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Management of guillain-barré syndrome patients with type 2 respiratory failure in RSUD Gambiran

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ABSTRACT

Guillain-Barré syndrome (GBS) is an autoimmune disorder that affects the peripheral nervous system. Most patients with GBS are clinically characterized by tetraplegia with or without sensory disturbances. In GBS this is due to a hyperreactive immune response including the release of anti-ganglioside antibodies, the formation of antibody-dependent immune complexes, and an increase in macrophages that cause demyelination and axonal degeneration. In severe cases GBS can appear clinically progressive with respiratory muscle involvement requiring mechanical ventilation and treatment in the intensive care unit. The patient we report is a 24 year old man with clinical type 2 respiratory failure with suspicion of GBS who was initially examined at the RSUD Kilisuci, then referred to the RSUD Gambiran and treated in the ICU at the RSUD Gambiran and received immunomodulatory therapy in the form of plasma exchange (PE). The patient was treated for 10 days in the ICU at RSUD Gambiran with the outcome being that the patient was able to move to the ward.

Keywords: Intensive Management, Plasma Exchange, Guillain-Barré Syndrome, Respiratory Failure, Case report.

INTRODUCTION

First identified by Guillain, Barré, and Strohl in 1916, Guillain-Barré syndrome (GBS) is an autoimmune disease that affects the peripheral nerve system (Hughes et al., 2016). The annual incidence of GBS ranges from 0.81-1.89 cases per 100,000 people worldwide with the majority of patients are male (Shang et al., 2020). A report recruiting more than 900 Guillain-Barré syndrome patients worldwide found the average age of patients was 51 years with this figure peaking at 50-69 years. A population-based study in East Asia reported an incidence of Guillain-Barré syndrome of 0.44 cases per 100,000 people per year in Japan, and 0.67 cases per 100,000 people in China. While the incidence in Bangladesh is 1.5–2.5 cases per 100,000 people per year in adults, and 3.25 in children (Shahrizaila et al., 2021).

GBS usually occurs after the patient experiences an infectious disease infection in the upper respiratory tract or digestive tract (Hughes et al., 2014). Some of the most common pathogens that cause infections before GBS occur are Campylobacter jejuni (C. jejuni), followed by Cytomegalovirus (CMV), Epstein-Barr virus, Mycoplasma pneumoniae, Haemophilus influenzae, and influenza virus type A (Liu & Ma, 2020). Vaccination, surgery, trauma, and intravenous gangliosides can sometimes trigger disease (Shang, Zhu, et al., 2021).

Pathophysiologically, demyelination and axonal degeneration in GBS can be brought on by an overreactive immune response that includes the release of anti-ganglioside antibodies, the creation of immune complexes dependent on antibodies, and an increase in macrophages (Dhadke et al., 2013). The majority of GBS patients have tetraplegia, either in conjunction with or apart from sensory abnormalities. When GBS is severe, it can show clinical progression and involve the respiratory muscles, necessitating therapy in an intensive care unit and mechanical ventilation. (Manorenj et al.,

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2016). Various studies on the immunopathogenesis of Guillain-Barré syndrome show that this disease actually includes a group of peripheral nerve disorders, each of which is differentiated according to the distribution of weakness in the limbs or cranial nerves and innervated muscles and the underlying pathophysiology (Wakerley & Yuki, 2015).

This case report discusses Guillain-Barré syndrome as a whole and is expected to have the benefit of being learning material for considering GBS management in general to management in the ICU and comparing it with related literature.

RESEARCH METHOD

The present case report studies were conducted by analyzing patient medical records at Gambiran District Hospital and Kilisuci District Hospital with each Hospital permission while protecting the patient's identity.

