Pakistan Journal of Neurological Sciences (PJNS)

Volume 18 | Issue 3 Article 10

9-2023

Ataxic Variant Of Guillain Barre Syndrome: A Case Report

Haris Majid Rajput
Pakistan Institute of Medical Sciences, Islamabad, Pakistan

Malik Muhammad Adil
Pakistan Institute of Medical Sciences, Islamabad, Pakistan

Maryam Khalil
Pakistan Institute of Medical Sciences, Islamabad

Nayab Aslam
Pakistan Institute of Medical Sciences, Islamabad, Pakistan

Zaid Waqar
Pakistan Institute of Medical Sciences, Islamabad, Pakistan

https://doi.org/10.56310/pjns.v18i03.230

ATAXIC VARIANT OF GUILLAIN BARRE SYNDROME: A CASE REPORT

Haris Majid Rajput[1], Malik Muhammad Adil[1], Maryam Khalil[1], Nayab Aslam[1], Zaid Waqar[1]
[1]Pakistan Institute of Medical Sciences, Islamabad

Correspondence Author: Malik Muhammad Adil *Pakistan Institute of Medical Sciences, Islamabad* ***Email:*** *dr.adil31@gmail.com*

Date of submission: *March 3, 2023* ***Date of revision:*** *August 29, 2023* ***Date of acceptance:*** *September 10, 2023*

ABSTRACT
A 17-year-old girl was admitted after acute onset of unsteady gait succeeding acute gastroenteritis. Neurological examination reported normal power in all four limbs, impaired finger-nose, heel-shin tests, areflexia and ataxic gait. We eliminated other diseases with cerebellar symptoms; for example, Wernicke encephalopathy, multiple sclerosis, cerebellar vascular disease , encephalitis in the brain stem and cerebellum. Blood serum collected from the patient during the acute phase showed no anti-ganglioside antibodies. As the patient presented with evident cerebellar ataxia without muscle weakness, ophthalmoplegia or proprioceptive sensory disruption a diagnosis of ataxic form of Guillain-Barré syndrome (GBS) after nerve conduction studies. Though ataxic GBS is not a settled impression, we should have to give heed to the potential existence of such a scarce GBS variant.

Keywords: GBS, ataxic, variant

INTRODUCTION
Guillain-Barré syndrome (GBS) is an acute inflammatory polyradiculoneuropathy causing sensory symptoms such as numbness followed by ascending symmetrical motor weakness, facial weakness, dyspnea, dysphagia, diplopia, dysarthria and autonomic dysfunction.[1] It is often preceded by febrile illness, surgery, trauma, vaccination. Cellular and humoral immune mechanisms constitute its pathogenesis.

Acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor-sensory axonal neuropathy (AMSAN), and Miller Fisher syndrome (MFS) are the most prevalent variants of GBS. Others include acute panautonomic neuropathy, pure sensory GBS, pharyngeal-cervical-brachial variant.

Here, we are going to report a rare variant of Guillain-Barré syndrome presented with features of severe ataxia of cerebellar type without opthalmoplegia and with intact proprioception.

CASE PRESENTATION
A 17-year-old female, having no previous co morbidities presented in emergency room (ER) of Pakistan Institute of Medical Sciences with complaint of difficulty in walking for 10 days. This was preceded by low grade undocumented fever and loose stools from which she recovered within four days. She did not have any breathing difficulty, nasal regurgitation, dysphagia, diplopia, dysarthria. There was no history of fall, trauma, surgery, vaccination or previous episode of such illness.

At time of presentation, vital signs were blood pressure of 110/70 mmHg, pulse 84 beats/min, temperature of 37 degree Celsius, respiratory rate of 18 breaths/min, oxygen saturation was 97% at room air, single breath count >20. Glasgow coma scale (GCS) 15/15, conscious, oriented, pupils bilateral equal and light-reactive, horizontal nystagmus was noted, extraocular movements were intact. Rest of cranial nerves functions were intact. According to Medical Research Council (MRC) grading of muscle power 5/5 proximally and distally in both upper and lower limbs. Finger nose and heel-shin test were affected. Deep tendon reflexes were found absent in both upper and lower limbs. Pin prick and joint position sensations both were intact. Gait was ataxic. Plantars were bilateral equivocal. Rest of the systemic examination were unremarkable. Modified Rankin Scale was 4.

Preliminary investigations include blood complete picture that showed total leucocyte count of 7010 cells/μl, hemoglobin 12.8g/dl and platelet count of 253000/μl. Serum chemistry including liver and renal function tests along with serum electrolytes and coagulation profile were normal. Hepatitis B and C screening were negative. Chest X-ray and electrocardiography were normal. Magnetic resonance imaging of brain with contrast were unremarkable. Cerebrospinal fluid routine examination showed albumin-cytological dissociation (cell count 1/μl, protein 72mg/dl). Nerve conduction studies showed abnormal sensory responses in both upper and lower limbs with intact motor responses (table 1). Electromyography (EMG) showed normal responses.

