

Congenital melanocytic nevi: a retrospective study of epidemiological and therapeutic aspects in a series of 45 patients

Nevo melanocítico congênito: estudo retrospectivo dos aspectos epidemiológicos e terapêuticos em uma série de 45 pacientes

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ABSTRACT

Introduction: Although relatively rare, congenital melanocytic nevi are lesions that require monitoring for a long period in order to prevent psychological sequelae and complications inherent to this pathology such as psychological stress, neurocutaneous melanosis, and malignant degeneration. **Methods:** This study presents epidemiological therapeutic data from a series of 45 cases treated at the Divisão de Cirurgia Plástica e Queimaduras, Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo.

Keywords: Nevi and melanomas. Nevus. Melanoma. Melanosis.

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RESUMO

Introdução: Os nevos melanocíticos congênitos, apesar de apresentarem uma incidência relativamente rara, são lesões que necessitam acompanhamento por longo período, a fim de se evitar sequelas psicológicas e complicações inerentes a essa patologia, como estresse psicológico, melanose neurocutânea e degeneração maligna. **Método:** O presente estudo demonstra os dados epidemiológicos e terapêuticos em uma série de 45 casos atendidos na Divisão de Cirurgia Plástica e Queimaduras do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo.

Descritores: Nevos e melanomas. Nevo. Melanoma. Melanose.

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INTRODUCTION

Congenital melanocytic nevi (CMN) are pigmented lesions present from the time of birth, and they result from the accumulation of melanocytes of neuroectodermal origin in ectopic locations¹. Some peculiar histological features define the congenital etiology of the nevi, such as the presence of nevus cell clusters and the presence of such cellular elements in glandular structures, blood vessels, and subcutaneous tissue².

The CMN incidence in the general population is 1 in 100 live births; when considering only giant nevi, the incidence is reduced to 1 in 20,000 live births³. Its body distribution is broad, being found more frequently on the trunk and face^{4,5}.

CMN are classified according to their size in different ways. The 3 most cited classifications take into account the nevus' larger diameter, its area, or the percentage of body surface it occupies; according to each classification method, nevi are considered giant when they are more than 20 cm in diameter, cover more than 100 cm² of the body surface, or cover more than 2% of the body surface, respectively⁶⁻⁸.

The presence of neurocutaneous melanosis, malignant degeneration into melanoma, and the psychological stress for families and children are the major motivations for CMN treatment⁹.

The actual incidence of melanoma in CMN remains controversial in the literature, and most recent case selections report the incidence to range from 2.8-8.5% of patients with congenital nevi⁴. A nevus that is >20 cm², the number of lesions, and age of <3 years are the clinical characteristics associated with the risk of malignant transformation^{7,10-14}.

Surgical treatment is the preferred option when appropriate. The following surgical modalities are used: resection followed by skin grafting, partial resection, and the use of tissue expanders in the adjacent healthy areas followed by resection of the lesions and advancement or rotation of the expanded flaps.

This paper aims to demonstrate the experience of a referral service for CMN treatment from the perspectives of epidemiology, clinical presentation, the employed surgical modality, histopathological reports, and the rate of malignancy in a series of 45 patients.

METHODS

This study is a retrospective study conducted by a medical history review of patients treated at the Divisão de Cirurgia Plástica e Queimaduras, Hospital das Clínicas, Faculdade de Medicina, between 1998 and 2010. CMN was diagnosed considering the related ICD-10 codes: D-28, I-78.1, and Q-82.5. The medical records of 45 patients were located and reviewed.

Epidemiological, clinical, and anatomopathological data were assessed. Epidemiological data included age, gender, ethnicity, and family history. The lesion location, size, and the presence of satellite lesions were reviewed clinically. The employed surgical modality as well as the number of required surgical approaches and complications were also assessed.

The anatomopathological studies of the resected specimens were reviewed to observe the histological types, macroscopic size, and incidence of neoplastic foci.

RESULTS

Of the 45 patients who were assessed, 62% were female and only 13% had a family history of congenital nevus. The ethnic distribution was as follows: 75% of patients were white; 16%, brown; 5%, black; and 4%, Asian (Table 1).

