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Case Series of Guillain Barre Syndrome with Plasmaparesis in The ICU Bandung Hospital

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ABSTRACT

Background: Guillain-Barré syndrome (GBS) is an autoimmune disease of the peripheral nervous system, which is clinically characterized by tetraplegia with or without sensory disturbances. Manifestations include weakness, progressive muscle areflexia and can cause weakness in the respiratory muscles. This causes sufferers to need mechanical ventilation assistance. ICU complications such as lung infections and sepsis occur if treatment is prolonged. Plasmaparesis in GBS is a therapy other than IVIG (IntraVenous ImmunoGlobulin) administration.
Case report: We compared 2 cases, 42 years old (man A) and 31 years old (man B), with respiratory failure due to GBS were treated with plasmaparesis for 5 cycles. Patient A showed faster clinical improvement results. It took 6 days to get off mechanical ventilation on patient A, while on day 11 patient B still needed mechanical ventilation, with persistent tetraparesis.
Conclusion: This case highlights the importance of using TCD and NIRS in the intensive care unit as guiding therapies in maintaining patient blood pressure, administering blood components, and early detection of complications such as cerebral vasospasm.

Keywords

Guillain Barre Syndrome, IntraVenous ImmunoGlobulin (IVIG), Plasmapheresis

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INTRODUCTION

Guillain-Barré syndrome (GBS) is one of the most common autoimmune diseases affecting the peripheral nervous system in the world.[1] The incidence of GBS disease worldwide is recorded at 1.3-4 per 100,000 cases every year.[1,2] GBS disease is more often occurs in the age range of young adults and the elderly (>50 years). Approximately 75% of the GBS population are men.[2] However, Indonesian region there is currently no specific registered data regarding the incidence of GBS. This disease is characterized by acute neuromuscular paralysis that ends in respiratory failure and approximately 25% of patients require mechanical ventilation assistance and treatment in the ICU.[1-3]

GBS generally occurs after the patient experiences a triggering infection such as an upper respiratory tract or digestive tract infection. Some of the pathogens that cause infection before GBS occur are C. Jejuni, followed by CMV, Epstein-Barr virus, Mycoplasma pneumoniae, Haemophilus influenzae, and influenza virus type A.1 In addition, vaccination, surgery, and trauma have been reported can trigger GBS.[3]

CASE DESCRIPTION

We present 2 cases report of a patient with GBS who experienced respiratory failure requiring treatment in the ICU. During treatment in the ICU the patient experienced significant clinical improvement with the choice of 3 cycles of plasmaparesis therapy so that the patient could be discharged from the ICU, however, different result were obtained in other patient.

Case A

A 42 year old man came to our hospital with complaints of weakness in four limbs. The patient was a referral from Hospital A Cimahi with a diagnosis of GBS, because the patient's Erasmus GBS Respiratory Insufficiency Score (EGRIS) was 4 and required further treatment in the ICU, the patient was referred to the main hospital. Complaints of not being able to move all four limbs started 4 days before entering the hospital. The underlying complaint was weakness in the limbs since the previous 4 days and difficulty swallowing. Ten days before treatment the patient had a history of upper respiratory tract infection. The weakness was felt to gradually get worse starting from the legs which then went up followed by weakness in both arms, the patient had difficulty walking and holding objects, complaints of nasal speech had also been reported since the previous 4 days.When the patient arrived at our hospital, the patient was fully conscious but there was an increase in respiratory effort where the respiratory rate was 36x/minute and oxygen saturation was 92% with the use of 15 L/m non-rebreather oxygen. Assessed as respiratory failure et causa GBS accompanied by leukocytosis at 11,870/uL. Our patient underwent intubation and mechanical ventilation. Management continues with broad spectrum antibiotic prophylaxis, gastric ulcer prophylaxis, with fluid and nutritional needs met according to the patient's profile. Then the patient also undergoes supportive therapy for thromboembolism prophylaxis, namely early physiotherapy.

On the first day of treatment in the ICU we gave continuous midazolam sedation at a dose of 5 mg/hour IV and antibiotics with Ceftazidime 3x2 gr IV and Levofloxacin 1x750 mg IV, we planned for the patient to receive plasmaparesis for GBS management. Treatment in the ICU continued and monitoring of the patient's chest X-ray showed it was within normal limits. IVIG administration was not carried out.

