CASE REPORT

Anesthesia Spearheading Perioperative Safety Efforts in a Patient with Inclusion Body Myositis: A Case Report

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ABSTRACT

Anesthesiologists have been at the forefront of initiatives addressing perioperative patient safety. As anesthesia has no direct therapeutic benefit, its risk must be minimized. At times the surgery is simple but the patient's condition complicates anesthetic management, increasing the risk for complications. This report describes the anesthetic management of an adult patient diagnosed with inclusion body myositis (IBM), a rare inflammatory degenerative myopathy, who initially presented with decreased motor function in both lower and upper extremities causing him to be bedbound for two years. Due to the progression of his disease, he eventually developed dysphagia, hence he was scheduled for esophagoscopy, cricopharyngeal Botox injection, and percutaneous endoscopic gastrostomy. As patients with IBM are at risk for exaggerated sensitivity to neuromuscular blockers and respiratory compromise, anesthesia was at the helm of a multidisciplinary team approach. The perioperative management centered on preoperative optimization, prevention of aspiration, avoidance of anesthetics that may trigger malignant hyperthermia, and prevention of postoperative pulmonary complication. The hospital course was uncomplicated and the patient was discharged well after one day. This report emphasizes how improvements in resources, technology, and healthcare delivery, especially in anesthesia, help prevent perioperative adverse events.

Keywords: patient safety, inclusion body myositis, malignant hyperthermia, case report

INTRODUCTION

Patient safety is the "avoidance, prevention and amelioration of adverse outcomes or injuries stemming from the process of healthcare." Despite continued efforts to reduce harm in the perioperative period, adverse events were estimated to occur in approximately 30% of hospital admissions, with 50% of them considered to be preventable.[1] Having a central role in perioperative care, anesthesiologists can substantially contribute to patient safety and clinical outcomes.

Inclusion body myositis (IBM) is a rare inflammatory-degenerative myopathy typically affecting adults after the 5th or 6th decade of life.[2] It has an estimated incidence of only 2 to 12 cases per million.[2] IBM can be classified as sporadic, familial or hereditary. Sporadic IBM is not an inherited Mendelian disease; however, studies have shown that several genetic risk factors involving the human leukocyte antigen region are still contributory. Reports of familial distribution of the disease revealed an autosomal dominant pattern. On the other hand, hereditary IBM follows either autosomal recessive or dominant inheritance. Several mutations of different genes have been identified.[3] It is characterized by progressive bilateral but asymmetrical weakness commonly

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involving axial muscles such as oropharyngeal muscles, diaphragm, and limb girdle. As such, these patients may present with dysphagia, decreased respiratory function, and loss of ambulation, which may require surgical intervention. To ensure perioperative safety, the following must be addressed: (1) risk of aspiration due to impaired clearance of secretions, (2) susceptibility for malignant hyperthermia (MH), (3) exaggerated sensitivity to neuromuscular blockers, and (4) risk for postoperative pulmonary complications which may require invasive or noninvasive ventilatory support.[2]

There are very few existing literatures on the anesthetic management of a patient with inclusion body myositis.[2,4-8] This case report highlights how risk reduction and prevention of adverse events guaranteed patient safety in a patient with IBM who received general anesthesia for esophagoscopy, cricopharyngeal Botox injection, and percutaneous endoscopic gastrostomy.

CASE PRESENTATION

A 54-year-old male, diagnosed with IBM since 2014, was scheduled for esophagoscopy, cricopharyngeal Botox injection, and percutaneous endoscopic gastrostomy due to a one-year history of dysphagia, which was unresponsive to intravenous immunoglobulin. His dysphagia was severe enough to cause fatigue and inadequate food intake. He weighed 70 kg and had a body mass index of 22.7. He was maintained on Ivabradine due to a history of heart failure (functional class II-III). He had been bedbound since 2019 and requires assistance in activities of daily living. Previous surgeries prior to his diagnosis of IBM were unremarkable. Physical examination revealed atrophic muscles all over, contractures in the upper extremities, and hypotonia and decreased motor function in all extremities. Cognitive, cranial, and sensory exams were normal. Preoperative fiberoptic endoscopic evaluation of swallowing showed no aspiration; however, the patient still required several swallows to partially clear a spoon of food sample. Chest radiograph showed low lung volume, elevated left hemidiaphragm, and a possible infectious or inflammatory process versus confluence of vessels. Arterial blood gas (ABG) showed respiratory alkalosis with adequate oxygenation. Electrocardiogram was normal while 2D echocardiogram showed diastolic dysfunction with preserved systolic function. Blood counts and chemistries were unremarkable. A multidisciplinary meeting involving otolaryngology, anesthesiology, neurology, pulmonology, cardiology, gastroenterology and rehabilitation medicine was held before the surgery to discuss the perioperative management of the patient. The patient was deemed low risk for major cardiac events and postoperative acute kidney injury but high risk for postoperative pulmonary complications. Anesthesia-wise, the patient was at an increased risk for aspiration and malignant hyperthermia. Given these risks, the focus of the discussion was on the prevention of adverse events. The anesthesiologist gave a detailed perioperative plan after consolidating inputs from other services. Because the patient was assessed to be at high risk for developing postoperative pulmonary complications, delayed extubation and possible transition to bilevel positive airway pressure (BiPAP) ventilation were agreed upon.

