Status Epilepticus in a 3-Month-Old Male: A Case Report of Cerebellar Infarction

Bibek Shrestha[1], Vivek Karn[2], Priyesh Shrestha[2], Shiva Ale Magar[1], Pradeep Regmi[2], Sarthak Neupane[3], and Laxmi Shah[1]

[1]Tribhuvan University Institute of Medicine
[2]Tribhuvan University Teaching Hospital
[3]Tribhuvan University Institute of Medicine Maharajgunj Medical Campus

January 15, 2025

Title Page

Manuscript type: Case Report

Status Epilepticus in a 3-Month-Old Male: A Case Report of Cerebellar Infarction

Authorship

1. Bibek Shrestha, Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Kathmandu Nepal, Email: shresthabibek85iom@gmail.com
2. Vivek Karn, Department of Pediatrics, Tribhuvan University Teaching Hospital, Kathmandu Nepal, Email: drvivekkarn@gmail.com
3. Priyesh Shrestha, Department of Radiology, Tribhuvan University Teaching Hospital, Kathmandu Nepal, Email: priyeshshrestha7@gmail.com
4. Shiva Ram Ale Magar, Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Kathmandu Nepal, Email: shivaramalemagar@gmail.com
5. Pradeep Raj Regmi, Department of Radiology, Tribhuvan University Teaching Hospital, Kathmandu Nepal, Email: pradeepregmi@iom.edu.np
6. Laxmi Shah, Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Kathmandu Nepal, Email: Shahlaxmi203@gmail.com
7. Sarthak Neupane, Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Kathmandu Nepal, Email: sarthakneupane47@gmail.com

Statement of Contribution

Bibek Shrestha played a central role in the research, contributing to conceptualization, data curation, formal analysis, and methodology. He was responsible for project administration and took the lead in writing the original draft, as well as in the review, editing, and visualization processes. Priyesh Shrestha, Vivek Karn,

Shivaram Ale Magar, Sarthak Neupane, Laxmi Shah and Pradeep raj regmi were involved in the supervision, validation, and investigation phases of the project, ensuring the study's accuracy and credibility.

Corresponding author

Bibek Shrestha, Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Kathmandu Nepal, Email: shresthabibek85iom@gmail.com

Disclosure

None

Data availability statement

None

Funding statement

None

Conflict of interest

None

Patient consent statement

Written informed consent was obtained from the parents of the patient for publication of this case image and accompanying images, complying with the requirements as mentioned in Wiley's CCR Consent Form.

Ethical Approval

The institutional review board (IRB) of Institute of Medicine, Maharajgunj Medical Campus does not mandate ethical approval for Case Image.

Acknowledgments

The authors have nothing to report.

Status Epilepticus in a 3-Month-Old Male: A Case Report of Cerebellar Infarction

Key clinical message

Cerebellar infarction is an exceptionally rare cause of status epilepticus in infants, often presenting with non-specific symptoms that mimic other conditions such as febrile seizures, metabolic disorders, or infections. This case highlights the importance of thorough evaluation in infants presenting with seizures, particularly when associated with atypical clinical features, a significant family history of neurological conditions, or consanguinity. Early neuroimaging, particularly MRI, is critical for identifying structural causes like cerebellar infarction and associated vascular anomalies, which may predispose infants to ischemic events. Elevated inflammatory markers, such as CRP, may indicate a proinflammatory or hypercoagulable state contributing to vascular injury. Management requires prompt seizure control, often with antiepileptic drugs like levetiracetam, along with supportive care addressing nutritional deficiencies and systemic inflammation. Genetic counseling and advanced testing should be considered in similar cases to explore potential hereditary predispositions, particularly in resource-limited settings where diagnostic challenges may arise.

Introduction

Global cerebellar infarct is a relatively uncommon type of ischemic stroke, representing about 2% of all cases. It can present with a wide range of symptoms, including vertigo, headache, vomiting, gait imbalance, and dysarthria. [1] Cerebellar infarction, although rare, can also present with seizures in neonates. It has been documented that such infarctions can occur due to various factors, including traumatic birth and vascular insults, leading to neurological deficits and seizures [2]. Status epilepticus is a neurological emergency characterized by prolonged or repetitive seizures lasting over 5 minutes without recovery of consciousness between episodes. It can be convulsive or non-convulsive, with the latter often requiring electroencephalogram monitoring for detection [3]. Common causes include cerebrovascular disorders, brain trauma, infections, and low antiepileptic drug levels in epileptic patients. Noncompliance with anticonvulsant regimens is the most frequent cause, accounting for 28% of all cases. Less common causes, such as inflammatory disorders and inborn errors of metabolism, have been identified through systematic review [4]. Cerebellar lesions, though rare, can also manifest solely as seizures in children, potentially indicating subcortical epilepsy as status epilepticus [5]. This report describes a 3-month-old male infant who presented with status epilepticus and was ultimately diagnosed with cerebellar infarction. The case highlights the diagnostic challenges in infants with seizures, the utility of neuroimaging in identifying structural etiologies, and the importance of a detailed clinical history and family background in guiding investigations and management.

