CASE REPORT

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Neurocutaneous melanosis with giant congenital melanocytic nevus: a case report

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ABSTRACT

Background: Congenital melanocytic nevus syndrome is a rare neurocutaneous disorder characterized by congenital melanocytic nevus with extracutaneous involvement, most commonly neurocutaneous melanosis (NCM). Symptomatic NCM usually presents early in life with seizures, hydrocephalus, or developmental delay, and carries a poor prognosis. This study aimed to present a unique case of NCM in a child with giant congenital melanocytic nevus accompanied by a Dandy–Walker variant and unusual multisystem involvement.

Case presentation: We report a 5-year-old girl with a giant congenital melanocytic nevus who presented with status epilepticus. Brain magnetic resonance imaging (MRI) revealed hyperintense lesions in both amygdalae and the left pons consistent with NCM, accompanied by a Dandy–Walker variant. Skin biopsy confirmed congenital melanocytic nevi (CMN). Systemic evaluation demonstrated pancytopenia, chronic kidney disease stage V, and hypoparathyroidism with vitamin D deficiency. The patient was managed with intravenous and oral antiepileptic drugs, electrolyte correction, packed red cell transfusion, vitamin D supplementation, and multidisciplinary monitoring.

Conclusion: This case illustrates a rare presentation of neuricur with seizures at the age of five, multifocal NCM, and multisystem involvement. Prognosis remains poor in multifocal disease where surgical intervention is not feasible, emphasizing the importance of optimal medical therapy, systemic correction, and long-term multidisciplinary management to improve quality of life.

Keywords: congenital melanocytic nevus, Dandy–Walker variant, neurocutaneous melanosis, pediatric neurology, status epilepticus.

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INTRODUCTION

Congenital melanocytic nevus syndrome is a rare neurocutaneous disorder defined by the presence of congenital melanocytic nevi (CMN) associated with extracutaneous involvement, most commonly neurocutaneous melanosis (NCM).[1,2] Congenital melanocytic nevi are pigmented lesions present at birth, arising from nevomelanocytes of neural crest origin. Based on projected adult size, they are classified into small (<1.5 cm), medium (1.5–20 cm), and large/giant (>20 cm).[3] The incidence of congenital melanocytic nevi is approximately 1% of newborns, while giant CMN occurs in only 1:20,000 to 1:500,000 live births.[4] NCM is even rarer, reported in 2–3% of patients with large or giant CMN.[5]

Neurocutaneous melanosis is characterized by abnormal proliferation of melanocytes in the skin and central nervous system. Its pathogenesis is thought to be related to disturbances in neural crest development during early embryogenesis. Normally, melanocyte precursor cells migrate from the neural crest to the skin, leptomeninges, and other structures. However, in NCM, dysregulated proliferation or differentiation of melanocytes occurs during this migration, leading to abnormal melanocytic accumulation in the skin as congenital melanocytic nevi and in the leptomeninges or brain parenchyma. Melanocytic infiltration of the leptomeninges may disrupt cerebrospinal fluid circulation, causing hydrocephalus and, in some cases, malignant transformation into melanoma. Genetic factors, such as somatic mutations in neuroblastoma RAS viral oncogene homolog (NRAS) or B-Raf proto-oncogen (BRAF), have also been reported to trigger abnormal melanocytic proliferation, explaining the link between cutaneous and neurological manifestations.[6] This mechanism indicates that NCM is not merely a cutaneous disorder, but rather a systemic condition originating from aberrant neural crest development. Clinical manifestations of NCM typically appear in infancy or early childhood, usually before the age of two years, and include seizures, hydrocephalus, signs of raised intracranial pressure, and developmental delay.[7,8] The prognosis is generally poor once neurological

symptoms develop, with many patients deteriorating within two to three years due to intractable epilepsy, hydrocephalus, or malignant transformation.[9]

Diagnostic criteria for NCM were first proposed by Fox in 1972, requiring the presence of a large or multiple CMN with meningeal melanosis or melanoma, in the absence of cutaneous melanoma or other primary CNS melanoma.[10] Advances in MRI have improved early detection, particularly when T1-weighted hyperintensities are identified in the amygdala, hippocampus, or brainstem, which are known epileptogenic regions.[11] Additional structural malformations, such as the Dandy–Walker complex, may coexist and further worsen neurological outcomes.[12]

Although congenital melanocytic nevus syndrome is primarily considered a cutaneous and neurological disorder, rare reports describe associations with hematologic, renal, endocrine, and musculoskeletal abnormalities, suggesting a broader systemic spectrum linked to neural crest-derived tissues.[13] This case is unique because status epilepticus was the initial neurological presentation of NCM in a child with giant congenital melanocytic nevus, accompanied by a Dandy–Walker variant and unusual multisystem involvement, expanding the known clinical spectrum of this rare syndrome.

