Acta Med. Nagasaki 67: 27－30

Case Report

Anesthetic management with remimazolam in a patient with inclusion body myositis: a case report

Ryosuke Shintani[a], Hanako Ura[a], Taku Teshima[b], Keiko Tashiro[b], Yuri Miyazaki[a], Tatsuhito Tanaka[b], Takuji Maekawa[b], Tetsuya Hara[a]

[a] Department of Anesthesiology and Intensive Care Medicine, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan
[b] Department of Anesthesiology, Sasebo City General Hospital, Sasebo, Japan

Background: Inclusion body myositis (IBM), an inflammatory muscle disease, sometimes leads to swallowing impairment. The anesthetic management of patients with IBM should be carefully performed to prevent the development of postoperative pulmonary complications (PPCs). Prompt recovery of consciousness from general anesthesia may contribute to the prevention of PPCs. Remimazolam, a newly developed general anesthetic, can provide early and reliable awakening; however, its use in patients with IBM has not been reported.
Case presentation: A 76-year-old man with IBM experienced swallowing impairment, for which laparoscopy-assisted gastrostomy was planned. Total intravenous anesthesia was performed with remimazolam, remifentanil, and rocuronium. Upon awakening from anesthesia following flumazenil administration, he completely recovered consciousness. After extubation, he was able to protrude his tongue, cough, and breathe comfortably. There were no PPCs, and the patient's postoperative course was uneventful.
Conclusions: PPCs should be prevented in patients with IBM, and anesthetic management with remimazolam may help with this.
ACTA MEDICA NAGASAKIENSIA 67: 27−30, 2023

Key words: Inclusion body myositis, general anesthesia, remimazolam, postoperative pulmonary complications

Background

Inclusion body myositis (IBM) is an inflammatory muscle disease that sometimes leads to swallowing impairment [1]. Care should be taken to prevent postoperative pulmonary complications (PPCs) in patients with IBM. IBM is rare, and the optimal method for the anesthetic management of patients with IBM remains to be established. In 2020, Japan became the first country to approve the use of remimazolam, a benzodiazepine sedative, for use as a general anesthetic. Remimazolam is a short-acting drug, and its metabolites have no sedative effect. Since flumazenil, an antagonist, can be administered, early and reliable awakening can be expected [2]. Prompt recovery of consciousness from general anesthesia prevents PPCs, including aspiration pneumonia. The use of remimazolam for general anesthesia in patients with IBM has not been reported previously.

Here, we present a case of a patient with IBM in whom remimazolam was used for general anesthesia for laparoscopy-assisted gastrostomy.

Case presentation

A man in his 70s with a height of 159 cm and weight of 55 kg. The patient was receiving lixiana, verapamil, azelnidipine,

Address correspondence: Ryosuke Shintani, MD
Department of Anesthesiology and Intensive Care Medicine, Nagasaki University Graduate School of Biomedical Sciences,
1-7-1 Sakamoto, Nagasaki 852-8501, Japan
Phone: +81-95-819-7370, Fax: +81-95-819-7373, E-mail: r-shintani@nagasaki-u.ac.jp

Received June 29, 2022; Accepted May 11, 2023

olmesartan, and furosemide for chronic atrial fibrillation and hypertension. He has been a smoker for several decades. He had been diagnosed with IBM 9 months previously based on symptoms of progressive muscle weakness in the extremities and swallowing impairment. His swallowing function worsened despite the administration of steroid pulse therapy and intravenous immune globulin, and he requested gastrostomy. Since the abdominal wall was in contact with the greater curvature, a high risk of bleeding due to vascular injury could be expected if gastroesophageal endoscopy–assisted gastrostomy was performed; therefore, laparoscopy-assisted gastrostomy was planned. Preoperative pulmonary function test results showed obstructive lung disease, with a vital capacity of 3.75 L (118%) and forced expiratory volume in the first second of 1.95 L (56%). The serum creatine phosphokinase level was 555 U/L (normal range: 43–272 U/L). The results of other preoperative examinations were normal.