RESULT AND DISCUSSION

Case Report

24 year old male patient, referred from Kilisuci District Hospital with a diagnosis of type 2 respiratory failure suspected GBS. The patient came with complaints of headache followed by a tingling sensation in the tips of both feet and hands since 2 weeks before entering the hospital. After one day of treatment, the patient complained of difficulty swallowing and vomiting 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 was treated 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. The patient was referred to RSUD Gambiran for further intensive treatment.

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, namely 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 in the form of Pantoprazole 40 mg/24 hours. The patient also received corticosteroids in the form of intravenous methylprednisolone 125 mg/8 hours. The patient's fluid and nutritional needs are met during intensive treatment, both parenterally and enterally. On supporting examinations, there was no electrolyte imbalance or disturbances in other organ systems. The patient also underwent physiotherapy to prevent thromboembolism.

The patient was consulted to the Department of Clinical Pathology and planned for serial plasma exchange (PE) 3 times, namely on day 3, day 6, and day 8 of intensive care. During the third serial PE, the patient's hemodynamics and clinical stability were stable, and after PE on the 8th day of intensive care, the patient's condition improved both clinically and laboratoryly, so it was decided to extubate. After extubation, the patient's condition was stable with continued nasal cannula oxygenation at 4 lpm. After 1 x 24 hours free from mechanical ventilation, on the 10th day of intensive care, because the patient's condition is clinically and laboratory stable, the patient can be transferred to the ward with transfer of care to a neurology colleague.

Discussion

The case reported is a 24 year old man, referred from RSUD Kilisuci with a diagnosis of type 2 respiratory failure suspected of GBS who underwent further treatment in the intensive care unit of RSUD Gambiran.

The initial signs of Guillain-Barré syndrome are paresthesia, weakness, numbness, and limb discomfort, or any combination of these. The primary feature is bilateral limb weakness that progresses and is largely symmetrical; it can take anything from 12 hours to 28 days for the weakness to peak (Shang et al., 2020; Shang, Feng, et al., 2021) .Patients usually experience hyporeflexia or generalized areflexia. Additionally, it's typical to have experienced diarrhea or upper respiratory tract infection symptoms three days to six weeks prior to onset. The presence of distal paresthesia supports the possible diagnosis of Guillain-Barré syndrome. If no involvement of sensory function is found, other diseases such as poliomyelitis, myasthenia gravis, electrolyte disturbances, botulism, or acute myopathy should be considered. If paralysis appears suddenly accompanied by prominent symptoms of urinary retention,

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magnetic resonance imaging (MRI) of the spine should be considered, this is to rule out the possibility of spinal cord compression lesions as the cause (Wakerley & Yuki, 2015).

The reported patient was diagnosed with GBS with the first symptom, namely tingling in the tips of both feet and the tips of both hands progressively over a period of 1 day. Upper respiratory tract infection or diarrhea was denied by the patient and family. The patient has no electrolyte balance, so the patient is diagnosed with GBS

Patients are at risk of complications and severe nerve damage during the acute phase, which typically lasts for the first two weeks after the disease beginning. It is recommended that patients with autonomic nerve disorders and possible respiratory failure receive treatment in a high-dependency unit so that their illness development can be closely monitored. Patients are susceptible to indirect consequences such as deep vein thrombosis, pneumonia, and aspiration throughout the progressive phase. Supportive interventions including deep vein thrombosis prophylaxis, frequent respiratory physiotherapy, and enteral tube feeding can help avoid these problems. The earliest feasible start for physiotherapy is recommended. Pain and exhaustion symptoms need to be treated appropriately, and it may be helpful to speak with coworkers who have similar experiences. Close monitoring must be continued because the highest number of deaths due to Guillain-Barré syndrome occurs during the recovery phase. The cause of death is usually respiratory, cardiovascular or autonomic complications (Shahrizaila et al., 2021; Wakerley & Yuki, 2015).

