Critical Care Management of Myasthenic Crisis: A Case Report

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ABSTRACT

Background: The annual incidence of myasthenia gravis is 1-2 cases in 100,000 population. 30% of all cases will develop bulbar or respiratory muscle weakness. About 15-20% of myasthenia gravis patients will develop myasthenia crisis. Case presentation: An 18-year-old female patient complained of difficulty swallowing two days ago difficulty swallowing solid and liquid food, and this complaint caused the patient to cough and choke. Complaints were accompanied by difficulty opening the eyelids, especially in the afternoon. Complaints were also accompanied by double vision, especially when looking to the right. There was a nasal sound, and the patient had difficulty pronouncing vocal letters. The patient was admitted to ICU, In the ICU patient was intubated and given plasma exchange therapy. Conclusion: Myasthenic crisis is a neurological emergency that requires prompt diagnosis and treatment. Adequate intensive care, judicious weaning, and extubation decisions, along with specifically targeted therapy, will improve the outcome of myasthenia gravis.

1. Introduction

Myasthenia gravis (MG) is an autoimmune disorder with disruption of the neuromuscular junction caused by antibodies targeting the postsynaptic membrane. The prevalence of MG worldwide is estimated to be around 150 cases in 1 million population.1,2

In myasthenic crisis, the most severe problem is neuromuscular dysphagia, which rapidly progresses to loss of swallowing function and is often accompanied by respiratory muscle weakness and type 2 respiratory failure. This is a clinical emergency that requires intensive treatment.1

The annual incidence of myasthenia gravis is 1-2 cases in 100,000 population. 30% of all cases will develop bulbar or respiratory muscle weakness. About 15-20% of myasthenia gravis patients will develop myasthenia crisis. The number of women with myasthenic crisis is twice as high as men. The reported mortality rate due to myasthenic crisis is about 5% of cases.3,4

Approximately 15%-20% of people with myasthenia gravis will experience at least one crisis attack during their lifetime. Patients in crisis require endotracheal intubation. As many as 18% of patients admitted in myasthenic crisis require rehabilitation after discharge. The majority of patients are adults; however, an increased incidence in children under the age of 15 years has been reported in some Asian regions.5,6

Myasthenic crisis is more common in under-treated MG patients or those who have recently presented and are on treatment but whose presentation progresses more rapidly. The patient develops severe muscle weakness, including weakness of the respiratory muscles, usually preceded by severe bulbar weakness with dysphagia, with or without palatal weakness and nasal discharge.1

Respiratory support is essential in the management of myasthenic crisis. 2/3 to 90% of crisis patients require intubation and mechanical
ventilation. More than 20% require intubation during evaluation in the emergency room, and nearly 60% are intubated after admission to the ICU.5

The mortality rate from myasthenic crisis decreased from 40% to 5% recently due to the development of new drugs and improved intensive care techniques, especially those related to ventilation management.7

2. Case Presentation

An 18-year-old female patient was consulted from the Neurology department with myasthenia gravis. The patient complained of difficulty swallowing two days ago difficulty swallowing solid and liquid food, and this complaint caused the patient to cough and choke. Complaints were accompanied by difficulty opening the eyelids, especially in the afternoon, and did not improve even after resting. Complaints were also accompanied by double vision, especially when looking to the right. There was a nasal sound, and the patient had difficulty pronouncing focal letters. There was no fever, no cough, no vomiting, or loose stools. The patient's past medical history had been known to be a sufferer of myasthenia gravis for the past six months, with complaints of fluctuating difficulty in opening both eyes. History of administration of mestinone 4x60mg, methylprednisolone 1x4mg, calcium 1x1, and lansoprazole 1x30mg. The patient had been treated for 18 days. The patient was treated in the ICU for 16 days with a ventilator for five days, and a tracheostomy was performed on the sixth day of treatment in the ICU.

