Diagnostic challenge and management of Guillain-Barre syndrome in an infant: a case report

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European Journal of Medical Case Reports
Volume 9(5):102–106
DOI: 10.24911/ejmcr.9-1607

ABSTRACT

Background: Guillain-Barre syndrome (GBS) is an acute, immune-mediated post-infectious polyradiculoneuropathy usually presenting with symmetrical ascending weakness, diminished deep tendon reflexes, and nonspecific sensory symptoms. GBS is, in essence, an autoimmune disorder, and the underlying mechanism is thought to result from so-called molecular mimicry. This hypothesis is further supported by approximately two-thirds of the patients having a preceding infection. In most cases, the infectious trigger occurs in the gastrointestinal or respiratory tract, with the disease manifesting within 4 weeks. Even though it most commonly affects children aged 1-5 years, there are rare cases reported in neonates and infants.

Case Presentation: We report a case of a 6-month-old infant with GBS following a respiratory infection. The diagnosis was confirmed through cerebrospinal fluid (CSF) analysis, electromyoneurography, spine MRI, and clinical assessment. Positive human herpes virus 6 (HHV-6) in CSF suggested a potential infectious trigger. The infant was treated with intravenous immunoglobulin and ganciclovir, requiring intensive care and mechanical ventilation. Recovery involved gradual neurological improvement and restored motor function over 30 days.

Conclusion: GBS is a rare disorder in children, especially when associated with HHV-6 infection. It requires multidisciplinary management to prevent complications and improve the prognosis of patients. Upon arrival at the emergency department, all patients should be carefully evaluated, looking for autonomic and respiratory dysfunction signs. Generally, pediatric patients have a better prognosis compared to adults. Initiation of treatment in the early stages of the disease leads to a faster recovery and, consequently, fewer sequelae.

Keywords: Guillain-Barré syndrome, post-infectious polyradiculoneuropathy, acute flaccid paralysis, pediatric, infant, HHV-6, case report.

Type of Article: CASE REPORT **Specialty:** Neurology

Received: 25 November 2024
Revised (1): 26 May 2025
Accepted: 28 May 2025

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Background

Guillain-Barré syndrome (GBS) is an acute, immune-mediated, post-infectious polyradiculoneuropathy usually presenting with symmetrical ascending weakness, diminished deep tendon reflexes, and nonspecific sensory symptoms. Diagnostic criteria [1] for GBS are shown in Table 1. Inflammation results from immune-mediated injury to the peripheral nerves, wherein the generation of autoantibodies leads to the activation of the complement system and the invasion of myelin by macrophages and autoreactive cytotoxic T cells [2]. The autoantibodies target components of the Schwann cell's plasma membrane known as gangliosides; these anti-ganglioside antibodies include anti-GM1 and anti-GD1a, among others [2]. The underlying mechanism of GBS is thought to result from molecular mimicry. This hypothesis is further supported by approximately two-thirds of the patients having a preceding infection [3].

In most cases, the infectious trigger occurs in the gastrointestinal or respiratory tract, with the disease manifesting within 4 weeks [1,4]. The worldwide incidence of GBS for all ages has been estimated to be within the range of 0.4-2.4 per 100,000 individuals [5]. The syndrome generally affects children aged 1-5 years, however, there are rare cases reported in neonates and infants [6]. According to reports, GBS shows a slight male predominance, with a male-to-female ratio of 1.3-1.5:1 [7]. Low incidence and non-specific symptoms make the diagnosis of GBS in infants particularly challenging (low suspicion index).

This report documents a case of GBS in a 6-month-old infant, outlining the clinical course, diagnostic approach, therapeutic interventions, and prognosis.

Table 1. *Diagnostic criteria for Guillain-Barre syndrome (modified from Asbury and Cornblath) [1].*

1. FEATURES NEEDED FOR DIAGNOSIS IN CLINICAL PRACTICE
a. Progressive weakness in legs and arms (sometimes initially only in legs)
b. Areflexia/hyporeflexia in affected limbs
2. ADDITIONAL SYMPTOMS
a. Progressive phase lasts days to weeks (often 2 weeks)
b. Relative symmetry
c. Mild sensory symptoms or signs
d. Cranial nerve involvement, especially bilateral facial palsy
e. Autonomic dysfunction
f. Pain (common)
g. Absence of fever at onset
3. LABORATORY STUDIES
a. CSF: elevated CSF proteins (>0.45 mg/dl) after the first week of symptoms, presence of albumin-cytological dissociation (ACD)
b. Abnormal neurophysiological parameters on nerve conduction studies
4. FEATURES CASTING DOUBT ON THE DIAGNOSIS
a Marked, persistent asymmetry of weakness
b. Persistent bladder or bowel dysfunction
c. Bladder or bowel dysfunction at onset
d. More than 50 mononuclear cells/ µl in CFS
e. Sharp sensory level

