma accounts for approximately five percent; fifty percent of melanomas develop before age two, and eighty percent develop before age seven. The risk of transformation to malignant melanoma is early in giant melanocytic nevus cases and adolescence in small and medium-sized nevi cases. Other complications like transformation to neurocutaneous melanosis consisting of the spread of melanoma to the central nervous system and leptomeninges are also dreadful. Hence, early removal is recommended. Here, we have reported a rare case of a six-year-old patient with a medium-sized nevus on their face with hairy tufts in between. On biopsy, the diagnosis was made as a junctional nevus. After which, the patient was managed with subsequent lesion excision and split skin grafting with an uneventful postoperative period. Our case report aims to report a rare case of congenital melanocytic nevus and to determine the importance of timely diagnosis and intervention.

Keywords: Hairy cell nevus, junctional nevus, skin grafting, nevus, congenital nevus.

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I. INTRODUCTION

Congenital melanocytic nevus is a brown to black mole appearing since birth and is usually found on the back and thighs.¹ Giant congenital melanocytic nevus, which is more than twenty centimeters in diameter, is infrequent and is found among one in five lakh neonates. Giant congenital melanocytic nevus has fuzzy and irregular margins that are dark brownish to blackish in colour, with a surface which is verrucous, and it may have the presence of satellite lesions extending beyond the periphery of the main lesion.² They may be dispersed across the entire surface of the skin.³ Depending on where they are seen, these lesions are known as 'bathing trunk,' 'coat sleeve,' or 'stocking naevi'.⁴ The presence of giant nevus on the head and neck may be linked to leptomeningeal melanocytosis and neurologic illnesses such as neurofibromatosis and seizures.⁵ Congenital melanocytic nevus is more commonly found in females compared to males. It is well recognized that the lesion's displeasing nature might have psychological and social ramifications, negatively affecting the patient's self-esteem, particularly females.⁶ Although congenital melanocytic nevus is mostly clinically diagnosed, the histopathological investigation is required to confirm the diagnosis and rule out malignant changes. The likelihood of transformation of the giant nevus to malignant melanoma accounts for approximately five percent, and fifty percent of melanomas develop before age two. Eighty percent develop before the age of seven.⁷ The risk of transformation to malignant melanoma is early in cases of giant melanocytic nevus and adolescence in small and medium-sized nevi cases.⁸ Melanoma is more common in children with a congenital melanocytic nevus. Melanomas in Giant Congenital Nevus typically develop before puberty in the first 5 years of life, as opposed to small and medium nevi, where they commonly develop after puberty.⁹ The size and location of the congenital melanocytic nevus, as well as its linkage with multiple satellite nevi, appear to have an impact on malignancy and melanoma development. The likelihood of melanoma development from a CMN is proportional to the extent of the nevus.¹⁰ It has also been

found to be linked to the satellite lesions present in the patient. Other complications like transformation to neurocutaneous melanosis consisting of the spread of melanoma to the central nervous system and leptomeninges are also dreadful.¹¹ Hence, early removal is recommended. Timely intervention and proper treatment of congenital melanocytic nevus are advisable. The treatment plan may be surgical removal or laser. It should be determined by looking at the depth of involvement of the nevus. If it involves only the superficial layers, we prefer excision and grafting as it is simple and provides better outcomes. Patient cooperation is also better if we go for grafting instead of the flap, and regrafting can be done if there is graft loss in primary intervention. Here, we have reported a rare case of a six-year-old male child presenting with a medium-sized congenital melanocytic nevus on the face with hairy tufts in between. A biopsy was done, which was consistent with our clinical diagnosis. The patient was managed with subsequent lesion excision followed by split skin grafting. The study aimed to report a rare case of congenital melanocytic nevus and to determine the importance of timely diagnosis and intervention.

2. CASE PRESENTATION

- **History and clinical examination:** A six-year-old male child brought by their parents came complaining of black discoloration of the skin over the left side of the face, which has been present since birth. No significant family history of such lesions was reported. The physical examination revealed an extensive pigmented patch over the face's left side, covering almost fifty percent of the area over the left cheek and around the left eye. In addition, tufts of coarse and lustreless hair of 2-3 cm in length were scattered over the lower part of the lesion (Figure 1).
- **Medical History:** No significant medical or surgical history was found.
- **Family History:** No relevant family history was found.



