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REVIEW ARTICLE

Congenital melanocytic nevus – What are the potential complications?

Nevo melanocítico congénito – Quais as possíveis complicações?

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Abstract

Congenital melanocytic nevi (CMN) are benign cutaneous lesions resulting from post-zygotic somatic mutations. The estimated incidence in newborns ranges from 1% to 6%. Although most cases follow a benign course, those of larger size are associated with an increased risk of severe complications, such as cutaneous melanoma, leptomeningeal melanoma, and neurocutaneous melanosis. The psychosocial impact of these often disfiguring lesions has significant repercussions on the quality of life of children and their families. The aim of the present work is to review the existing literature on the management of CMN, with particular attention to the various complications that may arise. Review articles, systematic reviews, and case series, in Portuguese and English, published since 2010, were included. The diagnosis of a CMN includes clinical examination, and eventually, dermatoscopy, and histopathology. In large nevi, dermatological and neurological monitoring is essential, with recommended regular photographic documentation and brain and spinal cord magnetic resonance imaging before 6 months of age, to screen for possible neurological anomalies in high-risk patients. The detection of alarm signs, such as rapid growth, bleeding, pain, ulceration, or development of nodules, should be immediately evaluated, preferably by a dermatologist. The therapeutic approach should be individualized and multidisciplinary, aiming to reduce the risk of melanoma and mitigate its esthetic impact.

Keywords: Congenital melanocytic nevus. Complications. Melanoma. Neurocutaneous melanosis. Proliferative nodules.

Resumo

Os nevos melanocíticos congénitos são lesões cutâneas benignas resultantes de mutações somáticas pós-zigóticas. A incidência estimada em recém-nascidos varia entre 1% e 6%. Apesar de a maioria ter uma evolução benigna, os de maiores dimensões estão associados a um risco elevado de complicações graves, como melanoma cutâneo, melanoma leptomenígeo e melanose neurocutânea. O impacto psicossocial destas lesões, muitas vezes desfigurantes, tem repercussões importantes na qualidade de vida das crianças e das suas famílias. O objetivo do presente trabalho é rever a literatura existente sobre a abordagem aos nevos melanocíticos congénitos, com particular atenção às diferentes complicações que neles podem surgir. Foram incluídos artigos de revisão, revisões sistemáticas e séries de casos, de língua portuguesa e inglesa, publicados desde 2010. O diagnóstico de um nevo melanocítico congénito engloba a clínica, que pode ser variável, a dermatoscopia e a histopatologia. Nos nevos de maiores dimensões a monitorização dermatológica e neurológica é fundamental, sendo

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recomendada a documentação fotográfica regular e a realização de ressonância magnética cerebral e da medula espinhal antes dos seis meses de vida para rastrear possíveis anomalias neurológicas em doentes de elevado risco. Sinais de alarme, como crescimento rápido, hemorragia, dor, ulceração ou desenvolvimento de nódulos devem ser imediatamente avaliados por um dermatologista. A abordagem terapêutica deve ser individualizada e multidisciplinar com o objetivo de reduzir o risco de melanoma e atenuar o seu impacto estético.

Palavras-chave: Nevo melanocítico congénito. Complicações. Melanoma. Melanose neurocutânea. Nódulos proliferativos.

Introduction

Congenital melanocytic nevi (CMN), present in 1-6% of newborns, arise from clonal proliferation and migration of neural crest-derived melanocytes (NC), secondary to sporadic post-zygotic somatic mutations, most commonly involving Neuroblastoma RAS viral oncogene homolog (*NRAS*) and B-Raf proto-oncogene, serine/threonine kinase (*BRAF*) oncogenes.[1]

Large and giant CMN are associated with an increased risk of complications, including cutaneous melanoma (CM), leptomeningeal melanoma, and neurocutaneous melanosis (NCM).

Proliferative nodules (PN) may develop within CMN and can clinically and histologically mimic melanoma. Histopathology, immunohistochemistry, and molecular analysis are essential for accurate differentiation.

A multidisciplinary approach is recommended, including dermatological, neurological, and psychosocial follow-up. However, variability in management recommendations has led to a lack of standardized guidelines.

The aim of this study is to review CMN, focusing on associated complications, and to provide an updated and clinically relevant synthesis.

Materials and methods

The PubMed electronic database was searched using a query composed of MeSH descriptors related to melanocytic nevi and their complications.

The selected articles included review articles, systematic reviews, and case series, published in Portuguese and English, since 2010. Isolated case reports, articles without full-text access, and those lacking relevant content for the topic under analysis were excluded from the study.

