CASE REPORT

Diagnostic Challenges of Acute Motor Axonal Neuropathy in Pediatric: A Case Report

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ABSTRACT

Acute motor axonal neuropathy (AMAN) is a variant of Guillain-Barrй Syndrome (GBS) that poses diagnostic and therapeutic challenges in pediatric patients. This case report details a previously healthy preschool girl who developed acute flaccid paralysis (AFP), initially diagnosed as having transverse myelitis and varicella myelitis. A definitive diagnosis of AMAN was reached after a complex diagnostic process. Despite the parents' refusal to permit a lumbar puncture, treatment was initiated, resulting in significant but incomplete recovery. This case underscores the importance of accurate diagnosis, timely intervention, and the multifaceted impact on family dynamics.
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INTRODUCTION

Acute Motor Axonal Neuropathy (AMAN) is characterized by a rapid onset of motor deficits without sensory involvement, leading to a quick progression toward severe disability or life-threatening complications, such as respiratory failure. GBS and its variants are estimated to affect 0.6 to 4 people per 100,000 globally yearly, with incidences lower in children than adults (1). The prognosis of GBS varies widely among patients. Most patients regain independent function within six months to a year, although about 20-30% may still experience some weakness after three years (2). This case study explores diagnostic dilemmas and their impacts on patients' recovery and family psychosocial dynamics.

CASE REPORT

A 5-year-old girl with no significant medical history experienced acute weakness in both lower limbs over two days, initially presenting with an abnormal gait. Her mobility deteriorated to crawling and unable to stand by the second day. She also reported right calf pain and intermittent fever, but denied bowel or urinary dysfunction. Born at term via emergency cesarean section for fetal distress, she weighed 3.8 kg at birth. Her vaccinations were complete by 18 months, and her development was age-appropriate. Family history was unremarkable.

Lower limb examination revealed bilateral generalized hypotonia, hyporeflexia, muscle power 3/5, and a limping gait when supported, with normal sensory. Cognition, speech, cranial nerves, bulbar function and upper limb examination were normal.

Laboratory investigations showed elevated C-reactive protein (CRP) of 9.2 mg/L, with normal full blood count, blood culture, neuromyelitis optica (NMO) antibody, viral serology. Serum creatine kinase (CK), CK-MB, and lactate dehydrogenase (LDH) were normal, ruling out muscle pathology and cardiac involvements. A limited nerve conduction study (NCS) was performed as the patient was uncooperative. The result showed prolonged F wave latencies for age, variable CMAP and normal conduction velocities. suggesting an early phase of GBS. A contrasted whole-spine magnetic resonance imaging (MRI) was performed and revealed a patchy long-segment hyperintensity in the thoracic spinal cord from T2 to T10, as shown in Figue 1. Figure 2 revealed the hyperintensity of the spinal cord in axial view.

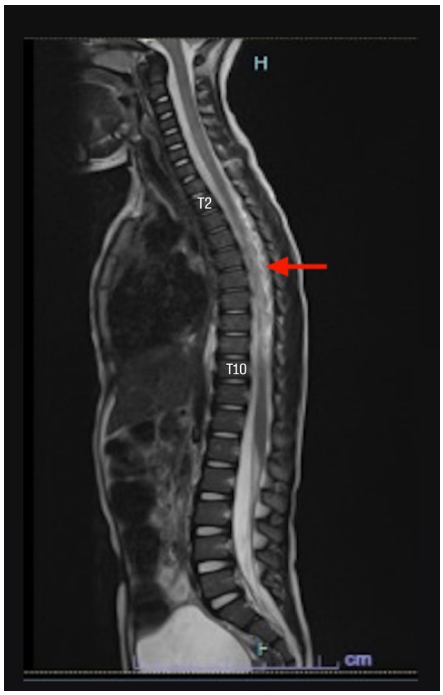

Figure 1: T2 weighted MRI Spine Sagittal View: Patchy long segment hyperintensity from T2 until T10/11.