**Fig 1: A- Pre-operative image of congenital hairy naevus
B-Healing of skin graft after skin grafting (Post-operative day 9)**

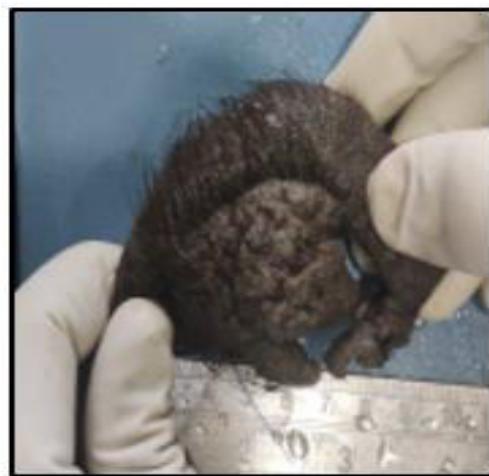


Fig 2: Gross: Irregular blackish skin tissue piece measuring 9 x 5 x 0.2 cm with hair attached

- **Special tests and investigations:** A biopsy from the representative area was taken.
- **Diagnosis:** The section from the biopsy showed histopathological features consistent with a congenital junctional melanocytic nevus.
- **Management:** The lesion was excised and sent for histopathological examination. Grossly, we received an

irregular blackish skin tissue measuring 9 x 5 x 0.2 cm. (Figure 2). The final diagnosis was reported to be congenital junctional nevus (Figure 3). After excision, successful split skin grafting was done.

- **Follow-up:** The patient was followed up, and the postoperative period was uneventful.

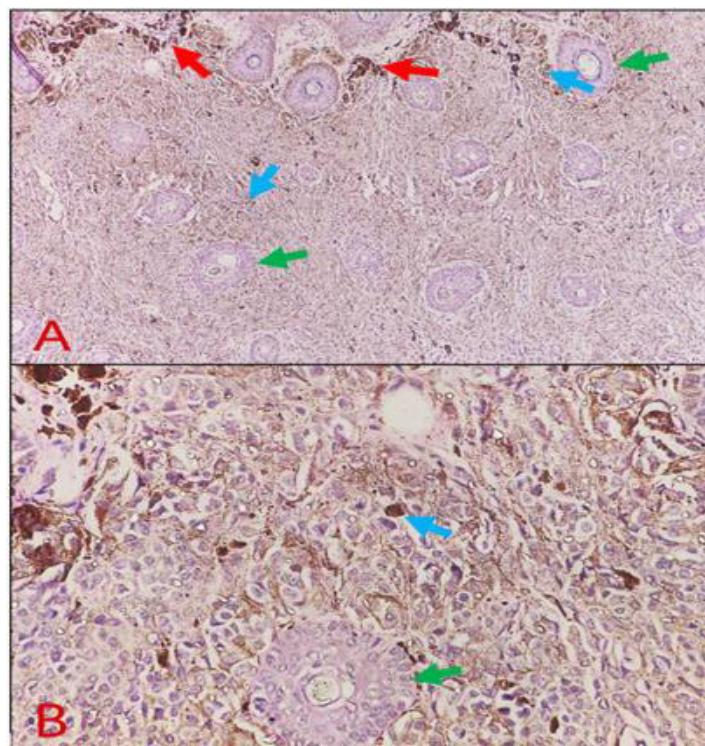


Fig 3: Microscopy

1. Figures show round cells with small round nuclei with brown pigment (melanin) in cytoplasm. The cells are located at the dermo-epidermal junction (red arrow).
2. Few hair follicles also are seen in some places (green arrow).
3. Elongation of epidermal ridges, hyperkeratosis, and hyperplasia, and an increase in the number of melanocytes is seen (blue arrow).

A – Photomicrograph at low power view (10x)
 B – Photomicrograph at high power view (40x)

3. DISCUSSION

Congenital melanocytic nevus is a pigmented skin lesion formed by melanocytic cells derived from the epidermis and

dermis. They can appear at birth or after a few weeks of life.⁵ The lesion's presence from birth, inclination to develop in size and potential for malignant transformation characterize it from the acquired type of melanocytic naevus.⁶ Many authors have