Congenital melanocytic nevus

Between the 8th and 10th weeks of embryonic development, melanocytes from the NC migrate from the embryo to various areas of the body, including the skin, mucosae, uveal layer of the eye, inner ear, adrenal glands, and central nervous system (CNS). Within the latter, they are located at the level of the leptomeninges, preferentially distributed at the base of the skull, ventral brainstem, and cervical spinal cord.[2,3]

If mutations in the NRAS and BRAF oncogenes occur during an early stage of embryonic development, they may affect a multipotent progenitor cell, leading to the formation of multiple cutaneous CMN and, in some cases, involvement of other systems such as the CNS.[4]

Mutations in other genes have also been described, namely: *MC1R, TP53, KRAS, APC, MET, PIK3CA*, and *GNAQ*.[5,6]

The etiology is predominantly sporadic, although some risk factors have been described, such as a family history of CMN and neurofibromatosis type 1.[7]

Large or giant CMN are associated with higher morbidity and have an estimated incidence of approximately 1 in 20,000 births and 1 in 500,000 births, respectively.[8]

Classification

Classification according to projected adult size (PAS) is currently the most widely used. It is crucial for prognosis.[9] According to this classification, CMN can be subdivided into: small (< 1.5 cm), medium (M1: 1.5-10 cm; M2: > 10 and ≤ 20 cm), large (L1: > 20 and ≤ 30 cm; L2: > 30 and ≤ 40 cm), and giant (G1: > 40 and ≤ 60 cm; G2: > 60 cm).[1]

Multiple medium-sized CMN are defined as the presence of three or more medium-sized nevi without an associated larger primary nevus. This classification also includes the number of satellite lesions (S0: no satellite lesions; S1: < 20; S2: > 20 and < 50; S3: > 50) and other characteristics, such as anatomical location, surface rugosity, color heterogeneity, presence of subcutaneous nodules, and hypertrichosis.[1,9,10]

Diagnosis

The diagnosis of CMN is based on clinical, dermoscopic, and histopathological evaluation. On skin

examination, the trunk and extremities are the most common locations, although lesions may also occur on the face or scalp.[11,12] They may present as solitary, multiple, or confluent lesions.[13]

Lesions may be flat (macules and patches), palpable (papules, plaques, and nodules), or both,[5] and may evolve into plaques or nodules.[1] More rarely, they may present as verrucous or cerebriform lesions.[14] Variations in pigmentation may be observed, particularly in multiple lesions.[5,14] Pigmentation intensity tends to decrease, especially during the 1st year of life.[13] The growth of CMN parallels somatic growth, reaching maximum size in adulthood.[1,15]

Warning signs include peripheral growth along the contour of the main nevus or vertical growth, poorly defined borders, and areas of hypo- or hyperpigmentation that differ from the baseline coloration of the nevus.[14] Ulcerated lesions, vascular patterns, and rapidly growing nodules, particularly when adherent to deeper structures, are features suggestive of melanoma.[1]

There is a correlation between lesion size and the depth of melanocytic infiltration; therefore, larger nevi show a greater tendency to involve the deep dermis, adnexal structures, and neurovascular elements.[14,16]



Figure 1. Proliferative nodule in a newborn with a giant congenital melanocytic nevus.



Figure 2. Giant congenital melanocytic nevus in a newborn with a superimposed raised erythematous dome-shaped nodule.

Complications

Proliferative nodules

These lesions correspond to benign proliferations of melanocytes, arising mainly in large, giant (Figs. 1 and 2), or multiple CMN.[14] Their prevalence in giant CMN ranges from 2.9% to 19%.[1] They may be present at birth or appear at any stage of childhood and typically exhibit rapid growth over several weeks, followed by stabilization. Subsequently, they may remain stable or increase in size and/or undergo hyperpigmentation. Their natural course is most often characterized by spontaneous regression.[1,4]

Histologically, PN may occur in the superficial or deep reticular dermis and is characterized by increased cellularity. They do not exhibit necrosis, cytological atypia, pagetoid spread in the upper epidermis, or increased proliferative activity, with a Ki-67 proliferative index typically ≤ 10%.[4,14] Atypical mitoses are absent, and mitotic figures are rare.[14] However, PN composed of small cells may be associated with higher mitotic indices and an increased Ki-67 index, and cases of PN with cytological atypia have been reported.[4,17] In such cases, the cells are generally homogeneous and exhibit low mitotic activity. PN with rapid growth, ulceration, and/or hemorrhage, or high mitotic activity on histopathology, is particularly challenging.[1]

Immunohistochemical analysis comparing histologically benign PN and atypical PN has demonstrated differences in the expression of Ki-67 and phosphohistone H3, with higher expression observed in atypical PN, reflecting increased proliferative activity.[4]

A study conducted by Tan et al. evaluated PRAME, an antigen predominantly expressed in melanomas, as a diagnostic tool. The results showed that anti-PRAME antibody expression was diffusely positive in melanoma and negative in PN.[6]

Congenital melanocytic nevus syndrome

Children with CMN may have extracutaneous manifestations (ECM), namely neurological, endocrine, skeletal abnormalities, or dysmorphisms.[4,12,13,18] Neurological involvement is the most common ECM.

