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Case Report

Anesthetic Considerations in A Patient With Bulbar Onset Myasthenia Gravis (MG) For Paraesophageal Hiatal Hernia Repair: A Case Report And Brief Review Of Literature.

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ABSTRACT:

Introduction: Myasthenia Gravis (MG) poses a huge challenge for any anesthetist in different terms of management. These patients require special terms of care in the pre, peri and post operative period. In this study we are addressing the anesthetic considerations for an MG patient with variable clinical symptoms and several medical conditions during general anesthesia (GA).

Aim and Objectives: The main aim of this case report is to present our approach for GA in an MG patient taking into consideration the peri-operative care and challenges. We also describe how an interprofessional team manages MG during GA, briefly provide an explanation for the pathophysiology of the disease, examine the risk factors and clinical outcomes and summarize peri-operative evaluation of the disease.

Conclusion:

MG is an autoimmune neuromuscular junction disorder, characterized by skeletal muscle weakness that worsens with activity. Respiratory muscle function, medications, and medical history are important factors in assessing the risk of postoperative respiratory failure. Neuromuscular blocking agents (NMBAs) should be avoided as possible in such patients. A clear understanding of the disease allows for an individualized anesthetic approach.

Case presentation:

A middle-aged female patient with bulbar onset MG, controlled asthma and suspected allergy to fentanyl was posted electively for laparoscopic paraesophageal hiatal hernia repair under GA. Preoperative evaluation was done and the patient was cleared from neurology side. American Society of anesthesiologists (ASA) basic standard monitoring was applied. A test dose of remifentanil was administered. During induction, she received remifentanil, ketamine, propofol and lidocaine intravenously (IV). Desflurane and continuous remifentanil infusion were used for GA maintenance. The emergence from GA was uneventful. Postoperative analgesia was covered with paracetamol, ketorolac and meperidine. Patient was transferred to post anesthesia care unit (PACU) post operatively for observation. On the 6th day postoperatively, she was discharged home safely.

Key words: myasthenia gravis, Bulbar manifestation, General Anesthesia, neuromuscular monitoring, sugammadex

INTRODUCTION:

Myasthenia Gravis (MG) is the most common neuromuscular junction (NMJ) disorder. It is an autoimmune disease that attacks the NMJ and affects neuromuscular transmission [1]. 3-7 people out of 100,000 can fall victims to MG which can present with variable types of muscle weakness particularly after exercise [2]. General anesthesia (GA) in patients with a previous diagnosis of MG are considered high risk and are extremely challenging if no special consideration were taken. Medications such as induction agents, muscle relaxants, antibiotics as well as surgical stress can deteriorate the symptoms and worsen the prognosis. MG patients show an increased sensitivity to nondepolarizing muscle relaxants with a high chance for persistent and prolonged postoperative paralysis [2].

Case presentation:

A 57-year-old woman, 71.5 kg, 153 cm, BMI 30.5, previously diagnosed with bulbar onset MG, controlled asthmatic with suspected fentanyl allergy, was scheduled for laparoscopic surgical repair of paraesophageal hiatal hernia under GA.

Since 2013, the patient was diagnosed with Osserman

stage IIb MG and has been receiving regular treatment with pyridostigmine bromide 180 mg twice daily, prednisolone 5 mg daily, budesonide nebulizer and ipratropium-albuterol as needed. The patient has a surgical history of hysterectomy, upper gastrointestinal endoscopy, colonoscopy and sigmoidoscopy, during which she experienced itching following the procedure that was attributed to fentanyl; however, no skin test was conducted. On pre-anesthesia evaluation, she reported to be in her optimum condition. Physical examination revealed bilateral facial weakness, lip ptosis with tongue movement and swallowing difficulties. She was able to rise from a seated position but experienced arm and leg weakness bilaterally where finger flexors and hip flexors had grades of 3+ and 4+ in the muscles grading scale. Her activities of daily living (ADL) score was 6 and had a New York Heart association (NYHA) class II A. Spirometry findings showed a forced vital capacity (FVC) of 3.3 L and a forced expiratory volume in one second (FEV1) of 2.7 L and an FEV1/FVC ratio of >70%. Neurology consultation was done, and the patient was fit for surgical intervention from neurology side. The patient was scheduled for surgery under GA. She was categorized as American Society of Anesthesiologists classification of stage III (ASA III). Critical care bed was requested for possible postoperative ventilation. The patient received her morning regular medications as usual. Prior to induction, standard intraoperative monitoring (ASA monitoring) were applied including three-lead ECG, noninvasive blood pressure, body temperature and end-tidal carbon dioxide (EtCO2) monitoring.