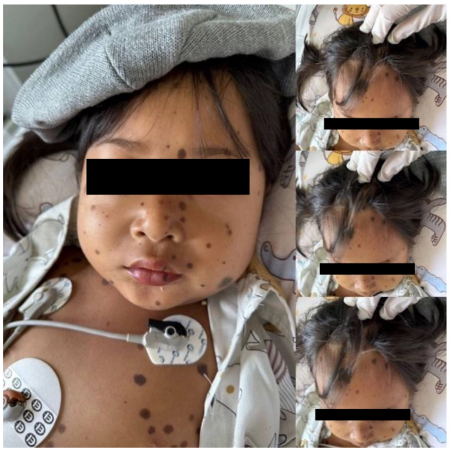


Figure 1. The skin lesions in patient with neurocutaneous melanocytosis (NCM) and congenital melanocytic nevus (CMN)

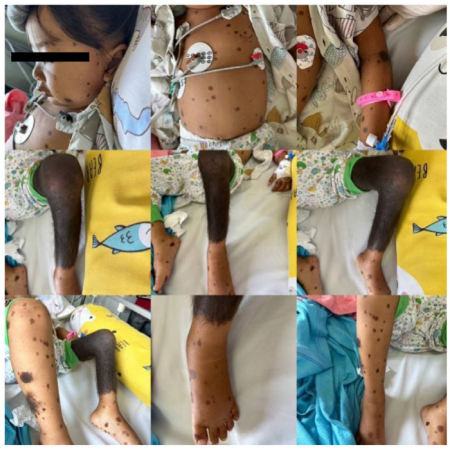


Figure 2. Nevus was scattered discretely or in groups, and some with fine hair on them.

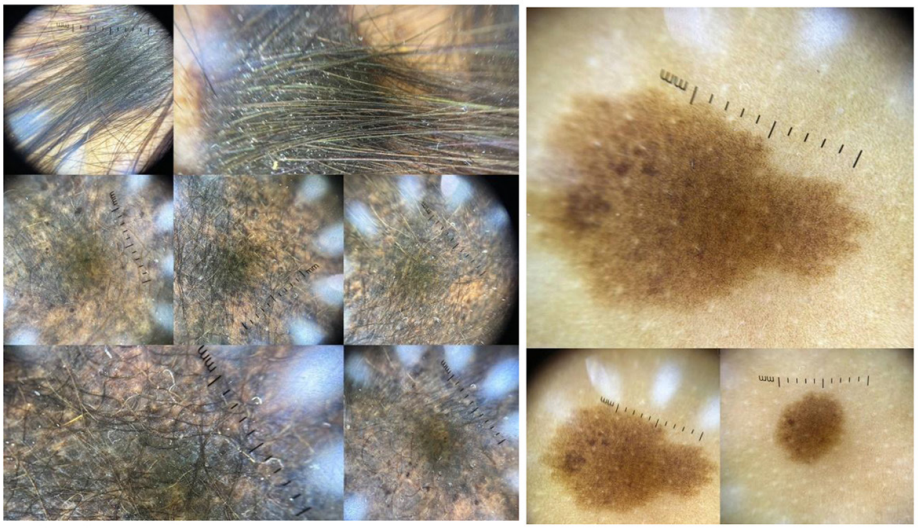


Figure 3. The Dermoscopic Findings

CASE REPORT

A 5-year-old girl was referred to the Emergency Department of Prof. Dr. I. G. N. G Ngoerah Hospital due to a prolonged seizure. The event was characterized by focal tonic–clonic movements involving the left upper and lower extremities with associated upward eye deviation. The seizure lasted for approximately 30 minutes and was accompanied by loss of consciousness, consistent with status epilepticus. Upon arrival, the patient was unresponsive to verbal and painful stimuli. She had received diazepam suppository and intravenous phenobarbital prior to referral, with partial improvement. Stabilization was continued in the ED, after which she was admitted to the pediatric high care unit for further monitoring and management.

The child is the third of four siblings, all born to healthy, non-consanguineous parents. Both older and younger siblings are healthy with no similar medical conditions reported. She was delivered by cesarean section at term with a birth weight of 3200 gram and a length of 49 cm. No history of birth asphyxia or perinatal complications was reported. She had achieved normal developmental milestones until the onset of illness. Immunizations were up to date according to the national schedule. There was no family history of similar skin lesions, seizures, or neurological disorders.

Since birth, the patient had extensive blackish-brown pigmented skin lesions involving the trunk, extremities, and scalp, which progressively increased in number and extent with age. Atrophy of the left foot has also been noted since infancy. There was no history of trauma, bleeding, or infection associated with the lesions.