Case Presentation

A 6-month-old male infant presented to the Pediatric Clinic (the Clinical Centre University of Sarajevo) with a 3-day history of generalized weakness, absent sucking and swallowing reflexes, diminished deep tendon reflexes, and signs of respiratory distress. A history of a current respiratory infection was evoked, and the patient was previously started on antibiotic treatment.

Pregnancy, labor, and neonatal history were unremarkable. The infant reached typical motor milestones, had age-appropriate vaccination status, and had no significant family medical history. He was exclusively breastfed. Upon initial examination, the infant was in respiratory distress with paradoxical breathing. He appeared irritable, with generalized weakness, reduced muscle strength, spontaneous movements, and areflexia. Plantar grasp and Babinski were absent, with no signs of meningeal irritation. Shortly after admission, the patient went into respiratory insufficiency and was transferred to the pediatric intensive care unit, where he was intubated. From a post-diagnosis perspective and according to the GBS Disability Score, his level of disability would be scored 5.

Laboratory investigations were practically within normal limits, with only mild anemia and a hemoglobin level of 9.9 g/l. The cerebrospinal fluid (CSF) analysis performed initially was unequivocal, but due to a high index of suspicion for GBS and clinical reasoning, the lumbar puncture was repeated on the 10th day. This is because CSF protein levels in GBS may be negative during the first week and rise thereafter [1]. The second analysis revealed albumin-cytological dissociation with an elevated protein level (5.2 g/l) and a normal cell count. Antiganglioside antibodies were negative. Interestingly, the patient had slightly elevated anti-acetylcholine receptor antibodies (IgG fraction 1.2 nmol/l – ref. range <0.5 nmol/l). CSF microbiological studies were positive for human herpes virus 6 (HHV-6). Brain MRI results (T1-weighted, T2-weighted, FLAIR, diffusion, and gadolinium injection sequences) were unremarkable; however, his spine MRI (T1-weighted, T2-weighted, STIR, diffusion, and gadolinium injection sequences) demonstrated marked enhancement of the anterior spinal nerve roots in the region of the cauda and conus medullaris (Figures 1 and 2). Neurophysiological parameters obtained on 1 month after the initial symptoms demonstrated a demyelinating polyneuropathy. These findings were consistent with the presumptive diagnosis of GBS.

He was immediately started on intravenous immunoglobulin (IVIG) therapy at a dose of 2 g/kg over 5 days, as per standard protocol for pediatric GBS. Since the patient had a positive PCR for HHV-6, indicating active replication within the central nervous system (CNS) (but without clear evidence of acute encephalitis), intravenous ganciclovir was started in consultation with an infectious disease specialist. He was extubated after 25 days, with a gradual recovery. The neurological function progressively recovered, with remaining muscle weakness, albeit with better motor strength and increased spontaneous