On days 3-5 of ICU care, the patient received 3 cycles of plasmaparesis therapy and achieved significant improvement in respiratory parameters and weaning from mechanical ventilation was successful. The patient was successfully removed from mechanical ventilation on the 5th day of ICU treatment, and then moved to the HCU room for further therapy.

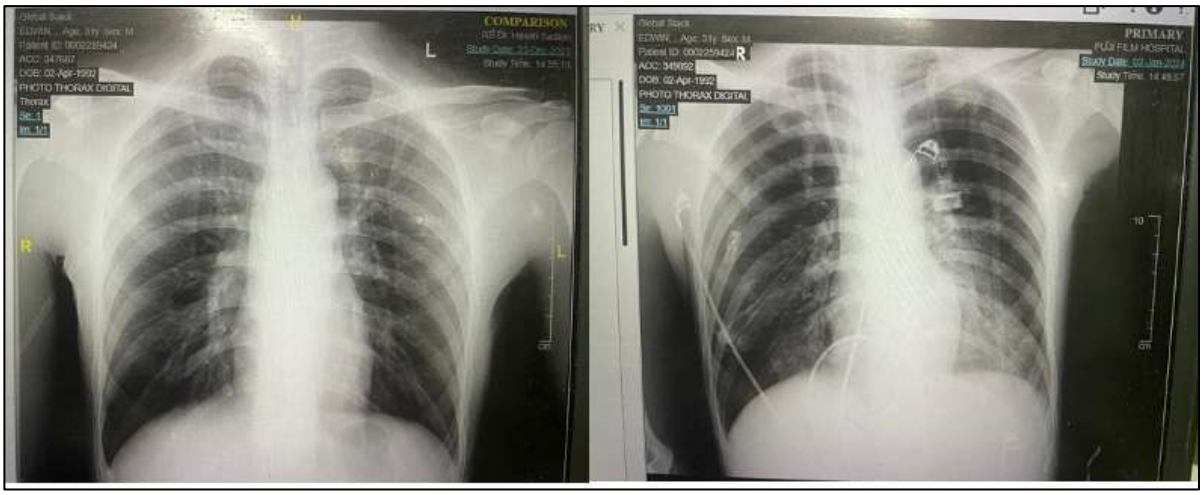

Figure 1. Comparison of thorax photos of patient B on treatment days 1 and 9 which showed the impression of pneumonia

Case B

A 31 year old man came to the hospital with complaints of weakness in four limbs. The patient is a referral from Al-Ihsan Hospital. The presence of distal paresthesia supports the possible diagnosis of GBS. Both legs and arms since 2 weeks before entering the hospital swallowed and vomited when eating or drinking, then the patient had difficulty expelling phlegm with increased saliva production, the patient also experienced difficulty breathing so the patient was intubated followed by mechanical ventilation, and treatment was carried out in the intensive unit. The patient's complaints were not accompanied by fever, weakness on one side of the body, blurred vision, seizures or previous micturition and defecation disorders. A history of aspiration after eating was also obtained in this patient. The patient was referred to RSUP for further intensive treatment.

