



Guillain Barre Syndrome: A Rare Presentation Mimicking Acute Stroke

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Authors' contributions

This work was carried out in collaboration between all authors. Author AY led the conceptual design. Authors SA and HA assisted in the data collection and writing the case summary.

Authors GA, MH and GB managed the reference, literature review, case discussion and wrote the first draft. Author NA reviewed the literature, edited the case discussion and refined the final draft of the manuscript. All authors read and approved the final manuscript.

Case Study

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ABSTRACT

Guillain Barre Syndrome (GBS) is an acute neuromuscular weakness and paralysis associated with areflexia and often spontaneous recovery, but carries the potential risk of respiratory depression owing to muscle weakness. Worldwide, 1 to 3 cases/100,000 are reported. The syndrome is most commonly reported as symmetrical ascending weakness in arms and legs accompanied by hyporeflexia or areflexia. Sensory disturbances are not required for diagnosis, but may or may not be present. Acute inflammatory demyelinating poly-radiculoneuropathy (AIDP) is the most common variant, but acute motor and sensory axonal neuropathy (ASMAN) is more severe and usually leads to partial or slow recovery. We present a case of GBS presenting with asymmetric weakness and sensory disturbance in a patient with bloody diarrhea of unknown etiology. This patient had asymmetrical paralysis mimicking stroke, but the physical findings, laboratory studies, normal CT and MRI of the brain, Electromyogram (EMG) and the patient's improvement with Intravenous Immunoglobulin (IVIG) support the diagnosis of GBS. People with inflammatory bowel disease are at increased risk of

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developing GBS. Persons with antecedent *Campylobacter jejuni* infections are 77 percent more likely to contract GBS compared to the general population, and Cytomegalovirus (CMV) and Epstein Barr virus (EBV) are also implicated risk factors.

Keywords: Guillain-barre syndrome; acute inflammatory demyelinating polyradiculoneuropathy (AIDP); acute motor and sensory axonal neuropathy (ASMAN); amyotrophic lateral sclerosis; mono-neuritis multiplex; campylobacter jejuni.

1. CASE PRESENTATION

49 years old male with past medical history of gastritis, ulcerative colitis and Deep Vein Thrombosis (DVT) presented to Emergency Department (ED) with severe crampy lower abdominal pain and bloody diarrhea for 7 days. The patient was in no acute distress and he was afebrile, and had respiratory rate 20 breaths/min, heart rate 97/minutes, blood pressure 100/75mmHg and O₂ saturation of 97% on ambient air. He had tenderness in the left lower quadrant (LLQ), and focused neurological examination was unremarkable. Laboratory studies showed hemoglobin 10.5 g/dl, hematocrit 31.6%, WBC 7700/uL and Platelet 396,000/uL. The patient was admitted to the regular floor for treatment with prednisone, mesalamine and metronidazole for ulcerative colitis flare up.

Next day, he almost fell on his way to the bathroom due to left leg weakness. On further questioning, it was discovered that he began to have this progressively worsening weakness 3 weeks ago. Detailed neurological examination revealed that muscle strength was 2/5 on the left side and 4/5 on the right. All the deep tendon reflexes were absent. Sensory sensations were impaired more on the left side than right side. Cranial nerves were intact and there were no cerebellar signs. Given the asymmetry of neurological symptoms, the possibility of acute stroke was considered. However, CT of the brain without contrast found no evidence of intracranial hemorrhage or acute stroke. A brain MRI without contrast showed no evidence of infarction or any other abnormality.

MRI lumbar spine revealed mild degenerative findings with a small left L5-S1 foraminal disk herniation. MRI of cervical spine showed mild degenerative findings with no significant stenosis and no cervical cord compression. EMG and nerve conduction study (NCS) of both lower extremities revealed bilateral poly-neural peripheral demyelinating disease with slowed conduction time highly suggestive of a demyelinating disorder most likely GBS.