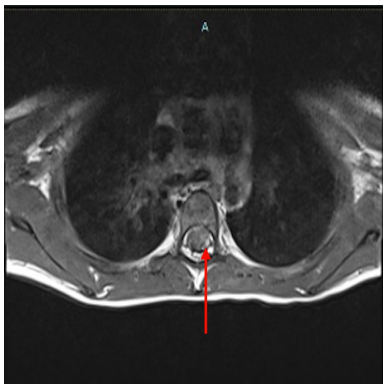

Figure 2: MRI Spine Axial View: Hyperintensity of the spinal cord.

She was treated as transverse myelitis with intravenous methylprednisolone 150 mg three times daily (TDS) for 7 days. On day 3 of treatment, she developed varicella infection with upper limb weakness, leading to revised diagnosis to varicella myelitis. Due to worsening varicella lesions, methylprednisolone was withheld, and started IV Immunoglobulin (IVIG) therapy 15g over 2 hours for 2 days, IV acyclovir 300 mg four times daily for 2 weeks and pregabalin 15 mg three times daily. Despite treatment, she developed persistent fever with no neurological improvement. Lumbar puncture (LP) was recommended to confirm the diagnosis and guide further management but declined by her parents due to procedural worries. After 20 days of hospitalisation, she was discharged following resolution of varicella infection, though lower limb weakness and areflexia persisted.

One week post-discharge, another NCS was done and confirmed Acute Motor Axonal Neuropathy (AMAN), with evidence of significant reduction in CMAP amplitude across all nerves, mild conduction velocity reduction, and normal distal latencies, predominantly affecting the left median and ulnar nerves. The interval between first and second NCS was 27 days. She was readmitted for IVIG 15g over 12 hours for 2 days. The interval between first and second IVIG was 20 days. Repeated MRI spine showed reduction in previously observed patchy long-segment hyperintensity in the thoracic spinal cord. Her 4-day hospitalisation was uneventful, and she completed IVIG therapy without complications.

One month post-discharge, she could stand and walk with support but not fully weight-bearing, with distal lower limb strength 3/5. and improved fine motor skills. Three months later, she could climb chairs and walk with a limping gait, with improved distal muscle strength at 4/5. The follow-up plans include continuing intensive rehabilitation and scheduling the next appointment in six months to assess further progression.

DISCUSSION

Guillain-Barré Syndrome (GBS) is a rare but significant cause of acute flaccid paralysis in children. A Malaysian study found that among GBS patients, 74.2% presented with demyelinating subtype, 12.9% had AMAN, and 9.7% were unclassifiable (3). AMAN is more common in Asian populations, including Malaysia, compared to Western countries (4). AMAN primarily affects motor neurons, leading to more severe axonal damage as compared to acute inflammatory demyelinating polyneuropathy (AIDP). Regarding prognosis, most pediatric patients with AMAN often regain independent mobility within a year. A previous study by Salehiomran et al. (2016) indicates that approximately 80% - 94% of children with GBS achieve complete recovery within two years, although some experience incomplete recovery. In this case, despite a challenging course complicated by a concurrent varicella infection, her condition showed marked improvement with treatment, reflecting the good prognosis seen in pediatric GBS cases.

Diagnosing pediatric GBS is challenging due to non-specific initial presentations, which can be misattributed to benign conditions like viral myositis or musculoskeletal injuries. MRI spine is useful to differentiate GBS and transverse myelitis. However, our patient's first MRI raised suspicion of transverse myelitis, delaying the diagnosis of AMAN. Parental concerns and refusal of lumbar puncture (LP) may complicate the diagnostic process. In addition, performing NCS can be challenging, particularly in less cooperative pediatric patients.

