

Guillain-Barré syndrome with hyperreflexia precipitated by Lyme's disease

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DOI: 10.29322/IJSRP.9.06.2019.p9044

<http://dx.doi.org/10.29322/IJSRP.9.06.2019.p9044>

Abstract: Guillain-Barré syndrome (GBS), though associated with progressive motor weakness and areflexia, there have been case reports of GBS presenting as hyperreflexia and maintained reflexes. Early detection and treatment is of paramount importance as delay in diagnosis portends a poor prognosis to the patient. There is clear evidence of an association of an antecedent infectious disease associated with the development of GBS, therefore if proper diagnostic studies are not considered early in the course of treatment, the potential antecedent etiology may remain elusive. Here we report an atypical case of GBS with hyperreflexia precipitated by Lyme's disease.

Keywords: Autoimmune response; Hyperreflexia; Molecular mimicry; Neuroborreliosis

Introduction:

Guillain-Barré syndrome (GBS) is an acute autoimmune polyradiculoneuropathy, presenting as areflexic, flaccid paralysis with variable sensory disturbances, and elevated cerebrospinal fluid (CSF) protein without pleocytosis.¹ The 2 features essential for a diagnosis of GBS are progressive motor weakness and areflexia. There are 2 distinctive pathologic subtypes of GBS: demyelinating and axonal. Recently, there have been several descriptions of reflex preservation and hyperreflexia in axonal GBS in Chinese, Japanese, and European populations.^{1,2} Although this variant is not common in the Indian subcontinent, a few cases have been reported.^{3,4} A high index of suspicion is therefore needed to diagnose this rare presentation of GBS. It has been recently understood that an antecedent infection may predicate an immune response resulting in the development of GBS with *Campylobacter jejuni* and Cytomegalovirus being the commonly reported causative agents.⁵ Tick borne diseases such as *Borrelia burgdorferi*, the causative agent of Lyme have been identified as a causative agent in Lyme disease in humans. Lyme disease may have protean immune modulating manifestations involving multiple organ systems including the skin, joints, heart, and brain. Neuroborreliosis, a common presentation of CNS Lyme disease is characterized by peripheral and cranial neuropathies, which may also be a presenting symptom of GBS.⁶ This particular case, reports a possible association of *B. burgdorferi* as an inciting agent for GBS.

CASE REPORT:

A 50 year old male, non-diabetic, non hypertensive presented with the chief complaints of numbness of the palms and soles, slurring of speech since last 7 days. He had difficulty in gripping objects with his hands and difficulty in walking without support with slippage of his slippers over these last 7 days along with backache. He had a history of fever 7 days back preceding this episode of weakness which lasted for only 3 days. There were no history of any rashes or joint pain. The patient was a vegetarian by diet and had a history of occasional alcohol intake. CNS examination revealed bilateral LMN type facial nerve palsy (Fig 1), normal tone of all four limbs, power of 5/5 in all four limbs, brisk deep tendon reflexes on the right side and absent on the left side, sensations were intact and plantars were bilateral flex or other systemic examinations were within normal limits.

Investigations revealed a normal hemogram, normal electrolytes, liver and kidney function tests. Thyroid function test, Vit B12 and Folate levels were within normal limits. CXR, NCCT head and MRI brain were within normal limits, serum ACE levels were normal. A lumbar puncture was done and CSF study revealed albuminocytological dissociation with nil cells, protein-100mg/dl, sugar-70mg/dl. An NCV was done which was suggestive of decreased CMAP amplitudes of bilateral ulnar and left tibial nerves with mildly delayed distal latency with rest of the nerves being non-recordable, features suggestive of polyneuropathy, axonal > demyelinating (Fig 2). So a provisional diagnosis of GBS (AMSAN with bilateral facial nerve palsy) was

made and patient was initiated on IVIg infusion at 2g/Kg body weight over 5 days, eye padding was done while sleeping. Lyme's, Campylobacter jejuni and Cytomegalovirus serologies were sent.

Patient responded to IVIg consequently, his gait had improved, there was appearance of the deep tendon reflexes on the left side but his numbness still persisted, so did the lagophthalmos. Lyme's serology (IGM) came out to be positive while Campylobacter and CMV serologies were negative. Patient was initiated on IV Ceftriaxone 2g/day for 14 days. He showed improvement in his residual weakness, disappearance of lagophthalmos (Fig 3), his numbness had subsided as well. So this was an atypical case of GBS with hyperreflexia precipitated by Lyme's disease.



