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Case Report

Guillain Barre Syndrome

Guillain Barre Syndrome - Acute Inflammatory Demyelinating Polyneuropathy with Unusual Descending Paralysis

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Abstract: Guillain-Barré syndrome (GBS) is a rare autoimmune, acute fulminant polyradiculoneuropathy. Incidence is between 1- 4 cases per 100,000 per year. It can affect age groups such as both young and old. The distribution depending on age-specificity appears to be bimodal, and has high rates in young adults as well as the elderly. Mostly manifests as rapidly evolving areflexic motor paralysis with a usual ascending type of paralysis. Autonomic involvement is common with usual manifestations of loss of vasomotor control such as BP fluctuations, postural hypotension, and arrhythmias. Deep tendon reflexes are diminished or disappear in the first few days of disease onset. Several cases have been reported with descending type of paralysis, but they were patients of Miller Fischer syndrome variant of GBS. Only 15% of the cases were reported regarding descending type of paralysis in AIDP variant of GBS. Our patient presented with unusual descending paralysis, starting with the upper limbs and progressively involving weakness of the lower limbs. The patient did not have any comorbid illnesses rather than recent gastrointestinal infections. In the progress of the disease, he developed respiratory complications for which he needed ventilator support and with all supportive care and keen monitoring, the patient was weaned off from the ventilator, a tracheostomy done, and improved. Thus presenting a case of Guillain Barre syndrome with an unusual descending type of paralysis and with all other typical symptoms of this condition, who have been treated with high dose IV immunoglobulin, creates awareness in view of diagnosing the disease even with unusual presentation would save the life of the patient, instead presenting the usual case scenario or rare disease case.

Keywords: AIDP, Paralysis, GBS, Immunoglobulin and Reflexes

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1. INTRODUCTION

Guillain-Barré syndrome (GBS) is an autoimmune, frequently severe, acute, and fulminant polyradiculoneuropathy. Its incidence is between 1- 4 cases per 100,000 per year; in US approximately 5000–6000 cases annually. Males are at higher risk than females, and likewise in Western countries, adults are at higher risk than children.[1] GBS has been identified in families in a few European and Asian countries (42 GBS patients found from 20 families). In a South Indian hospital-based study of 150 GBS cases recorded over ten years, two cases belonged to the same family[2]. The incidence and the GBS risk is increasing progressively in the urban centers than in rural zones in Northern Italy as per studies.[3] The most common are respiratory infections, which are identified in about 40% of cases within one month of the onset of the disease. Gastroenteritis is the antecedent cause in around 20% of cases.[4] It can also be triggered by any surgery or vaccine administration. The initial symptoms are weakness and tingling sensations usually starting in the legs and can involve upwards to arms and face.[5] Neurological examination including diminished or absent deep tendon reflexes. A lumbar puncture and CSF analysis can be done. Electromyography and nerve conduction studies are being done to diagnose this condition. Most of the patients need hospitalization and around 30% of the patients need ventilatory support. Once the patient's clinical worsening stops, the disease's progression is unlikely. Guillain Barre syndrome is ascending type of paralysis. In 10–15 percent of cases, descending paralysis develops, with symptoms starting in the cranial nerves or arms and spreading to the legs.[6] Immunoglobulin given intravenously (0.4 gram per kilogram body weight for five days) is also the recommended treatment for GBS. It is a relatively safe treatment, but there are some side effects and contraindications to be aware of. Contraindications following a prior intravenous immunoglobulin infusion, anaphylaxis may occur due to selective IgA deficiency. While there is no cure for Guillain-Barre syndrome, it can be sped up by using plasma exchange, a high dose of intravenous immunoglobulin to suppress the destructive antibodies that cause the disease. Anticoagulants are used to avoid thrombosis, and pain relievers are also used. Several factors affect the prognosis, including the aetiology, clinical characteristics, electrophysiology, and biochemistry. However, the initial finding of non-excitable nerves and decreased compound muscle action potentials has been linked to a poor prognosis. Atypical AIDP presentations, such as the one described in this case study, can cause delays in diagnosis and care. It is important to remain aware of the existence of atypical manifestations and their variations to avoid delays in the care of patients with AIDP. Thus, presenting a case of Guillain Barre syndrome with an unusual descending type of paralysis and all other typical symptoms of this condition, which has been treated with high dose IV immunoglobulin, creates awareness in view of diagnosing the disease even with unusual presentation. We have addressed the presenting signs, progression of the condition, diagnosis, treatment procedures, and the patient response to the given treatment in this case of GBS with acute inflammatory demyelinating polyradiculopathy with a rare descending type of paralysis.