Case History/ Examination

A 3-month-old male infant was brought to the emergency department with a two-day history of poor feeding, lethargy, and recurrent abnormal body movements. These movements were described as tonic-clonic seizures affecting all four limbs, lasting a few minutes per episode, and were not followed by postictal drowsiness. The mother reported prior episodes of fever and respiratory distress. On further inquiry, perinatal complications were noted, including delayed crying at birth, for which the infant received a 14-day course of antibiotics. The neonatal period was also marked by jaundice and neck swelling. He also had a significant family history of born from consanguineous parents (third-degree relatives). The infant had achieved normal developmental milestones until the current illness. However, a significant family history of consanguinity between the parents and a maternal uncle with a history of seizures was noted. The uncle reportedly died in his early twenties due to unknown causes. On admission to the Pediatric Intensive Care Unit (PICU), the infant

required intubation and mechanical ventilation due to recurrent seizures and severe metabolic acidosis. The neurological examination revealed an initial Glasgow Coma Scale (GCS) score of 13/15, which later deteriorated to 9/15. Pupillary reflexes were bilaterally reactive, and there were no signs of bradycardia, meningismus, or papilledema. Respiratory findings included a respiratory rate of 25 breaths per minute, symmetrical air entry, and oxygen saturation of 100% on room air. Cardiovascular examination revealed normal heart rate, blood pressure, and peripheral pulses, with a capillary refill time of less than two seconds. Gastrointestinal and genitourinary examinations were unremarkable, with normal bowel sounds and adequate urine output (1.25 mL/kg/hour).

Differential Diagnosis, investigation and treatment

Laboratory investigations revealed elevated total leukocyte count (17,400/mm^3), with neutrophilia (59%) and anemia (hemoglobin 8.8 g/dL). Biochemical parameters were largely normal, except for low Vitamin D levels (13.1 ng/mL, normal range 30–50 ng/mL) and an elevated C-reactive protein (CRP) of 142 mg/L (normal range: 0–6 mg/L), indicating systemic inflammation. Screening for metabolic disorders, including the Watson-Schwartz, Benedict's, and ferric chloride tests, was negative ruling out metabolic causes of epilepsy. Electroencephalogram (EEG) findings were normal, ruling out non-convulsive status epilepticus. (Table 1) Neuroimaging was pivotal in establishing diagnosis. Initial computed tomography (CT) scan of the head revealed diffuse hypodensity in the cerebellum with loss of differentiation of grey and white matter and periventricular trans ependymal seepage, consistent with a global cerebellar infarction. (Figure 1 and 2) Magnetic resonance imaging (MRI) with angiography (MRA) and venography (MRV) identified congenital vascular anomalies, including hypoplasia of the right anterior cerebral artery (A1 segment) and attenuated right vertebral artery. (Figure 3, 4 and 5) MRV findings were unremarkable, ruling out venous thrombosis. However, genetic testing could not be pursued due to the family's financial constraints. Management initially focused on controlling the seizures and addressing the metabolic derangements. The child was started on levetiracetam, phenytoin, and midazolam to control seizures, followed by a micronutrient "cocktail therapy" with intravenous phenytoin (20 mg twice daily at 8 mg/kg/day) and levetiracetam (200 mg twice daily at 30 mg/kg/dose). Once the seizures were controlled, oral levetiracetam was continued at a dose of 200 mg twice daily for maintenance therapy. Additional supportive treatments included riboflavin (700 mg daily), pyridoxine (500 mg daily), folic acid (20 mg daily), calcium (4 mL twice daily), biotin (10 mg daily), thiamine (200 mg daily), vitamin C (four times daily), L-carnitine (2 mL twice daily), and coenzyme Q (30 mg daily).