On the day of the surgery, the necessary measures to avoid malignant hyperthermia were done. The Dräger Fabius anesthesia machine was prepared by replacing the carbon dioxide absorbent, removing the vaporizers, and mounting a new breathing circuit. The machine was then flushed for 1.5 hours with the following settings: oxygen flow rate at 10 L/min, tidal volume of 600 mL and rate of 15/min. A full oxygen tank and a backup ventilator with the capacity for BiPAP ventilation were made available. All medications and supplies which will be used in case of malignant hyperthermia were prepared, particularly dantrolene, sodium bicarbonate, dextrose, insulin, calcium gluconate, and cold intravenous fluids. All drugs that may trigger malignant hyperthermia were avoided.

Before surgery, the patient received omeprazole, paracetamol and antibiotic prophylaxis. The patient was placed in a head up position upon transfer to the operating room bed. The following monitors were attached to the patient: electrocardiogram, pulse oximeter, noninvasive blood pressure, peripheral nerve stimulator, and core body thermometer. Instead of using the anesthesia machine, the anesthesiologist decided to use the available oxygen tank and do manual ventilation using a Jackson-Rees circuit since the procedure was short. A sampling line for the capnogram was attached to the breathing circuit. The patient was preoxygenated and a rapid sequence induction was done using fentanyl (3 mcg/kg), propofol (2 mg/kg), and rocuronium (1.2 mg/kg). The patient was then intubated and manually ventilated. Anesthesia was maintained with propofol and remifentanil infusions at 50-120 mcg/kg/min and 0.05-0.2 mcg/kg/min, respectively. Additional doses of rocuronium at 0.1 mg/kg were given based on neuromuscular blockade monitoring. Fluid therapy was restricted. Surgery was unremarkable and lasted for 1 hour and 4 minutes. At the end of surgery, neuromuscular blockade was reversed with sugammadex (2 mg/kg) and a 100% train-of-four value was achieved. Throughout the procedure, there were no noted hyperthermia, unexplained sinus tachycardia or tachyarrhythmia, or marked increase in end tidal CO_2.

The patient was then transferred to the postanesthesia care unit (PACU), fully awake but still intubated, with the remifentanil infusion continued. An ABG was done at PACU after committing the patient to mechanical ventilation. Results showed normal acid-base status and adequate oxygenation. About an hour after surgery, spontaneous breathing trial was successful, permitting extubation. BiPAP ventilation was no longer employed as the patient was able to tolerate oxygen support via face mask. The patient was transferred to his room and his condition remained stable. Feeding was progressed by gastroenterology, while bedside

therapy was initiated by rehabilitation medicine. The patient was eventually discharged the next day.

DISCUSSION

Reports on patients with IBM who underwent surgery often described the risks and complications related to the disease itself or to anesthesia, rather than those arising from surgery. A case series by Mortenson et al. reviewed the perioperative outcomes of patients with IBM who underwent general anesthesia, with emphasis on respiratory complications. Of the 18 cases included in the study, five had delayed extubation – three of whom died while one underwent tracheostomy for long-term ventilatory support. Outcomes were dependent primarily on conditions related to advanced IBM.[4] In a case report by Igari et al., a patient for video-assisted thoracic surgery received total intravenous anesthesia consisting of propofol, remifentanil and reduced doses of rocuronium (due to concerns for postoperative respiratory depression). At the end of the surgery, the authors were unable to extubate their patient despite reversal of neuromuscular blockade. This was attributed to the underlying pulmonary dysfunction related to IBM.[7] In another case report, an elderly man with IBM who underwent jejunostomy was intubated without the use of a neuromuscular blocker. The patient developed postoperative aspiration pneumonia.[8]

Since patients with IBM are at risk of pulmonary complications, the utility of a preoperative pulmonary function test (PFT) was raised during the multidisciplinary meeting. Some suggested getting a baseline pulmonary status with pulmonary function testing.[6,9]

However, given that a preoperative PFT is not predictive of postoperative pulmonary complications and of the need for postoperative ventilatory support,[10,11] the team decided not to delay the surgery to obtain a pulmonary function test. In addition, PFT is more useful in monitoring treatment response.[11] As the patient is not in respiratory failure and the surgery to be done is not a form of treatment for respiratory dysfunction, obtaining a PFT was deemed not crucial. Instead, the perioperative management for this case focused on the prevention of adverse events that result from the disease itself and from anesthesia. Patients with IBM are at risk of the following: aspiration, malignant hyperthermia, exaggerated sensitivity to neuromuscular blockers, and postoperative pulmonary complications.

