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Case report

Guillain Barre Syndrome with Hemiplegic Presentation: An Atypical Rare Presentation

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Abstract

Guillain Barre Syndrome (GBS) is an autoimmune disorder that is thought to be a post-infectious polyneuropathy, involving mainly motor but also sensory and sometimes autonomic nerves. The classical description of GBS is that of demyelinating neuropathy with ascending weakness, usually begins in the lower extremities and progressively involves the trunk, the upper limbs and finally the bulbar muscles. Many clinical variants with atypical presentations have been documented and variants involving the cranial nerves or pure motor involvement and axonal injury have also been described. Here we report a rare case of GBS presenting as acute hemiplegia. The report suggests the clinicians that GBS can be used as a differential diagnosis of acute hemiplegia, which broadens the ideas of clinicians.

Keywords: Guillain Barre Syndrome, Acute hemiplegia, Clinical Variants.

Abbreviations: GBS-Guillain Barre Syndrome, IVIG-Intravenous Immunoglobulin, AIDP-Acute Inflammatory Demyelinating Polyradiculoneuropathy, AMAN-Acute Motor Axonal Neuropathy, AMSAN-Acute Motor and Sensory Axonal Neuropathy, MFS-Miller Fisher syndrome, CSF-Cerebral Spinal Fluid, CNS- Central Nervous System.

Introduction

Guillain-Barre Syndrome (GBS) is the most common cause of acute flaccid paralysis in the developed and developing countries [1]. The overall incidence is 0.6-4 per 100000 each year in the population younger than 18 years [1]. GBS is an autoimmune disorder that is thought to be a post-infectious polyneuropathy, involving mainly motor but also sensory and sometimes autonomic nerves. This syndrome affects people of all ages and is not hereditary. Most patients have demyelinating neuropathy but few patients have primary axonal degeneration [2].

The classical description of GBS is that of demyelinating neuropathy with ascending weakness, usually begins in the lower extremities and progressively involves the trunk, the upper limbs and finally the bulbar muscles and it is a symmetrical weakness [2]. Many clinical variants with atypical presentations have been documented and variants involving the cranial nerves or pure motor involvement and axonal injury have also been described [3]. Here we report a case of GBS presenting as acute hemiplegia which is a rare atypical presentation.

Case Report

12 years boy, presented with chief complaints of weakness on left side of body over a period of 24 hours and weakness started from left lower limb and progressed to left upper limb without any facial involvement. There was no history of fever, headache, seizures, altered level of consciousness and head injury. There was history of viral upper respiratory tract infection two weeks back.

With the clinical suspicion of left sided hemiplegic stroke, this boy was referred to DY Patil Medical College and hospital and research Institute, Kolhapur. On clinical examination, he was conscious, well oriented, afebrile, heart rate 82 beats per minute, blood pressure 120/80mm of mercury, respiratory rate 20 breaths per minute and SpO2 of 94%. On CNS (Central Nervous System) examination, all cranial nerves were normal. Motor system examination showed normal bulk of muscles, grade 3/5 power, deep tendon and planter reflex were absent on left lower and upper limbs. There was no bladder bowel involvement.

Sensory system examination was normal on left side. The neurological examinations on right upper and lower limbs were normal and there were no signs of meningeal irritation. After admission in the next 24 hours, his weakness progressed with power 2/5 in left lower and upper limbs (both proximally and distally), hypotonia and also developed truncal weakness. He was having difficulty to sit from lying position and also to stand. His fundoscopic examination was normal. Rest of the other system examinations including cardiovascular and respiratory were normal. His laboratory parameters like complete blood counts along with ESR, serum electrolytes and ANA (Antinuclear Antibodies) were normal.

Metabolic workup like serum lactate, serum homocysteine and serum B12 levels were normal. His brain MRI with contrast was normal and MRI spine showed extensive contrast enhancement of ventral nerve roots and thickened roots of the cervical and lower vertebral segments. CSF (Cerebral Spinal Fluid) showed albumino-cytological dissociation with 5 lymphocytes per HPF and 80 mg/dl proteins and normal glucose. CSF staining and cultures were negative.

Nerve conduction study showed slow conduction velocity. Stool culture for polio virus was negative. By excluding all other possibilities including infectious, vascular and metabolic causes, a diagnosis of GBS was confirmed and a 5 days course of Intravenous Immunoglobulin (IVIG) in the dose of 0.4 g/kg/day along with supportive care and physiotherapy during recovery phase was given. Our patient did not required ventilator care as he did not develop respiratory muscle weakness. He showed clinical recovery within 14 days with power of 4/5 in left upper and lower limbs and normal muscle tone and no truncal weakness. The patient was discharged on 20th day of admission without any residual deficit.