On examination, the patient appeared pale but was conscious and interactive after stabilization of the seizure. Anthropometric measurements showed a weight of 13 kg and height of 99 cm, corresponding to mild malnutrition. Head circumference was 49 cm, within normal limits for age. Vital signs were stable. Neurological examination revealed normal muscle tone and no focal deficits after seizure resolution. Fundoscopic examination showed no papilledema.

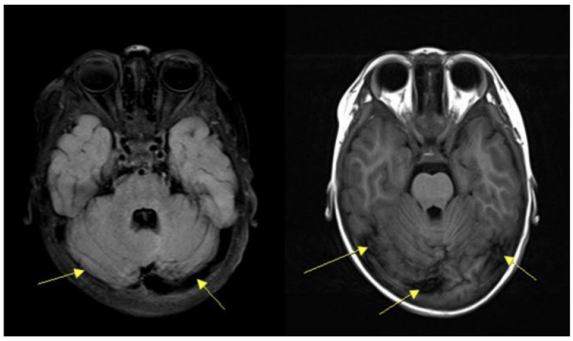
Dermatological examination revealed multiple, well-demarcated hyperpigmented macules and patches ranging in size from 0.3 × 0.8 cm to 22 × 10 cm, some associated with fine hair growth, diffusely distributed across the body. A large hyperpigmented plaque measuring approximately 22 × 10 cm extended from the left knee to the ankle, with overlying hypertrichosis, consistent with a giant CMN. Smaller satellite lesions were scattered over both dorsal feet and the trunk. No ulceration or signs of secondary infection were observed (**Figure 1** and **2**). Dermoscopic findings are shown in **Figure 3**.

Laboratory investigations revealed pancytopenia with hemoglobin 6.5 g/dL, leukocytes 3.37 × 10^3/µL, and platelets 108 × 10^3/µL. Peripheral blood smear showed normocytic normochromic anemia with leukopenia (lymphopenia) and thrombocytopenia, consistent with pancytopenia. Electrolyte profile showed hyponatremia (125 mmol/L) and severe hypocalcemia (4.4 mg/dL). Renal function tests demonstrated markedly elevated BUN (168.5 mg/dL) and serum creatinine (12.02 mg/dL), consistent with chronic kidney disease (CKD) stage V. Endocrine profile revealed a markedly suppressed intact parathyroid hormone (PTH) level (1.578 pg/mL, reference 9–59 pg/mL), confirming hypoparathyroidism, and vitamin D deficiency (8 ng/mL).

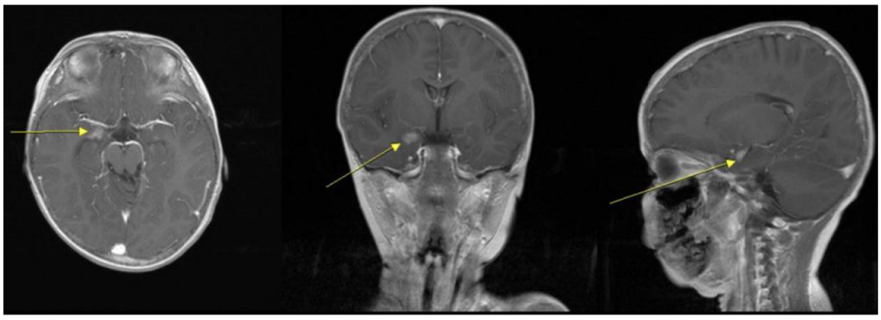
Neuroimaging with brain MRI revealed hyperintense T1 lesions in the bilateral amygdala and left pons, consistent with neurocutaneous melanosis (NCM). Additional findings included a Dandy–Walker variant and multiple soft-tissue nodules of the scalp involving connective tissue, aponeurosis, and periosteum (**Figure 4A**, **4B**, **5**, and **6**).

Histopathological examination of the skin biopsy demonstrated nests and cords of nevus cells extending into the deep dermis with infiltration around adnexal structures such as hair follicles, sebaceous glands, and arrector pili muscles. The nevus cells appeared small, round to oval, with uniform nuclei and finely pigmented cytoplasm. No nuclear atypia, mitotic activity, or necrosis was observed. These features were consistent with a congenital melanocytic nevus.

The patient was managed acutely with intravenous antiepileptic drugs (phenobarbital and phenytoin) and supportive therapy. Electrolyte correction was provided with intravenous calcium gluconate, and packed red cell transfusions were administered to address anemia. She was subsequently maintained on oral



4A



4B

Figure 4. Brain MRI findings of neurocutaneous melanosis. **(A)** Axial T1-weighted MRI shows hyperintense lesions in the left pons and cerebellum (arrow), consistent with parenchymal involvement of neurocutaneous melanosis. **(B)** Axial and coronal T1-weighted MRI demonstrates bilateral hyperintense lesions in the amygdala (arrows), a typical feature of neurocutaneous melanosis and a potential epileptogenic focus associated with recurrent seizures.