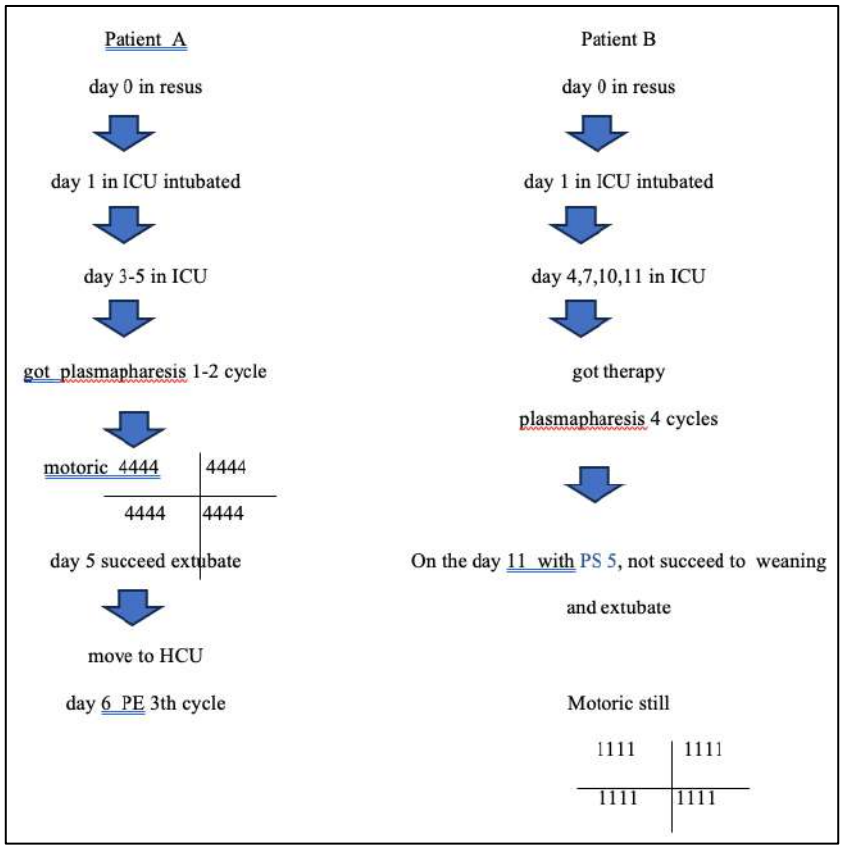

Figure 2. Timeline comparison of patients A and B during ICU treatment

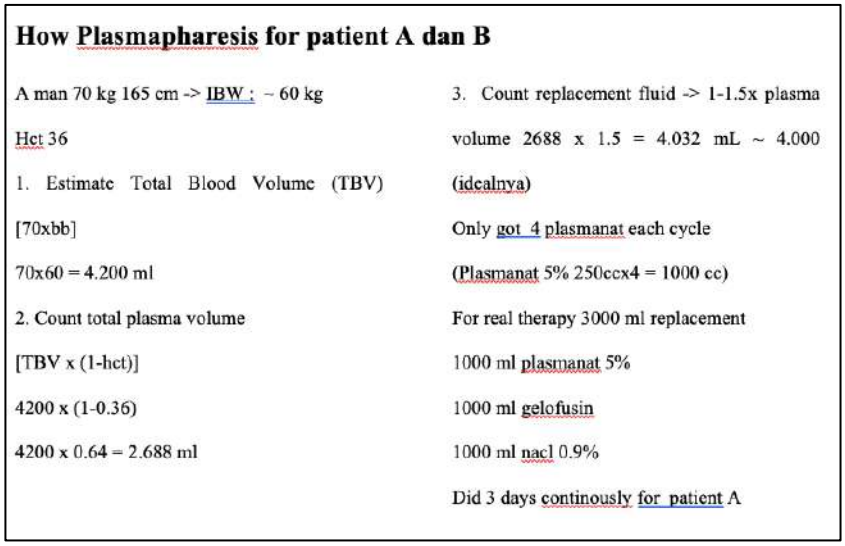

Figure 3. How Plasmapharesis for patient A dan B

On physical examination, vital signs were stable, the patient did not require vasopressor or inotropic drugs during treatment in the intensive unit. During treatment, the patient received broad spectrum antibiotic therapy; Ceftriaxone 1 gr/12 hours, with escalation of Cotrimoxazole 960 mg/24 hours on the 7th day of treatment according to culture, the patient also received gastric ulcer prophylaxis Omeprazole 40 mg/24 hours. The patient's fluid and nutritional needs are met during intensive treatment, both parenterally and enterally. On supporting examination, there was no electrolyte imbalance or disturbances in other organ systems. The patient also underwent physiotherapy to prevent thromboembolism. During the third serial PE, the patient's hemodynamics were stable until on the 11th day. Tetraparesis persisted in this patient. The patient was planned for tracheostomy due to expected long treatment with a ventilator.

DISCUSSION

Guillain-Barré syndrome (GBS) is a type of peripheral neuropathy and is the cause of acute flaccid paralysis which is characterized by symmetrical limb weakness, hyporeflexes or areflexes and increased cerebrospinal fluid protein without accompanying cellular reactions (pleocytosis) which can become increasingly severe in 4 weeks.[1,4] GBS is an inflammatory disease of the peripheral nerves and is the most common cause of acute flaccid paralysis, with an annual global incidence of approximately 1-2 per 100,000 people annually.[1-5] GBS occurs more frequently in men than women and the incidence of GBS is increasing with age, although all age groups can be affected. This syndrome is classified as an autoimmune which is characterized by the presence of autoimmunity which plays a role in its pathogenesis.[6] GBS is an autoimmune disease of the peripheral nervous system that causes progressive muscle weakness and areflexia. Upper respiratory tract and gastrointestinal infections that occur 2 weeks before onset of weakness are the two main diseases that trigger GBS.[6,7]

