



Plasmapheresis in Myasthenic Crisis

Alegra Rifani Masharto^{1*}, Bastian Lubis¹, Andriamuri Primaputra Lubis¹,
Rommy Nadeak¹

1. *Department of Anesthesiology and Intensive Care, Faculty of Medicine, North Sumatra University - Adam Malik General Hospital, Medan, Indonesia*

**corresponding author*

DOI: 10.55497/majanestricar.v4i1i3.290

ABSTRACT

Introduction: Myasthenic crisis is the most lethal complication of myasthenia gravis. Referral to an intensive care unit is crucial in managing the myasthenic crisis.

Case illustration: Female, 30 years old, weighed 60 kgs, with myasthenia crisis. The patient came to an emergency department and was then intubated before being admitted to the intensive care unit. The physical diagnostic was normal and laboratory findings were leukocytosis. The patient was treated with normal saline, antibiotics, high-dose corticosteroids, and pyridostigmine. The patient was done plasmapheresis with synchronized intermittent mandatory ventilator mode. The patient was examined every 30 minutes. The physical examinations were relatively normal. The plasmapheresis procedure was ended in 12 hours. From the literature, plasmapheresis was found to have significant results for myasthenia gravis compared to conventional therapy because of its blood separation technique to remove autoantibodies. The next day patient was extubated with normal physical examinations and normal laboratory findings. The patient then moved from the intensive care unit to the normal ward and outpatient on the third day of hospital stay. The patient was given oral medicine that included antibiotics, corticosteroids, and pyridostigmine.

Conclusion: From this case, we can see that plasmapheresis therapy has a really good outcome compared to other conventional therapy, although until now therapy with plasmapheresis is still relatively expensive.

Keywords: Intensive Care Unit; myasthenic crisis; myasthenia gravis; plasmapheresis; treatment efficacy.



Plasmaferesis pada Krisis Miastenia

Alegra Rifani Masharto^{1*}, Bastian Lubis¹, Andriamuri Primaputra Lubis¹,
Rommy Nadeak¹

1. Departemen Anestesiologi dan Terapi Intensif, Fakultas Kedokteran, Universitas Sumatera Utara - RSUP H. Adam Malik, Medan, Indonesia

*penulis korespondensi

DOI: 10.55497/majanestcricar.v41i3.290

ABSTRAK

Pendahuluan: Krisis miastenia adalah komplikasi yang mengancam jiwa pada penyakit miastenia gravis. Rawatan pada unit rawat intensif sangat penting dalam tatalaksana krisis miastenia.

Ilustrasi Kasus: Perempuan, 30 tahun, berat badan 60 kg, terdiagnosis dengan krisis myasthenia gravis. Pasien datang ke unit gawat darurat dengan tanda gawat nafas kemudian diintubasi sebelum dirawat di ruang intensif. Pemeriksaan fisik dalam batas normal dan pemeriksaan laboratorium dijumpai leukositosis. Pasien diterapi dengan cairan fisiologis, antibiotik, kortikosteroid dosis tinggi, dan piridostigmin. Pasien lalu dilakukan plasmapheresis dengan modus ventilator synchronized intermittent mandatory ventilator mode. Pasien dilakukan pemantauan setiap 30 menit hingga plasmapheresis selesai. Pemeriksaan fisik selama tindakan relatif normal sampai prosedur selesai selama 12 jam. Dari literatur didapati bahwa plasmapheresis memberikan hasil akhir yang signifikan untuk pasien myasthenia gravis dibandingkan dengan terapi terdahulu karena kemampuannya untuk memisahkan autoantibodi didalam sirkulasi darah. Keesokan harinya, pasien sudah dapat diekstubasi dengan pemeriksaan fisik dan laboratorium dalam batas normal. Pasien kemudian dipindahkan dari ruang rawat intensif ke ruang rawat inap biasa di hari ketiga rawatan. Pasien lalu diberikan terapi oral berupa antibiotik, kortikosteroid, dan piridostigmin sebelum dipulangkan.

Simpulan: Dari kasus ini kita dapat melihat bahwa terapi plasmapheresis memberikan hasil yang signifikan baiknya dibandingkan terapi konvensional lainnya, walaupun sampai saat ini terapi dengan plasmaferesis masih tergolong mahal.

Kata Kunci: Intensive Care Unit; krisis miastenia; miastenia gravis; plasmaferesis; efektivitas pengobatan.

INTRODUCTION

Myasthenic crisis is a complication of myasthenia gravis marked by deteriorating muscular wasting that leads to respiratory failure and necessitates intubation and mechanical ventilation.¹ Myasthenic crisis develops in around 15% to 20% of patients with generalized myasthenia gravis.² The most common precipitant of myasthenic crisis is infection, although no obvious triggers can also be found in up to 30 % to 50% of cases.^{1,2} There are two mainstay treatments in myasthenic crisis, namely intravenous immunoglobulin (IVIG) and plasmapheresis. Based on the recommendation, plasmapheresis should be used as the first-line therapy in all myasthenic crisis patients, considering its fast and predictable recovery.² Here we reported a case of a myasthenic crisis patient who underwent plasmapheresis, which showed marked improvement.