A test dose of remifentanil was given under close monitoring and after 10 minutes the patient did not exhibit any signs and symptoms of allergy/anaphylaxis. The patient was preoxygenated for 3 minutes followed by induction of GA with propofol 1.5 mg/kg, ketamine 20 mg, lidocaine 1 mg/kg, and remifentanil 0.5µg/kg. No NMBAs were used. Hydrocortisone 100 mg and dexamethasone 8 mg were given on induction. Vital signs during induction are summarized in **table I.**

Intubation was uncomplicated and uneventful. Immediately after the intubation, a transesophageal temperature probe was placed as well as a radial arterial line for invasive arterial monitoring.

Maintenance of GA was achieved using remifentanil in a continuous intravenous (IV) infusion of 0.05-0.1µg/kg/min during the surgery and inhalational anesthesia using desflurane in a low gas flow air/oxygen mixture.

During anesthesia, mechanical ventilation parameters were adjusted based on standard anesthetic evaluation. Doses of anesthetic agents were modified according to the clinical assessment and depth of anesthesia. The management described above resulted in acceptable circulatory stabilization (Table I).

The patient underwent laparoscopic repair of paraesophageal hiatal hernia without implantation of mesh. Normal laparoscopic insufflation pressure was required for optimum working conditions (less than 15 mmHg). Blood loss was minimal, and the procedure took over 4 hours to complete.

	Heart rate (beats/minutes)	NIBP (mm Hg)
Induction	72	159/91
Start of surgery	69	100/61
End of surgery	71	112/68
Extubation	120	160/110

Table 1. Vital signs throughout surgery.

After the completion of skin suturing, emergence from GA was initiated. Remifentanil infusion was discontinued, and a 20 mg of meperidine was administered. The patient regained efficient respiration, and full neurological contact was re-established.

Upon emergence and extubation, the patient was transferred to the post anesthesia care unit (PACU) after ensuring full verbal contact and recovery of consciousness level as baseline were achieved and maintained. The patient's respiratory function and circulatory status were uncompromised with 5 liters (L) of supplemental oxygen. Postoperative analgesia was adequate with 1 gram paracetamol, 30 mg ketorolac and further meperidine injection with a total dose of 50 mg.

The patient stayed in PACU for two hours before being transferred back to the surgical ward. During the first 24 hours, she was kept on 2 L of Oxygen therapy via nasal cannula. She received her regular treatment during her hospital stay and was discharged on the 6th day postoperatively.

DISCUSSION:

MG is a concerning disease for anesthesiologists as it is associated with a high risk for postoperative weakness and respiratory failure, hence it is extremely important for MG patients to continue their prescribed medications in order to prevent further respiratory or bulbar weakness. Our patient was advised to continue her regular daily medications, especially on the day of surgery during the preoperative fasting period. For optimal management of MG cases, it is crucial to understand both the primary medical condition and any coexisting illnesses. Therefore, preoperative preparation for elective surgery should be coordinated with the patient's neurologist, as was done in our case. Additionally, whenever possible, these patients should be in their optimized condition and preferably scheduled for morning procedures, when their muscle power is at its highest, to minimize the risk of postoperative complications [3].

In addition to the standard preoperative evaluation, patients with myasthenia gravis require thorough assessment for bulbar symptoms, including dysphagia, dysarthria, nasal speech, and low-intensity speech, as these manifestations may increase the risk of aspiration. Comprehensive history that includes any prior episodes of myasthenic crisis and the necessity for endotracheal intubation is paramount as well as a thorough evaluation of respiratory function, focusing on indicators of respiratory muscle weakness, shortness of breath, and dyspnea. Our patient

exhibited signs and symptoms of bulbar involvement early in the disease however, respiratory involvement was minimal and pulmonary function tests were reassuring [4].

In literature, preoperative and postoperative risks of myasthenia crisis after Surgery were addressed interchangeably, reported preoperative factors include bulbar symptoms, history of preoperative myasthenic crisis, preoperative serum level of anti- acetylcholine receptor antibody>100 nmol/L, and intraoperative blood loss>1000 mL. Osserman stage IIb + III + VI , perioperative usage of pyridostigmine of more than >240 mg, abnormal pulmonary function test (PFT) reporting VC <2.9 L and disease duration of more than 2 years [5,6,7,15].