The difference in clinical results for patients A and B was obtained due to the different types of polyneuropathy disease progression. Patient A came with milder motor complaints than patient B, so the clinical improvement response to 3 cycles of plasmaparesis therapy was also different. Other comorbidities can also influence these differences in outcomes. In patient B there was no history of previous infections such as respiratory or gastrointestinal infections which were found in patient A. Prolonged ventilation in patient B was associated with a poor prognosis for GBS, however patients who required prolonged ventilation may show a slow but continuous recovery during years before achieving the ability to walk independently.

Autoimmunity is an immune response against self-tissue antigens involving T cells, macrophages, various cytokines and chemokines, autoantibodies, and complement. In GBS, there are autoantibodies formed by B cells and attacking ganglioside antigens in peripheral nerves which are similar to the cell membrane components of the *C. Jejuni bacteria. Guillain-Barré syndrome (*GBS) is a post-infectious disorder. Two-thirds of patients reported symptoms of respiratory or gastrointestinal infections before the onset of GBS. About half of GBS patients experience a specific type of infection. The occurrence of GBS was preceded by an infection an average of 6 weeks previously, two thirds of cases were preceded by an infection of the respiratory tract or gastrointestinal tract, most often caused by *Campilobacter jejuni*.[1-3] Many infections have been associated with GBS, the most common being gastrointestinal and respiratory diseases.[6-10]

Every year, it is estimated that 100,000 patients worldwide will suffer from GBS, with a quarter of sufferers experiencing respiratory problems. Respiratory failure caused by respiratory muscle weakness is a major complication in 20-30% of patients. The onset and progression of GBS is rapid and almost 80% of patients become unable to walk, 20% experience total paralysis requiring mechanical ventilation, 5-10% die and 12% of sufferers cannot walk after 1 year.[1,2,6]

Typical symptoms of Guillain-Barré syndrome (GBS) are bilateral, symmetrical, progressive, rapid motor weakness and a combination of areflexia or hyporeflexia. Weakness is classically described as ascending and usually begins in the distal lower extremities, but can begin more proximally in the legs or arms.[11-13] GBS is highly variable with respect to the presence, distribution and extent of cranial nerve deficits, sensory symptoms, weakness, ataxia, pain, autonomic dysfunction, and disease course. About half of patients have cranial nerve deficits, especially bilateral facial weakness, difficulty swallowing or sometimes extraocular motor dysfunction.[6,7]

The diagnosis of GBS is based on the patient's history and neurological, electrophysiological and cerebrospinal fluid examination. A fairly high percentage (54-89%) of patients with GBS experience pain, including painful paraesthesia, back pain, and muscle pain that can even precede the onset of muscle weakness. Approximately 25% of patients experience respiratory insufficiency requiring mechanical ventilation.[1,3,6]

Specific management of GBS includes TPE and intravenous immunoglobulin (IVIG). If the patient becomes unable to walk without assistance or if he shows a significant reduction in vital lung capacity or signs of severe oropharyngeal weakness, TPE or IVIG should be performed immediately. This usually occurs on the

fifth to tenth day after the appearance of the first symptoms but may be as early as one day or as late as 3 weeks from the initial onset.

Plasma exchange therapy is the first treatment proven to be effective for GBS, if given within 4 weeks of the onset of weakness, but the effect is most optimal when started early (within 2 weeks). TPE procedure requires the placement of an intravenous line in one arm to withdraw blood, and a second line in the other arm for blood return. This procedure takes 2-3 hours. For patients with GBS, three to five cycles of TPE are performed over a period of 7-14 days with a total exchange rate of 120-200 mL/kg (40-50 mL/kg/day).[6-8]

Intravenous Immunoglobulin (IVIg) is derived from the pooled plasma of 10,000 to 100,000 blood and plasma donors. Intravenous immunoglobulin may contain more than 10,000,000 specific antibodies. Natural antibodies and anti-idiotypic autoantibodies are prominent, but other immunological molecules, such as cytokines, soluble receptors, and soluble CD4, CD8, and HLA molecules may also be contained in some IVIGs. In controlled trials, there are two treatment options that are currently considered the standard of care in GBS, namely IVIG and TPE. IVIG is given 2 grams / kilogram divided over 5 days.[6,9]

Clinical trials have shown an effect of IVIg therapy when started within 2 weeks of the onset of weakness, whereas PE therapy has shown an effect when started within 4 weeks. Outside this time period, available evidence of efficacy is still lacking.[1,4] In these two patients PE therapy was started in the first week of disease onset, this may be why patient B has not yet experienced significant clinical improvement.