The reported patient entered the acute phase where the onset of the disease occurred within the first 2 weeks. The patient experienced difficulty swallowing and difficulty breathing, so to prevent aspiration and maintain airway patency, intubation was carried out and continued with mechanical ventilation before being referred. During treatment in the intensive unit of RSUP Dr. Sardjito, management continues with broad spectrum antibiotic prophylaxis, gastric ulcer prophylaxis, by meeting fluid and nutritional needs according to the patient's profile. Then the patient also undergoes supportive therapy for thromboembolism prophylaxis, namely early physiotherapy.

If the patient is unable to walk 10 meters on their own, immunomodulatory medication ought to be started (Kumar et al., 2015). Treatment should be considered even though there is little evidence of its effectiveness in patients who are still able to walk on their own. This is especially true if the patients exhibit other severe symptoms like autonomic dysfunction, respiratory insufficiency, or rapidly progressive weakness. Intravenous immunoglobulin (IVIg) therapy has been demonstrated in clinical trials to have an impact when started within two weeks after the onset of weakness, while PE therapy has been found to have an impact when started within four weeks. There isn't much proof of efficacy available after this point (Leonhard et al., 2019).

IVIg is a plasma product with a variety of distinct antibody types. IVIg inhibits complement factor-mediated macrophage activation, inhibits antibody binding to nerve targets, and blocks complement activation that could lead to additional nerve damage, among other pleiotropic immunomodulatory actions. Anti-ganglioside IgG antibodies can get dimerized by IVIg, which lowers their immunoreactivity in GBS. High dosages of IVIg, which vary from 1000 to 3000 mg/kg body weight, on the other hand, result in immunosuppressive phenotypes and anti-inflammatory effects, and are therefore widely utilized in the treatment of autoimmune illnesses like SGBS. (Leonhard et al., 2019; Shahrizaila et al., 2021).

IVIg is most frequently given in GBS because of its machine-independent qualities and straightforward process. IVIg, however, should not be administered to those who have a history of severe systemic or anaphylactic reactions to the product, are hypersensitive to the product's active ingredients, have anti-IgA antibodies, or have a specific IgA deficiency. Hemolytic anemia, aseptic meningitis, transfusion-related acute lung injury (TRALI), stroke, and venous embolism are among the side effects associated with IVIg. (Shahrizaila et al., 2021).

Administering IVIg was the next immunotherapy to be proven successful for GBS, after plasma exchange (PE). PE is still a useful treatment that helps GBS patients recover at the moment. About 4% of GBS patients globally undergo PE, with the exception of a few nations: 15% in the US, 33% in Malaysia, and 30% of GBS patients who are able to access and benefit from PE therapy.

According to a Shahrizaila research from 2021, immunomodulatory therapy with PE was chosen by 35% of GBS patients who had taken IVIg therapy without showing any clinical improvement. In practice, PE is highly recommended for GBS patients in the acute phase with impaired ability to walk

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independently or for patients who require mechanical ventilation assistance, while Plasma Exchange is contra indicated for a patients who can't tolerate the installation of a central line due to impaired coagulation function, unstable hemodynamics or allergies. against plasma or frozen albumin (Shahrizaila et al., 2021).

Plasma Exchange functions as a scavenging agent for pathogenic agents and antibody-autoimmune complexes contained in the patient's blood. Patients with GBS routinely benefit from standard Plasma Exchange administration, namely 5 sessions at a dose of 40-50 ml plasma/kg per session for 7-14 days. To enable the redistribution of pathogenic agents in the extravascular and intravascular compartments, PE is typically carried out on a daily basis. The effectiveness of PE is highly dependent on the rate of production and clearance of pathogenic agents, thus, regular administration of immunosuppressive drugs is considered as adjuvant therapy for PE (Shahrizaila et al., 2021).

It's intriguing to talk about how to optimize the PE technique; for instance, the number of PE sessions is set at four for moderate to severe SGB instances and two for light SGB cases. Instead of using fresh frozen plasma, PE can alternatively be carried out by giving out albumin and gelatin. However, extra care must be used when diluting antiinfective immunoglobulin when albumin or gelatin is used to substitute patient serum (Mustafa et al., 2019). It is advised that PE be done using a continuous flow machine rather than an intermittent one, in between sessions. The advantages of continuous flow devices for PE are still debatable, though.