CASE ILLUSTRATION

Female, 30 years old, weight 60 kg, with a myasthenic crisis that includes respiratory distress. The patient was admitted to the emergency department and then intubated before being admitted to the intensive care unit. The physical examination was within normal limits, and laboratory findings showed leukocytosis. The patient was treated with intravenous hydration, antibiotics, high-dose corticosteroids, and pyridostigmine, but there was no significant improvement in outcome. The patient underwent plasmapheresis with a normal base examination. The patient was already intubated in synchronized intermittent mandatory ventilator mode. The patient was evaluated every 30 minutes during the plasmapheresis for 12 hours. The next day, after plasmapheresis, the patient was able to be extubated with a normal physical examination, and laboratory findings were within normal limits. The next day, the patient was discharged from the intensive care unit to a normal ward.

DISCUSSION

Plasmapheresis is considered an option in critically ill autoimmune diseases. It can provide rapid improvement clinically and also provide fewer side effects, earlier patient independence,

safety, and efficacy that refer to improving patients' quality of life. But the downfall is the infusion time inflexibility, which is more invasive and less available compared to IVIG.³ Our patient was immediately extubated after plasmapheresis, as the main complaint, such as difficulty breathing and other limitations in muscular movement, has improved.

Although IVIG is believed to be more cost-effective, plasmapheresis could shorten hospital length-of-stay.⁴ A prospective cohort showed that plasmapheresis is associated with faster recovery, especially in intensive care units, although the clinical efficacy was comparable with IVIG after 1 month of treatment.⁵ Plasmapheresis also benefits in myasthenic crisis with sepsis because of its ability to remove a variety of plasma pathogens, such as antibodies and other immune complexes.⁶

In other studies, response rates to plasmapheresis were more than 50% compared to IVIG, including all changes from the myasthenic crisis severity scales. Studies have also found that pre-operative therapeutic plasma exchange in thymoma, such as plasmapheresis, is associated with better postoperative outcomes. It also showed significant response rates among juvenile patients. But every treatment should include glucocorticoids and pyridostigmine to augment the improvement and decrease the risk of exacerbation.⁷

A study in Japan showed that plasmapheresis can be applied to various neurological disorders such as myasthenia gravis, autoimmune encephalopathy, or Guillain-Barre syndrome, resulting in more than 60% improvement with a low frequency of adverse events.⁸

CONCLUSION

We reported a case of myasthenic crisis that underwent plasmapheresis, which showed marked improvement in the outcome. The patient was able to be treated in a normal ward and discharged safely with minimal issues. Unfortunately, this therapy is still considered expensive, especially in Indonesia. Most healthcare providers and insurance companies don't provide financial support. Hopefully, in the future, hospitals and healthcare providers can be supported for this treatment.

CONFLICT OF INTEREST

Penulis menyatakan tidak ada konflik kepentingan dalam penulisan artikel ini.

REFERENCES

1. Feng Wendell LC, Levine JM. Myasthenic Crisis. *The Neurohospitalist*. 2011;1(1):16-22.
2. Claytor B, Cho SM, Li Y. Myasthenic crisis. *Muscle and Nerve*. 2023;68(1):8-19.
3. Alcantara M, Barnett C, Katzberg H, Bril V. An update on the use of immunoglobulins as treatment for myasthenia gravis. *Expert Rev Clin Immunol*. 2022;18(7):703-15.
4. Heatwole C, Johnson N, Holloway R, Noyes K. Plasma exchange versus intravenous immunoglobulin for myasthenia gravis crisis: An acute hospital cost comparison study. *J Clin Neuromuscul Dis*. 2011;13(2):85-94.
5. Wang Y, Huan X, Jiao K, Jiang Q, Goh LY, Shi J, et al. Plasma exchange versus intravenous immunoglobulin in AChR subtype myasthenic crisis: A prospective cohort study. *Clin Immunol*. 2022;241:109058.
6. Imron A, Aditiansih D, George YW. The Role Of Plasmapheresis In Myasthenia Gravis With Sepsis. *JAI (Jurnal Anestesiologi Indones)*. 2013;5(3):210-6.
7. Ipe TS, Davis AR, Raval JS. Therapeutic Plasma Exchange in Myasthenia Gravis: A Systematic Literature Review and Meta-Analysis of Comparative Evidence. *Front Neurol*. 2021;12:662856.
8. Lin Y, Oji S, Miyamoto K, Narita T, Kameyama M, Matsuo H. Real-world application of plasmapheresis for neurological disease: Results from the Japan-Plasmapheresis Outcome and Practice Patterns Study. *Ther Apher Dial*. 2023;27(1):123-35.