CASE REPORT

Benefit of Modified Plasmapheresis in the Management of Myasthenia Gravis: A Case Report

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ABSTRACT

Background: *Myasthenia gravis is an autoimmune disease condition caused by the generation of antibodies against the acetylcholine receptor sites at the neuromuscular junction. The treatment modalities include anticholinesterase drugs, corticosteroids, immunotherapy, thymectomy and plasmapheresis. However, because of the poor financial state of our patients and the dearth of appropriate equipment in our centres modifications are made to standard treatment modalities including plasmapharesis.*

Method: *We report a case of myasthenia gravis who was on various occasions on neostigmine, pyridostigmine and prednisolone. After about 18 months of treatment, he developed myasthenic crises on two occasions. He was admitted in the ICU for respiratory support where he also had modified plasmapheresis.*

Results: *The patient had remarkable improvement following the modified plasmapheresis with reversal of symptoms of the myasthenic crises.*

Conclusion: *In the absence of facilities for standard plasmapheresis in this environment, the use of modified plasmapheresis is hereby recommended.*

KEYWORDS: *Myasthenia gravis; Modified Plasmapheresis; Benefit.*

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INTRODUCTION

Myasthenia Gravis (MG) is an autoimmune disorder of the neuromuscular junction characterized by fluctuating weakness and fatigue of the voluntary muscles. This is as a result of the development of autoantibodies against the acetylcholine receptors (Ach-R)[1,2]. This leads to a reduction in the number of the receptors available for the stimulation, decreased amplitude of stimulation and fiber activation, and of cause, weakness that is worse with muscle activity, while there is improvement with rest. The treatment modalities for MG, which is largely dependent on the severity of the disease, involve the use of anticholinesterase agents [neostigmine, pyridostigmine], corticosteroids [prednisolone], immunotherapy and plasmapheresis. Routine thymectomy is carried out in most patients with MG. Patients with MG may require intensive care unit(ICU) management to support the respiratory system. This is especially so when the disease entity involves the bulbar/airway muscles, thoracic/chest muscles, and the diaphragm. The adequacy of the respiration should be assessed by the close monitoring of the respiratory rate, tidal volume, forced expiratory volume, peak expiratory rate, the arterial blood gases and arterial oxygenation. The modified plasmapheresis (MPP) had earlier been found to be useful in the management of Waldenstrom's Macroglobulinaemia[3] in this environment. This communication highlights the beneficial role of modified plasmapheresis (MPP) in place of the standard plasmapheretic process[4,5,6,7] which is not yet available at the University College Hospital, Ibadan, Nigeria. We hereby report our experience with MPP in a patient with Myasthenia gravis.

CASE REPORT

A 33-year old male Negroid laboratory scientist presented at the Department of Medicine, University College Hospital, Ibadan, Nigeria with a 6-month history of recurrent drooping of his left upper eyelid and blurring of vision. The weakness got worse with exertion and improved with rest or on waking in the morning.

He was diagnosed as a case of MG following a positive response to the edrophonium (Tensilon) test within 30 seconds of administration. There were no facilities for electromyography, Ach-R antibody assay and repetitive nerve stimulation in this institution at the time of this study.

He was treated with either Neostigmine 15mg thrice daily or pyridostigmine 60mg thrice daily; depending on what was available. However, non-availability and the relatively high cost of the drugs hampered effective management of the patient. Prednisolone 30mg daily was added to his therapy while slow K was given because of the relatively low potassium of 2.2-3.0 mmol/L. Patient appeared relatively stable for about 18months. Thereafter he was noticed to have developed generalised weakness with bilateral ptosis and easy fatiguability. He developed hoarseness of

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voice, difficulty with swallowing and recurrent upper and lower limb weakness. He could not see clearly. He had difficulty completing a sentence and the response to neostigmine/pyridostigmine was minimal. Attempt to add azathioprine was impossible, as the patient could not afford it. The patient got progressively weak with affectation of the respiratory muscles. He was subsequently admitted into the intensive care unit (ICU) of the hospital for endotracheal intubations and ventilatory support on two occasions because of myasthenic crisis.

During these two admissions, he was offered a modified plasmapheresis (Appendix 1) as part of his management. He had two sessions in the first admission. The patient showed remarkable improvement. This was evident by the ability to demonstrate objective improvement in muscle strength and sustainability of muscle activity. On the second ICU admission, he had 5 sessions of modified plasmapheresis and showed good response. He was able to once again open the eyelids, make sentences and engage in conversations. He was ambulant and his breathing improved. Thymectomy was not performed for logistic reasons. Computerised tomographies of the chest, as well as the thyroid function tests were not done because of the poor financial status of the patient. He was transferred to the general medical ward and was subsequently commenced on breathing exercises. After about a week, he was placed on oral neostigmine/ pyridostigmine. He was subsequently planned for more sessions of plasmapheresis, whenever necessary, prior to referral to a centre with facilities for immunotherapy and thymectomy, as soon as he is able to get the necessary financial assistance.

Intensive Care Unit Monitoring

	DAY 1	DAY 2	DAY 3
PH	7.0	7.3	7.42
PC02(mmHg)	22	24	26
P02 (mmHg)	20.6	60	90
HC03 (mmol/L)	27.9	27.5	27.8
FI02	0.45	0.32	0.45
Serum Potassium (mmol/L)	2.2	3.3	4.0

DISCUSSION

Medical practitioners often easily make the diagnosis of MG, which is largely dependent on history and clinical examination. The problem has always been with the confirmation of the diagnosis with the Tensilon test (edrophonium), which often is not available. Neostigmine could be helpful in confirming the diagnosis. By far the major set back commonly encountered in Nigeria in the management of MG is the paucity of management options. Immunotherapy, plasmapheresis and often thymectomy are not possible for lack of facilities and logistics.

While Immunotherapy is virtually non-existent in our level of practice, routine thymectomy can not be encouraged because of lack of adequate postoperative back up. However thymectomy in generalized MG[6] has been associated with long-term benefits. There is need to exclude the possibility of a cholinergic crisis whenever a patient with MG deteriorates. Where this is not immediately possible, with a tensilon test, stoppage of anticholinenesterase and placement under respiratory support is recommended. Modified plasmapheresis produced clinical improvement in this patient, and may be life-saving in our environment, especially as facilities for standard plasmapheresis techniques[4-7] are not available . FEV1, a sensitive indicator of air flow was not documented in this patient's records. Significant improvement following MPP as evidenced by the ability to open the eye-lids widely, improved ability to make sentences and indeed engage in conversations, ambulation, as well as the disappearance of difficulty with breathing noted in this patient. Virtually all the pre-plasmapheresis difficulties were reversed after the procedures. This response is similar to the previous responses to the standard plasmapheresis techniques[4,5,6,7].

There is a need to acquire standard machines such as cell-separators which can be used for plasmapheresis and other procedures especially in tertiary centres like ours where patients are referred. Meanwhile, the MPP will continue to find relevance in the management of patients with MG as well as other autoimmune diseases in the developing countries, particularly the sub-Saharan states.

APPENDIX 1

MODIFIED PLASMAPHERESIS / PLASMA EXCHANGE

1. 500mls of normal saline is infused into the patient.
2. 450mls blood is phlebotomized into a CPD-A plastic double bag
3. 500mls of normal saline is infused into the patient
4. The blood removed in (2) is spun at 300 rpm for 15 minutes in a cold centrifuge machine; the plasma is

transferred into the second blood bag and discarded

5. The packed red blood cells in 4 are transfused back into the patient.
6. The procedures in 2 and 4 are repeated twice at each session.

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