According to Zhu et al (2021), gangliosides are primarily located in the outer layer of the neuronal cell membrane's bilayer structure, playing vital roles in cellular growth,

signal transduction and immune responses. When anti-ganglioside antibodies bind to their corresponding gangliosides, they trigger complement system activation, leading to neural damage, including axonal degeneration and myelin loss. In AMAN, common anti-ganglioside antibodies include anti-GM1 and anti-GD1a. Anti-GM1 antibodies are linked to the pure motor variant of GBS, which occurs without sensory impairment. Anti-GD1a antibodies are associated with conduction failure and have strong correlation with AMAN and pure motor GBS. Various techniques can be used to detect anti-ganglioside antibodies, including enzyme linked immunosorbent assay (ELISA), immunodot assays and thin-layer chromatography (TLC) overlay.

Management of GBS in pediatric patients involves early recognition, supportive care and timely intervention with immunomodulatory therapies. In Malaysia, as elsewhere, two primary treatments for GBS are intravenous immunoglobulin (IVIG) and plasmapheresis, both aimed at mitigating the autoimmune response driving the disease (5). According to Madaan et al. (2019), IVIG is typically preferred for pediatric patients due to its relative safety, ease of administration and efficacy. IVIG also has been proven by Kalita et al. (2022) to be particularly effective in AMAN, by neutralizing pathogenic antibodies and modulating the immune response. Supportive care is also a crucial component of GBS management, focusing on monitoring respiratory function, managing pain, providing nutritional support and rehabilitation. Intensive physiotherapy and occupational therapy were useful in this patient to improve her muscle strength, mobility, and functional independence. According to Gawande et al. (2024), early and aggressive rehabilitation has been associated with improved functional outcomes and a reduction in long-term disability.

AMAN is often associated with preceding *Campylobacter jejuni* infection (4), commonly seen in undercooked poultry or contaminated water. Good food handling protocols are important, including cooking meat thoroughly and washing hands regularly. Additionally, ensure all vaccinations are up-to-date to prevent diseases that could potentially trigger GBS, like polio and influenza vaccines.

The sudden onset of a severe illness like GBS can significantly impact the family. As in this case, her mother developed depression due to the sudden onset of her child's illness and stress of caregiving, highlighting the need for comprehensive care that addresses both medical and psychosocial needs. A study by Bernsen et al. (2006) has mentioned that caregivers have a higher risk of anxiety, depression and other psychological distress due to the uncertainty of the prognosis, disruption of normal routines and burden of caregiving.

CONCLUSION

The diagnostic challenges of AMAN in pediatric patients, compounded by overlapping symptoms with other neurological conditions underscore the importance of timely and accurate diagnosis. Despite significant recovery outcomes, this case illustrates the long-term effects of GBS, such as persistent weakness. Comprehensive chronic care, multidisciplinary coordination, and family-centered support remain essential in addressing both medical and psychosocial aspects of care for the patient and her family.

By implementing preventive measures and maintaining a vigilant approach to patient care, healthcare providers can help reduce risk of inadvertent uvula injury during anaesthesia.

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REFERENCES

1. McGrogan A, Madle GC, Seaman HE, de Vries CS. The epidemiology of Guillain-Barrй syndrome worldwide. A systematic literature review. Neuroepidemiology. 2009;32(2):150–63. doi: 10.1159/000184748
2. Hughes RAC, Cornblath DR. Guillain-Barrй syndrome. Lancet Lond Engl. 2005;366(9497):1653–66. doi: 10.1016/S0140-6736(05)67665-9
3. Goh KJ, Ng WK, Vaithialingam M, Tan CT. A clinical and electrophysiological study of Guillain-Barrй syndrome in Malaysia. Neurol Asia. 1999;4:67–72.
4. Bae JS, Yuki N, Kuwabara S, Kim JK, Vucic S, Lin CS, et al. Guillain-Barrй syndrome in Asia. J Neurol Neurosurg Psychiatry. 2014;85(8):907–13. doi: 10.1136/jnnp-2013-306212
5. Hughes RA, Swan AV, van Doorn PA. Intravenous immunoglobulin for Guillain-Barré syndrome. Cochrane Database Syst Rev. 2014;2014(9):CD002063. doi: 10.1002/14651858.CD002063.pub6