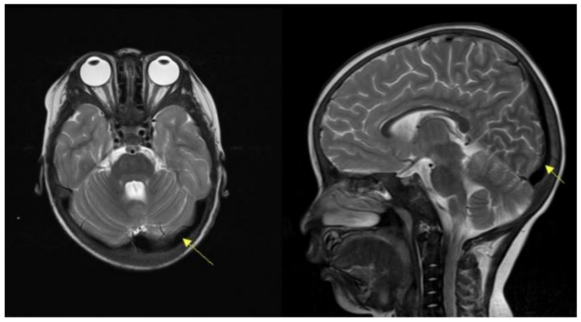


Figure 5. Sagittal T1-weighted MRI demonstrates a posterior fossa cyst with hypoplasia of the cerebellar vermis (arrow), consistent with a Dandy–Walker variant.

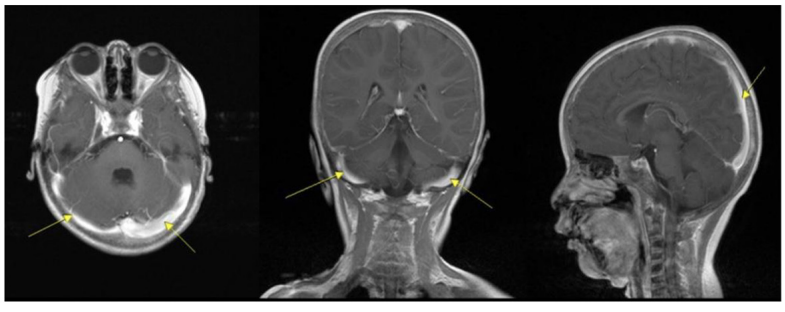


Figure 6. Sagittal post-contrast MRI reveals multiple soft tissue nodules in the scalp involving the right frontal, parietal, and left occipital regions (arrows), consistent with extracranial involvement of congenital melanocytic nevus.

phenytoin for seizure prophylaxis and vitamin D supplementation.

During hospitalization, seizures were successfully controlled with intravenous antiepileptic drugs and metabolic correction. The patient experienced one brief seizure during hemodialysis, which improved after acute therapy, and was subsequently discharged on oral phenytoin and vitamin D supplementation. In the post-discharge period, the patient remained seizure-free despite discontinuation of antiepileptic medication. However, chronic kidney disease stage V persisted, accompanied by hypoparathyroidism and severe vitamin D deficiency, which were managed with supplementation. On long-term follow-up, the patient continued regular hemodialysis twice weekly. She remains under multidisciplinary follow-up, focusing on seizure prevention, metabolic stabilization, and quality-of-life improvement.

This case highlights a rare presentation of neurocutaneous melanosis associated with a giant congenital melanocytic nevus and a Dandy–Walker variant, further complicated by multisystem involvement including chronic kidney disease, pancytopenia, and endocrine abnormalities. Such a constellation of findings underscores the broad clinical spectrum of congenital melanocytic nevus syndrome and the importance of multidisciplinary management for optimizing long-term outcomes.

DISCUSSION

Congenital melanocytic nevus syndrome is a rare condition characterized by the presence of multiple or large congenital melanocytic nevi with extracutaneous involvement, most notably neurocutaneous melanosis (NCM). Congenital melanocytic nevus occurs in approximately 1% of newborns, while giant congenital melanocytic nevus is much rarer, with an estimated incidence of only 1:20,000 to 1:500,000 live births.[4] In general, congenital melanocytic nevus is classified as small (<1.5 cm), medium (1.5–20 cm), and large/giant (>20 cm).[3] Compared to small or medium, giant CMN carries a higher risk of serious complications, including malignant transformation into melanoma and central nervous system involvement through NCM.[4,5] These lesions originate from neural crest melanoblasts, and abnormal migration of these cells into the central nervous system leads to NCM. Magnetic resonance imaging (MRI) is the primary diagnostic tool, with melanin typically appearing hyperintense on T1-weighted sequences, most often affecting the amygdala, temporal lobes, cerebellum, and brainstem.[6,7] Furthermore, congenital melanocytic nevus syndrome may be associated with other neurodevelopmental anomalies, such as Dandy–Walker malformation, characterized by cerebellar vermis hypoplasia, fourth ventricle dilatation, and enlargement of the posterior fossa.[8] The association between NCM and Dandy–Walker malformation (DWM) is extremely rare, occurring in only about 10% of NCM patients. Most reported cases present during the neonatal or infantile period, with poor prognosis and high mortality. For instance, Lookzadeh et al. described a neonatal case with NCM and Dandy–Walker complex that rapidly progressed to severe neurological complications.[14] In this case, a 5-year-old girl presented with a giant congenital melanocytic nevus measuring 22 × 10 cm on the lower extremity with numerous satellite lesions across the body, fulfilling the criteria for giant CMN. MRI of the brain demonstrated T1 hyperintense lesions in both amygdalae and the left pons, consistent with NCM, accompanied by a Dandy–Walker variant. The coexistence of CMN, multifocal NCM, and structural CNS malformation is exceedingly rare and underscores the complexity of neurological involvement in this patient. Moreover, our case is unique because neurological symptoms manifested only at the age of five years in the form of status epilepticus, much later than the majority of reported cases.