Dysmorphic features, such as frontal bossing, hypertelorism, and a prominent premaxilla, may be attributed to the neuroectodermal origin of the osteocartilaginous structures of the face.[1,19]

Screening for neurological complications

Neurological and neurodevelopmental assessment is crucial, as children with CMN may present a broad spectrum of neurological abnormalities. In cases of multiple CMN, according to Dutch guidelines, annual neurological evaluation is recommended, at least until 5 years of age.[20]

Indications for brain and spinal cord magnetic resonance imaging (MRI) vary. Some authors argue that this examination is not essential,[1,7] whereas others recommend that it should be performed before 6 months of age in all children with two or more CMN.[5,7,21] However, the universal implementation of this approach is limited by disparities in available resources across healthcare systems.[1,5] Some authors recommend MRI only in high-risk patients, including: giant CMN; > 20 satellite lesions at birth; multiple medium-sized CMN; and large CMN located in the posterior axial region.[1,15,22,23]

The detection of congenital CNS abnormalities on MRI is associated with an increased risk of neurological complications and melanoma, representing a statistically more significant prognostic predictor than the cutaneous phenotype.[1,5-7,21]

The most frequently identified imaging findings include neural melanosis, with or without leptomeningeal involvement, CNS malformations, primary CNS tumors, arachnoid cysts, and hydrocephalus.[1,5] Both symptomatic and asymptomatic children with imaging abnormalities should be followed in pediatric neurology and/or neurosurgery.

Neurocutaneous melanosis

NCM is a rare congenital neuroectodermal dysplasia characterized by the presence of large, giant, and/or multiple CMN associated with melanocytic deposits in the CNS, such as the brain parenchyma or the leptomeninges.[3,6,11,24-26]

The estimated incidence in children with large CMN ranges from 2.5% to 45%.[6]

Multiple CMN constitute the main risk factor for NCM.[6,10] Other risk factors include male sex, PAS > 20 cm, CMN located on the head, neck, or posterior midline, and ≥ 10 satellite lesions.[2,6,7,10,18,27-31] Familial cases have also been described.[3,24,32]

Isolated intraparenchymal melanosis is the most frequently observed abnormality on MRI, with melanin foci predominantly located in the amygdala, thalamus, cerebellum, and pons.[1,3,5,6,10,23,33] In asymptomatic cases, MRI repetition is not indicated; however, neurodevelopment should be assessed annually until school age. Neurodevelopmental disorders, focal seizures, and epilepsy are the most common clinical manifestations.[4,6,21,34] Epilepsy is strongly associated with melanosis in the temporal lobe, particularly at the level of the amygdala, and in some cases may be refractory to treatment.[34]

Leptomeningeal involvement may include various primary melanocytic lesions, the imaging distinction of which may be challenging.[2,35] Melanocytic proliferation may be benign (meningeal melanocytosis or melanocytoma) or malignant (melanomatous involvement or meningeal melanoma), with either a nodular growth pattern (melanoma and melanocytoma) or a diffuse pattern (melanomatosis or melanocytosis).[2,28,18,33,36-39] Leptomeningeal melanocytosis consists of a localized or diffuse proliferation of melanocytes with benign histological features within the leptomeninges, particularly in the mesencephalon, pons, and spinal cord.[10,22,25,36] When associated with neurological symptoms, it carries a poor prognosis, even in the absence of malignant progression.[1,19,22,25,39]

Although rare, numerous CNS and spinal cord abnormalities associated with NCM have been described in the literature, including: Dandy-Walker malformation, Arnold-Chiari malformation type I, lissencephaly, agenesis of the corpus callosum, primary CNS tumors (cerebellar astrocytoma, choroid plexus papilloma, and ependymoma), arachnoid cysts, tethered spinal cord, syringomyelia, spinal dysraphism, among others.[1,2,10,11,22,18,38,40-42]

Clinical presentation and diagnosis

Approximately two-thirds of patients with NCM have a giant nevus, whereas the remaining present multiple small or medium-sized nevi without an associated larger lesion.[24,27,32,35,38]

Most patients with NCM are asymptomatic.[1,6,15,16,18] However, melanocytic proliferation within the CNS may lead to a wide range of complications, including compression of vital structures and/or obstruction of

cerebrospinal fluid (CSF) drainage, resulting in hydrocephalus manifested by increased intracranial pressure.[7,16,22,24-26] In this context, children may present a broad spectrum of neurological symptoms depending on the location of melanosis or tumor, its size and extent of invasion, and the presence of mass effect.[24,34,43]

