

Case report

Giant Congenital Melanocytic Naevus: The Effect of Patient-Centered Multidisciplinary Approach on Quality of Life

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Abstract

Giant Congenital Melanocytic Nevus, a rare skin lesion associated with a significantly increased relative risk of childhood melanoma compared to the general population. This report details the successful management of a five-year-old boy with Giant Congenital Melanocytic Nevus through serial tissue expander skin excision, overcoming complications and showcasing the efficacy of this approach, with perceived Quality of Life (QoL) measured, revealing a small to medium effect. The correlation between QoL and satisfaction with surgical outcomes is explored, highlighting the impact of insufficient pre-procedural information on dissatisfaction, and emphasizing social factors, such as peer perceptions, contributing to low social acceptance in Giant Congenital Melanocytic Nevus cases. A proposed biopsychosocial model aims to address challenges through tailored counselling for parents and children, emphasizing the importance of effective communication, especially for children facing bullying.

Keywords: Paediatric; Dermatology; Nevus; Quality of life; Serial tissue expander skin excision; Biopsychosocial model

Abbreviations

1. Giant Congenital Melanocytic Nevus (GCMN)
2. Congenital Melanocytic Nevus (CMN)

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3. Quality of Life (QoL)

Introduction

Giant Congenital Melanocytic Nevus (GCMN) can be defined as a proliferation of melanocytes within the skin that develops in utero, is present at birth and reaches a diameter larger or equal to 20 cm in adulthood. The incidence of GCMN is estimated to be less than 1:20,000 making it an exceedingly rare condition [1]. Congenital melanocytic nevi (CMN) are significantly associated with an increased relative risk of developing melanoma during childhood compared to the general population ranging from 0.7% [2] to 2.3% [3]. However, both aforementioned studies indicated a higher incidence with increased size of the nevi.

GCMN is one of the many skin conditions that can cause physical and mental stress during childhood leading to impaired Quality of Life (QoL). Children's Dermatology Life Quality Index (CDLQI), validated for ages 4 to 16, was introduced in 1995 [4] and since then has been deployed for a large spectrum of childhood skin conditions. Furthermore, the Family Dermatology Life Quality Index (FDLQI) was created to measure the secondary impact of skin conditions on the family of the patient [5].

Due to the complexity of GCMN, the treatment options require a multidisciplinary, individualized approach that ranges from non-interventive monitoring to novel topical treatment and surgical excision [6,7]. Interestingly, one of the most discussed surgical interventions in literature is serial tissue expander skin excision [8]. With an ever-growing knowledge of surgical techniques, we would like to shed light on the importance of a multifaceted approach incorporating the psychosocial aspect of both parent and child in the management on GCMN. The aim of the current report is to present the effect of GCMN diagnosis and management on the patient's and family's QoL.

Case Presentation

A five-year-old boy presented to hospital with multiple episodes of vomiting, a fever of 39.2°C and reduced feeding. He was admitted for IV fluid replacement with a working diagnosis of viral gastroenteritis. Upon physical examination a large nevus was observed on the patient's back.

Medical history revealed normal vaginal delivery at term with birth weight of 3.9kg, following an uncomplicated pregnancy. Both parents are non-consanguineous, and healthy with no history of genetic syndromes. His mother has a history of two melanocytic nevi resected in the past, with insignificant pathology reports. At birth, a lesion, 28cm at its largest point, was noted on his back and buttocks area (Figure 1). The lesion had a darker area in the right lower quadrant and additional two melanocytic nevi below his left knee, in the mid femoral region and a hyperpigmented lesion on left elbow.

Soon after birth, the boy was initially seen by a paediatric dermatologist, who investigated potential central nervous system malformations, without revealing any structural abnormalities. Genetic



Figure 1: 28cm Giant Congenital Melanocytic Nevus located on the posterior trunk and buttocks taken post-partum.

assessment concluded the cause to be a somatic mutation of the NRAS gene with potential complication including malignant transformation to melanoma, neurocutaneous melanosis and other central nervous system malformations. Three plastic surgeons were consulted, suggesting various treatments including curettage, conservative management, and serial tissue expander skin excision. After thoughtful deliberation and shared-decision making approach the latter method was followed for both aesthetic reasons and melanoma risk reduction. (Figure 2).



Figure 2: Granuloma formation on the suture line of the first skin excision surgery using tissue expanders, at 3 years 5 months old.

The patient underwent his first surgery at the age of 23 months and at the age of 2 years and 4 months a tissue expander was placed. The father of the patient who is a nurse was responsible for inflating the expanders. Five months later, partial excision of the naevus was performed, with biopsy revealing a mixed naevus. Another tissue expander surgery took place at the age of 3 years and 5 months (Figure 3). A repeat surgery was conducted a week later, due to pus collection around the left tissue expander. At 3 years and 11 months old, surgery was performed to further excise the lesion.



Figure 3: The patient prior to most recent skin excision surgery with tissue expanders fully inflated, at 5 years 9 months old.