On 6th day of admission, the patient began complaining of difficulty in breathing. The patient was transferred to the Intensive Care Unit (ICU) for GBS with impending respiratory failure, due to vital capacity (VC) of 1.010Liter and a negative inspiratory force (NIF) of -12cm H₂O. Orthostatic hypotension was also noted. An EMG and NCS found the left leg to have slowed conduction time suggestive of a demyelinating disorder most likely GBS. There was no albuminocytological dissociation on CSF analysis done on day 11 and 17 (Table 1).

Table 1. Laboratory data

CSF examination					
Parameter	Normal value	Day 2	Day 11	Day 17	Day 25
Glucose	40-70 mg/dL		99	97	
Total Protein	15-45 mg/dL		38	30	
Site			CSF	CSF	
Color			Colorless	Colorless	
Character			Clear	Clear	
RBC-F/uL			810	192	
WBC-F/uL			5	1	
Mononuclear %			10	100	
Polymononuclear %			90	0	
Serum					
Quantiferon			Negative		
HIV-1 RNA by PCR	Positive if >20 copies/mL			<20	
HIV1 Antibody	Negative if <1.00	<1.00			
HIV1 Antibody confirm	Negative if Non reactive	Non Reactive			
Parvo B19 IgG	0.0 to 0.8	3.20			
Parvo B19 IgM	0.0-0.8	0.10			
EBV Antibody Viral Capsid AG IgG	Negative if 0.0 to 0.8				>8.0
EBV Antibody Viral Capsid AG IgM	Negative if 0.0 to 0.8				0.30
CMV, IgG Antibody	Negative if 0.0 to 0.8				9.60
CMV IgM Antibody	Negative if 0.0 to 0.8				<0.9
HBS Antigen		Negative			
Hepatitis A Antibody (IgM)		Negative			
Hepatitis B Core IgM Antibody		Negative			
Hep C Antibody	0.0 to 0.9	<0.1			
Antinuclear Antibody (IgG)			Negative		
Serum Angiotensin converting Enzyme (ACE)	12 to 68 U/L		30.00 U/L		
Rapid HIV1 Antibody					
Sedimentation Rate	1 to 20 mm/hr	20	10		
Rheumatoid Factor	0 to 13.9 IU/mL		5.6		
Albumin	3.2 to 5.6 g/dL				

Treatment with IVIG started and patient started showing gradual progressive improvement in respiratory and neurological status, VC, NIF and muscle strength improved and patient started ambulating without any difficulty. Patient was discharged home without any residual neurological deficit after 6 weeks of hospital stay including 2 weeks of stay in ICU.

2. DISCUSSION

Differential diagnosis of asymmetric weakness and sensory disturbance includes but not limited to Amyotrophic lateral sclerosis (ALS), Mononeuritis multiplex (MNM), multiple sclerosis (MS), Acute stroke and Guillian-Barre syndrome.

People with inflammatory bowel disease are at increased risk of developing GBS [1].

The incidence is less than 3 per 100,000 persons, with a higher incidence in males and increasing risk with aging [2,3]. Persons with antecedent *Campylobacter jejuni* infections are 77 percent more likely to contract GBS compared to the general population, and EBV and CMV infection are also implicated risk factors [4,5,6]. Our patient had history of IBD and presented with bloody diarrhea, and hence had the risk factors for developing GBS.

ALS can involve both the upper and lower motor neurons. Patient will have symptoms of muscle atrophy and or wasting, dysphasia, and skeletal muscles cramping and twitching. Typically ALS is common in elderly male. There is no specific test for diagnosis of ALS, and this is a clinical diagnosis of exclusion. Patients will have normal blood tests, nerve conduction studies, MRIs and CSF analysis.

The patient will present with a slow onset of combination of the UMN and LMN symptoms [7]. Characteristically ALS does not exhibit any sensory disturbances [8].

