Acute hemiplegia as a rare presentation of infantile Guillain-Barré syndrome

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ABSTRACT

Guillain-Barré syndrome (GBS) usually presents in a symmetrical ascending fashion of weakness. We present a 6-month-old male infant who manifested acute hemiplegia and head lag after a febrile illness. The diagnosis of GBS was confirmed by laboratory, neurophysiologic, and neuroradiologic investigations. The clinical improvement following intravenous immunoglobulin (IVIG) treatment and physiotherapy was noted. A literature review for GBS presenting in infants as hemiplegia revealed no previous reports. Our objective in presenting this particular case is to highlight the importance of considering GBS in the differential diagnosis so that early effective treatment may be started.

Case Report. A previously healthy 6-month-old male infant presented to our emergency room with acute left hemi-body limb weakness and head lag 3 days after an acute febrile upper respiratory tract infection. There was no history of change in his sensorium, seizure, head trauma, or recent vaccination. His vital signs were stable and cranial nerves were intact. He had remarkable head lag and very much reduced tone, power, and deep tendon reflexes (DTRs) in the left upper and lower limbs in comparison with normal motor examination on the right. A brain CT completed a few hours after presentation to the hospital was normal. Lumbar puncture was performed, and cerebrospinal fluid (CSF) showed high protein (1.31 g/L) (normal values: 0.15-0.45 g/L) and all other CSF parameters were negative including PCR studies and culture. Nerve conduction study (NCS) showed a reduced motor nerves compound muscle action potential (CMAP), and slow conduction velocity (Table 1, Figure 1). Brain MRI and

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MR spectroscopy (MRS) were unremarkable. A spinal cord MRI (Figure 2) showed T2 high signal intensity affecting the ventral roots and thickened enhancing filum roots of the cervical and lower vertebral segments. A metabolic workup was normal and stool culture for poliovirus was negative. After confirming the diagnosis of GBS and excluding all other possibilities including infectious, vascular, and metabolic causes; a 5 day course of 0.4 g/kg/day IVIG in addition to intensive physiotherapy were initiated. He showed remarkable clinical and neurophysiological improvement, and 6 months later he was able to support his neck, stand with support, and use both hands symmetrically to grasp objects and play with toys.

**Discussion.** Acute inflammatory demyelinating polyradiculoneuropathy is commonly known as Guillain-Barré syndrome. It is an inflammatory, demyelinating disorder of the spinal nerve roots and peripheral nerves of acute to subacute onset,
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associated with a T-cell-mediated immune response. Guillain-Barré syndrome is a clinical diagnosis that typically causes an ascending pattern of symmetrical weakness and depressed DTRs. Several other children younger than 6 months of age with GBS have been reported, however, it is rarely reported in children under 2 years. Classically it presents in infants as symmetric weakness and hypotonia. The atypical asymmetric clinical presentation in our case made the diagnosis challenging.

The diagnosis of GBS was confirmed by: 1) The presence of cyto-albuminologic dissociation with high CSF protein and normal CSF white blood cells. 2) The electrophysiological evidence decreased CMAP amplitude and prolonged conduction velocity (Tables 2 & 3, Figure 3). 3) The spinal cord nerve roots enhancement on MRI. 4) The clinical improvement following IVIG treatment. A unilateral presentation of GBS in infants is rarely reported. The acute presentation of focal weakness in an infant warrants considering a broad differential diagnoses.

Acute hemorrhagic or ischemic neurovascular injury within the CNS is an important differential diagnosis in our case that was ruled out by brain imaging.