The most frequent location of the CMN was the head and neck region in 35% of cases, followed by the trunk in 27%, and the limbs in 9%. The simultaneous occurrence of nevi in 2 body segments occurred in 20% of patients and occurrence in 3 or more in 9% (Figure 1).

With respect to size, in 69% of the cases the nevi covered 2% or more of the body surface, indicating a giant congenital nevus. Excluding nevi covering less than 1% of the body surface, the arithmetic mean of lesion occurrence was 9.65% (2-50%).

Satellite lesions were present in 30% of the studied cases.

In this case series, 29 (64%) patients underwent some type of surgical treatment. The remaining 16 patients were either under clinical observation, refused surgical treatment, or were awaiting a surgical program.

In absolute numbers, in these 29 patients, 66 partial resections, 13 resections followed by skin grafting, and 18 tissue expansions followed by rotation of the extended flaps were

Table 1 – Epidemiological data of patients with CMN.

Variables	N (%)
Gender	
Female	28 (62%)
Male	17 (37%)
Family history	
Yes	6 (13%)
No	39 (57%)
Ethnicity	
White	75%
Brown	16%
Black	5%
Yellow	4%

CMN = congenital melanocytic nevi.

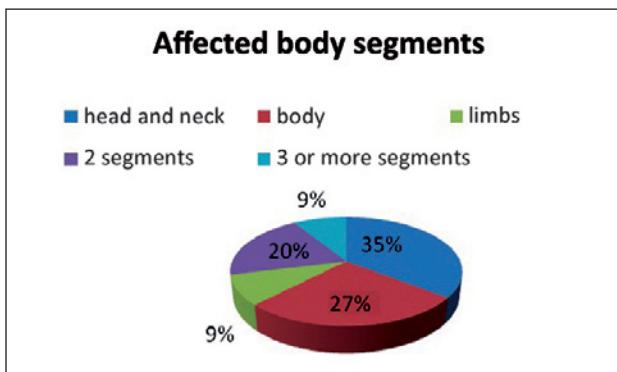


Figure 1 – Distribution of patients with CMN according to affected body segment.

performed (Table 2). Each child received an average of 2.57 surgical approaches (range, 1-9), and only 7 patients were discharged after completion of treatment. The other patients were either awaiting minor resections, refused to continue the treatment due to the high exposure of patients to surgical procedures, or were lost to follow-up.

Surgical wound or expander infections, small dehiscence, and wound healing disorders among others were considered as treatment complications. Of the 31% cases that had complications, most were infections (4/29); other complications were small wound dehiscence (2/29), perforation of expanders (2/29), and hypertrophic scarring (1/29) (Table 2).

The analysis of anatomopathological results showed that in 20 cases, there were reports of compound nevi, 12 intra-dermal nevi, and 2 junctional nevi. Of these, 16 reports mention pathognomonic characteristics of a congenital pattern. Patients with a diagnosis of blue nevus, nevus sebaceous of Jadassohn, and hamartoma of neural cyst as well as isolated cases of proliferative nodules in congenital nevi, typical hyperplasia, and Spitzoid melanoma were evidenced.

In this study, only 1 (2.2%) patient was diagnosed with melanoma. Although the revised report hypothesizes a differential diagnosis of Spitzoid pattern, a melanoma was considered for treatment and follow-up in this case. The patient was a female and had lesions on the trunk that occupied 24% of the body surface, without satellite lesions or neurocutaneous melanosis. As treatment, 7 surgical approaches had already been performed, including 5 partial resections and 2 tissue expansions.

Neurocutaneous melanosis was present in 3 (6.67%) cases, with lesions in meningeal and subcutaneous locations as well as the nails. All of these patients were white and had nevi in multiple segments; only 1 of them had satellite lesions.

With regard to the resected specimens, it was noticed that on an average 147.4 cm^2 of skin was resected in each patient, and the largest resection involved resection of 560 cm^2 of skin.

Figures 2 to 5 illustrate some cases in this case selection.

Table 2 – Surgical data of patients with CMN.

Variables	N (%)
Surgical procedures	
Partial resection	66 (68%)
Tissue expansion	18 (18.5%)
Resection + graft	13 (13.4%)
Complications	
Infection	4 (13.8%)
Perforation	2 (6.9%)
Wound dehiscence	2 (6.9%)
Hypertrophic scar	1 (3.4%)

CMN = congenital melanocytic nevi.

