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CASE REPORT ON GUILLAIN BARRE SYNDROME - ACUTE MOTOR AXONAL NEUROPATHY (AMAN)

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ABSTRACT: Guillain-Barre syndrome occurs when the body's immune system targets a portion of the peripheral nervous system. In addition to the nerves that convey pain, temperature, and touch sensations, the condition can also impair the nerves that regulate muscular movement. This may lead to muscular weakness and/or loss of feeling in the arms and/or legs. A 12-year-old boy was taken to the hospital with chief complaints of Weakness in bilateral upper and lower limbs, trouble in swallowing, inability to hold neck, mouth frothing, fever spikes. On physical examination, the patient has experienced weakness in bilateral upper and lower limbs, bulbar weakness is present, pain experiencing in both legs, S1 and S2 sound are present, Air entry is bilaterally equal, pupils are reflected to light, tone and power of upper and lower limbs are decreased", then treatment was initially started with immunoglobulin therapy, intravenous fluids, antibiotics, amino glycosides, glucocorticoids, anti-allergic as per physician orders. In this study, we primarily focus on professional management and summarize the clinical aspects of GBS, its current and immunotherapy which primarily focus on professional management.

INTRODUCTION: An extremely severe autoimmune fulminant polyradiculoneuropathy known as "Guillain-Barre syndrome (GBS)" causes this. "Guillain-Barre Syndrome" is the most common cause of acute or sub-acute widespread paralysis, and it was once only second to polio in terms of prevalence. Acute inflammatory demyelinating polyneuropathy (AIDP) and Landry-Guillain-Barre Strohl syndrome are two other names for "Guillain-Barre syndrome." According to estimates, there are between 1.3 cases per 100,000 people worldwide each year. Males are over 1.5 times as likely as women to suffer damage.

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90% of cases of Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP) in North America and Europe have this subtype [1]. The most prevalent cause of neuromuscular paralysis is Guillain-Barre syndrome. Subtypes of Guillain-Barre syndrome are acute inflammatory demyelinating polyneuropathy (AIDP) and acute motor axonal neuropathy (AMAN). The most prevalent symptoms are paralysis and areflexia, a weakening of the limbs. Miller Fisher Syndrome is one of the most common disorders and is similar to "Acute Motor Axonal Neuropathy and Acute Inflammatory Demyelinating Polyneuropathy." This sickness is immune-mediated.

This is treated by plasma exchange and immunoglobulin [2]. Guillain-Barre syndrome require close monitoring for disease progression, in particular for bulbar weakness, respiratory insufficiency, and autonomic dysfunction. Prognostic scales have been developed to predict patient outcome and to stratify treatment.

To date, intravenous immunoglobulin and plasma exchange are the only recognised immunotherapeutic drugs that can accelerate recovery in Guillain-Barre syndrome [3].

Case Presentation: 12 years old boy presented with history of prodromal symptoms and developed weakness of distal fingers of upper limb followed by lower limb of flue weakness of shoulder flue. Inability to speak breathing difficulty. He was initially treated like, pneumonia with acute RF, chest x-ray, right lower limb infiltration. He was extubated but couldn't maintain saturation and patient was reintubated on Ventilator support. Patient was then suspected to have GBS. Given IVIG totally 2gm/kg over 5 days. Administration of immunoglobulin therapy, intravenous fluids, antipyretic, multivitamins, potassium, antibiotics, amino glycosides, glucocorticoids, anti-allergic as per physician orders.

Physical Examination: On physical examination, the patient has experienced distal weakness of upper limbs which was followed by weakness of proximal muscles of upper limbs and inability to speak with breathing difficulty, In cardiovascular system, S1 and S2 sound are present, power of both upper and lower limbs are reduced and then treatment was started as soon as possible.