2. CASE REPORT

A 21-year-old male presented with complaints of weakness of left upper limb followed by lower limb for 2 days, which was sudden in onset, left upper limb weakness started from medial 2 digits to other regions. And the weakness gradually progressed to right upper limb, and also developed slight weakness in the right lower limb. History of loose stools 1 week back. Patient does not have a history of fever, bladder/ bowel incontinence. Patient had no comorbidities.

3. PAST MEDICAL HISTORY

Not a known case of Diabetes mellitus, Hypertension, Coronary artery disease, Bronchial asthma, Chronic kidney disease, Epilepsy and Tuberculosis. Not on any drug, not a smoker/alcohol consumer. No history of previous surgeries. No previous significant hospital admissions.

4. EXAMINATION

On examination, the patient was moderately built. On Cardiovascular system examination, S1 S2 were normal, no murmurs, no split were heard. Respiratory system examination revealed equal bilateral air entry, no added sounds. Abdomen was soft, non-tender, not distended. On Central nervous system examination, the power of Left upper limb and lower limb as 0/5 and 0/5. And Right upper limb and lower limb power was 0/5 and 2/5 respectively. Hypotonia on all four limbs. Bilateral plantar reflex was non-responsive. Deep tendon reflexes were absent.

5. INVESTIGATIONS

Nerve conduction studies were done and it showed bilateral severe sensorymotor polyradiculoneuropathy (demyelination changes). MRI whole spine screening was done and it showed a mild diffuse disc bulge with central disc protrusion indenting the anterior thecal sac at C3-C4 and C4-C5 levels, mild posterior disc bulge seen at L4-S1 level. Furthermore, all the necessary blood investigations, such as negative viral markers, blood culture sensitivity, no growth, coagulation activity markers, normal values, complete urine analysis, normal, urine culture sensitivity, no growth, and renal function test within normal limits were done. Complete blood showed total counts- 18030, neutrophils-86.7%, lymphocytes- 8.7%, eosinophils – 0.0%, monocytes -3.7%, and basophils 0.3% respectively.

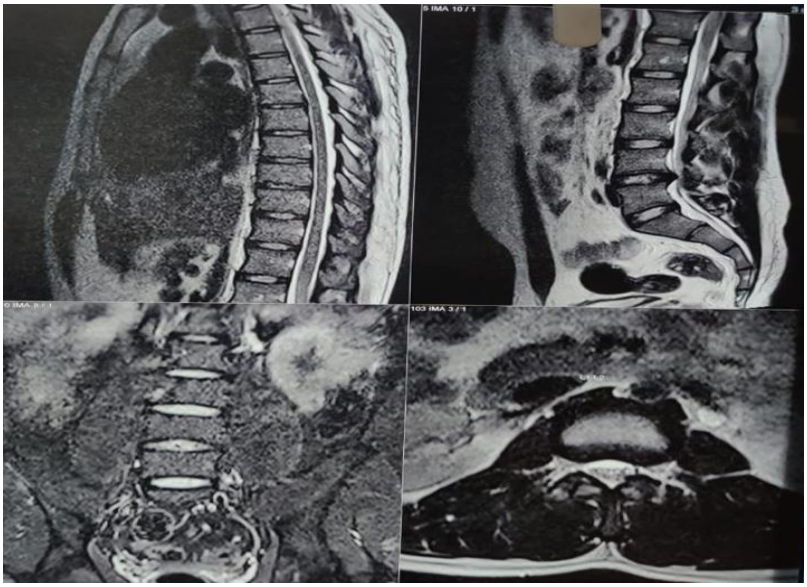

Fig 1: Illustrates the disc bulge over the L5- S1 vertebrae in the MRI dorsal lumbosacral spine of the patient.