The development of neurocutaneous melanosis (NCM) and Dandy–Walker variant (DWV) can be traced back to early embryogenesis. Melanocytes, which later contribute to the formation of congenital melanocytic nevi, originate from neural crest cells, whereas posterior fossa structures, including the cerebellar vermis and fourth ventricle, derive from the hindbrain during closely overlapping periods of organogenesis. Under normal conditions, these processes occur in a synchronized and coordinated manner. However, when disturbances such as somatic mutations in the NRAS or BRAF genes arise, melanocyte proliferation may become dysregulated while simultaneously disrupting signaling mechanisms essential for the development of posterior brain structures. This combined defect may lead to the accumulation of abnormal melanocytes in the leptomeninges or brain parenchyma (NCM), as well as morphological anomalies of the posterior fossa characteristic of DWV. Thus, the coexistence of NCM and DWV is not coincidental but rather reflects overlapping embryogenic abnormalities, explaining why these conditions may occur together in a small subset of patients.[2,7,8] In this case, sagittal T1-weighted MRI revealed a posterior fossa cyst with partial hypoplasia of the cerebellar vermis, consistent with a Dandy–Walker variant. Pathophysiologically, this structural malformation disrupts the normal development of the fourth ventricle and posterior fossa, resulting in cystic dilatation and impaired cerebrospinal fluid (CSF) circulation. On the other hand, neurocutaneous melanosis contributes through melanocytic infiltration of the leptomeninges in the posterior fossa. This infiltration thickens the leptomeninges, further aggravating CSF outflow obstruction and interfering with local neural tissue development. These dual mechanisms CSF circulation impairment due to DWV and meningeal burden from melanocytic infiltration in NCM act synergistically to increase the risk of hydrocephalus, elevated intracranial pressure, and cortical hyperexcitability. Such a pathophysiological process may account for the late-onset seizures observed in our patient, particularly as the amygdala and brainstem, which are commonly involved in NCM, are highly susceptible to becoming secondary epileptogenic foci.

Histopathological evaluation is essential to differentiate CMN from melanoma. CMN is characterized by nests of nevus cells extending into the dermis and subcutaneous tissue, often involving

adnexal structures, but lacking atypia or mitotic activity.[15,16] In this case, skin biopsy confirmed a diagnosis of benign congenital melanocytic nevus without evidence of malignancy, supporting the interpretation that neurological manifestations were due to benign melanocytic infiltration of the CNS rather than malignant transformation.

Seizures are the most common neurological manifestation of NCM, reported in 50–70% of symptomatic patients.[7,9] Amygdala involvement is highly epileptogenic and strongly associated with recurrent or refractory seizures.[6] Structural abnormalities such as the Dandy–Walker variant may further exacerbate epileptogenesis by disrupting cerebrospinal fluid dynamics and cortical excitability.[8] Beyond structural substrates, metabolic disturbances such as hyponatremia and hypocalcemia can further lower the seizure threshold and trigger prolonged convulsions.[17-19] In most reports, neurological manifestations of NCM occur within the first two years of life, typically presenting as seizures or signs of increased intracranial pressure. In contrast, this patient developed neurological symptoms at the age of five years, which is considerably later than commonly reported. This underscores the importance of long-term neurological surveillance in children with giant CMN, even in the absence of early symptoms. She presented with status epilepticus requiring benzodiazepines and barbiturates. Lesions in the amygdala and pons provided clear epileptogenic substrates, the Dandy–Walker variant increased seizure susceptibility, and severe hyponatremia (125 mmol/L) together with hypocalcemia (4.4 mg/dL) acted as additional triggers, resulting in a multifactorial etiology of seizures.

