A CASE REPORT OF INTENSIVE CARE MANAGEMENT OF REFRACTORY MYASTHENIA GRAVIS WITH ACUTE RESPIRATORY FAILURE: DIAGNOSIS, TREATMENT AND MANAGEMENT

INTRODUCTION:

Myasthenia gravis (MG) is an autoimmune neuromuscular disorder characterized by fluctuating muscle weakness, often involving ocular, bulbar, and respiratory muscles. A myasthenia crisis (MC) is a life – threatening condition involving respiratory failure due to diaphragmatic and intercostal muscle weakness, requiring mechanical ventilation and intensive care support.

It affects a substantial number of individuals, leading to considerable morbidity due to its unpredictable nature and potential for severe complications. The clinical implications are profound, as the fluctuating nature of muscle weakness may lead to respiratory complications, exacerbating patient care challenges. As the incidence of MG continues to rise, this case study serves as an essential resource for medical professionals detailing the intensive care management strategies and multidisciplinary approaches applied to a patient experiencing an acute exacerbation in enhancing patient outcomes. This case study presents the management of a 24-year-old female with refractory MG in crisis, highlighting the challenges in airway management, plasma exchange therapy (PLEX) and its complications, immunosuppression, and secondary infections in the ICU[1].

CASE PRESENTATION:

A 24-year-old female, with a known case of non thymoma myasthenia gravis presented to us with chief complaints of shortness of breath, inability to swallow oral secretions and fever for 1 day.

She is on anticholinesterase inhibitor pyridostigmine 60 mg thrice a day and mycophenolate mofetil 500 mg once daily with a minimal dose of wysolone 5 mg once daily.

Her past history was significant with admission for myasthenia crisis for which she was treated with IVIG (0.4 g / kg / day x 5 days) 2 months back.

She is also a known case of carcinoma ovary for which she underwent Oophorectomy and received chemotherapy in 2010.

On physical examination, patient had a BP of 130/90 mmHg in right u-per limb in supine position, HR: 110 bpm, RR: 35/min, SpO2 – 88% RA, with single breath count of 5. She died not have a sustained neck lift for more that 5 seconds, had a significant proximal muscle weakness of bilateral upper and lower limbs [3/5]. Rest all osteotendinous reflexes were normal. Systemic examination of respiratory system and cardiovascular systems were insignificant.

ICU Management Strategy

In view of repiratory distress, patient was intubated with 7.5 mm ETT under opioid sedation. She was initiated on mechanical ventilation and placed on volume control mode. Laboratory findings revealed Hb of 10.0 g/dL with wbc count of 10,000 and normal renal and liver function test. Her acetylcholine receptor antibody levels were quantified and was found to be more than 8 mmol/L {normal<0.40}. Patient was initiated on pulse dose of glucocorticoids with injection methylprednisolone 1 gm iv od for 3 days and tapered slowly. A broad-spectrum antibiotic was initiated empirically.

Post pulse dose of steroids, patient did not show any improvement in muscle power of upper and lower limbs with absent sustained neck lift. Neurologist opinion was obtained and suspecting steroids unresponsive myasthenic crisis, it was decided to initiate plasma exchange [PLEX] for 5 cycles, with each cycle on alternate days[2].

After informed consent, right internal jugular vein was cannulated with 13 cm, 16 fr triple lumen HD catheter. With patients' weight being 60 kg and hematocrit of 31, Total plasma volume (TPV) of 2691 ml was obtained. It was decided to exchange 1-1.5 times of TPV in each cycle over alternate

days. On day 3 of ICU admission, patient developed increased ET tube secretions with a radiological opacity seen in left lower lobe on chest x ray. There was an increase in total white cell count to 14,000. Endotracheal tube culture was sent which came out as growth of Aceinetobacter baumanii which was carbapenem sensitive. Patient's antibiotics were escalated to meropenem. After the 3rd cycle of PLEX, patient started showing improvement in muscle power of both the limbs UL 5/5, LL 3/5, sustained neck lift >5 sec.

Weaning trial was initiated and patient was placed on pressure support ventilation. On 4th cycle of PLEX, after successful weaning trial; she was extubated to NIV. The last cycle of PLEX was continued on the next day. Towards the next few days, patient total count started decreasing, radiological regression of the opacity was noticed, her SBC improved to 20, she was mobilized to chair with the help of physiotherapist and shifted to ward. She was discharged on maintenance dose of tablet wysolone 5 mg, pyrodistigmine bromide 60 mg 6th hrly and mycophenolate moefetil 500 mg once daily.

Three weeks after the current admission, patient presented to us again with shortness of breath, unable to swallow secretions and respiratory distress. Her MG-ADL (Activities of Daily Living) score was around 18. In view of respiratory distress, patient was electively intubated and started on PLEX again. The PLEX cycles were continued for 5 cycles, alternating every day with 1.5 TPV to be exchanged in every cycle[3,7,8].

Considering frequent myasthenic relapse, elective tracheostomy was performed. Patient was initiated on injection Rituximab 1000 mg iv stat over 4 hours; 2nd dose repeated after 4 weeks. Post rituximab, patient was discharged with tracheostomy tube in situ. Plan to decannulate tracheostomy was made after 4 weeks based on the disease remission.

Discussion:

Refractory MG was defined as lack of response to treatment with steroids and at least 2 immunosuppressants with an inability to withdraw treatment

without relapse in the last 12 months, or intolerance to treatment resulting in adverse reactions[6]. The incidence of refractory MG is approximately 14 – 20 cases per 100,000 individuals, with a prevalence increase noted among women under 40 and men over 60[10].

This case poses significant challenge for healthcare providers due to its unpredictable nature and potential for severe exacerbations, leading to critical illness or respiratory failure.

Effective management is imperative, as exacerbations, particularly during crisis, can lead to respiratory failure and necessitate intensive care interventions. This case study illustrates the critical need for prompt, multidisciplinary strategies to enhance patient outcomes in MG crisis, underscoring its relevance for medical professionals and researchers dedicated to improving care protocols.

CONCLUSION

Non-thymomatous myasthenia gravis refractory to steroids with frequent relapses poses a significant challenge to the practitioner[6]. This case exemplifies how prompt interventions, multidisciplinary care, and vigilant ICU monitoring can significantly improve outcomes in MG crisis[9].

This case emphasizes the critical role of effective interventions, such as plasma exchange and newer immunosuppressants like rituximab in managing autoimmune respiratory crisis[3,4,5].

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