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Case Report

Internal Medicine Section

Unusual Presentation of *Legionella* as an Acute Flaccid Quadriparesis in a Case of Guillain-Barre Syndrome

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ABSTRACT

Guillain-Barré Syndrome (GBS) is a rare polyradiculoneuropathy of the peripheral nerves and, occasionally, the cranial nerves, causing dysfunction, segmental demyelination, and/or axonal degeneration. GBS is considered an autoimmune disease because the body's own immune system attacks its own tissues. There is a strong association between GBS and preceding acute infectious illness. Aetiology is unknown in most cases. GBS has been less commonly reported in association with *Legionella*. Furthermore, unusual GBS-related clinical signs might lead to confusion and incorrect diagnoses, like transverse myelitis, spinal cord compression, stroke, infections, myasthenia, and periodic paralysis. To establish the diagnosis of GBS, patient history, electrophysiological assessments, neurological evaluations, and Cerebrospinal Fluid (CSF) analyses are equally important. Hereby, the authors present a case report of 75-year-old male patient who presented with a history of fever spikes and diarrhoea 10 days prior, followed by the acute onset of ascending paralysis and hypertension. A systemic examination revealed right eye ptosis, loss of power in the bilateral lower limbs, areflexia, and diminished sensory function. Further evaluation through nerve conduction studies showed findings suggestive of demyelinating axonal sensory-motor polyneuropathy involving all four limbs. Based on the clinical suspicion of an acute infectious illness history followed by ascending paralysis, the patient was investigated for the cause of GBS, which was found to be *Legionella*. The patient was treated with plasmapheresis and physiotherapy and is now on regular follow-up.

Keywords: Albumino-cytological dissociation, Polyneuropathy, Polyradiculopathy

CASE REPORT

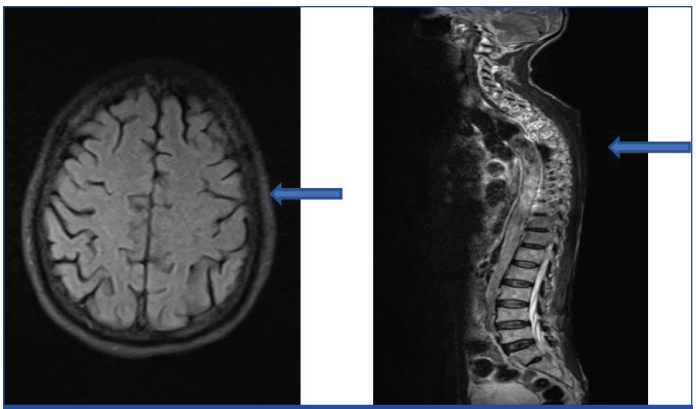
A 75-year-old male, with a history of travel to a rural area for 15 days, presented with one fever spike of 101 degrees Fahrenheit and 2-3 episodes of diarrhoea that were watery in consistency and non blood-tinged, occurring 10 days prior to admission. He reported weakness and a tingling sensation in both the upper and lower limbs, which was sudden in onset and progressive. There was no history of chest pain, palpitations, breathlessness, vomiting, headache, dizziness, seizures, loss of consciousness, or urinary and faecal incontinence. The patient had a known history of hypertension for three years and chronic obstructive pulmonary disease for five years but was not on any medication at the time.

On general examination, the pulse was 96 beats per minute, regular, and the blood pressure was 180/90 mmHg in the right arm in the supine position. Fundus examination was within normal limits. Neurological assessment revealed right eye ptosis, while the other cranial nerves were within normal limits. Flaccid quadriparesis was present. Bilateral triceps, supinator, and knee reflexes were absent, while biceps and ankle reflexes were 1+. Bilateral plantar responses were mute. Muscle power was rated at 4/5 in the bilateral upper limbs and 0/5 in the bilateral lower limbs. Sensation was diminished in both the upper and lower limbs. Nystagmus and cerebellar signs could not be evaluated, and involuntary movements were absent. Other systemic examinations were within normal limits.

