General Anesthesia Management in a Patient with Mitochondrial Encephalomyopathy Undergoing MitraClip® Implantation

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Abstract

Background: Because mitochondrial encephalomyopathy carries a risk of malignant hyperthermia, volatile inhalation anesthetics are contraindicated, but propofol can also carry the risk of propofol infusion syndrome. In recent years, increasing reports have shown that the use of volatile inhalation anesthetics can be safely managed for anesthesia. We report that sevoflurane made it possible to safely manage anesthesia in a patient with mitochondrial encephalomyopathy undergoing MitraClip® implantation.

Case presentation: The patient was an 82-year-old woman with mitral regurgitation (MR) and mitochondrial encephalomyopathy discovered by genetic testing. She also had rheumatoid arthritis and was taking steroids. Preoperative transthoracic echocardiography revealed severe MR due to left atrium dilatation. Due to her high risk for surgery, she was scheduled to undergo MitraClip® implantation. She was placed under general anesthesia, which was induced with midazolam, and neuromuscular monitoring was used for administration of a muscle relaxant. Anesthetic management was maintained with remifentanil and sevoflurane. After MitraClip® implantation, mitral regurgitation was reduced.

Conclusion: MitraClip® implantation in a patient with mitochondrial encephalomyopathy could be safely managed under monitoring using an inhalation anesthetic.

Key words

Mitochondrial encephalomyopathy, MitraClip®, propofol infusion syndrome, malignant hyperthermia

Background

Mitochondrial encephalomyopathy is a rare disease with multi-organ lesions and diverse symptoms, and there are no guidelines for anesthesia management. It should be managed under local anesthesia if possible, but local anesthesia is difficult when MitraClip® (Abbott, Lake bluff, IL USA) implantation is performed because of the need for intraoperative breath-holding and no patient movement when the device is deployed. In addition, shallow anesthesia is stressful and should be avoided. We present a case of a patient with mitochondrial encephalomyopathy who underwent MitraClip® implantation managed with general anesthesia using an inhalational anesthetic agent.

Case presentation

The patient was an 82-year-old woman (height, 150 cm; weight, 37 kg) with mitral regurgitation (MR), and she was short of breath and had repeated heart failure. She had sensorineural deafness, diabetes, and cardiomyopathy, had suffered multiple cerebral infarctions, and was diagnosed as having mitochondrial encephalomyopathy discovered by genetic testing. She also had rheumatoid arthritis and was taking steroids. Her electrocardiogram showed atrial fibrillation.

Preoperative transthoracic echocardiography revealed an indication of severe MR due to left atrium dilatation. There was no pericardial effusion. Her aorta dimension/left atrial dimension (AoD/LAD)
was 25/52; left ventricular dimension diastolic/systolic (LVDD/LS) was 37/26; interventricular septum thickness/left ventricular posterior wall thickness (IVS/PW) was 9/9; and her left ventricular ejection fraction (LVEF) was 60%.

After the patient entered the operating room, her level of consciousness was normal, her blood pressure was 174/52 mmHg, her pulse rate was 76 bpm, she was in atrial fibrillation, and her oxygen saturation (SpO₂) was 96% on room air. After insertion of an arterial pressure line, she was placed under general anesthesia, which was induced with 2 mg of midazolam and 10 mg of rocuronium (with reference to a muscle relaxant monitor), and 5 mg of rocuronium was additionally administered. At train-of-four (TOF) electromyography count 87, 5 mg of rocuronium was added, and tracheal intubation was performed at TOF 54. Anesthetic management was maintained with 0.05 to 0.2 μg/kg/min of remifentanil, 0.6 to 3% sevoflurane, and 0.05 to 0.25 μg/kg/min of noradrenaline. A pulmonary artery catheter was inserted from the right internal jugular vein to monitor cardiac function. Monitoring of both anesthetic depth with a PSI (patient state index) monitor and cerebral blood flow (rSO₂) were also performed. Transesophageal echocardiography after anesthesia induction showed severe MR (Fig. 1). In addition to intraoperative blood pressure, heart rate, and SpO₂ monitoring and capnometry, we also monitored and managed bladder temperature to control body temperature (Fig. 2). During the operation, arterial blood was collected for measurement of blood glucose level, and a fast-acting insulin preparation was used for hyperglycemia. Lactic acidosis did not progress, and stable circulatory management was possible (Table 1). After the operation, mitral regurgitation was reduced (Fig. 3), the muscle relaxant was reversed with no residual muscle relaxant effect, and she was extubated in the operating room and moved to the Cardiac Care Unit. The symptoms of heart failure disappeared, and she was discharged without any problems such as postoperative neurological sequelae after the operation.