classified the lesion based on its size, as it has been observed that The likelihood of sequelae is proportionate to the naevus' largest diameter.⁷ Kopf et al. suggested three distinct types of congenital melanocytic nevus according to its largest diameter: small (less than 1.5 cm), medium (from 1.5-19.9 cm), and large or giant (more than or equal to 20 cm) which is the most widely accepted classification.⁸ Epidemiologically, approximately one percent of live births have congenital melanocytic nevus. It occurs more frequently in females with a female: male ratio ranging from 1.2:1 to 1.5:1.⁹ Our case is a male patient, which is uncommon for this lesion. Melanoma is more common in children with a congenital melanocytic nevus. Melanomas in Giant Congenital Nevus typically develop before puberty in the first 5 years of life, as opposed to small and medium nevi, where they commonly develop after puberty. The size and location of the congenital melanocytic nevus, as well as its linkage with multiple satellite nevi, appear to have an impact on malignancy and melanoma development. The likelihood of melanoma development from a CMN is proportional to the extent of the nevus. It has also been found to be linked to the satellite lesions present in the patient.¹⁰ The risk of transformation to malignant melanoma is greatest before adolescence.¹¹ Malignant transformation manifests clinically as hyper- or hypopigmentation, a papule or nodule, and occasionally ulceration. Melanoma in GCN is most commonly found at the dermal-epidermal junction, but it can also be found in the dermis and subcutaneous tissue.¹² Histological examination reveals high mitotic activity, cytonuclear atypia, transformation to atypical melanocytes in the epidermis, epidermis flattening or ulceration, and short maturation of the melanocytes at the lesion's base.¹³ In our case, no other satellite lesion was found, and the existing melanocytic nevus was medium. Our case showcased increased melanocytes at the dermo-epidermal junction, but they lacked nuclear atypia. The risk of conversion into malignant melanoma stresses the importance of timely diagnosis and excision of congenital melanocytic nevus. The pathogenesis is driven by genetic mutations, specifically the NRAS gene, which regulates cell division, and BRAF gene mutations.¹⁴ Thus, between the 4th and 6th weeks of pregnancy, these gene abnormalities cause a morphological defect in the neuroectoderm, resulting in the unrestrained proliferation of melanocyte progenitor cells known as melanoblasts, which leads to the creation of congenital melanocytic nevus.⁵ The lesion is usually asymptomatic but is cosmetically displeasing to some patients. Physical examination results vary, but infected individuals typically show brown to black, nodular or flat, well-defined lesions associated with excessive hair growth at the lesion.¹⁵ It is most commonly located on the trunk. The common terms used are 'bathing trunk' when it is present in the sacral and perineal areas and 'turtle child' when the whole back is involved. The face is an uncommon site of presentation as in our case. Clark noted that perhaps the complex cellular structure of some nodular overgrowths in congenital melanocytic lesions causes diagnostic challenges at the histopathological level. At birth or in the neonatal period, the major four histological patterns of proliferation have been evidenced in CMN: 1) simulants of superficial spreading melanoma having several large epithelioid melanocytes; 2) simulants of nodular melanoma which have black nodules of epithelioid melanocytes; 3) Neurocristic hamartomas (Nodular proliferative) 4) Biologically malignant melanomas.¹⁶ In our case, no nodular overgrowth was demonstrated, but it showed a classic flat-pigmented nevus with coarse hair tufts

present on the surface of the nevus. Giant melanocytic congenital nevus may have either organic or inorganic consequences. Indeed, it is well recognized that the lesion's displeasing nature might have psychological and social ramifications, negatively affecting the patient's self-regard, particularly females.¹⁷ As was nearly the case with our patients, psychological stress can lead to social marginalization. In less than 5% of instances, the most serious organic consequences include neurocutaneous melanosis and the progression to cutaneous melanoma. Our case belonged to the medium category of classification. Also, it was noted in the other studies that the giant cutaneous melanocytic nevus (>40 cm diameter) and those which have more than twenty satellite lesions have a risk of progression.¹⁸ Although not seen in our case, multiple congenital disabilities are frequently related to the lesion, which may worsen the prognosis: Occult spina bifida, meningocele, club foot, neurofibromatosis, lipomatosis, and limb hypertrophy or atrophy.¹⁹ Although congenital melanocytic naevus is mostly clinically diagnosed, the histological investigation is required to confirm the diagnosis and rule out malignant changes. Our case's most typical histological findings include hyperkeratosis and hyperplasia, elongation of epidermal ridges, and an increase in the number of melanocytes. Ultimately, congenital melanocytic nevus procedures range from surgical excision to laser treatments. Treatment should be tailored to the patient's age, the dimension and placement of the lesions, the likelihood of melanoma, neurocutaneous melanosis, and other congenital disabilities or comorbidities. All of these factors influence the prognosis.²⁰

4. CONCLUSION

Hence, on personal experience and going through the literature, timely intervention and proper treatment of congenital melanocytic nevus are advisable. The treatment plan should be determined by looking at a depth of involvement of naevus. If it involves only the superficial layers, we prefer excision and grafting as it is simple and provides better outcomes. Patient cooperation is also better if we go for grafting instead of the flap, and regrafting can be done if there is graft loss in primary intervention. In case of delay in treatment, there are chances of the lesion turning into malignancy, so prompt surgical and medical treatment is necessary in such cases for preventing social stigma as well.

5. ETHICAL APPROVAL STATEMENT

Written and oral informed consent from parents was taken, and ethical approval was taken from the Institutional Ethical Committee.

6. AUTHORS CONTRIBUTION STATEMENT

Dr. Ketki Wajpeyi has collected information and prepared the manuscript, which has been thoroughly reviewed and approved by Dr. Kishor Hiwale. Dr. Anil Reddy has planned the treatment protocol for the patient. All the authors have read and agreed to the manuscript.

7. CONFLICT OF INTEREST

Conflict of interest declared none.

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