The electrocardiogram suggested sinus tachycardia. Ultrasonography of the abdomen and pelvis, 2-D echocardiography, and chest X-ray were all within normal limits.

Laboratory investigations showed haemoglobin at 14.3 gm/dL, Total Leucocyte Count (TLC) at 8100/mm^3, and platelets at 2.33 lakhs. Serum sodium was 124 mmol/L, while serum potassium and chloride levels were normal. Renal function tests, liver function tests, serum proteins, and urine routine and microscopy were all within normal limits. The fasting lipid profile was normal. Serology tests for dengue, Widal, rapid malaria test, and chikungunya were negative.

The neuroimaging of the brain and spine was normal, as it was performed to rule out stroke and spinal cord compression, respectively has been depicted in [Table/Fig-1,2].



[Table/Fig-1]: Shows Magnetic Resonance Imaging (MRI) brain suggestive of chronic ischaemic changes without any acute infarct/bleeding.
[Table/Fig-2]: Shows MRI whole spine screening. No significant spinal cord compression was seen. (Images from left to right)

The CSF was sent for analysis, which suggested albumino-cytological dissociation. The protein level was 145.70 g/L, the TLC was 2 cells with lymphocytic predominance of 100%, glucose was 69 mg/dL, and adenosine deaminase was 0.8. Culture and sensitivity, cartridge-based nucleic acid amplification tests, and malignancy tests were negative. The CSF dengue Polymerase Chain Reaction (PCR), Japanese Encephalitis Virus (JEV) IgM Enzyme-linked Immunosorbent Assay (ELISA), JEV PCR, Herpes Simplex Virus PCR, Enterovirus PCR, and Myelin Oligodendrocyte Glycoprotein/Neuro Myelitis Optica tests were all negative. Urine for *Legionella* and serum *Legionella* IgM were both positive.

Nerve Conduction Velocity (NCV) studies showed findings of decreased compound muscle action potential amplitude, prolonged distal latency, and decreased conduction velocity in the bilateral median, ulnar, and peroneal nerves. Decreased compound muscle action potential amplitude, normal distal latency, and decreased conduction velocity were observed in the bilateral tibial nerves. Sensory nerve action potentials were seen in the bilateral median, ulnar, and sural nerves. F-waves were observed as impersistent and prolonged in the right ulnar nerve, prolonged in the left ulnar nerve, and absent in the bilateral median, peroneal, and tibial nerves. All these findings suggested decreased nerve conduction velocities (motor and sensory) across the nerves, indicative of demyelinating axonal sensory motor polyneuropathy involving all four limbs, likely GBS.

The patient was started on antibiotic coverage and antihypertensive drugs, including Tablet Amlodipine 5 mg twice a day and Tablet Telmisartan 40 mg twice a day. Sodium correction was administered. Five sessions of Plasma Exchange (PLEX) were conducted, along with a total of 32 pints of fresh frozen plasma transfused. Physiotherapy was initiated. The patient improved with PLEX and physiotherapy and was discharged on the same antihypertensives, along with nutritional supplementation and physiotherapy, and is currently on regular follow-up. On subsequent follow-ups, the patient showed clinical improvement with power, reflexes, and sensory systems returning to normal, along with no neurological deficits.

DISCUSSION

Globally, the incidence of GBS is 1-2 per 100,000 persons annually, with a higher frequency in males than in females. GBS is the most frequent cause of acute flaccid paralysis, characterised by symmetrical limb weakness, hyporeflexia, and areflexia [1]. Although both humoural and cell-mediated immune processes are involved in this autoimmune condition, the precise aetiology of the illness is still unknown. Numerous microbes, including viruses and bacteria, are known to cause infections associated with GBS. Several microbiological pathogens, including *Campylobacter jejuni* and *Legionella pneumophila*, have been identified as triggers. Other pathogens that have been linked to this condition include the Epstein-Barr virus, Coronavirus Disease-19 (COVID-19), cytomegalovirus, and Haemophilus influenzae [2].