Neurological symptoms typically begin before 2 years and include signs and symptoms of intracranial hypertension in two-thirds of cases, neurodevelopmental delay, cranial and/or spinal nerve palsies, seizures, and/or focal neurological deficits.[7,16,19,22,31,32,43] In adolescents, clinical presentation tends to be less severe, with symptoms such as headaches and neuropsychiatric manifestations, including psychosis.[1,6,31,35] Spinal leptomeningeal involvement is observed in approximately 20% of cases, particularly in patients whose symptoms begin between the second and third decades of life.[33,34] Spinal cord compression may result in myelopathy, radiculopathy, and bowel or bladder dysfunction.[7,18,22,32,37]

Diagnosis requires evidence of meningeal or CNS melanosis or melanoma, confirmed by MRI and/or histology, in the presence of large and/or multiple CMN (≥ 3).[33,36] In the presence of CM, meningeal lesions must be histologically benign, and likewise, cutaneous lesions must be benign in cases of meningeal melanoma.[18,22,24,25,31,32,38]

Although definitive diagnosis is histological, gadolinium-enhanced MRI is essential, allowing identification and characterization of intraparenchymal melanosis and/or leptomeningeal melanosis, the latter evidenced by leptomeningeal enhancement within the CNS.[16,21,24] It also enables detection of other intracranial and intraspinal abnormalities, aiding in the assessment of the feasibility of neurosurgical interventions aimed at symptomatic relief.[15,22,27] Nevertheless, a normal MRI does not exclude the presence of histological abnormalities and therefore does not rule out the diagnosis.[31]

The presence of melanocytes in the CSF suggests NCM. The presence of malignant tumor cells may indicate leptomeningeal melanoma or leptomeningeal metastases.[22]

Prognosis and treatment

The presence of neurological symptoms is a poor prognostic predictor in patients with NCM, even in the absence of malignancy.[10,24]

Isolated intraparenchymal melanosis is associated with a benign course, except in cases with leptomeningeal involvement or malignant melanoma.[2,20] The presence of hydrocephalus is associated with accelerated disease progression and an unfavorable prognosis.[43]

Treatment of NCM is primarily symptomatic.[16,31] Ventriculoperitoneal shunting may be performed for the management of symptomatic hydrocephalus.[2,20,22,32]

Although antiepileptic therapy allows control of seizure activity, temporal lobectomy and hippocampectomy may be required in cases of refractory epilepsy.[34,38]

Neurosurgical intervention for lesion resection and subsequent symptomatic relief may be considered, also allowing for tissue biopsy.[16,22,24,38]

Melanoma on CMN

Melanoma on CMN is a rare entity, accounting for approximately 0.9% of all pediatric neoplasms.[44] It shows a bimodal distribution, with around 70% of cases occurring in childhood and the remainder in adulthood.[44]

In children, approximately 70% of melanomas on CMN are diagnosed before puberty, and up to 50% before the age of 5 years.[17,42]

Most cases are diagnosed at locally advanced stages,[17] and the mortality rate is high.[4,7,17,45]

In giant CMN, particularly when associated with multiple CMN, the lifetime risk of melanoma may reach 10-15%.[4,7]

Melanoma on CMN may be cutaneous or extracutaneous,[15,46] with cutaneous being the most common form. ECM most frequently manifests in areas of neural melanosis within the CNS, presenting as leptomeningeal melanoma or melanoma within the brain parenchyma.[6,13,15,42]

Cases of melanoma have also been described in other locations, including lymph nodes, latissimus dorsi muscle, vagina, retroperitoneum, bone marrow, and the gastrointestinal mucosa.[4,6,15,42,47,48] In approximately 24% of melanoma cases, metastases are already present at diagnosis, and the primary origin cannot be identified.[1,5,7,42,48]

CM typically presents as a *de novo*, rapidly growing nodule or mass arising in the deep dermis or subcutaneous tissue. It is more easily detected by palpation than by visual inspection, as the surface of these nevi is often rough or nodular, making changes difficult to observe.[10,15,42,44,49] Breslow thickness is usually high at presentation.[4,17] The deeper location facilitates early dissemination through lymphatic and blood vessels, leading to early metastasis.[1]

In contrast, CM on smaller CMN frequently occurs at the periphery of the lesion and exhibits features similar to sporadic melanomas, namely an origin at the dermoepidermal junction.[1,29,49,50] Changes in the appearance of the nevus, such as the development of hypo- or hyperpigmented areas, alterations in lesion shape, and the presence of ulcerated, necrotic, or hemorrhagic areas, are indicators of malignancy.[1,7,16,17] Unlike CM on large or giant CMN, dermoscopic evaluation may be useful in these cases.[4,17]

Clinical suspicion of CM should prompt urgent excisional biopsy, with histopathological evaluation by two specialists.[4,10,40,44] Genetic analysis is an important adjunct to clinical, radiological, and histopathological assessment.[4,6,44]

CNS melanoma

Primary CNS melanoma accounts for approximately one-third of melanomas occurring in patients with CMN.[1,4] It may present as a solid tumor within the brain parenchyma or, more frequently, as leptomeningeal melanoma.[4]