Discussion

GBS was first reported by Landry in 1859 and later detailed by Guillain, Barre and Strohl, in 1916. The Disease has become well known internationally under the name of Guillain Barre Syndrome [4]. GBS is an inflammatory, demyelinating disorder of spinal nerve roots and peripheral nerves of acute to subacute onset associated with a T cell mediated immune response [3]. An antecedent presumably viral infection triggers inflammation and demyelination, as in our case there was viral upper respiratory tract infection two weeks prior to acute hemiplegic GBS presentation [3]. GBS can now be divided into at least four subtypes. The most important is Acute Inflammatory, Demyelinating Polyradiculoneuropathy (AIDP). The other subtypes include Acute Motor Axonal Neuropathy (AMAN) and Acute Motor and Sensory Axonal Neuropathy (AMSAN). The fourth and rarest subtype, representing 1% of cases, is the Miller Fisher syndrome of ataxia, areflexia and ophthalmoplegia [3,5].

There are unusual childhood clinical variants of GBS and which include:

- A ‘pure ‘ataxic form.
- A ‘pure ‘sensory form.
- A pharyngeal cervical brachial syndrome, in which acute oropharyngeal, neck and shoulder weakness occurs in the absence of significant limb weakness.
- A paraparetic form.
- Bifacial weakness with parasthesias.
- Miller Fisher syndrome (MFS) of ataxia, external ophthalmoplegia and areflexia, with minimal limb weakness.
- Bickerstaff brainstem encephalitis, in which features of MFS and hypersomnolence.
- A form resembling brain death with total paralysis and fixed dilated pupils but a normal electroencephalogram.

In our case, acute ischemic, haemorrhagic or metabolic hemiplegic stroke was the differential diagnosis and was ruled out by MRI brain and metabolic workup studies. The diagnosis of GBS in our case was confirmed by

- The presence of albumino-cytologic dissociation with high CSF proteins and normal CSF white blood cells. The increase in CSF proteins is because of inflammation of nerve roots.
- The spinal cord nerve root enhancement on MRI Spine.
- Nerve conduction study demonstrated prolonged conduction velocity and lastly,
- The clinical improvement following IVIG treatment.

Similar to our case report, Muthaffar O.Y, et al. reported acute left sided hemiplegia as a rare presentation of infantile GBS in a 6 months infant who presented 3 days after febrile upper respiratory tract infection and showed remarkable improvement after IVIG use [6]. Also Kim EJ, et al. from Australia reported an unusual case of GBS who presented with left hemiparesis in the acute phase and right hemifacial weakness at the recovery phase of the hemiparesis [7].

Khattak S, et al. reported a case of GBS in a 50 years old patient who initially presented to the Emergency room with hemiparesis and cranial nerve palsies simulating a cerebrovascular event and the diagnosis of GBS was made, based on neurological examination, CSF analysis and needle EMG findings [8]. Acute hemiplegia as a clinical presentation of GBS could be confidently added to the category of atypical presentation of pediatric GBS which constitutes 11.2%-24.3% of the whole pediatric GBS presentation reported previously [8]. But the exact underlying cause and pathogenesis of acute hemiplegia in GBS is not well known. Early diagnosis and treatment of GBS have a better prognosis in children as compared with adults, though recovery may still take 6-12 months [9].

Severe or rapidly progressive muscle weakness is treated with IVIG in the dose of 0.4g/kg/day for 5 consecutive days or 1g/kg/day for 2 days. Treatment with IVIG is widely used in children with GBS and can alter the autoimmune reaction which will fasten the recovery. Plasmapheresis and or immunosuppressive drugs are alternative if IVIG is ineffective [2]. Supportive care, such as respiratory support, prevention of pressure sores, nutritional support, and pain management, prevention of deep vein thrombosis and treatment of secondary bacterial infections is also important [2].

Conclusion

Many clinical variants of GBS have been documented and this report suggests the clinicians that GBS can be used as a differential diagnosis of acute hemiplegia, which broadens the ideas of clinicians.

References

1. Hughes RA and Rees JH. Clinical and epidemiological features of Guillain Barre Syndrome (1997) J Infect Dis 176: 92-98. https://doi.org/10.1086/513793
2. Ryan MM. Guillain Barre Syndrome Kliegman RM (eds) Nelson Textbook of Pediatrics, Elsevier, Netherlands.
3. Smith SA and Ouvrier R. Acute inflammatory demyelinating polyradiculopathy (Guillain Barre Syndrome) Swaimman’s Pediatric Neurology (2012) Elsevier, Netherlands.
4. Hughes RA and Comblath DR. Guillain Barre Syndrome (2005) Lancet 366: 1653-1666.
5. Wakerley BR and Yuki N. Mimics and chameleons in Guillain-Barre and miller fisher syndromes (2015) Pract Neurol 15: 90-99. https://doi.org/10.1136/practneurol-2014-000937
6. Muthaffar OY, Mahmoud AA and AL-Salman AS. Acute hemiplegia as a rare presentation of infantile Guillain Barre Syndrome (2014) Saudi Med J 35: 861-864.
7. Kim EJ and Yuki N. Hemiparetic Guillain Barre syndrome (2016) J Neurol Sci 361: 131-132.
8. Khattak S, Nabi S, Khattak I and Badshah M. An unusual presentation of GBS: case report and literature review (2016) Pakistan J Neuro Sci 11: 40-43.
9. Akbayram S, Dogan M, Peker E, Saytn R, Aktar F, et al. Clinical features and prognosis with Guillian Barre syndrome (2011) Ann Indian Acad Neurol 14: 98-102. https://doi.org/10.4103/0972-2327.82793