One of the most frequently reported associations of neurocutaneous melanosis (NCM) is with the Dandy–Walker complex, which was also present in our case. The coexistence of this additional malformation has been recognized as a poor prognostic factor, as it increases the risk of symptomatic NCM. Interestingly, the literature also describes rare and unusual associations with NCM, such as type 1 diabetes mellitus that resolved following treatment of CNS melanoma, Hirschsprung's disease in a neonate, and acute disseminated encephalomyelitis (ADEM) in a child. These reports reinforce the understanding that NCM represents a wide clinical spectrum and may involve multiple organ systems, depending on the extent and direction of abnormal neural crest cell migration.[13,14] This is consistent with our case, in which the combination of multifocal NCM, a Dandy–Walker variant, and multisystem involvement highlighted the extreme clinical complexity. Hematologically, the patient presented with pancytopenia (Hb 6.5 g/dL, leukocytes 3.37 $\times 10^3$/µL, platelets 108 $\times 10^3$/µL), most likely related to chronic disease and end-stage CKD. This condition reduced cerebral oxygenation due to anemia and increased bleeding risk from thrombocytopenia, both of which exacerbated seizures. Renally, stage V CKD (BUN 168.5 mg/dL; creatinine 12.02 mg/dL) resulted in severe uremia and electrolyte imbalance, conditions known to lower neuronal membrane stability and promote seizures. Endocrinologically, the patient exhibited an unusual profile—hypoparathyroidism with low PTH levels, in contrast to the secondary hyperparathyroidism typically seen in advanced CKD. Together with severe vitamin D deficiency (8 ng/mL), this further aggravated hypocalcemia, enhancing neuronal excitability and lowering the seizure threshold. Therefore, in this patient, seizures were not only driven by epileptogenic substrates from NCM and the Dandy–Walker variant but were also compounded by hematological, renal, and endocrine dysfunction. The convergence of these multisystem abnormalities rendered status epilepticus multifactorial and significantly worsened the prognosis.

The management of *neurocutaneous melanosis* (NCM) is essentially supportive, as no definitive curative therapy currently exists. Several therapeutic approaches have been reported in the literature, although outcomes remain variable. Surgical resection of intracranial or parenchymal lesions may be considered when the lesions are localized, operable, and cause significant mass effect; however, such indications are rare.[20] In patients with hydrocephalus, placement of a ventriculoperitoneal shunt can reduce intracranial pressure and improve neurological symptoms.[21] More recently, the potential role of targeted molecular therapy has been highlighted, particularly in patients with somatic NRAS or BRAF mutations. Mitogen-activated protein kinase (MEK) and B-Raf proto-oncogen (BRAF) inhibitors, such as trametinib and dabrafenib, have been anecdotally reported to provide clinical responses in cases of leptomeningeal melanocytosis harboring NRAS mutations.[22,23] In contrast, conventional radiotherapy and chemotherapy have generally demonstrated little long-term benefit and are not recommended as standard treatment.[24] Therefore, the management of NCM remains focused on seizure control, correction of metabolic disturbances, and long-term multidisciplinary follow-up aimed at maintaining the patient's quality of life. In this patient, acute seizures were managed with intravenous antiepileptic drugs (phenobarbital and phenytoin), followed by oral phenytoin as prophylaxis. Electrolyte disturbances particularly severe hypocalcemia and hyponatremia were corrected with intravenous calcium gluconate and vitamin D supplementation, in accordance with the presence of hypoparathyroidism and severe vitamin D deficiency. Anemia and pancytopenia were managed with packed red cell transfusions under hematology supervision. Chronic kidney disease stage V required regular hemodialysis twice weekly, accompanied by strict fluid and electrolyte management. Given the multifocal distribution of NCM lesions (bilateral amygdala and pons), epilepsy surgery was not feasible. Seizure control was therefore focused on optimization of medical therapy and metabolic stabilization.

The prognosis of symptomatic neurocutaneous melanosis (NCM) is generally poor, with most patients experiencing neurological deterioration within three years of symptom onset. Recurrent and refractory seizures are the main determinants of morbidity and mortality, and to date, no curative therapy is available. Management therefore focuses on supportive approaches, including seizure control, correction of

metabolic disturbances, and long-term monitoring. To describe prognostic factors and therapeutic options in epilepsy associated with neurocutaneous melanosis (NCM), the distribution pattern of parenchymal melanosis is the strongest predictor of epilepsy outcomes in NCM patients. Patients with isolated amygdala involvement have a higher likelihood of achieving seizure control, even seizure freedom, making epilepsy surgery a potential option. In contrast, multifocal lesions significantly increase the risk of drug-resistant epilepsy, and surgical intervention is usually not beneficial due to the absence of a single resectable focus.[25] In relation to our patient a 5-year-old girl with bilateral amygdala and pontine involvement, presenting with status epilepticus, chronic kidney disease stage V, hypoparathyroidism, vitamin D deficiency, and pancytopenia these findings support that epilepsy surgery is not a realistic option given the multifocal lesion pattern. The best strategy is to optimize medical therapy (antiepileptic drugs), correct metabolic abnormalities, and provide multidisciplinary care. This approach aligns with the evidence from Pellino et al.[25] which emphasizes that failure to control seizures is strongly associated with worse long-term neurological and cognitive outcomes.