Conclusion and Results

The diagnosis of the 3-month-old male infant was cerebellar infarction associated with congenital vascular anomalies, including hypoplasia of the right anterior cerebral artery (A1 segment) and attenuated right vertebral artery. Follow-up recommendations include regular neurological assessments to monitor for potential developmental delays or recurrent seizures. Periodic neuroimaging may be considered to evaluate vascular changes or further ischemic events. Nutritional supplementation should continue to address identified deficiencies, particularly Vitamin D, while maintaining a balanced diet to support overall health. Genetic counseling and testing are advised to explore hereditary predispositions, especially given the family history of consanguinity and seizures. Additionally, long-term coordination with a multidisciplinary team, including pediatric neurologists and developmental specialists, is essential to optimize the child's neurodevelopmental outcomes. This case highlights the diagnostic challenges posed by cerebellar infarction in infants, particularly when presenting as status epilepticus. Early recognition of structural causes through neuroimaging is crucial for appropriate management. Genetic counseling and testing should be considered in cases with consanguinity or a family history of neurological conditions, as such factors may indicate an underlying predisposition. The importance of addressing nutritional deficiencies and systemic inflammation in the overall management of such cases cannot be overstated. Despite financial and resource limitations, a multidisciplinary approach

enabled a favorable outcome in this patient.

Discussion

Cerebellar infarction in infants is a rare but serious condition with potential long-term consequences. It can occur in both preterm and term infants, with various etiologies including hypoxic-ischemic events and migraine complications [2]. Cerebellar infarction is uncommon in infants and is rarely associated with status epilepticus as a presenting symptom. The patient's clinical history, including perinatal complications, recurrent infections, and a significant family history of seizures, adds a unique dimension to this case. Notably, the history of consanguineous marriage and a family history of seizures in the uncle suggest a possible genetic predisposition that could not be confirmed due to financial constraints. Initial differentials included febrile seizures, metabolic or mitochondrial disorders, and structural abnormalities. Febrile seizures were unlikely due to the absence of fever at presentation and the prolonged seizure duration [6]. Negative metabolic tests ruled out major metabolic disorders, although low Vitamin D and elevated CRP suggested possible inflammatory or nutritional contributions [7]. Neuroimaging, particularly MRI, plays a crucial role in detecting cerebellar infarcts due to their often subtle or atypical clinical presentation and the low sensitivity of CT scans. Posterior fossa subtle hypodensity can be missed in non-contrast CT because of the beam-hardening artifact from the temporal bones. Thus, MRI detect infarctions within the first 24 hours and provide superior anatomical detail without bony artifacts. MRI can also reveal small cerebellar infarcts occurring in typical spatial patterns [8]. In this case, CT of head was done and showed diffuse hypodensity of cerebellum with loss of grey-white differentiation likely due to global subacute cerebellar infarct and MRA and MRV showed right A1 anterior cerebral artery hypoplasia, attenuated right vertebral artery. The neuroimaging findings of right A1 anterior cerebral artery hypoplasia and attenuated right vertebral artery indicate congenital vascular anomalies as probable contributors to the cerebellar infarction. Studies have shown that vascular anomalies, combined with perinatal hypoxia, can predispose infants to ischemic injury. Elevated CRP levels further suggest a proinflammatory or prothrombotic state, which may exacerbate vascular vulnerability [9]. The management strategy focused on seizure control, stabilization of metabolic derangements, and supportive care. Intubation and mechanical ventilation in severe metabolic acidosis present significant challenges. While traditionally discouraged due to concerns about compromising compensatory hyperventilation, intubation may be necessary in cases of altered consciousness or refractory seizures [12, 13]. In cases of severe, refractory acidosis with multiple contributing factors, additional interventions such as sodium bicarbonate administration may be considered alongside intubation. The decision to intubate and ventilate in severe metabolic acidosis should be personalized, weighing individual risks and benefits [13]. For status epilepticus, phenytoin and phenobarbital are widely used, but valproic acid and levetiracetam are emerging as safe and effective alternatives [10]. Recent randomized controlled trials show equal efficacy for parenteral phenytoin, levetiracetam, and valproic acid as second-line agents [11]. In this case, the patient was initially managed with intravenous (IV) Phenytoin and Levetiracetam. Once the abnormal body movements were controlled, the treatment was transitioned to oral Levetiracetam. This case highlights the importance of thorough investigation in infants presenting with status epilepticus, particularly in the context of family history and atypical clinical features. Early neuroimaging is indispensable for identifying structural etiologies like cerebellar infarction, which may be overlooked in the differential diagnosis of seizures. Genetic counseling and targeted diagnostic testing should be considered in similar cases, particularly where there is consanguinity or a family history of neurological conditions. The primary limitation of this case was the unavailability of genetic testing and advanced metabolic assays, which could have provided a more definitive diagnosis. Despite the limitations, this case report emphasizes the importance of considering rare structural causes in infants presenting with status epilepticus and highlights the utility of neuroimaging in resource-limited settings. In the future, advancements in genetic testing and metabolic profiling could aid in the early identification of at-risk populations, particularly in consanguineous families. Furthermore, this case underscores the need for enhanced awareness and capacity-building efforts to integrate advanced diagnostic and therapeutic modalities in pediatric neurology. Long-term follow-up studies would also provide crucial insights into the

prognosis and efficacy of tailored management strategies in similar cases.