In addition to standard monitoring, the bispectral index (BIS) and electromyographic muscle relaxation were monitored. Anesthesia was induced with remimazolam, remifentanil, and rocuronium after adequate aspiration of oral secretions. Remimazolam was administered at 6 mg/kg/h, and the patient lost consciousness approximately 120 s later. Rocuronium was administered in doses of 10 mg each, and the train-of-four count was zero after a total dose of 30 mg had been administered. No adverse events occurred during endotracheal intubation. Anesthesia was maintained with remifentanil (0.1–0.25 μg/kg/min) and remimazolam (1 mg/kg/h) to maintain the BIS value at 40–60. The train-of-four count at 40 min after tracheal intubation was one. Therefore, an additional 10 mg of rocuronium was administered for muscle relaxation; no additional administration was required thereafter. At the end of the surgery, rectus sheath block was performed, followed by administration of sugammadex (100 mg) and flumazenil (0.5 mg) for anesthesia reversal. Administration of sugammadex increased the train-of-four ratio from 26% to 100%. (Table 1) Extubation was performed 28 minutes after the end of surgery and 4 minutes after the administration of flumazenil. After extubation, the patient was able to protrude his tongue, cough, and breathe comfortably. The operation time was 78 min, and the anesthesia time was 146 min. The postoperative course was uneventful; no respiratory complications, such as aspiration pneumonia, occurred. The patient was discharged home on the ninth postoperative day.

Written informed consent was obtained from the patient for the publication of this case report.

Table 1 : Summary of drug administration for each phase of anesthesia

	Induction	Maintenance	Emergence
Remimazolam*	6 mg/kg/h	1 mg/kg/h	Antagonized by flumazenil 0.5 mg
Remifentanil*	0.5μg/kg/min	0.1-0.25μg/kg/min	End of administration
Rocuronium**	30 mg (10 mg each)	10 mg (just once)	Antagonized by sugammadex100mg

* Drug doses were titrated so that BIS values of 40 – 60.

** To achieve a train-of-four count of zero, 10 mg of rocuronium was administered.

Discussion

IBM is characterized by painless muscle weakness and atrophy, mainly in the quadriceps and forearm flexors [3–5]. The disease is chronic and progressive, and other muscles may also be affected [3–5]. Compared to other inflammatory muscle diseases, IBM is also characterized by atrophy of muscles related to swallowing, such as the pharyngeal muscle and the upper esophageal sphincter [6]. In fact, dysphagia occurs in about 50% of IBM patients, and aspiration pneumonia is an important cause of death in these patients [1]. In the perioperative period, residual anesthesia and somnolence after general anesthesia further impair swallowing function. Therefore, during the anesthetic management of patients with IBM, it is necessary to pay attention to PPCs associated with swallowing impairment [6].

However, owing to its rarity, there are few reports and no guidelines on the anesthetic management of patients with IBM. Mortenson et al. summarized the clinical course of 18 patients with IBM at their institution, which is the largest number of cases reviewed to date [6]. They reported that the anesthetic management of patients with IBM with moderate disease severity could be safely performed with no perioperative pulmonary complications. In contrast, Igari et al. reported a case of prolonged postoperative requirement of ventilatory support in a 40-year-old man with IBM who underwent thoracoscopic partial pneumonectomy for spontaneous pneumothorax [7]. In that patient, the disease progressed to the point of involvement of the respiratory muscles, which is rare in patients with IBM. Nakano et al. reported a case of aspiration pneumonia on the second postoperative day in a 76-year-old man with IBM who underwent an open jejunostomy [8]. The patient's swallowing function was impaired to the extent that he required jejunostomy.

Our findings and those from previous cases indicate that care should be taken to prevent PPCs in patients with IBM

undergoing general anesthesia, especially in those with respiratory muscle affection and severe swallowing impairment. Prompt recovery of consciousness from general anesthesia may help with this. This is because prolonged somnolence after general anesthesia further increases the risk of PPC in IBM patients, who are already at high risk of aspiration. At least, the level of consciousness should be restored to a point where the patient can extend the tongue and cough and take deep breaths. To achieve this, we used remimazolam to induce and maintain general anesthesia, which was successfully reversed using flumazenil, and the patient had clear consciousness at the time of extubation.

Propofol has a suppressive effect on pharyngeal muscle strength, and no specific antagonist exists [9]. Midazolam contains a sedative metabolite and its use predisposes patients to postoperative delirium, which can lead to impaired postoperative consciousness [10]. Remimazolam, on the other hand, is a short-acting drug, and its metabolites have no sedative effect. Since flumazenil, an antagonist, can be administered, early and reliable awakening can be expected [2]. For these reasons, we consider remimazolam the most preferred general anesthetic drug for IBM patients.