All patients with neurological symptoms or changes in baseline neurological status, regardless of age or previous MRI findings, should undergo contrast-enhanced brain and spinal cord MRI, given the high risk of CNS melanoma.[5-7] According to some authors, if MRI findings are normal or unchanged compared to screening MRI performed within the first 6 months of life, regular neurological follow-up should be maintained, and in the event of clinical progression, the threshold for repeating MRI should be low.[4]

Conversely, the detection of new leptomeningeal involvement or unexplained hydrocephalus should prompt repeat MRI after 2 weeks. In cases of radiological progression or evidence of a solid intraparenchymal tumor, lesion biopsy should be performed, with histopathological analysis and genotyping for NRAS and BRAF mutations.[4,21]

In all cases of leptomeningeal disease, repeated MRI is recommended until clinical and radiological stabilization is achieved, due to the high risk of progression to malignancy.[4]

Currently, there are no specific therapeutic guidelines for melanoma, either cutaneous or CNS, in patients with CMN.[4]

Malignant and benign tumors

The origin of giant CMN in the NC confers a predisposition to a variety of neoplastic transformations, including melanocytic tumors, as well as mesenchymal and neural tumors, with an associated risk of malignant transformation,[51] such as rhabdomyosarcoma, liposarcoma, and undifferentiated neural tumors.[4,11,13,29,51-53]

CMN may also be associated with benign tumors, including hemangiomas, lipomas, schwannomas, ganglioneuroma, and PN.[1,52]

Management of CMN

The management of CMN should be individualized and multidisciplinary, with regular dermatological, neurological, and psychosocial monitoring.[7]

Referral to pediatric dermatology should be recommended in cases of multiple, large, or giant CMN, particularly when associated with multiple CMN, when located on the midline, in CMN with atypical presentation or associated complications, esthetic implications, and the presence of neurological deficits or other comorbidities.[5,11]

Some authors recommend that, in the aforementioned situations, follow-up visits should be conducted every 3 months during the 1st year of life, followed by annual follow-up.[5] Assessment during puberty is also relevant, considering the greater potential for nevus changes during this stage of development.[5,49]

Caregivers should regularly inspect and palpate the nevus. They may use photographic documentation at different time points to monitor its evolution.[5] The detection of warning signs, such as rapid growth, bleeding, pain, ulceration, or the development of nodules, should be promptly evaluated.[5]

Cutaneous complications result from impairment of the skin barrier and include skin fragility, hypertrichosis, pruritus, and dysfunction of eccrine and sebaceous glands.[5]

Regarding bathing, some authors recommend a frequency of 2-3 times/week, using only water or a pH-neutral cleanser, followed by the application of an emollient cream.[5]

In cases of hypohidrosis, anhidrosis, and xerosis, resulting from functional impairment of eccrine and sebaceous glands, the main recommendation is to avoid overheating.[5,42,46]

Regarding ultraviolet radiation protection, children with CMN should follow general pediatric sun protection recommendations.[5]

From a psychosocial perspective, increasing awareness in the field of mental health has influenced the management of patients with CMN, particularly when large or located in exposed areas, which impacts

physical appearance and self-image.[10,42] Low self-esteem, anxiety, and depression are common comorbidities, often exacerbated by social isolation.[10] Children frequently experience difficulties in forming emotional bonds, resulting in excessive dependence on parental protection.[6]

In addition to providing clear information and addressing all concerns, it is essential that the child and their family have access to a psychological assessment.

Treatment

The main objectives of CMN treatment are to reduce the risk of CM and to minimize esthetic implications.[6,44,54,55]

Partial or full-thickness excisions, laser therapy, curettage, dermabrasion, chemical peels, and cryotherapy are treatment options described in the literature.[56] Surgery and laser therapy, either alone or in combination, are the most commonly used therapeutic approaches.[6]

The choice between a surgical or conservative approach should be individualized.[5,6,11,15,16] Currently, conservative management strategies, focusing on quality of life and long-term esthetic improvement, are prioritized.[6,12,57]

Prophylactic excision of CMN does not reduce the risk of CM.[5,45] The persistence of residual nevus cells after removal, particularly in larger lesions, makes complete excision unfeasible.[5,10,42,44,55,58,59]

Infection and wound-healing disturbances are possible post-operative complications.[5,10,12,55,57] The risk of undesirable esthetic scarring and repigmentation should be discussed, considering the possibility of gradual lightening of the nevus over the first decade of life, particularly in fair-skinned patients.[15,49]

Furthermore, the benefits of surgery during the 1st year of life, due to greater tissue elasticity, should be weighed against the risks associated with general anesthesia, which may have deleterious effects on the developing brain.[10]

Suspicion of malignancy constitutes an absolute indication for surgical excision.[12]

Functional impairment, stigma, and the presence of lesions or chronic pruritus refractory to medical treatment, particularly when adequate clinical follow-up cannot be ensured, constitute relative indications.[12,42]

In patients with symptomatic NCM, CNS melanoma, or in cases where surgery carries a risk of functional loss or results in more significant scarring than the nevus itself, surgical intervention is not indicated.[12,15,18,38]

Laser therapy aims to improve esthetic outcomes in cosmetically sensitive areas and is indicated for smaller CMN.[1,6,60,61]

Conclusion

The presence of a CMN, especially when large or located in exposed areas, is associated with physical, psychological, functional, and esthetic consequences, with a significant impact on quality of life.