Our patient suffered from MG for more than 10 years with Osserman stage IIb, requiring 360 mg of pyridostigmine per day along with bulbar manifestations which made her a candidate for postoperative mechanical ventilation and peri operative complications.

MG patients are often on long-term corticosteroid therapy and may be susceptible to adrenal crisis after abrupt withdrawal of glucocorticoids. Stress-dose may be administered depending on the surgical procedure and other stress factors [8].

Our patient was taking 5 mg/day of prednisolone for over 6 months and known case of bronchial asthma so we decided to administer 100 mg of hydrocortisone prior to induction in addition to her usual corticosteroid regimen.

Although their use can ensure good surgical condition especially during laparoscopic surgeries neuromuscular blocking agents (NMBA) should be avoided if possible, to prevent further reduction in postsynaptic acetylcholine receptors. Since MG patients have unique sensitivity to nondepolarizing NMBAs and are resistant to depolarizing NMBAs. Depolarizing NMBA like Succinylcholine is not recommended for MG patients because it has a slower onset of action and a delayed recovery. Moreover, neostigmine may be ineffective in reversing the residual effects of non-depolarizing NMBAs if acetylcholinesterase is already fully inhibited by pyridostigmine. We have avoided the use of NMBAs in our patient. Both induction and maintenance of anesthesia were accomplished by intravenous and inhalational agents.

Many anesthetists prefer the use of rocuronium-sugammadex when NMBA is required as assumed it is safe and effective; however few studies in literature have proved failure of such regimen in some cases. MG patients need to be reviewed on regular basis and caution before any supplemental NMBA dose as response may be unexpected [1].

Propofol is advantageous in these patients due to its short action and lack of effect on neuromuscular transmission.

According to multiple reports, the use of etomidate, ketamine, and thiopental has not led to any incidents [9].

Opioids, while not impairing neuromuscular transmission may cause central respiratory depression henceforth, short-acting opioids like remifentanil with its rapid elimination and brief half-life, are particularly suitable [10].

Our concern was the patient's history of alleged allergy to fentanyl, however there is limited literature on the cross-reactivity between fentanyl and remifentanil but documented cases do exist. Hence a test dose was given with caution in our patient with close monitoring to observe for any signs of allergic reaction which did not develop [11,12].

Studies indicate that volatile anesthetics may impair neuromuscular transmission in terms of muscle relaxation or sensitivity to NMBAs as in our case. However, among inhalational anesthetics, desflurane may have a lesser impact compared to sevoflurane and isoflurane, due to its rapid onset and clearance, as what was utilized in our case. GA was maintained with remifentanil infusion and desflurane in a low gas flow air/oxygen mixture [13].

Using desflurane with remifentanil and low gas flow provides a balanced approach to anesthesia management, reducing the risk of residual paralysis while maintaining a stable anesthetic depth. A low gas flow mixture supports conservation of anesthetic agents, reducing cost and environmental impacts whilst maintaining patient's safety [14].

It is crucial for MG patients to verify adequate spontaneous ventilation before extubation. In our case the criteria include sufficient return of consciousness, tidal volume ≥ 5 ml/kg, $PaCO2 \leq 50$ mmHg, $PaO2 \geq 90$ mmHg, and a respiratory rate ≤ 30 breaths/min [16].

Pain management in MG patients is crucial to prevent stress-induced myasthenic crisis that can lead to intensive care admission. A comprehensive, individualized and multidisciplinary approach to pain management can significantly improve patient functional outcomes and satisfaction. If opioids are required, small doses of short-acting injections are preferred until lasting pain relief is achieved to avoid central respiratory depression. Additionally, nonsteroidal anti-inflammatory drugs (NSAIDs) may help manage pain in MG patients by decreasing opioid requirements. Moreover, adjuvant therapy (such as gabapentin and pregabalin) can help manage the pain without affecting respiratory function [17].

CONCLUSION:

MG, the most common neuromuscular junction disorder, is characterized by skeletal muscle weakness that worsens with use due to autoimmune destruction of acetylcholine receptors. Respiratory muscle function, pharmacotherapy, and disease history are key factors in assessing postoperative respiratory failure risk. A clear understanding of the disease allows for tailored management during pre-, peri-, and postoperative stages.