Table 1: Nerve conduction study

MOTOR NERVE	**Latency. Ms**	**Duration. ms**	**Amplitude mV**	**Segment**	**Distance mm**	**NCV m/s**
Median	Right					
Wrist	2.8	5.8	10.1	Wrist		
Elbow	7.8	6.2	9.4	Wrist-Elbow	250	50.0
Ulnar	Left					
Wrist	2.2	6.7	5.3	Wrist		
Elbow	5.8	6.7	4.7	Wrist-Elbow	200	55.6
Ulnar	Right					
Wrist	2.4	6.4	7.3	Wrist		
Elbow	6.4	6.1	8.2	Wrist-Elbow	230	58.2
Peroneal	Left					
Ankle	5.8	4.5	3.9	Ankle		
Head of Fibula	12.3	5.9	3.6	Ankle-Head of Fibula	290	45.0
Tibial	Right					
Ankle	5.0	5.7	8.7	Ankle		
Popliteal	14.9	6.4	7.0	Ankle-Popliteal	370	37.6
SENSORY NERVE	**Latency.1 Ms**	**Latency.2 ms**	**Amplitude µV**	**Segment**	**Distance mm**	**NCV m/s**
Median	Left					
Wrist	2.4	3.2	8.2	Wrist	130	53.7
Median	Right					
Wrist	2.2	3.0	3.9	Wrist	130	58.0
Ulnar	Right					
Wrist	2.5	3.0	0.9	Wrist	130	52.8

Anti- neuronal antigen antibodies panel encompassing anti- GM1, anti- GM2, anti- GM3, Anti-GQ1, anti-GT1b and anti-GD1b antibodies were negative.

Other tests including thyroid function test and serum vitamin B12 levels found to be normal. Antinuclear antibody (ANA) was negative. Erythrocyte sedimentation rate was 10/hr.

Keeping the diagnosis of variant of Guillain-Barré syndrome, immunotherapy was considered. Family and patient were counseled in detail regarding the use of intravenous immunoglobin (IVIG) and plasma exchange. Risks, benefits, pros and cons of both treatments along with expected cost were explained. They opted for plasma exchange. Five sessions of plasma exchange were performed on alternate days. During hospitalization, patient remained hemodynamically stable. General nursing care in terms of bowel and bladder care, prevention of bed sores, deep vein thrombosis prophylaxis and physiotherapy were offered. At time of discharge, her symptoms of limb and gait ataxia improved markedly with Modified Rankin Scale of 3. She was discharged on Pregabalin due to complaint of paraesthesia with referral to rehabilitation centre for physiotherapy and follow up after two weeks.

DISCUSSION

When Guillain-Barre syndrome (GBS) first manifests, some individuals have severe ataxia but not ophthalmoplegia or significant sensory loss. The condition may be a GBS variation based on distal paresthesias, areflexia, CSF albumino-cytologic dissociation, and sensory neuropathy. The majority of these people have some weakness, and as the illness worsens and becomes classic GBS, the ataxia becomes less noticeable. Richter first suggested an ataxic variant of GBS in patients who demonstrated severe ataxia without opthalmoplegia or sensory disturbance. This variant has features of distal paraesthesiae, areflexia, and increased CSF protein concentrations.[2] Ataxic Variant of GB syndrome forms a continuum with patients with Miller fisher syndrome (MFS) who present with ataxia, areflexia and opthalmoplegia, although there are also incomplete forms of MFS in which opthalmoplegia or ataxia is absent the differentiating feature from other forms of GB syndrome is that ataxia is cause of motor disability rather than true motor paralysis.[3,4] Pathology of ataxic variant isn't completely understood but autopsy revealed remarkable degradation of Clarke's column's fiber system in spinal cord.[1] Anti GQ1b antibody is a strong diagnostic marker for MFS, however only a few cases of anti-GQ1b–negative MFS cases were documented.[3] Research revealed that similar autoantibodies were also present in some individuals who had acute self-limited ataxia without ophthalmoplegia. There is a raised serum anti –GD1b IgG antibody in some cases of Guillain-Barre syndrome with cerebellar symptoms. [5]

In our case, both anti GQ1b and anti-GD1b antibodies came negative. Symptoms of ataxia improved with immunotherapy making diagnosis of seronegative ataxicvariant of Guillain-Barre syndrome without opthalmoplegia. Given that the effectiveness of IVIG and plasma exchange for GBS has been demonstrated by randomised controlled trials, both interventions may be beneficial in the treatment of ataxic GB syndrome.[6] However, as no randomised controlled trials have been conducted, their effectiveness in treating is yet undocumented.[7]

REFERENCES

1. Hughes RA, Rees JH. Clinical and epidemiologic features of Guillain-Barre syndrome. J Infect Dis. 1997;176 Suppl2:S92-8.
2. Richter RB. The ataxic form of polyradiculoneuritis (Landry-Guillain-Barre syndrome). J Neuropathol Exp Neurol. 1962;21:171-184.
3. Lee KY. Anti-GQ1b-negative Miller Fisher syndrome after Campylobacter jejuni enteritis. Pediatr Neurol. 2012;47(3):213-5.
4. Ito M, Matsuno K, Sakumoto Y, Hirata K, Yuki N. Ataxic Guillain–Barré syndrome and acute sensory ataxic neuropathy form a continuous spectrum. J Neurol Neurosurg Psych. 2011;82(3):294-9.
5. Araki T, Kusunoki S, Arai Y, Terashi A. [Guillain-Barré syndrome with cerebellar symptoms and elevated serum anti-GD1b IgG antibody]. RinshoShinkeigaku. 1999;39(5):527-30. Japanese.
6. Hughes RA, Swan AV, Raphaël JC, Annane D, van Koningsveld R, van Doorn PA. Immunotherapy for Guillain-Barré syndrome: a systematic review. Brain. 2007;130(9):2245-57.
7. Overell JR, Hseih ST, Odaka M, Yuki N, Willison HJ. Treatment for Fisher syndrome, Bickerstaff's brainstem encephalitis and related disorders. Cochrane Database Syst Rev. 2007(1).

Conflict of interest: Authors declare no conflict of interest.
Funding disclosure: Nil

Author's contribution:
Haris Majid Rajput; concept, case management, manuscript writing
Malik Muhammad Adil; case management, manuscript writing
Maryam Khalil; case management, manuscript writing
Nayab Aslam; case management, manuscript revision
Zaid Waqar; case management, manuscript revision
All the authors have approved the final version of the article, and agree to be accountable for all aspects of the work.