Examination of the patient's vital signs showed that the general condition was moderately ill, cooperative consciousness, blood pressure 128/62 mmHg, HR 78 x/minute, RR: 24 x/minute, T: 36.8 and SpO2: 98% on nasal canule 3 liters/minute. Patient with GCS 15 (eye 4, motor 6, verbal 5) pupil isocore 3 mm ODS, light reflex +++, right eye movement limited laterally, plica nasolacrimalis symmetrical. Dysphonia was found, motor 555/555 RF ++/+ RP -/- Wartenberg test (+) counting test (+). Physical examination of the patient did not reveal anemia, and the sclera was not icteric. Physical examination of the lungs was obtained, static and dynamic inspection, and the chest was symmetry. The movement of the right chest wall was the same as the left. Palpation revealed that the right fremitus was the same as the left. On sonor percussion and auscultation, bronchovesicular breath sounds rhonchi were present, and wheezing was absent.

The patient was admitted to the hospital and was treated in the HCU neurology room for three days, and then the patient was consulted by an intensivist to receive ICU treatment. In the ICU patient was intubated. Laboratory results on the first day in ICU showed Hb: 13 Ht: 38 Leukocytes: 16,000 Platelets: 36,000. Procalcitonin: <0.05. AGD result was obtained: pH 7.30, pCO2 54.7, pO2 105, HCO3 30.3, BE 2.5, SatO2 98.2 with the impression of respiratory acidosis. The result of tumor marker LDH: 159, BHCG: < 0.1, AFP: 1.1, CEA: < 0.5, all of which was normal. On the patient's chest X-ray on the first day in the hospital, there was fluid in the right lung fields with the impression of pneumonia and improvement compared to the chest X-ray five days later. The results of blood culture and urine culture showed no growth. The chest X-ray two weeks after hospitalization found an infiltrate in the right lung. The patient was accepted for ICU with oxygenation NRM 15 lpm, compos mentis consciousness, BP: 150/97 mmHg, MAP 115 hr: 92 x/min rr: 35 x/min SpO2: 98%, then intubated with ventilator setting PCBPAP PC=17, PS=10 I:E 1: 2.6 RR 12 FIO2: 40% PEEP 5 patients had an NGT installed, a urinary catheter and diagnosed with myasthenia gravis, thrombocytopenia ec. Susp. ITP and respiratory disorders and suspect pleural effusion.

The therapy from the intensivist was given Ca gluconate 1 gram, antibiotic ceftriaxone 2x1 mg, omeprazole 120 mg/24 hours, methylprednisolone 4x125 mg, and down titration of prostigmin: SA 5 amp in NaCl 0.9% 2cc/hour, Terbutaline 3x 0.3 cc paracetamol 1 gr
(needed), IV ranitidine 2x50mg, Ventolin four time in a day and flumucyl two times in a day. Specific management for this patient is plasma exchange.

Laboratory results on the sixth day in ICU showed Hb: 14.3 Ht: 39 Leukocytes: 15.220 Platelets: 73.000. Procalcitonin: <0.05. AGD results were obtained: pH 7.4, pCO₂ 39, pO₂ 85, HCO₃⁻ 24.2, BE -0.6, SatO₂ 98.2, Lactat 2.5 with the impression of metabolic acidosis, ureum/creatinin: 45/0.5, Na 135, K 3.5, Cl 101, Ca 9.2 The patient's chest X-ray on fifth days in ICU showed atelectasis. Intubated with ventilator setting PC BIPAP PC=17, PS=10 I:E 1:2.6 RR 12 FIO₂: 80% PEEP 6 diagnosed with myasthenia gravis, thrombocytopenia ec. Susp. ITP and respiratory disorders and pleural effusion improvement. The therapy antibiotic ceftriaxone 2x1 mg, omeprazole 120 mg/24 hours, methylprednisolone 4x125 mg and down titration of prostigmin: SA 5 amp in NaCl 0.9% 2cc/hour, Terbutaline 3x0.3cc, IV ranitidine 2x50mg, Ventolin four time in a day and flumucyl two times in the day. Plasma exchange is done five times.

3. Discussion

This study discussed an 18-year-old female patient with difficulty swallowing two days ago difficulty swallowing solid and liquid food, and this complaint caused the patient to cough and choke. Complaints were accompanied by difficulty opening the eyelids, especially in the afternoon, and did not improve even after resting. Complaints were also accompanied by double vision, especially when looking to the right. There was a nasal sound, and the patient had difficulty pronouncing focal letters. The patient was diagnosed with myasthenia gravis, respiratory failure and pleural effusion.

