Case Report

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A Classic Presentation of Guillian Barre Syndrome

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Abstract

Guillain Barré Syndrome (GBS), is an acute polyneuropathy characterized by immune–mediated peripheral nerve myelin sheath destruction. GBS is an uncommon diagnosis in the emergency department (ED), usually present with complaints of weakness. We report a case of 13-year old female who was presented to the emergency department with initial complaints of headache and sent home with a diagnosis of migraine and she returned to ED with weakness and ultimately found to be GBS.

Keyword: Guillain Barré Syndrome (GBS); Polyneuropathy.

Case Report

A 13-year-old female presented to the emergency department (ED) with a chief complaint of headache since one week and weakness of the upper and lower extremities, increasing over the three days prior to arrival. The patient has no associated symptoms like numbness, paresthesias, fever, neck stiffness, nausea, vomiting and visual changes. Patient came to ED 1 day back with complaints of headache since 5 days and sent home with a diagnosis of migraine and she returned to ED with weakness. Past medical history was not significant. There was no positive family history.

The physical exam was normal with the exception of mild dysarthria, bifacial LMN paresis, neck flexor weakness, moderate truncal weakness, SBC-1020 , BHT 45 sec power: UL- proximal 3/5; distal 4/5 hand grip 90%; LL—proximal 3/5; distal 4+/5, generalized areflexia and plantars- flexor.

Our laboratory results are as follows:

WBC: 13.7*10^9/L

N-64/L-28/E-04/M-04

Hb: 14.6 gm/dl

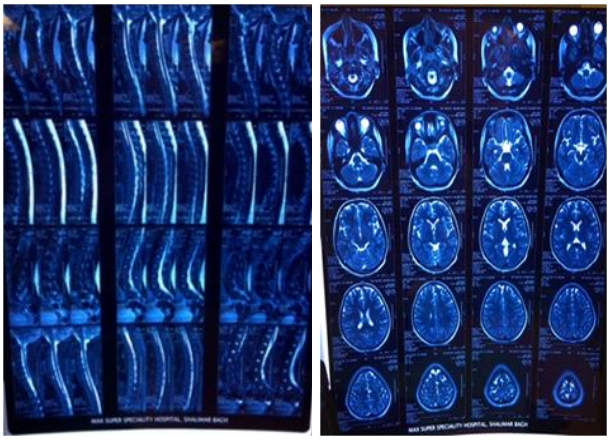
Platelet count- 327*10^9/L

LFT-T.protein-7, albumin-4.3, globulin 2.7, t.bilirubin-0.6, direct-0.1, indirect-0.5, SGOT-21, SGPT-11 ALPO4-167, GGT-14

RFT- BUN-35mg/dl, s.creatinine-0.42mg/dl, s.Na-138, s.K-4.1, s.CL-105

The differential diagnosis included Guillain Barré Syndrome. A neurology consultation was sought . After taking history and performing physical examination they recommended for electromyography (EMG), MRI whole spine and MRI brain with contrast.

MRI Whole Spine and MRI Brain with Contrast – Normal



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An EMG was also done, which showed severe demyelinating + axonal motor polyradiculo-neuropathy. The patient was started on intravenous immunoglobulin (IVIg) and regular limb physiotherapy.

Discussion

Guillain-Barré syndrome (GBS) can be described as a collection of clinical syndromes that manifests as an acute inflammatory polyradiculoneuropathy with resultant weakness and diminished reflexes. GBS is a postinfectious, immune-mediated disease. Most patients report an infectious illness in the weeks prior to the onset of GBS. GBS usually involves a prodromal event such as an upper respiratory infection, vaccination, surgery, or a gastrointestinal infection. It is thought that up to 88% of those affected by GBS have a prodromal infection. The most commonly linked organism with GBS is *Campylobacter jejuni* [1,2]. Many of the identified infectious agents are thought to induce production of antibodies that cross-react with specific gangliosides and glycolipids, such as GM1 and GD1b, that are distributed throughout the myelin in the peripheral nervous system [3].

Table 166-2 Diagnostic Criteria for Classic Guillain-Barré Syndrome

Required
Progressive weakness of more than one limb
Areflexia
Suggestive
Progression over days to weeks
Recovery beginning 2–4 wk after cessation of progression
Relative symmetry of symptoms
Mild sensory signs and symptoms
Cranial nerve involvement (Bell's palsy, dysphagia, dysarthria, ophthalmoplegia)
Autonomic dysfunction (tachycardia, bradycardia, dysrhythmias, wide variations in blood pressure, postural hypotension, urinary retention, constipation, facial flushing, anhydrosis, hypersalivation)
Absence of fever at onset
Cytoalbuminologic dissociation of cerebrospinal fluid (high protein and low white cell count).
Typical findings on electromyogram and nerve conduction studies

Four subtypes of peripheral neuropathy are classified under the umbrella of GBS; these are acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor and sensory axonal neuropathy (AMSAN), and the Miller Fisher Syndrome (MFS). GBS has been reported throughout the world. AMAN and AMSAN occur mainly in northern China, Japan, and Mexico, making up only 5-10% percent of GBS cases in the United States. AIDP accounts for up to 90% of cases in Europe, North America, and the developed world. GBS also has a slightly greater male preponderance with a ratio of 1.25–1.5:1 [4].

Differential diagnosis can be poliomyelitis, myasthenia gravis, electrolyte disturbance, botulism, acute myopathy, diphtheria, vasculitis, porphyria, tick paralysis, and toxic neuropathy.

Treatment of GBS includes plasmapheresis or intravenous immunoglobulin (IVIg), as well as respiratory support when needed.

Table 166-3 Managing Respiratory Failure in Guillain-Barré Syndrome

Indications for Intubation
Vital capacity <15 mL / kg
PaO_2 <70 mm Hg on room air
Bulbar dysfunction (difficulty with breathing, swallowing, or speech)
Aspiration
Indications for admission to intensive care unit
Autonomic dysfunction,
Bulbar dysfunction
Initial vital capacity <20 mL/kg
Initial negative inspiratory force <–30 cm of water
Decrease of >30% of vital capacity or negative inspiratory force
Inability to ambulate
Treatment with plasmapheresis

Approximately 80% patients with GBS walk independently at 6 months, and about 60% of patients attain full recovery of motor strength by 1 year. Recovery in approximately 5-10% of patients with GBS is prolonged, with several months of ventilator dependency and a very delayed, incomplete recovery.

We report on this patient because she provides a good example of the difficulty that continues to exist in the ED with respect to the diagnosis of GBS. GBS

should be considered in any patient with neurological complaints for example headache in this patient. A complete neurologic examination including tone, active and passive motor power, all sensations, reflexes, coordination and anal tone should be checked, especially in the ED.

In situations where the diagnosis is unclear but where GBS is a part of the differential diagnosis—it is important for the EP to give clear discharge instructions with signs to look out for and close follow up as an outpatient. GBS is the leading cause of acute neuromuscular paralysis and however rare the presentation, we as EPs need to have a keen eye for the presentation characteristics. We have the potential to diagnose and intervene early in the disease progression, both of which benefit the patient's ultimate outcome.

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