List of Figures

List of Tables

References

1. Hashmi AMA, Aaron S, Sinani AA, Pancharatnam D. Clinical and radiological profile of patients presenting with isolated acute cerebellar infarct: A Single Tertiary Center 3 years Retrospective Study from Central Stroke Unit, Muscat, Oman. Journal of Stroke Medicine [Internet]. 2021 Sep 29;4(2):95–9. Available from: https://doi.org/10.1177/25166085211038210
2. Mühlbacher T, Bohner G, Bührer C, Dame C. Cerebellar Infarction: Unusual Manifestation with Facial Palsy, Focal Seizures, and Secondary Generalization. Neonatology [Internet]. 2017 Sep 25;113(1):33–6. Available from: https://doi.org/10.1159/000480423
3. Hawkes MA, Hocker SE. Status Epilepticus. In: Oxford University Press eBooks [Internet]. 2021. p. 473–8. Available from: https://doi.org/10.1093/med/9780197512166.003.0057
4. Tan RYL, Neligan A, Shorvon SD. The uncommon causes of status epilepticus: A Systematic Review. Epilepsy Research [Internet]. 2010 Aug 15;91(2–3):111–22. Available from: https://doi.org/10.1016/j.eplepsyres.2010.07.015
5. Işık U, Dinçer A, Sav A, Özek MM. Seizures as the sole manifestation of cerebellar lesions in three children. Journal of Pediatric Neurology [Internet]. 2009 Jan 1;7(2):193–8. Available from: https://doi.org/10.3233/jpn-2009-0286
6. Martin ET, Kerin T, Christakis DA, Blume HK, Gospe SM, Vinje J, et al. Redefining Outcome of First Seizures by Acute Illness. PEDIATRICS [Internet]. 2010 Nov 23;126(6):e1477–84. Available from: https://doi.org/10.1542/peds.2010-1138
7. Amirkhizi F, Hamedi-Shahraki S. Association of vitamin D status with metabolic and inflammatory factors in adults with obesity. Medical Journal of Mashhad University of Medical Sciences [Internet]. 2021 Aug 23;64(3). Available from: https://mjms.mums.ac.ir/article_18847.html
8. De Cocker LJL, Lövblad KO, Hendrikse J. MRI of Cerebellar Infarction. European Neurology [Internet]. 2017 Jan 1;77(3–4):137–46. Available from: https://doi.org/10.1159/000455229
9. Grad E, Danenberg HD. C-reactive protein and atherothrombosis: Cause or effect? Blood Reviews [Internet]. 2012 Dec 23;27(1):23–9. Available from: https://doi.org/10.1016/j.blre.2012.12.001

10. Verrotti A, Ambrosi M, Pavone P, Striano P. Pediatric status epilepticus: improved management with new drug therapies? Expert Opinion on Pharmacotherapy [Internet]. 2017 May 8;18(8):789–98. Available from: https://doi.org/10.1080/14656566.2017.1323873
11. Swarnalingam E, Woodward K, Esser M, Jacobs J. Management and prognosis of pediatric status epilepticus. Zeitschrift Für Epileptologie [Internet]. 2022 Nov 1;35(4):332–44. Available from: https://doi.org/10.1007/s10309-022-00538-0
12. Fernando SM, Charbonneau V, Rosenberg H. Hypercapnea and Acidemia despite Hyperventilation following Endotracheal Intubation in a Case of Unknown Severe Salicylate Poisoning. Case Reports in Critical Care [Internet]. 2017 Jan 1;2017:1–3. Available from: https://doi.org/10.1155/2017/6835471
13. Vidanapathirana M. Sodium bicarbonate and intubation in severe diabetic ketoacidosis: are we too quick to dismiss them? Clinical Diabetes and Endocrinology [Internet]. 2024 Apr 15;10(1). Available from: https://doi.org/10.1186/s40842-024-00171-y

Hosted file

`Table 1.docx` available at https://authorea.com/users/823487/articles/1259844-status-epilepticus-in-a-3-month-old-male-a-case-report-of-cerebellar-infarction