So far, the patient went through 3 cycles of serial tissue expander skin excision that lasted from 4 to 6 months and has more surgeries scheduled in the years to come. The most recent image of the nevus is presented in (Figure 4). Some of the complications seen throughout management included expander deflation during physical activity, as well as, post-operatively back oedema, low-grade fever, granulomatous lesion, and a subcutaneous pus collection requiring paracentesis. The patient was admitted to hospital and received antibiotics accordingly on multiple occasions due to the treatment complications.



Figure 4: The patient after third cycle of lesion excision surgery, with the nevus covering the upper quadrants of the left buttock and the upper inner quadrant of the right buttock, spanning to the anterior superior iliac spine bilaterally, stretching to the iliac crest posteriorly and bordered the linear surgical scar.

At the age of 5 years and 6 months old, the patient and his parents completed the CDLQI and FDLQI respectively. The CDLQI score was 6 points out of 30. Aspects such as itchiness/pain, embarrassment, playing, sport avoidance, bullying and treatment perception, were reported as having “little” impact on the boy’s life. The FDLQI score, on the other hand was 8 points out of 30, featuring emotional

distress as “quite a lot” of impact, while personal relationships, physical well-being, problems with peoples’ reactions, social life, time spent as a carer and effect on job have been affected only a “little”. There was no reported effect on the family member’s leisure activities, extra-household work or extra expenditures. This indicates that CGMN had a “small” effect on patient’s QoL and “medium” effect on his family’s QoL.

Discussion

This case report details the long-term management of a five-year-old boy with GCMN, undergoing successful serial tissue expander resections. Serial excision with assisted balloon expansion, popular for its natural-looking results, was associated with higher satisfaction in a 2021 case series. Additionally, caregivers and surgeons unanimously considered surgical intervention at a younger age as the best choice for optimal long-term outcomes [8]. Similar findings were reported in a cohort study comparing the expander technique with other surgical methods [2].

While surgical management is the preferred approach for GCMNs, it imposes significant psychological stress on paediatric patients and their families. Factors like lower socioeconomic status, older age, malignant melanoma presence, neurological issues, skin discomfort, and lesion visibility are linked to diminished Health-Related Quality of Life and psychological adaptation in children and teenagers with GCMN [6]. This case report quantifies the perceived QoL of the patient and parents using QoL indices, revealing a small to medium effect respectively. An analysis of 135 CMN reports indicated varied impacts on children’s and adolescents’ QoL, with 75% of parents reporting “no” or “small” and 19% a “moderate” impact on their child’s QoL. While 46% of adolescents reported “no” or a “small impact”, 43% experienced a “moderate” impact on their QoL [9].

The correlation between the QoL and satisfaction with surgical outcomes is well-established in the literature. However, a study from 2017 indicated that a primary cause of dissatisfaction with therapeutic results often stemmed from perceived shortcomings in pre-procedural information [2]. In our case, this issue did not significantly impact satisfaction, possibly due to the patient’s father being a nurse, providing a more comprehensive understanding of the disease from the outset and inflating the expander balloons himself at home. However, it is crucial to recognize that this circumstance may not be universally applicable to all cases. The father of our patient emphasized that the families with children affected by GCMN that he met over the years frequently grapple with numerous unanswered questions and have not received sufficient information.

In addressing challenges associated with GCMN management, healthcare professionals should consider implementing a biopsychosocial model, recognizing the interplay of biological, psychological, and social factors influencing an individual’s health. Providing tailored counselling to both parents and children is essential within this framework [10]. Emphasizing social factors, including self-esteem and mental health, is crucial, given the significant impact of peers’ perceptions on individuals with the condition [6]. It’s noteworthy that children with skin conditions often experience social isolation due to differing physical appearances [10]. A study in 2021 highlighted that GCMN is perceived with only 0.65% social acceptance, while 94.12% consider scars acceptable [11]. The case of a 5-year-old patient facing bullying at a young age, underscores the importance of early counselling.

Within the context of emphasizing a favourable long-term aesthetic outcome, healthcare professionals should prioritize effective communication between patients and doctors. Achieving optimal outcomes in GCMN management necessitates a comprehensive approach, involving a multidisciplinary team comprising a plastic surgeon, paediatric dermatologist, geneticist, radiologist, and paediatrician [12,13]. As treatment progresses, the inclusion of a psychologist in the healthcare team could further enhance the holistic well-being of the child.

Conclusion

Management of GCMN is a challenging and long-term process with considerable impact on the patient’s and family’s daily life. Given the absence of standardized treatment guidelines for GCMNs, a multidisciplinary approach and shared decision making are crucial for positive medical and psychosocial outcomes. The serial tissue expander skin excision surgery proved to be successful in this case and despite its complications the family is happy with the overall outcome so far. The parents wish the full excision to happen before the patient enters primary school to avoid further psychological pressure and social isolation.

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Competing Interests

The authors declare that they have no competing interests.

Availability of data and materials

The original contributions presented in the case report are included in the article, further inquiries can be directed to the corresponding author.

Consent for publication

The father gave informed consent to publication and signed informed consent regarding publishing his child’s data.

Ethical approval

The ethical approval was obtained from the Cyprus National Bioethics Committee for this case report.

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