The increased risk for aspiration is due to the impaired clearance of secretions from the progressive weakness and atrophy of oropharyngeal muscles.[2,5] To decrease the risk of aspiration, the patient in this case was given omeprazole, was placed in a head up position, was induced with rapid sequence induction, and was intubated.

At present, there is still no definitive evidence for a direct correlation between IBM and MH. Nevertheless, the avoidance of triggering agents [by using total intravenous anesthesia (TIVA) and nondepolarizing muscle relaxants] is prudent because patients with IBM typically have elevated creatinine kinase (CK) levels, which may indicate an increased susceptibility to MH.[12] A study by the Mayo Clinic revealed MH susceptibility (through caffeine-halothane contracture test on muscle biopsies) in 49% of patients with elevated CK.[12] Despite this concern and contrary to what was used in other published studies, the case series by Mortenson et al. reported no adverse events in those patients who received inhalational anesthesia.[4]

The anesthesia used for this case was TIVA using propofol and remifentanil infusions, and intermittent boluses of rocuronium to avoid malignant hyperthermia. Given that the estimated duration of surgery was short, manual ventilation using a Jackson-Rees circuit connected to an oxygen tank was used instead of an anesthesia machine to further reduce the risk. Availability of the necessary equipment, supplies and medications, particularly dantrolene, was ascertained in case of malignant hyperthermia.

Some published articles report the use of reduced doses or avoidance of muscle relaxants in patients with IBM because of the possibility of exaggerated sensitivity to these drugs and prolonged duration of action in inflammatory myopathies including IBM.[7,8,13,14] However, there are no conclusive data regarding the pharmacokinetics, pharmacodynamics, and safety of neuromuscular blockers in IBM. In fact, the case series by Mortenson et al. reported 16 patients with IBM who underwent surgery and received either depolarizing or nondepolarizing agents. All had uneventful perioperative outcomes except those who were anticipated to require postoperative mechanical ventilation.[4]

The use of succinylcholine in patients with primary muscle diseases is discouraged because of the possibility of an exaggerated hyperkalemic response. Nonetheless, succinylcholine has been used in some cases of IBM with no obvious evidence of hyperkalemia.[4]

For this case, rocuronium was used to avoid a hyperkalemic response. Administration was guided by neuromuscular blockade monitoring to avoid excessive dosing. To reverse the effects of rocuronium, sugammadex was used, given its association with a lower incidence of major pulmonary complications.[15]

IBM may involve respiratory muscles, leading to diaphragmatic dysfunction and diminished respiratory effort. This increases the risk for postoperative pulmonary complications such as aspiration pneumonia and respiratory failure, needing prolonged ventilatory support in some cases.[5,16]

Aside from the aforementioned lowering of aspiration risk, several approaches were considered in this case to decrease the risk of postoperative pulmonary complications. A restrictive fluid strategy was employed. A spontaneous breathing trial was done to ensure readiness for extubation. Although not used, BiPAP ventilation was considered if extubation was not tolerated. Postoperative rehabilitation was also facilitated.

CONCLUSION

This report describes the successful prevention of perioperative complications, namely aspiration, malignant hyperthermia, exaggerated sensitivity to neuromuscular blockers, and postoperative pulmonary complications, in a high-risk patient for a low-risk surgery. With multiple services involved in the care of this patient, anesthesia took the lead in synthesizing all inputs and coordinating all aspects of perioperative management. With the aid of improved resources, technology, and healthcare delivery, initiatives centered on the avoidance of anesthesia-related adverse events and postoperative pulmonary complications assured patient safety and a successful outcome in the conduct of general anesthesia for a patient with inclusion body myositis for palliative surgery. Since this report mainly focused on prevention of adverse effects, further studies are needed to establish the safety of anesthetic agents such hypnotics, analgesics, and neuromuscular blockers in order to expand the options for anesthesia management in patients with inclusion body myositis.

Informed Consent

The patient has signed PGH Form No. P-31005(a) and PGH Form No. Q-310051 (Broad Consent Form for Use of Health Information) upon patient admission. These two forms have received institutional legal approval.

Statement of Authorship

Both authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

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