The relationship between nevus size and the risk of severe complications, including malignant melanoma and NCM, is well established.

It is essential to inform patients and caregivers about the risks and benefits of each treatment, thereby promoting shared decision-making, based on the best available scientific evidence and the individual characteristics of each child, to minimize suffering and social stigma.

A limitation of this study is that only review articles, narrative reviews, and case series were included. This limitation is justified by the rarity of the described complications. Future studies with higher levels of evidence and greater methodological robustness are needed, including children with CMN from birth, to enable the development of universally applicable clinical guidelines.

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Conflicts of interest

None.

Ethical considerations

Protection of human and animal subjects. The authors declare that no experiments involving human or animal subjects were conducted for this study.

Confidentiality, informed consent, and ethical approval. The authors followed the confidentiality protocols of their institution, obtained informed consent from the patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

References

1. Farabi B, Akay BN, Goldust M, Wollina U, Atak MF, Rao B. Congenital melanocytic naevi: an up-to-date overview. Australas J Dermatol. 2021;62:e178-91.
2. Kim SJ, Kim JH, Son B, Yoo C. A giant congenital melanocytic nevus associated with neurocutaneous melanosis. Clin Neuroradiol. 2014;24(2):177-84.
3. Varela-Poblete J, Vidal-Tellez A, Cruz-Quiroga JP, Montoya-Salvadores F, Medina-Escobar J. Melanocytic lesions of the central nervous system: a case series. Arq Neuropsiquiatr. 2022;80:153-60.
4. Kinsler VA, O'Hare P, Bulstrode N, Calonje JE, Chong WK, Hargrave D, et al. Melanoma in congenital melanocytic naevi. Br J Dermatol. 2017;176:1131-43.
5. Jahnke MN, O'Haver J, Gupta D, Hawryluk EB, Finelt N, Kruse L, et al. Care of congenital melanocytic nevi in newborns and infants: review and management recommendations. Pediatrics. 2021;148:e2021051536.
6. Tan S, Hu H, Xin X, Wu D. A clinical and biologic review of congenital melanocytic nevi. J Dermatol. 2024;51:12-22.
7. Moustafa D, Blundell AR, Hawryluk EB. Congenital melanocytic nevi. Curr Opin Pediatr. 2020;32:491-7.
8. Fledderus AC, Franke CJJ, Eggen CA, Van Etten-Jamaludin FS, Van der Horst CM, Brinkmann SJ, et al. Outcomes and measurement instruments used in congenital melanocytic naevi research: a systematic review. J Plast Reconstr Aesthet Surg. 2020;73:703-15.
9. Krengel S, Scope A, Dusza SW, Vonthein R, Marghoob AA. New recommendations for the categorization of cutaneous features of congenital melanocytic nevi. J Am Acad Dermatol. 2013;68:441-51.
10. Schaffer JV. Update on melanocytic nevi in children. Clin Dermatol. 2015;33:368-86.
11. Warner C, Dinulos JG. Core concepts in congenital melanocytic nevi and infantile hemangiomas. Curr Opin Pediatr. 2014;26:130-5.
12. Ott H, Krengel S, Beck O, Böhler K, Böttcher-Haberzeth S, Cangir Ö, et al. Multidisciplinary long-term care and modern surgical treatment of congenital melanocytic nevi: recommendations by the CMN surgery network. J Dtsch Dermatol Ges. 2019;17:1005-16.
13. Yang C, Gru AA, Dehner LP. Common and not so common melanocytic lesions in children and adolescents. Pediatr Dev Pathol. 2018;21:252-70.
14. Zayour M, Lazova R. Congenital melanocytic nevi. Clin Lab Med. 2011;31:267-80.
15. Krengel S, Marghoob AA. Current management approaches for congenital melanocytic nevi. Dermatol Clin. 2012;30:377-87.
16. Lyon VB. Congenital melanocytic nevi. Pediatr Clin North Am. 2010;57:1155-76.
17. Merkel EA, Mohan LS, Shi K, Panah E, Zhang B, Gerami P. Paediatric melanoma: clinical update, genetic basis, and advances in diagnosis. Lancet Child Adolesc Health. 2019;3:646-54.
18. Islam MP. Neurocutaneous melanosis. Handb Clin Neurol. 2015;132: 111-7.
19. Van Engen-van Grunsven AC, Kusters-Vandevelde HV, Groenen PJ, Blokx WA. Update on molecular pathology of cutaneous melanocytic lesions: what is new in diagnosis and molecular testing for treatment? Front Med (Lausanne). 2014;1:39.
20. Fledderus AC, Widdershoven AL, Lapid O, Breugem CC, Pasmans SG, Van der Horst CM, et al. Neurological signs, symptoms and MRI abnormalities in patients with congenital melanocytic naevi and evaluation of routine MRI-screening: systematic review and meta-analysis. Orphanet J Rare Dis. 2022;17:95.
21. Di Stasi M, Mankad K, Carney O, Loebel U, Biswas A, Sudhakar S, et al. Congenital melanocytic naevus syndrome and Dandy-Walker malformation: a mistaken association: case report and literature review. Neuroradiology. 2023;65:1077-86.
22. Rahman RK, Majmundar N, Ghani H, San A, Koirala M, Gajjar AA, et al. Neurosurgical management of patients with neurocutaneous melanosis: a systematic review. Neurosurg Focus. 2022;52:E8.
23. Markovic I, Milenkovic Z, Jovanović M, Jakubi BJ, Sekulovic-Stefanovic L. Early diagnosis of asymptomatic neurocutaneous melanosis: 60-month follow-up. Pediatr Int. 2016;58:403-5.