Routine MRI screening in patients with CMN remains controversial. The prevalence of neurological involvement is approximately 6–7%, with developmental delay and seizures being the most common clinical manifestations, while CNS melanocytosis and hydrocephalus were the most frequent MRI findings.[6] However, current evidence was deemed insufficient to support strong recommendations for routine MRI in all CMN patients, as its clinical benefits remain uncertain. The authors therefore emphasized the need for standardized study protocols and large-scale, multi-center prospective research to clarify the role of MRI in long-term surveillance. For the time being, clinical centers are advised to determine their own policies by weighing the advantages and disadvantages of MRI screening, although MRI remains recommended whenever new neurological signs or symptoms appear. Our present case where neurological manifestations only emerged at the age of five further underscores the importance of long-term monitoring and a flexible approach in determining the timing of MRI in patients with Giant CMN.[6]

In summary, this case illustrates that the coexistence of Giant CMN, multifocal NCM, Dandy–Walker variant, and multisystem involvement can result in a highly complex clinical presentation with poor prognosis. The unusual feature of late-onset seizures underscores the importance of long-term follow-up and integrated multidisciplinary management. This report not only broadens the understanding of the clinical spectrum of NCM in children but also highlights the necessity of individualized management focused on symptom control, metabolic correction, and quality-of-life preservation.

CONCLUSION

This case describes a rare presentation of congenital melanocytic nevus syndrome with giant CMN, multifocal neurocutaneous melanosis (NCM), and a Dandy–Walker variant, further complicated by multisystem involvement including hematologic, renal, and endocrine abnormalities. Unlike most reports where neurological symptoms appear in the neonatal or infantile period, our patient developed late-onset seizures at the age of five, representing an uncommon clinical course. Multifocal melanosis involving the bilateral amygdala and pons, together with metabolic disturbances, contributed to recurrent and refractory seizures, highlighting the multifactorial pathogenesis. Prognosis in symptomatic NCM remains poor, particularly in multifocal disease where surgical intervention is not feasible, underscoring the importance of optimal medical therapy, systemic correction, and long-term multidisciplinary care to preserve quality of life.

CONFLICT OF INTEREST

All authors declare there is no conflict of interest.

FUNDING

None.

AUTHOR CONTRIBUTION

All authors had contributed in manuscript writing and agreed for the final version of manuscript for publication.

ETHICAL CONSIDERATION

The patient's legal guardian had signed written informed consent regarding publication of medical data in scientific medical journal with confidentiality to personal information.

GENERATIVE AI USAGE DISCLOSURE

The authors explicitly state that the writing of this scientific case report did not use any generative AI technology.

REFERENCES

1. 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(3):441-51. Available from: doi: 10.1016/j.jaad.2012.05.043.
2. Polubothu S, Kinsler VA. Final congenital melanocytic naevi colour is determined by normal skin colour and unaltered by superficial removal techniques: a longitudinal study. Br J Dermatol. 2019;182(3):721–8. Available from: http://dx.doi.org/10.1111/bjd.18149
3. Schaffer J V, Bolognia JL. The treatment of hypopigmentation in children. Clin Dermatol. 2003;21(4):296–310. Available from: http://dx.doi.org/10.1016/s0738-081x(03)00045-2
4. Krengel S, Hauschild A, Schäfer T. Melanoma risk in congenital melanocytic naevi: a systematic review. Br J Dermatol. 2006;155(1):1–8. Available from: http://dx.doi.org/10.1111/j.1365-2133.2006.07218.x
5. Agero ALC, Benvenuto-Andrade C, Dusza SW, Halpern AC, Marghoob AA. Asymptomatic neurocutaneous melanocytosis in patients with large congenital melanocytic nevi: A study of cases from an Internet-based registry. J Am Acad Dermatol. 2005;53(6):959–65. Available from: http://dx.doi.org/10.1016/j.jaad.2005.07.046
6. Fledderus AC, Widdershoven AL, Lapid O, Breugem CC, Pasmans SGMA, van der Horst CMAM, 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(1):1-9. Available from: http://dx.doi.org/10.1186/s13023-022-02234-8