Plasma exchange therapy is thought to work by eliminating pathogenic antibodies, humoral mediators, and complement proteins involved in GBS pathogenesis. TPE is generally given as exchange volume over five sessions. TPE and IVIG have been shown to be equally effective. Effects appear if either treatment is given within 4 weeks, but stronger effects may be present if treatment is given within two weeks.[8-10]

Plasma exchange (PLEX) plasmapharesis is a plasma exchange that removes circulating antibodies and immune complexes by removing plasma from the blood through a centrifuge. The plasma is then re-injected intravenously along with albumin infusion to compensate for the loss of filtrate. PE is performed on around 4% of GBS patients worldwide, except in several countries such as 15% in the United States, 33% in Malaysia, and 30% of GBS patients who have access and receive PE therapy.

A study by Shahrizaila in 2021 stated that 35% of GBS patients who received IVIg therapy without clinical improvement switched to immunomodulatory therapy with PE. In practice, PE is highly recommended for GBS patients in the acute phase with impaired ability to walk independently or for patients who require mechanical ventilation assistance, while PE is contraindicated for patients who cannot tolerate the installation of a central line due to impaired coagulation function, hypocalcemia, ACE inhibitor therapy. (stopped for at least 24 hours), unstable hemodynamics (sepsis) or allergic to plasma or frozen albumin.[1,4] Moderate evidence suggests higher efficacy of PE over supportive care alone in adults with GBS, without a significant increase in serious side effects.[9,10] Despite this, sometimes reported side effects of PE include Chateter Related Infection, Deep Vein Thrombosis (DVT) , hypotension, septicemia, anaphylaxis, and hemolysis.[10] Common complications include headache, chills, myalgia, noncardiac chest pain, and aseptic meningitis.

When do GBS patients need ICU care?

Reasons for patient admission to the ICU are as follows: respiratory distress accompanied by respiratory insufficiency, severe autonomic cardiovascular dysfunction (e.g., arrhythmia or hypotension/hypertension), severe swallowing dysfunction or reduced cough reflex, and development of paralysis.[9,10]

Acute respiratory insufficiency is defined as clinical signs of respiratory distress, including shortness of breath at rest or when speaking, inability to count to 15 in one breath, use of accessory respiratory muscles, increased respiratory or heart rate, vital capacity. <15– 20 ml/kg or <1 liter, or abnormal blood gas analysis or pulse oximetry. In both patients A and B, there were indications for mechanical ventilation due to type 2 respiratory failure. Because 22% of GBS patients require mechanical ventilation within the first week of hospital admission, patients at risk of respiratory failure should be identified as early as possible. EGRIS prognostic tool was developed for this purpose and calculates the probability (1–90%) that a patient will require

ventilation within 1 week of assessment. For patient A, the EGRIS score was 4, while B was higher 5. Risk factors for prolonged mechanical ventilation include inability to raise the arm from bed within 1 week after intubation, and axonal or nerve subtypes that cannot be stimulated in electrophysiological studies. Early tracheostomy should be considered in patients who have these risk factors.

CONCLUSION

GBS is a post-infectious autoimmune disease that causes nerve cell destruction. Severe muscle weakness can result in respiratory muscle weakness resulting in respiratory failure which results in the need for mechanical ventilation therapy. In patients A and B who require 5x TPE, the total cost of TPE is more economical than administering IVIG at the ideal dose. Current therapeutic options include treatment with intravenous immunoglobulin or plasmapharesis aimed at neutralizing, and eliminating antibodies circulating in the bloodstream, in addition to supportive measures to maintain motor function. These two therapies are equally effective in treating GBS. Primary therapy is carried out as early as possible. FASTHUG is important for repeated assessment and intervention to avoid prolonged treatment and unwanted complications in GBS patients. GBS is a post-infectious autoimmune disease that causes nerve cell destruction. Severe muscle weakness can result in respiratory muscle weakness resulting in respiratory failure which results in the need for mechanical ventilation therapy. Plasmaparesis therapy was preferred compared to IVIG administration in these two GBS patients.

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