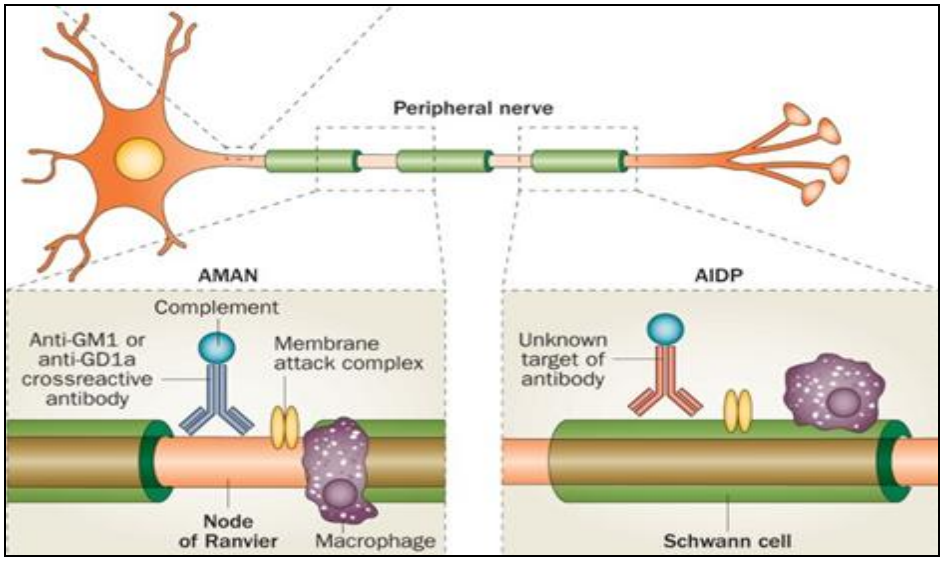
Diagnostic Assessment: Blood test: Hb-9.8%, Total RBC count-5.11 millions/ cu.mm, total WBC count- 11890/cu.mm, Total platelet Count-6.21l acs/cu.mm. Creatinine-0.44mg/dl, sodium-137 mmol/L, potassium-4.47mmol/L and phosphorous-104 mmol/L. He persisted to have tachycardia, which was initially attributed to autonomic instability in GBS. Since he developed fever spikes. Blood culture was sent. Initial set of blood culture was sterile. In view of persistent fever spikes, with worsening of distress and production of thick tracheal secretions, BAL culture was sent and started on IV antibiotics. BAL culture showed growth of GNB. Currently he is undergoing physiotherapy and adequate nutritional support and so discharged home. In view of respiratory distress, he was started on HFNC and later intubated and mechanically ventilated in view features of respiratory failure on 30/12/23. He was extubated on 2/1/23 which was not tolerated and reintubated within few hours. Due to a suspicion of GBS, lumbar puncture was done which showed features suggestive of Guillian Barre Syndrome. He received one dose of lvlg. During the hospital stay, he developed VAP (Ventilator associated pneumonia) which was treated with IV antibiotics. He was shifted to PICU for further care on 16/01/23. On arrival, he was hemodynamic ally stable, sensorium - normal. Hence a trial of spontaneous breathing was attempted. He failed in view of poor respiratory efforts. Nerve conduction studies showed features of AMAN (Acute motor exonal neuropathy). He advised to do plasma exchange. On inserting vascath, he developed right hem pneumothorax with features of obstructive shock. ICD insertion was done and 300ml of serosanguinous fluid was drained. After 48 hours of hemodynamic stabilization, he underwent plasmapheresis. A total of 5 cycles of plasma exchange was done. Procedure was uneventful. There was minimal improvement in the muscle power. MRI brain and spinal cord screening showed features of GBS. He was given second dose of lvIg. He was improving well in spontaneous and HME filter with regular breathing exercises.

DISCUSSION: The mechanism by which immune checkpoint inhibitors induce Guillain-Barre syndrome is not well understood, but it is possible that the abrogation of self-tolerance could activate cytotoxic T lymphocytes, along with a reduced suppression of antibody-producing B lymphocytes. Notably, neurological complications following immune checkpoint inhibitors do not have autoantibodies associated with the related conditions, suggesting a T-cell-mediated pathogenesis [4].

Acute mortality in GBS was mostly due to the poor respiratory care of patients and infective complications, but disability and probably late mortality were due to the axonal type involvement of nerve injury. Arami MA *et al.*, found a higher incidence of GBS in our society in the cold seasons, but the study period was too short to determine an absolute seasonal correlation with the prevalence of GBS [5]. Viral infections are very common in the community and close contacts are a usual route for epidemics of upper respiratory tract infections [6].

Currently, intravenous immunoglobulin (IVIg) and plasma exchange are proven effective treatments for GBS. Immunoabsorption, as an alternative to plasma exchange, is occasionally used as a treatment for patients with GBS, and may be equally effective [7]. IvIg treatment may inhibit Fc-mediated activation of immune cells, binding of antiganglioside antibodies to their neural targets or local complement activation [8]. Serum IgG Fc glycosylation in patients with GBS seems to be associated with disease severity and could influence the immunomodulatory effects of IVIg [9]. Plasma exchange is thought to remove neurotoxic antibodies, complement factors and other humoral mediators of inflammation [10]. The choice of treatment depends on both patient-related and socioeconomic factors. For instance, plasma exchange requires special equipment and is not always available in all hospitals.

In addition, plasma exchange can be difficult to perform in young children, and care should be taken in patients with autonomic cardiovascular instability because of the large volume shifts involved in the plasma exchange procedure. However, the direct costs of IvIg treatment can be more than twice those of plasma exchange, making this treatment less attractive in low-income countries. It is imperative to consider the role of the blood-nerve barrier in GBS pathogenesis and therapeutic development, and we advocate pathogenic leukocyte trafficking as a biologically relevant mechanistic target with translational potential for disease-specific immune modulatory therapy using function-neutralizing antagonists such as humanized monoclonal antibodies that do not require blood nerve barrier permeability and retention within peripheral nerve/ nerve root endoneurium [12].



CONCLUSION: Current immunotherapy for GBS is limited to ameliorating the disease via non-specific systemic pathogenic antibody modulation by PE and IVIg. It is imperative to consider the role of the blood-nerve barrier in GBS pathogenesis and therapeutic development. Immunoglobulin IV, plasma exchange and palliative care are all options for treatment. Assisted ventilator and tracheostomy may be needed in some patient with respiratory paralysis. Where muscle weakness persists after the acute phase of the illness, patients may require rehabilitation services to strengthen their muscles and restore movement. But treatment with autoimmune nature of the disease, its acute phase is typically treated with immunotherapy, such as plasma exchange to remove antibodies from the blood or intravenous immunoglobulin. When IVIG is given "early in the course", it improves recovery. Plasmapheresis may be a cost-effective treatment option for patients who have had a poor response to IvIg.

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CONFLICTS OF INTEREST: None declared

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