In the case of *Campylobacter jejuni*, the underlying mechanism established is molecular mimicry [3]. *Legionella pneumophila* is a Gram-negative bacterium that is aerobic, non spore-forming, and flagellated; it is the primary human pathogen in the genus *Legionella* [4].

As compared to the studies conducted by Yzerman EP et al., and Vigneri S et al., present patient presented with fever, diarrhoea, and hyponatraemia [5,6]. The usual presentation of pneumophila includes fever and chills, headache, cough, myalgia, and can lead to pneumonia, with diarrhoea and signs of hyponatraemia like mental confusion, occurring very occasionally. Diagnostic testing for *Legionella* infections can be obtained using culture, serology, and molecular methods. The most commonly prescribed test for diagnosis is *Legionella* Urine Antigen Testing (UAT), which has a sensitivity of about 70-80% and a specificity of over 99%. It typically remains positive for days to weeks beyond the start of effective therapy. In our case report, urine testing for *Legionella* was used to diagnose the cause of GBS [5,6].

In our case report, the patient exhibited symmetrical limb weakness, sensory loss, areflexia, ptosis, and hypertension. In the studies conducted by Seneviratne U et al., and Alanazy MH et al., the clinical features of GBS include sensory symptoms (paraesthesia, numbness, loss of joint position sense, touch, and vibration), motor symptoms (symmetrical limb weakness, areflexia, respiratory muscle weakness, neck muscle weakness, and cranial nerve palsies), and autonomic dysfunction (fluctuations in pulse rate and blood pressure, urinary sphincter disturbances, anhidrosis, and flushing [7,8].

The major subtypes of GBS are based on clinical features, pathophysiological, and NCV findings. The Acute Motor-sensory Axonal Neuropathy (AMSAN) type of GBS was observed in our case report. However, the most common GBS subtypes include Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), which affects the myelin sheath of the nerves and leads to slowed nerve conduction; Acute Motor Axonal Neuropathy (AMAN), which primarily affects the motor nerves and involves axonal damage rather than demyelination; and AMSAN, which involves both motor and sensory nerves, as seen in the studies by Estridge R and Iskander M, and Mahesh B et al. The most commonly involved cranial nerve is the facial nerve, followed by bulbar weakness, ophthalmoplegia, and tongue weakness [9,10].

Neuroimaging is necessary to rule out focal lesions in the brain and spine. Authors found that neuroimaging revealed no gross lesions or abnormalities in the brain and spine of our patient. Based on the patient's history and presentation, spinal cord lesions were ruled out due to the absence of a discrete spinal cord lesion on spinal neuroimaging or bowel and bladder involvement. Spinal cord compression secondary to intramedullary primary spinal cord tumours, lymphoma, and leptomeningeal malignancy were excluded based on the absence of corresponding findings on spinal neuroimaging [11].

In studies by Leonhard SE et al., and Verboon C et al., treatment for L. pneumophila includes antibiotic coverage with fluoroquinolones and macrolides [12,13]. The only proven effective treatments for GBS, as reported in the studies by Leonhard SE et al., and Verboon C et al., are Intravenous Immunoglobulin (IVIG) and PLEX. In our case report, the patient was treated with PLEX. About half of patients with GBS require supportive care for an extended period, as they do not improve within four weeks after the initiation of IVIg or PLEX. Our patient improved with IVIg and other supportive care measures, such as physiotherapy. To regain strength and function, physical, occupational, and speech therapy are of utmost importance [12,13].

CONCLUSION(S)

The GBS is a rare disorder in which an impaired immune system leads to nerve damage. The atypical presentations of GBS can cause significant confusion for clinicians, potentially leading to misdiagnoses such as transverse myelitis, spinal cord compression, stroke, infections, myasthenia gravis, and periodic paralysis, among others. The present case report highlights the importance of accurate diagnosis and the timely initiation of appropriate management strategies in managing GBS, highlighting the efficacy of PLEX therapy as a first-line intervention for GBS patients. Early and prompt diagnosis is crucial, as GBS can be life-threatening, particularly with involvement of the cardiovascular and respiratory systems. Hence, frequent reassessments and early diagnosis are of extreme importance for both patients and clinicians.

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