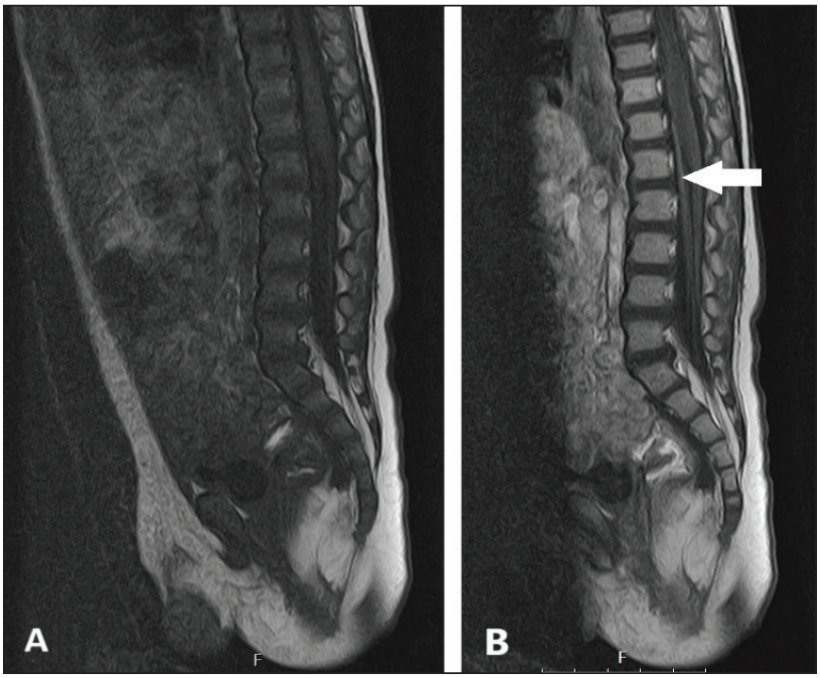

Figure 1. *Axial FSE T1-weighted MR image in native series (A) and after Gadolinium contrast injection (B) demonstrates post-contrast enhancement (white arrow) of the anterior spinal nerve roots.*

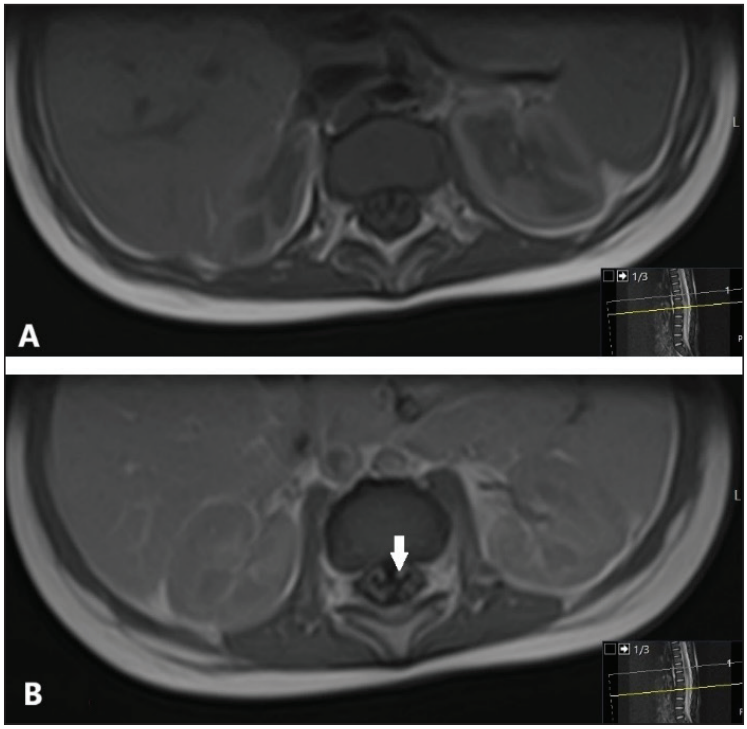

Figure 2. *Sagittal FSE T1-weighted MR image in native series (A) and after Gadolinium contrast injection (B) demonstrates post-contrast enhancement (white arrow) of the anterior spinal nerve roots.*

movements. By the time he was discharged, he could grab his feet, grab toys, rotate, and keep his head raised when in the prone position. On discharge, rehabilitation therapy was recommended, and close outpatient follow-up was planned. In particular, a re-evaluation of acetylcholine antibody titers was planned, considering that myasthenia gravis is one of the many disease mimics in this case.

Discussion

GBS is an acute peripheral polyneuropathy considered to be of autoimmune origin. It is caused by an aberrant response of the patient's immune system, mainly induced by an infectious trigger. Approximately 75% of patients with GBS show signs of infection preceding the diagnosis [8]. Two-thirds of patients present with respiratory

or gastrointestinal tract infection symptoms before neurologic symptoms. *Campylobacter jejuni* is detected in at least one-third of these patients [9]. Other pathogens identified as causes of GBS are Epstein-Barr virus, cytomegalovirus, *Mycoplasma pneumoniae*, *Haemophilus influenzae*, and influenza A virus [9].

It is known that major complications of primary HHV-6 infection involve the central nervous system [10]. In our case, the PCR test for HHV-6 in CFS was positive, suggesting a possible trigger for GBS. Although there have not been many cases of GBS reported in infants, Miyake et al. [10] reported the case of a 7-month-old female infant who developed GBS around 3 weeks after primary symptomatic HHV-6 infection [10].

Even though there is a lack of data considering HHV-6 infection and GBS in pediatric patients, Pereira et al. [11] conducted a comparative study analyzing the clinical presentation of GBS in adults with positive CSF PCR tests for HHV-6 versus those who tested negative for HHV-6. Their findings suggest that HHV-6 may contribute to a more severe clinical course of GBS, potentially due to its replication during the disease's progression. The study observed a high prevalence of motor deficits as initial symptoms and hypotension, indicating possible autonomic nervous system involvement.