7. Salgado CM, Tomás-Velázquez A, Reyes-Múgica M. Congenital melanocytic neoplasms: clinical, histopathological and recent molecular developments. Virchows Arch. 2025;486(1):165-176. Available from: doi: 10.1007/s00428-024-04011-3.
8. Barkovich MJ, Li Y, Desikan RS, Barkovich AJ, Xu D. Challenges in pediatric neuroimaging. Neuroimage. 2019;185:793–801. Available from: http://dx.doi.org/10.1016/j.neuroimage.2018.04.044
9. Coughlin CC, Polubothu S, Kinsler VA. Neurocutaneous melanosis: clinical features, management and outcomes in a large cohort of children. Br J Dermatol. 2020;183(4):718–26.
10. Fox H. Melanosis of the leptomeninges associated with congenital pigmented naevi. Brain. 1972;95(2):289–300.
11. Di Rocco F, Sabatino G, Koutzoglou M, Battaglia D, Caldarelli M, Tamburrini G. Neurocutaneous melanosis. Child's Nerv Syst. 2003;20(1):23–8. Available from: http://dx.doi.org/10.1007/s00381-003-0835-9
12. Cho IY, Hwang SK, Kim SH. Dandy-walker malformation associated with neurocutaneous melanosis. J Korean Neurosurg Soc. 2011;50(5):475-7. Available from: doi: 10.3340/jkns.2011.50.5.475.
13. Mena-Cedillos CA, Valencia-Herrera AM, Arroyo-Pineda AI, Salgado-Jiménez MA, Espinoza-Montero R, Martínez-Avalos AB, Perales-Arroyo A. Neurocutaneous melanosis in association with the Dandy-Walker complex, complicated by melanoma: report of a case and literature review. Pediatr Dermatol. 2002;19(3):237-42. Available from: doi: 10.1046/j.1525-1470.2002.00073.x.
14. Lookzadeh M, et al. Neonatal neurocutaneous melanosis associated with Dandy–Walker complex: a case report and literature review. Childs Nerv Syst. 2022;38(11):2293–8.
15. Sereke S. WHO classification of paediatric skin tumours. In: Radiopaedia.org. Radiopaedia.org; 2025. Available from: http://dx.doi.org/10.53347/rid-223183
16. Hurt MA. Book review: Weedon D. Weedon's Skin Pathology. 3rd ed. London: Churchill Livingstone Elsevier, 2010. Dermatology Pract & Concept. 2012;79–82. Available from: http://dx.doi.org/10.5826/dpc.0201a15
17. Kinsler VA, Aylett SE, Coley SC, Chong WK, Atherton DJ. Central nervous system imaging and congenital melanocytic naevi. Arch Dis Child. 2001;84(2):152-5. Available from: doi: 10.1136/adc.84.2.152.
18. Sterns RH. Disorders of Plasma Sodium — Causes, Consequences, and Correction. N Engl J Med. 2015;372(1):55–65. Available from: http://dx.doi.org/10.1056/nejmra1404489
19. Bilezikian JP, Brandi ML, Cusano NE, Mannstadt M, Rejnmark L, Rizzoli R, et al. Management of Hypoparathyroidism: Present and Future. J Clin Endocrinol & Metab. 2016;101(6):2313–24. Available from: http://dx.doi.org/10.1210/jc.2015-3910
20. Tsirikos AI, et al. Neurocutaneous melanosis: clinicopathological features, neuroradiological findings and treatment. J Neurosurg. 2004;100(4 Suppl Pediatrics):356–361.
21. 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(8):1471-1477. Available from: doi: 10.1007/s00381-018-3851-5.
22. Kinsler VA, Thomas AC, Ishida M, Bulstrode NW, Loughlin S, Hing S, Chalker J, McKenzie K, Abu-Amero S, Slater O, Chanudet E, Palmer R, Morrogh D, Stanier P, Healy E, Sebire NJ, Moore GE. Multiple congenital melanocytic nevi and neurocutaneous melanosis are caused by postzygotic mutations in codon 61 of NRAS. J Invest Dermatol. 2013 Sep;133(9):2229-36. doi: 10.1038/jid.2013.70. Epub 2013 Feb 7. Erratum in: J Invest Dermatol. 2016 Nov;136(11):2326. Available from: doi: 10.1016/j.jid.2016.09.009.
23. Küsters-Vandevelde HVN, et al. NRAS mutations in primary melanocytic tumors of the central nervous system. Acta Neuropathol. 2014;128(6):897–909
24. Kadonaga JN, Frieden IJ. Neurocutaneous melanosis: definition and review of the literature. J Am Acad Dermatol. 1991 May;24(5 Pt 1):747-55. Available from: doi: 10.1016/0190-9622(91)70115-i.
25. 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. Available from: doi: 10.1016/j.yebeh.2020.107061.