DISCUSSION

Although relatively rare, congenital nevi are lesions that require multidisciplinary monitoring for a long period in order to avoid psychological sequelae and complications inherent to its pathology.

In the presented case selection, female pediatric patients were predominant, although no consensus was found in the literature regarding gender dominance. With regard to ethnicity, the data corroborate a higher prevalence in white individuals.

Our results identify the head and neck regions as the most frequent areas of CMN, which corroborates the results published by Turkmen et al.⁵. However, other studies consider the trunk to be the most frequently affected area¹⁵. It is important to highlight that in this study, in 29% of the cases, a large proportion of nevi attained dimensions that exceeded the boundaries of body segments and affected more than 2 segments.



Figure 2 – A nevus occupying 50% of the body surface in a patient with CMN and neurocutaneous melanosis.



Figure 3 – Photo illustrating satellite lesions in CMN.



Figure 4 – Example of a patient with a congenital nevus on the face who underwent partial resections.



Figure 5 – Example of tissue expansion and flap rotation for a giant congenital nevus on the back.

Arneja and Gosain² published data showing the prevalence of satellite lesions to be 80%, which contradicts the present results that show that only 30% of the cases had satellite lesions.

There is a consensus that surgical treatment is most effective when its indication is relevant, but the surgical procedure depends on the characteristics of the lesion, the service complexity, and the plastic surgeon's personal experience. The approaches used in this study were not in line with those proposed in the international literature and predominantly comprised of partial resection followed by tissue expansion⁵; the reason for this may be the ease of implementing partial resection because of its lower costs and morbidity rates. However, partial resection requires a greater number of approaches to properly resolve each case as compared to tissue expansion.

Infectious complications are more frequent in pediatric patients, and infection remains the major type of complication, especially with the use of expanders. The incidence varies from 4.4% to 9.3% of cases¹⁶⁻²¹. The incidence was 13% in our case selection; furthermore, as already proposed in the literature, the occurrence of infection is not indicative of the interruption of the expansion program or its failure. In contrast, extrusion is another important complication and treatment involves early removal of the expander²¹.

Malignant degeneration into melanoma is considered an important complication of CMN because of the high morbidity and mortality associated with the pathology of CMN. Early published works present varying incidences of melanoma, ranging from 0-42%²²⁻²⁴. Recent studies show that the incidence is lower (around 2.8-8.5% of CMN cases) than what was previously believed^{7,9}. The present results are concordant with this since we obtained an incidence of only 2.2%. Although we identified only 1 case of malignant degeneration in our case selection, we observed that this patient presented with the risk factors proposed in the literature, i.e., large size (24% of the body surface) and age <3 years^{4,25}.

According to the latest diagnostic criteria for neurocutaneous melanosis, 3 patients presented with neurocutaneous melanosis in our case selection. Although all patients had large nevi, only 1 had satellite nevi, which are considered an important risk factor for neurocutaneous melanosis, especially when more than 20 satellite lesions are present²⁶. The occurrence of nevi on the midline, head, and neck is also considered an important risk factor for neurocutaneous melanosis²⁷, but this was not observed in our case selection. In our cases, the lesion was too large to be considered as restricted to one body segment or the midline of the body.

Depending on the age of incidence, neurocutaneous melanosis may be associated with clinical manifestations such as varying degrees of neuropsychomotor development delay, seizures, and weight and height growth restriction.

In this retrospective study, a preference for the treatment of patients with large nevi was noticed; these patients require repeated surgeries and follow-up. Clinical follow-up is critical for these patients in order to diagnose early malignancies and other complications.

Patients with giant CMN should receive special attention from multi-disciplinary staff, including occupational therapists, psychotherapists, and plastic surgeons. In these patients, psychological sequelae occur due to both the stigma associated with the pigmented lesions and the numerous hospitalizations and procedures required for total lesion excision. In order to prevent psychological sequelae and malignant degeneration in children with CMN, surgical treatment should be initiated as early as possible with the aim to complete treatment, if possible, when the child is of preschool age.

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