REFERENCES:

1. Fernandes, H., Ximenes, J.L.S., Nunes, D.I. *et al.* Failure of reversion of neuromuscular block with sugammadex in patient with myasthenia gravis: case report and brief review of literature. *BMC Anesthesiol* 19, 160 (2019). https://doi.org/10.1186/s12871-019-0829-0
2. Rudzka-Nowak, A., C Piechota, M. (2011). Anaesthetic management of a patient with myasthenia gravis for abdominal surgery using sugammadex. Archives of Medical Science, 2, 361–364. doi:10.5114/aoms.2011.22094
3. Jamal, B. T., C Herb, K. (2009). Perioperative management of patients with myasthenia gravis: Prevention, recognition, and treatment. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontics, 107*(5), 612-615.
4. Daum P, Smelt J, Ibrahim IR. Perioperative management of myasthenia gravis. BJA Educ. 2021 Nov;21(11):414-419. doi: 10.1016/j.bjae.2021.07.001. Epub 2021 Aug 19. PMID: 34707886; PMCID: PMC8520038.
5. Watanabe, A., Watanabe, T., Obama, T., Mawatari, T., Ohsawa, H., Ichimiya, Y., Takahashi, N., Kusajima, K., C Abe, T. (2004). Prognostic factors for myasthenic crisis after transsternal thymectomy in patients with myasthenia gravis. *Journal of Thoracic and Cardiovascular Surgery, 127*(3), 868.
6. Liu C, Liu P, Zhang XJ, Li WQ, Qi G. Assessment of the risks of a myasthenic crisis after thymectomy in patients with myasthenia gravis: a systematic review and meta-analysis of 25 studies. J Cardiothorac Surg. 2020 Sep 29;15(1):270. doi: 10.1186/s13019-020-01320-x. PMID: 32993739; PMCID: PMC7526111.
7. Leuzzi G, Meacci E, Cusumano G, et al. Thymectomy in myasthenia gravis: proposal for a predictive score of postoperative myasthenic crisis. Eur J Cardiothorac Surg. 2014;45:e76–e88. doi: 10.1093/ejcts/ezt641.
8. Fraser, C. G., Preuss, F. S., C Bigford, W. D. (1952). Adrenal atrophy and irreversible shock associated with cortisone therapy. *Journal of the American Medical Association, 14S*(17), 1542-1543.
9. Postevka E. Anesthetic implications of myasthenia gravis:a case report. AANA J. 2013;81:386-8.] [Abel M, Eisenkraft JB. Anesthetic implications of myasthenia gravis. Mt Sinai J Med. 2002;69:31-7.
10. Abe N, Kunisawa T, Sasakawa T, Takahata O, Iwasaki H. Anesthetic management using remifentanil target controlled infusion without muscle relaxants in two patients with myasthenia gravis. Masui. 2010;59:727-30.
11. Baldo B. A. (2023). Allergic and other adverse reactions to drugs used in anesthesia and surgery. *Anesthesiology and Perioperative Science, 1*(2), 16. https://doi.org/10.1007/s44254-023-00018-2
12. Baldo, B. A., C Pham, N. H. (2012). Histamine-releasing and allergenic properties of opioid analgesic drugs: resolving the two. *Anaesthesia and intensive care, 40*(2), 216–235. https://doi.org/10.1177/0310057X1204000204.]
13. Hagberg, C. A., C Benumof, J. L. (2002). "Anesthesia for Patients with Myasthenia Gravis." *Anesthesia & Analgesia*, 95(3), 771-779.
14. Baum, J. A., C Aitkenhead, A. R. (1995). Low-flow anaesthesia. *Anaesthesia, 50 Suppl*, 37–44. https://doi.org/10.1111/j.1365-2044.1995.tb06189.x
15. Blichfeldt-Lauridsen L, Hansen BD. Anesthesia and myasthenia gravis. Acta Anaesthesiol Scand. 2012;56:17-22.
16. Datt V, Tempe DK, Singh B, Tomar AS, Banerjee A, Dutta D, et al. Anesthetic management of patient with myasthenia gravis and uncontrolled hyperthyroidism for thymectomy. Ann Card Anaesth. 2010;13:49-52.
17. [Haroutiunian, S., Lecht, S., Zur, A. A., Hoffman, A., C Davidson, E. (2009). The challenge of pain management in patients with myasthenia gravis. *Journal of pain & palliative care pharmacotherapy, 23*(3), 242–260. https://doi.org/10.1080/15360280903098523]