Fig 1 showing bilateral Bell's phenomenon suggestive of bilateral LMN facial nerve palsy.

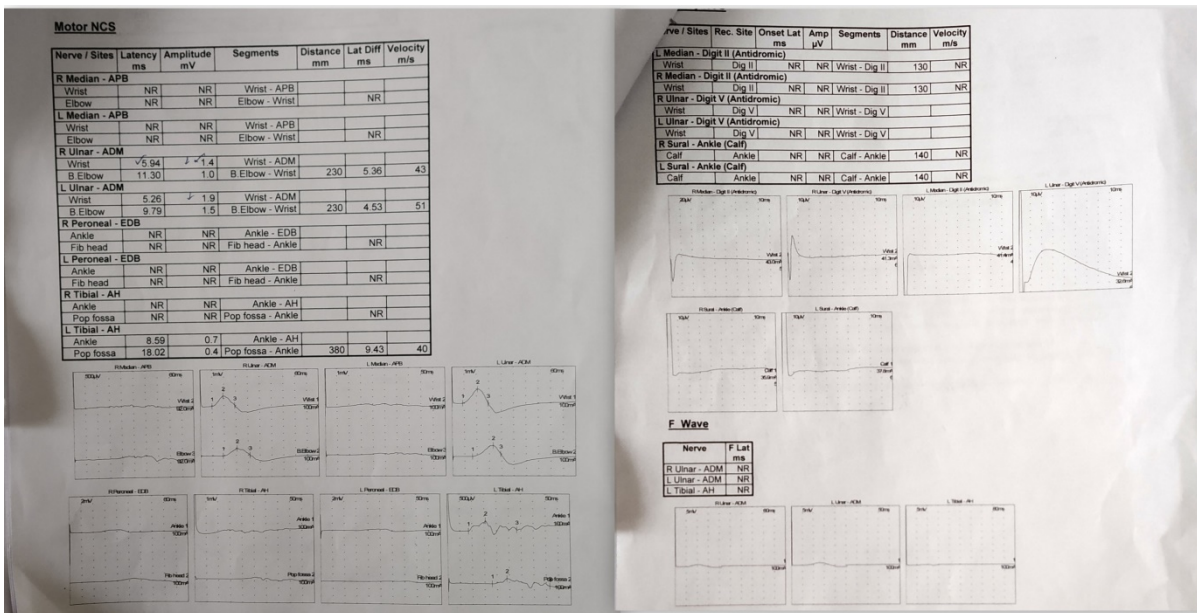


Fig 2 showing NCV of all 4 limbs showing decreased CMAP amplitudes of B/L ulnar and left tibial nerves with mildly delayed distal latency with rest of the nerves being non-recordable, suggestive of polyneuropathy, axonal > demyelinating-AMSAN variety of GBS.



Fig 3 showing disappearance of lagophthalmos on treatment.

Discussion

GBS is a heterogeneous disease consisting of many different clinical presentations to include: acute inflammatory demyelinating polyradiculopathy (AIDP), the most common type in the western world, acute motor axonal neuropathy (AMAN), acute motor and sensory neuropathy (AMSAN), as well as other less common variants.⁷ An antecedent infection has been noted in a significant number of patients with GBS with *Campylobacter* being the most commonly encountered, occurring in about two-thirds of patients prior to the start of symptoms associated with GBS.⁸ *Campylobacter* acting as an inciting agent for GBS originates from the principle of molecular mimicry which is described as a foreign antigen being perceived as a self-antigen due to the presence of similar structural units^{8,9}. It is hypothesized that when humans are infected with *Campylobacter*, autoantibodies are produced that target the *Campylobacter* ganglioside-like liposaccharides and these liposaccharides are similar to gangliosides which are important in the function of Schwann cells, that myelinate peripheral nerves^{8,9} and this results in the manifestation of GBS. It is a widely accepted hypothesis that *B. Burgdorferi* is capable of producing an autoimmune response⁹. It is therefore plausible that the spirochete or components of the spirochete associated with Lyme disease may also act as antigens or immune complexes, which facilitate production of anti-ganglioside antibodies predicated in the development of GBS in susceptible individuals although further research is warranted in this area¹⁰. Bilateral facial palsy is extremely rare with an incidence only 1 per 5 million patients and the most common infectious cause for this rare presentation is Lyme disease.¹¹ The exact mechanism that causes hyperreflexia in these cases is unknown and hyperexcitability of the motor neurons and dysfunction of the spinal inhibitory interneurons have been proposed as the possible mechanism.¹²

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