Malignant hyperthermia (MH), which has not been reported in patients with IBM but has been reported to be associated with several congenital myopathies, is another possible complication in patients with IBM [11–13] . It is recommended that patients with resting hyperCKemia should be monitored and considered to be at high risk of developing MH, as reports exist of them who have mutations in *RYR1*, the causative agent of MH [14]. In fact, the patient in this case had an elevated resting serum creatine phosphokinase level. Furthermore, patients with IBM may have increased sensitivity to muscle relaxants owing to muscle atrophy. Inhaled anesthetics potentiate the effects of muscle relaxants [15]. Consequently, intravenous anesthetics may be preferable to inhaled anesthetics for patients with IBM, and from this perspective, remimazolam may be a good general anesthetic for patients with IBM.

In conclusion, there are no guidelines for anesthetic management in patients with IBM; therefore, anesthesia should be planned on a case-by-case basis. Respiratory complications associated with dysphagia, malignant hyperthermia, and sensitivity to muscle relaxants should be considered, and the use of remimazolam may help to prevent these complications.

List of abbreviations

IBM: inclusion body myositis
PPC: postoperative pulmonary complication
BIS: bispectral index
MH: malignant hyperthermia

Declarations

Ethics approval and consent to participate

Ethical approval for case reports is not required at our institution.

Consent for publication

Written informed consent was obtained from the patient for the publication of this case report.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interests.

Funding

The authors have no funding sources to declare.

Authors' contributions

RS acquired the data and prepared the manuscript. HU, TTe, KT, YM, TTa, TM, and TH reviewed the manuscript. TH also supervised the writing of the report. The manuscript has been read and approved by all the authors.

Acknowledgments

We would like to thank www.editage.jp for English language editing.

References

1. Benveniste O, Guiguet M, Freebody J, et al. Long-term observational study of sporadic inclusion body myositis. *Brain* 134:3176–84, 2011
2. Kilpatrick GJ, McIntyre MS, Cox RF, et al. CNS 7056: a novel ultra-short-acting benzodiazepine. *Anesthesiology* 107:60–6, 2007
3. Beyenburg S, Zierz S, Jerusalem F. Inclusion body myositis: clinical and histopathological features of 36 patients. *Clin Investig* 71:351–61, 1993
4. Lotz BP, Engel AG, Nishino H, Stevens JC, Litchy WJ. Inclusion body myositis. Observations in 40 patients. *Brain* 112:727–47, 1989
5. Amato AA, Gronseth GS, Jackson CE, et al. Inclusion body myositis: clinical and pathological boundaries. *Ann Neurol* 40:581–6, 1996
6. Mortenson AR, Sprung J, Cavalcante AN, Watson JC, Weingarten TN. Inclusion body myositis and anesthesia: a case series. *J Clin Anesth* 31:282–7, 2016
7. Igari Y, Ito Y, Nagaya K. Anesthesia for pneumothorax surgery in a patient with type II chronic respiratory failure associated with inclusion body myositis. *Masui* 63:172–4, 2014 [Japanese]

8. Nakano N, Satsumae T, Mizutani T, Kimura M, Tokuwaka J, Tanaka M. Anesthetic management for a patient with inclusion body myositis. *J Jpn Soc Clin Anesth* 32:809–13, 2012 [Japanese].
9. Bryson HM, Fulton BR, Faulds D. Propofol -An update of its use in anaesthesia and conscious sedation- *Drugs* 50:513-59, 1995
10. Maldonado JR. Neuropathogenesis of delirium: review of current etiologic theories and common pathways. *Am J Geriatric Psychiatry* 21:1190–1222, 2013
11. Rosenberg H, Pollock N, Schiemann A, Bulger T, Stowell K. Malignant hyperthermia: a review. *Orphanet J Rare Dis* 10:93, 2015
12. Brislin RP, Theroux MC. Core myopathies and malignant hyperthermia susceptibility: a review. *Paediatr Anaesth* 23:834–41, 2013
13. Maggi L, Scoto M, Cirak S, et al. Congenital myopathies – clinical features and frequency of individual subtypes diagnosed over a 5-year period in the United Kingdom. *Neuromuscul Disord* 23:195–205, 2013
14. Sano K, Miura S, Fujiwara T, et al. A novel missense mutation of RYR1 in familial idiopathic hyper CK-emia. *J Neurol Sci* 356:142–7, 2015
15. Wulf H, Ledowski T, Linstedt U, Proppe D, Sitzlack D. Neuromuscular blocking effects of rocuronium during desflurane, isoflurane, and sevoflurane anaesthesia. *Can J Anaesth* 45:526–32, 1998