Mono-neuritis multiplex (MNM) is characterized as an asymmetrical progressive peripheral neuropathy in two or more nerves in separate parts of the body that is most common secondary to vasculitis. Serology markers including raised ESR, Rheumatoid Factor, and p-ANCA can be positive. This patient had sub-acute neurological deterioration and serum markers of raised ESR, Rheumatoid Factor, and p-ANCA were negative. Multiple sclerosis and acute stroke were excluded by negative brain MRI.

GBS is most commonly reported as symmetrical weakness starting in the legs [9]. The National Institute of Neurological and Communicative Disorders and Stroke (NINCDS) formulated a diagnostic criterion that addresses 85 percent of all GBS cases [10]. The diagnostic criteria for the syndrome include progressive weakness proximally and distally in more than 2 limbs for less than 4 weeks and areflexia [11]. Supportive signs and symptoms to diagnose GBS include sensory loss, elevation of sedimentation rate, hyponatremia secondary to inappropriate antidiuretic hormone release, and decrease in neuromuscular conduction, and normal cell count in CSF with elevated protein [11,12].

Respiratory failure secondary to respiratory muscle weakness occurs in 9 percent of patients [13]. Paresthesia in hands and feet occur in greater than 80 percent of patients, but sensory impairment is usually mild. Autonomic dysfunction can occur in 70 percent of patients; common manifestations are orthostatic hypotension, loss of sweating, urinary retention and ileus [14].

The most common form of GBS in the United States is acute inflammatory demyelinating poly-radicleoneuropathy (AIDP) accounting for 85 to 90 percent of the cases. AIDP classically presents with progressive symmetrical ascending paralysis and with areflexia. The mechanism is thought to be an autoimmune attack targeting nerve roots leading to demyelination.

A more severe form of GBS is acute motor and sensory axonal neuropathy (AMSAN). AMSAN has the same criteria as AIDP, but extends to sensory fibers in addition to motor fibers. It is secondary to axonal degeneration, which commonly leads to either a slower recovery or partial recovery [15].

The GBS is commonly preceded by a *Campylobacter jejuni* infection, but can also be associated with Cytomegalovirus and Epstein Bar virus [16]. *Campylobacter jejuni* infections are commonly characterized by low-grade fever, abdominal cramps and 8-10 watery or bloody bowel movements daily [17]. CMV most commonly presents with diarrhea, fever, gastrointestinal bleeding and abdominal pain in immunocompromised patients or those with inflammatory bowel disease [18].

Treatment options include intravenous immunoglobulin and plasma exchange [19]. Clinicians must monitor vital capacity and respiratory rate to predict respiratory failure, and blood pressure and heart rate to monitor autonomic dysfunction.

The difference between our patient and other documented cases was that our patient had profound unilateral motor and sensory deficits along with unilateral loss of sweating. However, the patient fits the GBS criteria; he had a loss of motor function in 2 limbs with areflexia. The patient also had many of the supporting criteria; he had sensory loss and the EMG studied revealed demyelination of peripheral nerves.

Recognition of acute stroke involves taking careful clinical history of abrupt onset of neurological deficit, and focal neurological signs might include unilateral weakness or numbness of face, arm, or leg, visual field defect or speech disturbance [20].

Reports of asymmetrical GBS exist but to our knowledge data does not exist discussing the frequency of asymmetrical presentation. We propose our patient developed atypical GBS. The patient did not present with symmetrical ascending paralysis, but the physical findings, laboratory studies, the EMG and the patient's improvement with IVIG support the diagnosis of GBS.

5. CONCLUSION

GBS, on rare occasions, can present with asymmetrical weakness and sensory disturbance mimicking acute stroke. People with inflammatory bowel disease are at higher risk of developing GBS. High index of suspicion is required for clinical diagnosis and standard tests including EMG help establish the diagnosis. Intensive monitoring for respiratory failure and autonomic dysfunction is very imperative. Standard treatment with IVIG or plasma exchange is the mainstay of therapeutic intervention.

CONSENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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