6. TREATMENT

The patient was started with high-dose intravenous immunoglobulins, anticoagulants (to prevent thrombosis during immobility), antibiotics, antihypertensives, and other supportive measures. The patient suddenly went into supraventricular tachycardia and intravenous adenosine was given and reverted. The patient general condition worsened and desaturated , for which he was intubated and mechanical ventilatory support was given. The patient was continuously monitored under intensive care. After three days, with the guidance of the Otorhinolaryngology team, an elective tracheostomy was done and their supportive care orders were followed. Patient condition was symptomatically improving, and power of the right upper limb and lower limb was 3/5 and 4/5 and of the left upper limb and lower limb was 3/5, respectively. Spigotting was initiated, and decannulation was done. The patient condition improved, and he was discharged.

7. FOLLOW UP

The patient is on regular follow-up every three months from discharge, and has no new complaints and complications further.

8. DISCUSSION

Guillain-Barre syndrome is a rare disease caused by the body's immune system attacking a peripheral nervous system component. Several cases have been reported with descending type of paralysis, but they were patients of Miller Fischer syndrome variant of GBS. Only 15% of the cases were reported regarding descending type of paralysis in AIDP variant of GBS. One such case was reported by Dr. Ala Mustafa from Mercy one North owa medical center, Mason city, IA, USA. According to epidemiological research from Europe the annual incidence in Australia and United States of the disease Guillain-Barré syndrome is around 1–3 for one lakh people.[1] GBS has been identified in families in a few European and Asian countries (42 GBS patients found from 20 families). In a South Indian hospital-based study of 150 GBS cases recorded over ten years, two cases belonged to the same family[2]. It can affect age groups such as both young and old. The distribution depending on age-specific, appears to be bimodal, which has high rates in young adults as well as the elderly.[3] The syndrome may affect the nerves that control muscle movement and transmit pain, temperature, and touch sensations. This can cause muscle weakness and loss of feeling in the legs and/or arms. In about two-thirds of patients, symptoms are accompanied by an antecedent condition. The most common are respiratory infections, which are identified in about 40% of cases within one month of the onset of the disease. Gastroenteritis is the antecedent cause in around 20% of cases[4]. It can also be triggered by any surgery or vaccine administration. The number of Guillain-Barre syndrome cases has risen in some way linked to Zika virus infection. Guillain-Barre syndrome is provoked by Zika virus infection, according to the theory. Infections with campylobacter, influenza, cytomegalovirus, Epstein Barr virus, Hepatitis virus, and covid 19 are all risk factors. Variations in pulse and blood pressure most often manifest dysautonomia. Acute or subacute symptoms may appear at any time. After a plateau, there is a gradual return to normalcy. It was also discovered that by four weeks after the onset, 98 percent of patients had reached the plateau level. [5] Acute motor axonal neuropathy or acute motor–sensory axonal neuropathy are more common in patients with C jejuni, whereas the variant of AIDP is less common. Symptoms normally last a few weeks, and most patients recover without any serious long-term neurological consequences. The first signs and symptoms are generally fatigue and tingling sensations in the legs, which can spread to the arms and face. The inability to walk or climb stairs is also a symptom, as is extreme crampy pain that worsens at night, an elevated heart rate, and low or high blood pressure. Chest muscles are also impaired in a small number of individuals, causing breathing difficulties. It can cause paralysis of the legs, arms, and facial muscles in some people. The ability to swallow and speak is also impaired in extreme cases, which are considered life-threatening and should be treated in an intensive care unit. The most common form of Guillain-Barré syndrome is acute inflammatory demyelinating polyradiculoneuropathy (AIDP). During the early stages, T cells have been seen infiltrating the endoneurium. [6] Acute Motor Axonal Neuropathy (AMAN)- action potential amplitudes of the compound muscles are decreased