24. Liu BC, Wang YB, Liu Z, Jiao Y, Zhang XF. Neurocutaneous melanosis with an intracranial cystic-solid meningeal melanoma in an adult: a case report and review of literature. World J Clin Cases. 2022;10:5025-35.
25. Küsters-Vandevelde HV, Willemsen AE, Groenen PJ, Küsters B, Lammens M, Wesseling P, et al. Experimental treatment of NRAS-mutated neurocutaneous melanocytosis with MEK162, a MEK inhibitor. Acta Neuropathol Commun. 2014;2:41.
26. Shah KN. The risk of melanoma and neurocutaneous melanosis associated with congenital melanocytic nevi. Semin Cutan Med Surg. 2010;29:159-64.
27. Levy R, Lara-Corrales I. Melanocytic nevi in children: a review. Pediatr Ann. 2016;45:e293-8.
28. Scattolin MA, Lin J, Peruchi MM, Rocha AJ, Masruha MR, Vilanova LC. Neurocutaneous melanosis: follow-up and literature review. J Neuroradiol. 2011;38:313-8.
29. Willard KJ, Cappel MA, Kozin SH, Abzug JM. Congenital and infantile benign skin lesions affecting the hand and upper extremity, part 2: nonvascular neoplasms. J Hand Surg Am. 2013;38:2284-92.
30. Chien JC, Niu DM, Wang MS, Liu MT, Lirng JF, Chen SJ, *et al*. Giant congenital melanocytic nevi in neonates: report of two cases. Pediatr Neonatol. 2010;51:61-4.
31. Vanood A, Lee YA, Leleszi E, Krishnan A. Symptomatic neurocutaneous melanosis: mild clinical onset in a teenager. BMJ Case Rep. 2020;13:e235772.
32. Sharouf F, Zaben M, Lammie A, Leach P, Bhatti MI. Neurocutaneous melanosis presenting with hydrocephalus and malignant transformation: case-based update. Childs Nerv Syst. 2018;34:1471-7.
33. Samara A, Gusman M, Aker L, Parsons MS, Mian AY, Eldaya RW. The forgotten phacomatoses: a neuroimaging review of rare neurocutaneous disorders. Curr Probl Diagn Radiol. 2022;51:747-58.
34. Pellino G, Gencarelli J, Bertelli S, Russo A, Fiumana E, Faggioli R. Epilepsy in isolated parenchymal neurocutaneous melanosis: a systematic review. Epilepsy Behav. 2020;107:107061.
35. Ma M, Ding ZL, Cheng ZQ, Wu G, Tang XY, Deng P, et al. Neurocutaneous melanosis in an adult patient with intracranial primary malignant melanoma: case report and review of the literature. World Neurosurg. 2018;114:76-83.
36. Matsumura M, Okudela K, Tateishi Y, Umeda S, Mitsui H, Suzuki T, et al. Leptomeningeal melanomatosis associated with neurocutaneous melanosis: an autopsy case report. Pathol Int. 2015;65:100-5.
37. Ruggieri M, Praticò AD. Mosaic neurocutaneous disorders and their causes. Semin Pediatr Neurol. 2015;22:207-33.
38. Flores-Sarnat L. Neurocutaneous melanocytosis. Handb Clin Neurol. 2013;111:369-88.
39. Küsters-Vandevelde HV, Küsters B, Van Engen-van Grunsven AC, Groenen PJ, Wesseling P, Blokx WA. Primary melanocytic tumors of the central nervous system: a review with focus on molecular aspects. Brain Pathol. 2015;25:209-26.
40. Mumtaz Hashmi H, Shamim N, Kumar V, Idrees S. Giant congenital melanocytic nevi in a Pakistani newborn. Cureus. 2021;13:e15210.
41. Dua R, Jain R, Momin M. Giant congenital melanocytic nevus with choroid plexus papilloma: case report and review of literature. J Neurol Surg A Cent Eur Neurosurg. 2012;73:337-40.
42. Viana AC, Gontijo B, Bittencourt FV. Nevo melanocítico congênito gigante. An Bras Dermatol. 2013;88:863-78.
43. Bianchi F, Tamburrini G, Colosimo C, Massimi L, Frassanito P. Neurocutaneous melanosis in infancy: always a dismal prognosis? Turk Neurosurg. 2020;30:476-82.
44. Belysheva TS, Vishnevskaya YV, Nasedkina TV, Emelyanova MA, Abramov IS, Orlova KV, et al. Melanoma arising in a giant congenital melanocytic nevus: two case reports. Diagn Pathol. 2019;14:21.
45. Tan S, Hu H, Li G, Zhao J, Wu D. Prevalence, incidence density and standardized morbidity rate of melanoma among patients with congenital melanocytic naevi: a systematic review. Clin Exp Dermatol. 2024;49: 765-73.
46. Slutsky JB, Barr JM, Femia AN, Marghoob AA. Large congenital melanocytic nevi: associated risks and management considerations. Semin Cutan Med Surg. 2010;29:79-84.
47. Vourc'h-Jourdain M, Martin L, Barbarot S. Large congenital melanocytic nevi: therapeutic management and melanoma risk: a systematic review. J Am Acad Dermatol. 2013;68:493-8.e14.
48. Onak Kandemir N, Bahadir B, Bektać S, Barut F, Yurdakan G, Doćan Gün B, et al. Malignant melanoma associated with congenital melanocytic nevus and diagnosed with intestinal metastases: two case reports. Turk J Gastroenterol. 2011;22:77-82.
49. Cotton CH, Goldberg GN. Evolution of congenital melanocytic nevi toward benignity: a case series. Pediatr Dermatol. 2019;36:227-31.
50. Scard C, Aubert H, Wargny M, Martin L, Barbarot S. Risk of melanoma in congenital melanocytic nevi of all sizes: a systematic review. J Eur Acad Dermatol Venereol. 2023;37:32-9.
51. Snyder JA, Khozeimeh N, Reyes C, Qureshi FG. Benign sacrococcygeal neoplasm arising at birth from a giant congenital melanocytic nevus. J Pediatr Surg. 2012;47:408-11.
52. Christman MP, Kerner JK, Cheng C, Piris A, Nepo AG, Sepehr A, et al. Rhabdomyosarcoma arising in a giant congenital melanocytic nevus. Pediatr Dermatol. 2014;31:584-7.
53. Chikhalkar S, Gutte R, Holmukhe S, Khopkar U, Desai S, Gupta S. Alveolar rhabdomyosarcoma arising in a giant congenital melanocytic nevus in an adult: case report with review of literature. Int J Dermatol. 2013;52:1372-75.