Many infectious etiologies that can affect brain parenchyma, anterior horn cells and nerves should be excluded in an infant who presents with acute flaccid paralysis. In our case, Epstein-Barr virus, cytomegalovirus, herpes simplex virus, varicella zoster virus, and, mycoplasma pneumonia was ruled out by CSF culture and PCR studies. Abnormalities in white blood cells and its differentials and/or glucose in CSF may raise the concern of meningitis as it can present with a similar picture that features fever and disturbed

<table>
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<th>Latency</th>
<th>Amplitude</th>
<th>Distance</th>
<th>Velocity</th>
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<tr>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
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<tr>
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</tr>
<tr>
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<tr>
<td>Knee</td>
<td>5.20</td>
<td>9.8</td>
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Table 2 - Nerve conduction study after 20 weeks.

<table>
<thead>
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<th>Min F Lat</th>
<th>Max F Lat</th>
<th>Mean F Lat</th>
</tr>
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<tbody>
<tr>
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<tr>
<td>R Tibial (Knee)- AH</td>
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<td>19.15</td>
<td>17.09</td>
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</table>

Table 3 - F-wave latencies after 20 weeks.

Figure 3 - Normal compound muscle action potential (NCS), conduction velocity and F-wave for age. APB- abductor pollicis brevis muscle, AH- abductor hallucis muscle. A) & B) upper limb, C) & D) lower limb.
sensorium. In particular, botulism can present in this age with a similar scenario of weakness but can be differentiated from GBS by the presence of bulbar involvement, constipation, history of contaminated food ingestion, and a specific EMG/NCS pattern (low-amplitude CMAP, tetanic or post-tetanic facilitation, and the absence of post-tetanic exhaustion).6

Each case of acute paralysis should be reported to local authorities for the possibility of poliomyelitis. Stool samples taken after 14 days of paralysis are usually the standard method to isolate poliovirus. In our infant, stool samples did not isolate poliovirus or *Campylobacter jejuni* (*C. jejuni*) as the later one can be a causative organism for GBS autoimmune response, especially if preceded by gastrointestinal symptoms. Testing for anti-GM1 ganglioside antibodies also could aid in the diagnosis of GBS as it can possibly being associated with *C. jejuni* infection.7

Neurometabolic causes of sudden weakness are rare, but should be considered in communities with a high rate of consanguineous marriage. Serum lactate and MRS were normal in our patient, however, mitochondrial diseases like mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes, should be suspected when there is a high lactate in serum, CSF, and on MRS that is concordant with stroke-like presentation at any age. Hypokalemia, organic acidurias, urea cycle defects, and homocystinuria can present as a similar picture, and measuring targeted metabolites in serum and urine levels would be indicated if suspected.8

Our patient had no history of trauma; however, thorough enquiry and clinical and radiological investigations for the possibility of both accidental and non-accidental trauma are necessary. Base line hematological work up can be carried out to explore causes like sickle cell disease and thrombotic abnormalities if suspected. These were negative in our patient. A similar clinical picture can occur in Hopkins syndrome, which is a rare form of acute flaccid paralysis of one or more limbs that usually occurs in the recovery period following bronchial asthma exacerbation; however, though Hopkins syndrome can show nonspecific motor neuron disease on NCS, it differs from our case as the CSF studies are normal and the spinal cord MRI rarely shows anterior horn T2 enhancement.9

Early diagnosis and treatment of GBS have a better prognosis in children as compared with adults, though recovery may still take 6-12 months.10 Treatment with IVIG is widely used in children with GBS and can hasten the recovery and alter the autoimmune reaction. Plasma exchange is another treatment modality, but sometimes clinicians prefer less invasive methods. Our patient received a course of IVIG 2 g/kg over 5 days with no complications and was discharged home in a stable condition. Intensive physical and occupational therapy is a key determinant of clinical improvement and led to a remarkable clinical improvement in our patient, who 6 months later was able to stand with support and to use both hands symmetrically.

In conclusion, GBS is a rare diagnosis in infants but is an important differential diagnosis in acute onset weakness as delayed diagnosis can increase morbidity and mortality. Cyto-albuminologic dissociation, electrophysiological tests, and spinal cord MRI are helpful tools in confirming the diagnosis and hence initiating the appropriate treatment.

References