In general, PE is effective in eliminating harmful substances from the patient's blood; nevertheless, the process is intricate and reliant on a machine. 2.5 In the patient that was reported, a patient with clinically severe GBS underwent PE therapy three times with an albumin regimen. Every PE therapy session a clinical and supporting evaluation is carried out on the patient and nothing is obtained There were side effects and complications in the patient, clinically improving progressively. After the 3rd session of PE, the patient was extubated and 1 day after extubation the patient was able to move to the ward.

For GBS, IVIg and PE work just as well (Sudulagunta et al., 2015). The potential for side effects with IVIg and PE is equivalent to their respective benefits. IVIg is typically the preferred treatment because it is easier to give and generally more accessible than PE. Other than PE and IVIg, no other treatments or drugs have been demonstrated to be successful in treating GBS.

Corticosteroids are expected to help reduce inflammation and the progression of the disease in GBS, but eight randomized controlled trials that looked at their efficacy for the disease did not find any evidence of a significant benefit; in fact, oral corticosteroid administration was found to have a negative impact on patient outcomes (Ulutaş et al., 2020). Furthermore, there is inadequate data to support the effectiveness of subsequent intravenous methylprednisolone treatment in patients receiving IVIg, and PE followed by IVIg is no more successful than medication therapy alone. In those who are currently receiving corticosteroids in the hopes of slowing the patient's severe GBS progression.

Intubation is necessary in patients with bulbar weakness or with poor cough reflex to avoid atelectasis caused by impaired clearance of secretions. In practice, mechanical ventilation assistance should be considered when at least one or two of the following criteria are met: (a) hypercarbia ([PaCO2] > 6.4 kPa); (b) hypoxemia ([PaO2] < 7.5 kPa); (c) decreased vital lung capacity <15 ml/kg; (d) intolerable respiratory distress. Mechanical ventilation is also indicated if oxygen saturation is lower than 90–94%, which is implicated in multiple organ hypoxic injury. Due to the dynamic clinical course of GBS, the speed and extent of disease progression is usually unpredictable, especially whether the disease will progress to a stage requiring mechanical ventilation.

The recommendation to initiate intubation and mechanical ventilation is if there are signs of respiratory fatigue and before clinical decompensation occurs. Intubation is also recommended in cases of dysphagia which may be accompanied by poor airway clearance or aspiration of saliva (Leonhart, 2019;Shang P, 2021). In patients who report complaints of dysphagia with increased saliva secretion accompanied by difficulty breathing, which results in the patient being decided to undergo immediate airway management. with intubation followed by mechanical ventilation.

Non-invasive ventilation(NIV) is not recommended in most patients with GBS due to: (a) persistent decline in GBS-related dysautonomia and prolonged respiratory muscle weakness; (b) GBS patients with dysautonomia are at high risk of emergency intubation, which can cause life-threatening complications including unstable blood pressure, arrhythmias, and fatal hyperkalemia after using

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succinylcholine as a muscle relaxant agent; (c) patients usually remain weak and require ventilation for a long time and respiratory failure can worsen dysautonomia making NIV an unsafe option or in patients with severe GBS. Generally, elective intubation should be implemented immediately in GBS patients with rapid progression of symptoms, marked bulbar weakness, and respiratory muscle insufficiency. Invasive ventilation in GBS patients is recommended to protect the airway from aspiration pneumonia and to facilitate clearance of bronchial secretions (Shang et al., 2020).