54. Sun J, Du F, Huang J, Long X. Staged excision of a cheek congenital melanocytic nevus: a case report and literature review. J Cosmet Dermatol. 2022;21:4605-8.
55. Ibrahimi OA, Alikhan A, Eisen DB. Congenital melanocytic nevi: where are we now? Part II. Treatment options and approach to treatment. J Am Acad Dermatol. 2012;67:515.e1-13.
56. ALMutairi HM, Al-hothali GI. The outcome of using different surgical modalities and laser therapy in the treatment of small- and medium-sized congenital melanocytic nevi: a systematic review. Int J Dermatol. 2020;59:535-42.
57. Gout HA, Fledderus AC, Lokhorst MM, Pasmans SG, Breugem CC, Lapid O, et al. Safety and effectiveness of surgical excision of medium, large, and giant congenital melanocytic nevi: a systematic review and meta-analysis. J Plast Reconstr Aesthet Surg. 2023;77:430-55.
58. Drakensjö IT, Hedblad MA, Wahlgren CF. Rapidly involuting congenital melanocytic naevi in two children. Acta Derm Venereol. 2012;92:605-6.
59. Funayama E, Sasaki S, Furukawa H, Hayashi T, Yamao T, Takahashi K, et al. Effectiveness of combined pulsed dye and Q-switched ruby laser treatment for large to giant congenital melanocytic naevi. Br J Dermatol. 2012;167:1085-91.
60. Zeng Y, Zheng YQ. Successful treatment of congenital melanocytic nevus on tragus with COć laser. J Dermatolog Treat. 2014;25:287-9.
61. Camargo CP, Saliba M, Saad EA, Milan M, Caldera JM. Treatments of palpebral congenital melanocytic nevus: a systematic review. Acta Cir Bras. 2023;38:e384823.