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Two-Stage Approach to Managing a Giant Congenital Melanocytic Nevus of the Scalp in an Infant

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Abstract

Congenital melanocytic nevus (CMN) is a benign skin condition that affects the epidermis and dermis. Large to giant CMNs are associated with a higher risk of malignancy over a lifetime, underscoring the importance of assessing and monitoring their potential for malignant transformation. We present a case of a large to giant CMN on an infant's scalp, emphasizing its potential risk for malignancy. The infant underwent a successful two-stage surgical procedure, resulting in excellent aesthetic outcomes.

Keywords

Congenital Melanocytic Nevus, Scalp, Skin Graft, Tissue Expansion, Case Report

Introduction

Congenital melanocytic nevus (CMN) is a benign skin condition present at birth that affects both the epidermis and dermis, occurring in approximately 1–2% of newborns without gender preference. CMNs can present with various surface textures, such as papular, rugose, pebbly, verrucous, or cerebriform, and often feature darker, thicker pigmented hairs [1]. Large to giant congenital melanocytic nevi carry an estimated lifetime melanoma risk of 3 to 11% [2]. Generally, treatment options such as full or partial excision, curettage, laser treatment, or a combination of these methods aim to reduce the risk of malignancy. However, there is no consensus on the optimal strategy for treating giant CMNs due to their varying sizes and locations, which can complicate partial or complete removal, particularly in infants. In this case, we report an infant with a large to giant CMN on the scalp who underwent a successful two-stage surgical procedure.

Case Presentation

An 11-month-old baby presented to our clinic with a giant congenital melanocytic nevus on the top of the forehead (**Fig-1A** and **Fig-1B**). The patient was born with black lesions on the scalp, which have progressively enlarged with age. The lesion is uneven and asymmetrical, with irregular borders, uneven pigmentation, and a prominent nodule at its center. The nevi have darkened over time without any spontaneous regression since birth, and there were no reported prenatal complications. Physical examination revealed normal development and nutrition, with a normal head circumference and no abnormalities in neurological, cardiovascular, musculoskeletal, or ophthalmic assessments. The family history was negative for congenital nevi or melanoma.

Considering the rapid progression of the lesion and the potential risk of malignancy, we planned a two-step surgical excision. In the first step, we completely excised the giant melanocytic nevus on the scalp. We then used a skin flap harvested from the right abdominal wall, prepared it into a medium-thickness mesh graft at a 1:2 ratio, and covered the residual scalp defect (**Fig-1C**). The donor sites were closed by direct suturing. Pathological diagnosis confirmed CMN with predominantly small pigment granules, significant nucleolar pleomorphism, and abundant cytoplasm, indicating a potential risk of malignancy.

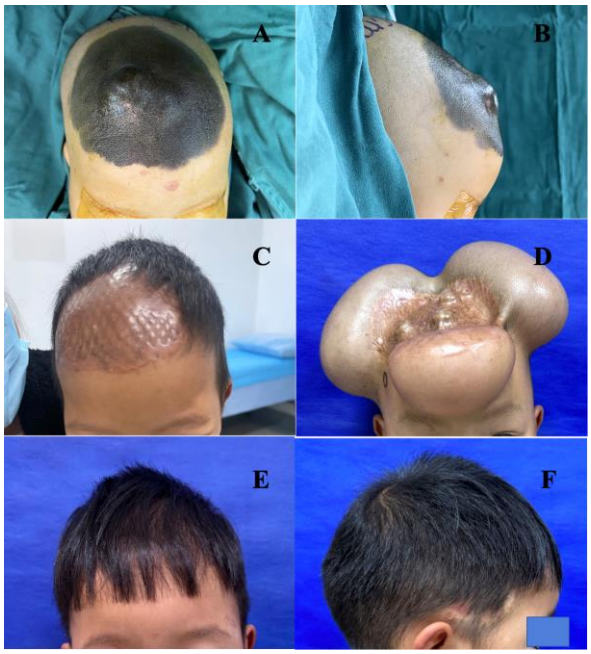

Fig-1A and Fig-1B: *A giant congenital melanocytic nevus on the scalp, characterized by an uneven and asymmetrical appearance, with irregular borders, uneven pigmentation, and a prominent central nodule.*

Fig-1C: *Scarring with hair loss formation on the top of the head.*

Fig-1D: *The expanders were inflated with normal saline twice a week for about 4 months to achieve the target volume.*

Fig-1E and **Fig-1F:** *After a one-year follow-up, the patient recovered well with no significant complications and achieved excellent cosmetic results.*

In the second step, when the patient was two and a half years old, we implanted three skin tissue expanders in his normal scalp. The expanders were inflated with normal saline twice a week for about 4 months to achieve the target volume (**Fig-1D**). We then excised the scar in the bald area and used the expanded normal scalp to cover the wound. After a one-year follow-up, the patient recovered well with no significant complications and achieved excellent cosmetic results (**Fig-1E** and **Fig-1F**).

Discussion

Melanocytic nevi, benign proliferations of melanocytes arranged in nests, are typically present at birth but can also develop within the first two years of life [3]. The prevalence of CMN ranges from 0.005% to 2.7%, depending on lesion size. They are caused by gain-of-function somatic mutations in BRAF or NRAS, which affect the MAPK pathway, leading to abnormal proliferation and dispersion of embryonic melanoblasts. While most cases are sporadic, some are familial [4]. The clinical presentation of CMN varies by size, location, and age. CMN are categorized into small (<1.5 cm), medium (1.5-19.9 cm), and large/giant (⩾ 20 cm). Size is crucial for assessing malignancy risk and determining cosmetic and therapeutic strategies.

Giant CMN are typically brownish lesions with distinct borders and hypertrichosis, larger and more heterogeneous than acquired nevi, with varying clinical presentations throughout life. In our case, the patient was born with black lesions on the top of his scalp that progressively enlarged, exhibiting asymmetry, irregular borders, uneven pigmentation, and a central prominent nodule. This nodule showed active proliferative characteristics, with reports suggesting potential invasion into subcutaneous tissue, periosteum, and even the skull and meninges [5]. The primary concern for this patient was the gradual enlargement of the lesion and its potential transformation into melanoma.

Although Bauer et al. reported the use of tissue expansion on the scalp and forehead for giant CMN in children as young as 6 months, there are inherent risks associated with this procedure [6]. Given that the patient's fontanelle had not completely closed, direct placement of skin tissue expanders posed potential risks. Moreover, due to the risk of temporary or permanent cranial deformation, tissue expansion in children under 18 months of age remains controversial. Therefore, we opted for a two-step surgical approach. In the first step, we excised the lesion and covered the wound with a skin graft obtained from the right abdominal wall. In the second step, when the patient was two and a half years old, we

implanted tissue expanders to enlarge the normal scalp and subsequently transplanted the expanded flap to replace the scarred bald area. This strategy aimed to achieve a normal cosmetic appearance before the patient reached school age, thereby improving both physical and psychological health.

Conclusions

A two-step surgical procedure, involving initial lesion excision and skin grafting followed by tissue expansion, proves to be an effective method for removing giant congenital melanocytic nevi on the scalp. It offers favorable cosmetic and oncological outcomes and is recommended at a young age.

Conflict of Interest

The author has read and approved the final version of the manuscript. The author has no conflicts of interest to declare.

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