**Discussion**

In anesthesia management of patients with mitochondrial encephalomyopathy, care must be taken to avoid tipping the balance of energy supply and demand, including checking baseline pH and lactate levels followed by perioperative monitoring. As these patients may have defects in energy production and use, their fasting times should be minimized and perioperative glucose supplementation monitored to avoid both hypo- and hyperglycemia. Metabolic demands should be minimized, including avoidance of stress, pain, nausea and vomiting, and hypoxemia, and normothermia should be maintained. In the present case, preoperative OS-1® water (Otsuka Pharmaceutical Factory, Tokushima, Japan) consisting of sodium (50 mEq/L), chloride (50 mEq/L), potassium (20 mEq/L), magnesium sulfate (2 mEq/L), lactate (31 mEq/L) and glucose (18 g/L) were administered to prevent long fasting time. Intraoperative infusion was administered by adding 5% glucose to Ringer’s bicarbonate solution. After induction, she was kept warm with an air type warmer.

Because patients with mitochondrial encephalo-
Table 1. Intraoperative Parameters

| Parameter   | Before anesthetic induction | 1h after anesthetic induction | 1.5h after anesthetic induction (At the end of the procedure) |
|-------------|----------------------------|-------------------------------|-------------------------------------------------------------|
| PH          | 7.448                      | 7.382                         | 7.374                                                       |
| PCO₂ (mmHg) | 32.0                       | 43.8                          | 37.9                                                        |
| PO₂ (mmHg)  | 90.4                       | 255.6                         | 387.9                                                       |
| HCO₃⁻ (mEq/L)| 22.4                      | 26.3                          | 22.3                                                        |
| BE (mEq/L)  | -1.8                       | 1.0                           | -3.1                                                        |
| Lac (mmol/L)| 1.4                        | 1.6                           | 1.4                                                         |
| BS (mg/dL)  | 195                        | 180                           | 170                                                         |

myopathy are at risk for malignant hyperthermia, volatile inhalation anesthetics are contraindicated. However, administration of succinylcholine, a depolarizing muscle relaxant, for malignant hyperthermia in myopathic patients may cause hyperkalemia or anesthesia-induced rhabdomyolysis and should not be used, and currently, it is rarely used. The risk of malignant hyperthermia in patients with mitochondrial encephalomyopathy is the same as that in healthy individuals.5

Although the safety of propofol use in patients with encephalomyopathy remains unclear, a single bolus dose is likely tolerated. However, propofol use in patients with encephalomyopathy may result in lipid metabolism and propofol infusion syndrome such as metabolic acidosis, hepatic failure, rhabdomyolysis, bradycardia, and heart failure1–4.

In recent years, there have been increasing reports that the use of volatile inhalation anesthetics can be safely managed for anesthesia in these patients, and we report that sevoflurane made it possible to safely manage anesthesia in a patient with mitochondrial encephalomyopathy undergoing MitraClip® implantation.

Since transesophageal echocardiography is essential for the Mitraclip® procedure and high peep is required, management with general anesthesia is required.

In the anesthesia management of patients under-
going a MitraClip® procedure, MR is underestimated due to the effects of general anesthesia7), so it is necessary to maintain afterload and manage it so that MR does not worsen after surgery.

During the operation, preload is maintained in consideration of the saline load due to the operation, but excessive fluid infusion can cause postoperative heart failure. Therefore, not only CVP8) but also our patient’s oxygen content fluctuation index (PVI® [pleth variability index]; Masimo Corporation, Irvine, CA, USA)9) was monitored. Peripheral circulation was monitored along with the oxygen reserve index (ORi™; Masimo Corporation, Irvine, CA, USA) during apnea associated with the procedure10), and management was possible without causing circulatory failure or the progression of acidosis.

Although there are some well characterized mitochondrial myopathic syndromes, some similar clinical presentations can be caused by different genetic mutations1,2). Moreover, massive parallel or next-generation genetic sequencing methodologies have emerged as the new gold-standard for accurate diagnosis of mitochondrial DNA disorders3). A history of muscle biopsy can add diagnostic clarity, but these tests might have limited sensitivity and specificity and add insufficient information to aid in the formulation of an anesthetic plan3). In recent years, inhalational anesthetics have been reported to be safely used in the anesthesia management of patients with mitochondrial encephalomyopathy6), but as clinical symptoms in these patients are diverse, treatment strategy should be determined for each case by the heart team, and more case reports are needed in the future.

**Conclusion**

We were able to manage anesthesia in a patient with mitochondrial encephalomyopathy undergoing MitraClip® implantation using inhalation anesthetics. In addition, we could infuse fluids and manage circulation without causing lactic acidosis by using circulation monitoring.

**Abbreviations**

AoD/LAD: aorta dimension/left atrial dimension; IVS/PW: interventricular septum thickness/left ventricular posterior wall thickness; LVDd/Ds: left ventricular dimension diastolic/systolic; LVEF: left ventricular ejection fraction; MR: mitral regurgitation; ORi: oxygen reserve index; PVI: pleth variability index; TOF: train-of-four

**Consent for publication**

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

**Conflicts of Interests**

The authors have nothing to disclose.

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