Provision of mechanical ventilation assistance generally begins with control mode ventilation with a tidal volume of around 10 ml/kg body weight, then switched to sychronized intermittent mandatory ventilation (SIMV) with pressure support for maintaining the adequacy of oxygenation and oxygen fraction in inspired air (FiO2 <0 .5, PO2 > 60 mmHg) after stabilization. Lower tidal volumes (<10 ml/kg BW) correlated with the development of atelectasis during the first 3 days of MV (Godoy & Rabinstein, 2015). However, lower tidal volumes combined with higher positive end-expiratory pressure (PEEP) administration are recommended in patients with GBS. Lower tidal volumes may be beneficial in obstructive lung disease and prevent ventilator-associated lung injury while higher tidal volumes may be beneficial in partial lung collapse.

Nevertheless, further studies need to be conducted to demonstrate the protective effect of lower tidal volume and higher PEEP in GBS patients dependent on mechanical ventilation. Nutritional therapy support, aseptic and antisepsis procedures, humidification of inspired air, chest physiotherapy, regular endotracheal tube (ETT) toileting, and repositioning further reduce the risk of atelectasis and other mechanical ventilation-related complications (Shang et al., 2020).

A tracheostomy should be explored right away if it is anticipated that the intensive unit's use of mechanical ventilation would last longer than three weeks over the course of treatment. For GBS patients, early tracheostomy offers various benefits, including increased comfort, earlier enteral feeding, proper oral hygiene, simpler oral communication, and mobilization away from the bed. Furthermore, there is a greater chance of fistula formation, laryngeal nerve, laryngeal mucosa, and voice cord damage if tracheostomy is postponed for longer than two weeks. However, due to perioperative hemorrhage, esophageal perforation, and pneumothorax, as well as complications from infection, tracheomalacia, tracheal stenosis, innominate tracheal artery fistula, and scar formation, tracheostomy performed in the latter phase may provide a challenging decision.

The challenges listed above can be overcome through interdisciplinary teamwork. For instance, ENTs and anesthesiologists can offer technical assistance to lessen tracheal stenosis following a tracheostomy and pre-tracheostomy hemorrhage. Additionally, the risk of bleeding, infection, and post-tracheostomy tracheal stenosis may be decreased by combining percutaneous dilatation tracheostomy with bronchoscopy or ultrasound. Antibiotic prophylaxis can also aid in preventing harmful side effects, such as bacterial colonization. When a patient does not recover quickly after receiving immunomodulatory medication in the form of PE or IVIg, early tracheostomy is advised (Shang, Feng, et al., 2021).

In the reported patient, when we received him in the ICU, an ETT was installed which aims to protect the airway from aspiration pneumonia and facilitate clearance of secretions, clinical consequences of dysphagia and salivation, as well as improving pulmonary gas exchange and preventing hypoxia and hypercarbia due to complaints of difficulty breathing in the patient . The expected complication while the patient was being treated in the intensive unit was pneumonia, however clinically and supportively the patient did not fall into a state of severe pneumonia and experienced progressive improvement after undergoing serial PE 3 times, so the patient was predicted not to require prolonged mechanical ventilation. The patient was extubated on the 8th day of treatment and was able to change wards on the 10th day.

CONCLUSION

A 26 year old female patient with progressive type 2 respiratory failure due to GBS underwent further treatment in the intensive care unit. During treatment, the patient received mechanical ventilation assistance with broad spectrum antibiotic prophylaxis, gastric ulcer prophylaxis, corticosteroids, and early physiotherapy to fulfill fluid and nutritional needs. The patient also received immunomodulatory therapy in the form of PE on the 3rd, 6th and 8th days of treatment with an albumin regimen. After the PE session on the 8th day, the patient improved progressively clinically and laboratoryly, so he could be extubated and moved to the ward. on the 9th day of treatment. Decision

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making for airway management with intubation and mechanical ventilation is very important to know in GBS patients to reduce patient mortality. The decision to provide immunomodulatory therapy requires interdisciplinary collaboration and must be decided immediately together to prevent clinical progression of GBS itself.

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