The management of myasthenia gravis may generally be divided into two, namely, symptomatic treatment in reducing neuromuscular symptoms and disease progression and immunosuppressive treatment that targets the improvement of pathological immune responses will be occurred.8

The myasthenia gravis treatment strategy is based on the severity of the disease, i.e. ocular disorders only or generalized, and whether the patient is currently in a state of exacerbation or myasthenia crisis. The goal of treatment is to achieve complete remission (no symptoms or signs of myasthenia).9

Myasthenia gravis is a B-cell-mediated autoimmune disease, in which antibodies bind to acetylcholine (ACh) receptors (AChRs) at the neuromuscular junction, or to functionally related molecules at the postsynaptic membrane. A decrease in AChR functional density results in a decrease in motor end plate potential amplitude and a failure in the initiation of muscle fiber contraction.10

The most commonly used symptomatic therapy is pyridostigmine. Pyridostigmine is an acetylcholine esterase inhibitor that blocks the degradation of acetylcholine at peripheral cholinergic synapses, including neuromuscular junctions. In early disease or with mild symptoms, Pyridostigmine allows for a significant and rapid increase in muscle strength.1,11

In its management, this patient also received corticosteroid therapy. Prednisolone or prednisone constitutes the main immunomodulatory therapy in the long-term management of patients with myasthenia gravis.1

Respiratory management, in this case, was prioritized to hypercapnia due to myasthenic crisis. The duration of mechanical ventilator was unpredictable in patients with myasthenia, especially those with myasthenic crisis. Weaning from mechanical ventilation and extubation depend on how quickly the plasma can be cleared of auto antibodies and the response to anticholinergic drugs and immunosuppressants. In addition to the management of myasthenia, patients may be managed according to the general principles of critical care, infection management, and nutritional attention, along with targeted therapy. During a myasthenia crisis, respiratory failure may be hypoxemic, hypercapnic, or both.3,12

The decision on the mode of assisted ventilation may be based on clinical judgment. Once intubated, patient was ventilated using the assisted Ventilation mode, with an ideal tidal volume of 8-10 mL/kg BW
and pressure support of 8-15 cmH₂O to prevent atelectasis and minimize the work of breathing. The level of support required depended on the patient's condition.⁵,¹³

Life-threatening hypoxemia (PaO₂ < 60 mmHg) occurs at the end of neuromuscular respiratory failure and generally improves with supplemental oxygen administration.¹⁴

In life-threatening hypoxemia, the ventilator mode that can be used is bilevel positive airway pressure (BiPAP) because the use of positive pressure, in addition to preventing alveoli collapse and preventing atelectasis, this mode can also help withstand increased airway resistance.³

Initial assisted ventilation may be directed at maintaining lung expansion and improving muscle fatigue. Target initial mode of assisted-controlled ventilation, positive end-expiratory pressure (PEEP) 5 cmH₂O, with low tidal volume (6-8 mL/kg), respiratory rate 12-16/min, and FiO₂ adjusted to achieve target oxygen saturation of 92% or PaO₂ > 70 mmHg.³,¹³

This patient was also given bronchodilator management. Bronchodilators work to overcome the bronchospasm. Inhaled ipratropium bromide is a bronchodilator option that can be used. Besides that, ipratropium bromide can also reduce bronchial secretions. Terbutaline, which is a β₂ adrenergic agonist, can be given as an effective adjunctive therapy in patients with myasthenia gravis.³

The specific treatment in these patients was plasma exchange. Plasma exchange involvesapheresis, where immunoglobulins, immune complexes, complement, cytokines and other inflammatory mediators circulating in the blood are removed. The normal process involves the exchange of about 1 to 1.5 times the blood volume. Plasma exchange is done on alternate days five to six times.⁶

The efficacy of plasma exchange for myasthenia gravis patients is through the direct removal of pathogenic autoantibodies and complement pathway components. Plasma exchange is currently one of the first-line acute treatment modalities (the other being IVIG) in myasthenia gravis crises.⁹

Most patients with myasthenia gravis can have a good prognosis with therapy. However, the disease can also have a worse prognosis if there are respiratory complications, such as pneumonia and weakness of the intercostal muscles or diaphragm.¹⁵

4. Conclusion

Myasthenic crisis is a neurological emergency that requires prompt diagnosis and treatment. Adequate intensive care, judicious weaning, and extubation decisions, along with specifically targeted therapy, will improve the outcome of myasthenia gravis.

5. References

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