electrophysiologically, but motor conduction velocities, F waves and nerve action potentials of sensory system are all within normal limits. Wallerian-like degeneration of motor axons has been found primarily in necropsy studies. [7,8]. Tendon reflexes may be preserved or exaggerated, with the latter being more common in AMAN cases. Around one-third of patient's experience hyperreflexia, which normally occurs during the early stages of rehabilitation and on rare occasions during the acute period. This result linked to the existence of anti-GM1 antibodies and the disease's severity. Degeneration of the axon was seen in one of the cases, but there was no demyelination or inflammation. [9] In Guillain-Barré syndrome, this has the immune-mediated axonal injury which is more serious. Miller Fisher Syndrome (MFS) is a GBS variant characterized by ophthalmoplegia (internal and external) and the other 2 features such as areflexia and ataxia—the classic triad of signs. MFS may also cause weakness of the limbs, bulbar and facial palsy and also ptosis. About a minimum of 5 percent of patients with GBS have this condition.[10] Axonal losses may be a secondary event, particularly in severe cases. Both humoral and cellular immunity seems to play a role in these pathological changes to varying degrees. Segmental demyelination is reflected in electrophysiological characteristics. Recovery is linked to subsequent remyelination. Clinical, laboratory and electrophysiological characteristics have been used to develop diagnostic criteria for Guillain-Barré syndrome.[11] The presence of progressive motor dysfunction and areflexia is a must for diagnosis. The only laboratory criterion is cerebrospinal fluid analysis. A high or increasing trend of levels of protein in the lumbar punctures consecutively, as well as 10 or less mononuclear cells per mm3, will confirm the diagnosis in CSF. Pleocytosis of the CSF is well-known in HIV associated GBS. When electrophysiological results are uncertain, magnetic resonance imaging may help make a diagnosis. It is a sensitive test but its non-specific. Guillain-Barré syndrome tends to be closely associated with selective anterior root enhancement. The cauda equina nerve roots were found to be enhanced in 83 percent of patients in a study.[12] Within first 2 weeks of the disease, plasma exchange is being started, which cuts down on hospital stays, time being on mechanical ventilation, and ambulatory time. Hypotension, septicemia, hypocalcaemia, and excessive clotting are all risks associated with plasma exchange. It may be particularly dangerous in patients who are hemodynamically unstable[13]. Immunoglobulin given intravenously (0.4 gram per kilogram body weight for 5 days) is also the recommended treatment for GBS. Contraindications following a prior intravenous immunoglobulin infusion, anaphylaxis due to selective IgA deficiency. Anticoagulants are used to avoid thrombosis, and pain relievers are also used. In this state, physiotherapy is also essential for a quick recovery, overcoming exhaustion, mobility, muscle endurance, and regaining strength. Several factors affect the prognosis, including the aetiology, clinical characteristics, electrophysiology, and biochemistry. Other researchers were unable to discover such a predictive value. It should be remembered, however, that the initial finding of non-excitable nerves and decreased compound muscle action potentials has been linked to a poor prognosis. [13]

9. CONCLUSION

Guillain Barre syndrome is a rare autoimmune condition with acute and fulminant polyradiculoneuropathy with a usual ascending type of paralysis. In 10–15 percent of cases, descending paralysis develops, with symptoms starting in the cranial nerves or arms and spreading to the legs. Tendon reflexes may be preserved or exaggerated, with the latter being more common in AMAN cases. Around one-third of patient's experience hyperreflexia, which occurs typically during the early stages of rehabilitation and on rare occasions during the acute period, which is essential to be excluded in the case of GBS. Atypical AIDP presentations, such as the one described in this case study, can cause delays in diagnosis and care. Therefore, it is important to remain aware of the existence of atypical manifestations and their variations in order to avoid delays in the care of patients with AIDP. We have addressed the presenting signs, progression of the condition, diagnosis, treatment procedures, and the patient response to the given treatment in this case of GBS with acute inflammatory demyelinating polyradiculopathy with a rare descending type of paralysis.

10. AUTHORS CONTRIBUTION STATEMENT

Dr. V. Padma, Professor in the Medicine department provided us a keen guidance in the manuscript design, concept and motivated us in publishing the article in order to educate the rare scenario. Dr. Sathyapriya and Dr. Shiny, senior residents supervised the whole work and helped in final drafting of the article. My colleagues Dr Sarath and Dr Saketh lended me support in collecting necessary data from the patient.

11. CONFLICT OF INTEREST

Conflict of interest declared none.

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