If HHV-6 DNA is present in CSF, it is a strong indication of active replication of the virus within the CNS [12]. Active HHV-6 infection in the CNS parallel with clinical presentation of GBS suggests that previous and current infections might be associated with GBS [12].

GBS is uncommon in children, even more so in those under 2 years of age [13]. In this age group, a high index of suspicion is essential due to a broad differential diagnosis, including acute cerebellar ataxia, transverse myelitis, spinal cord compression, tick paralysis, botulism, trauma, ADEM, myasthenia gravis, poliomyelitis, and other myopathies [1].

The pathophysiology of acute inflammatory demyelinating polyradiculoneuropathy (AIDP) is still unclear, but a direct connection has been found between acute motor axonal neuropathy and anti-ganglioside antibodies [8]. Recent studies regarding GBS demonstrated an association between gangliosides and autoimmune mechanisms [14].

Kim et al. [15] conducted a multicenter study on 119 GBS patients, finding anti-ganglioside antibodies (IgG or IgM) in 60 cases. Antibody-positive patients were mainly male and often had preceding gastrointestinal infections, a shorter interval to motor weakness, negative sensory signs, and lower CSF protein levels. In contrast, our patient tested negative for these antibodies, had no gastrointestinal symptoms, and was negative for *C. jejuni*, supporting the AIDP variant of GBS.

When considering a potential differential diagnosis, we should mention that our patient had slightly elevated anti-acetylcholine receptor antibodies, which can be seen in myasthenia gravis. However, the patient's age and clinical presentation do not fit this disease state. We believe that this is the result of the IVIG therapy that our patient has received, or it might be the result of general immune system dysfunction associated with autoimmune disorders.

Conclusion

This case highlights the need for a high index of suspicion for GBS in infants, despite its rarity in this age group. Acute flaccid paralysis in infancy poses a significant diagnostic challenge due to a wide range of differential diagnoses. Our case demonstrates that early recognition and timely treatment of GBS can lead to a favorable outcome and prevent serious complications, such as prolonged neuromuscular weakness and autonomic dysfunction.

A notable finding was the detection of HHV-6 DNA in the CSF, suggesting active viral replication as a potential trigger for GBS. While the link between HHV-6 and GBS in children is not well established, this case supports emerging evidence that viral agents beyond *C. jejuni* may play a role in GBS pathogenesis.

The absence of anti-ganglioside antibodies and negative *C. jejuni* serology supported the AIDP subtype of GBS. Mildly elevated anti-acetylcholine receptor antibodies were likely influenced by IVIG therapy, emphasizing the need for cautious interpretation of serologic results, especially with concurrent IVIG therapy.

Ultimately, this case underlines the critical importance of early diagnosis and intervention. Clinical vigilance and prompt management are essential to improving outcomes and minimizing long-term sequelae in infants, where GBS may be easily missed.

What is new?

Even though it is rare, GBS should be considered a differential diagnosis in an infant presenting with acute flaccid paralysis, especially after a preceding infection (respiratory or gastrointestinal). Considering the pattern of myelination, it is not surprising to find infants presenting with bulbar symptoms and signs, in addition to generalized weakness, hyporeflexia, and areflexia. Timely diagnosis and management significantly improve outcomes and reduce potential long-term consequences.

List of Abbreviations

AIDP	Acute inflammatory demyelinating polyradiculoneuropathy
CNS	Central nervous system
CSF	Cerebrospinal fluid
GBS	Guillain-Barre syndrome
HHV-6	Human herpes virus 6
IVIG	Intravenous immunoglobulin
PICU	Pediatric intensive care unit

Conflict of Interest
The authors declare that there is no conflict of interest regarding the publication of this article.

Funding
None.

Consent for publication
Written informed consent was obtained from the patient's parents.

Ethical approval
Our institution does not require ethical approval to publish an anonymous case report.

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Summary of the case

1	Patient (gender, age)	Male, 6 months
2	Final diagnosis	Guillain-Barre syndrome
3	Symptoms	Generalized weakness, absent sucking and swallowing reflexes, and signs of respiratory distress
4	Medications	IVIG, ganciclovir
5	Clinical procedure	Mechanical ventilation was initially required, but the infant was extubated shortly after. Started on IVIG 2 g/kg over 5 days, and ganciclovir i.v. 5 mg/kg bid
6	Specialty	Pediatric Neurology, Pediatric Intensive Care, Radiology