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CASE REPORT

A Patient With Muscular Dystrophy Posted For Mastoid Exploration: A Case Report

Kumar Shailly[1], **Sandill Surabhi**[2], **Baruah Udismita**[1]

[1]Department of Anaesthesia, VMMC & Safdarjung Hospital, Delhi, India.
[2]Department of Onco-Anaesthesia and Palliative Medicine, NCI, AIIMS, New Delhi, India.

*Corresponding author: Dr. Udismita Baruah E-mail: udismitabaruah@gmail.com

https://orcid.org/0000-0001-9036-2117

ABSTRACT

Muscular dystrophies are a group of genetic disorders that cause progressive weakness and loss of muscle mass. They are characterised by an impaired synthesis or regeneration of contractile protein. Patients with such disorders possess a high postoperative risk of hyperkalaemia, rhabdomyolysis and cardiovascular instability. We herein describe the anaesthetic management of a 45-year-old male, a known case of Duchenne muscular dystrophy, posted for left mastoid exploration. Using a multidisciplinary approach, a thorough preoperative evaluation was done. The patient was induced with intravenous anaesthetic agents, and maintenance was done using total intravenous anaesthesia. The peri-operative period was uneventful, and the patient was successfully discharged from the hospital.

Keywords: Muscular dystrophy, mastoid exploration, TIVA

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INTRODUCTION

Muscular dystrophies are inherited neuromuscular disorders characterized by progressive skeletal muscle weakness, with Duchenne muscular dystrophy being the most common subtype.[1] These patients pose significant perioperative challenges because of multisystem involvement and susceptibility to anaesthesia-related complications. Progressive muscle weakness, restricted neck mobility, and limited mandibular movement may complicate airway management.[2] Exposure to succinylcholine and volatile anaesthetic agents can precipitate rhabdomyolysis and malignant hyperthermia-like reactions. Therefore, meticulous preoperative assessment, avoidance of triggering agents, and the use of short-acting intravenous anaesthetics are essential to minimize perioperative morbidity and ensure safe recovery.[3]

CASE REPORT

A 45-year-old male patient (weight: 75 kg and height: 179 cm) presented with bilateral ear discharge and progressive hearing loss for the past six years. The ear discharge was not associated with otalgia, vertigo, tinnitus, trauma, swelling, nasal discharge, nasal obstruction, deviation of the angle of the mouth, or dysphagia.

The patient had been diagnosed with Duchenne muscular dystrophy at the age of five years using genetic testing. Early manifestations included difficulty in running and jumping. Progressive motor impairment developed over time; he experienced difficulty in standing by the age of 10 years and walking by the age of 15 years. For the past 25 years, he had been wheelchair-dependent. There was no history suggestive of respiratory muscle involvement. The patient had no other significant medical or surgical history.

A physical examination was conducted while the patient remained comfortably seated in his wheelchair. Facial features included an elongated face and a high-arched bifid palate. Contractures were present in both lower limbs. Muscle strength was graded as 3/5 in both upper limbs and 1/5 in both lower limbs. Sensory examination was normal, although muscle tone was reduced in all four limbs. Laboratory investigations, chest radiography, pulmonary function tests, and arterial blood gas analysis were unremarkable. Two-dimensional echocardiography demonstrated mild concentric left ventricular hypertrophy with preserved left ventricular ejection fraction. Preoperative evaluations were conducted by the cardiology, neurology, and respiratory medicine departments.

The patient and his relatives were counseled regarding the anaesthetic and surgical risks associated with the procedure, and informed consent was obtained.

Considering the elevated risk of rhabdomyolysis and malignant hyperthermia–like reactions in patients with muscular dystrophy, total intravenous anaesthesia (TIVA) was planned for the elective procedure.

The operating room was prepared with special precautions to prevent exposure to volatile anaesthetic agents. A new breathing circuit was used, the soda lime canister was replaced, and the vaporizers were removed from the anaesthesia machine. The machine was flushed with a fresh gas flow of 8 L/min for 20 minutes to eliminate any residual anaesthetic vapours. As dantrolene was not available, ice packs were kept readily available in the operating room as part of the emergency preparedness for malignant hyperthermia–like reactions. A difficult airway cart was prepared. Emergency

drugs and defibrillator were also checked and kept ready. The patient received intravenous pantoprazole 40 mg on the morning of surgery as aspiration prophylaxis. Premedication with intramuscular glycopyrrolate 0.2 mg was administered 30 minutes prior to induction of anaesthesia.

On arrival in the operating room, standard monitoring according to the American Society of Anaesthesiologists (ASA) guidelines was instituted, and baseline vital parameters were normal. An arterial line was placed for invasive blood pressure monitoring, and the non-invasive blood pressure cuff was removed to avoid potential muscle injury from repeated cuff inflation. Additional monitoring included end-tidal carbon dioxide ($EtCO_2$), neuromuscular transmission, core temperature, and urine output. Because lower limb contractures prevented adequate flexion or extension, a ramp was used to achieve optimal positioning in the supine position.

The patient was preoxygenated with 100% oxygen for three minutes. Anaesthesia was induced with intravenous fentanyl 130 µg, propofol 120 mg, and atracurium 40 mg, while ventilation was maintained with a mixture of oxygen and air in a 50:50 ratio. The initial attempt at tracheal intubation was unsuccessful due to difficulty in achieving optimal head positioning and limited visualization of the glottic opening. A second attempt was performed after repositioning the patient's head and using a smaller endotracheal tube. A cuffed endotracheal tube of 7.0 mm internal diameter was successfully inserted during the second attempt.

To minimize the risk of prolonged neuromuscular blockade and delayed recovery, additional doses of muscle relaxant were avoided after the initial induction dose. Volatile anaesthetic agents were completely excluded. Anaesthesia was maintained with TIVA using propofol infusion at 40 µg/kg/min, fentanyl at 10 µg/kg/h, and dexmedetomidine administered as a loading dose of 1 µg/kg over 10 minutes followed by an infusion of 0.5 µg/kg/h. Ventilation was maintained with a 50:50 mixture of oxygen and air. All pressure points were carefully padded. Active warming was provided using a forced-air warming device, and nasopharyngeal temperature was maintained between 36°C and 36.5°C.

The five-hour procedure was completed without haemodynamic instability. Intravenous anaesthetic infusions were discontinued 30 minutes before the end of surgery. The trachea was extubated after recovery of the train-of-four ratio to ≥ 0.9, adequate spontaneous ventilation, and return of airway reflexes. The patient was observed in the intensive care unit for 24 hours, remained free of respiratory or cardiac complications, and was discharged home on the second postoperative day.

DISCUSSION

Patients with muscular dystrophy require meticulous perioperative planning because of their susceptibility to anaesthesia-related complications, including rhabdomyolysis, hyperkalaemia, and malignant hyperthermia-like reactions following exposure to succinylcholine or volatile anaesthetic agents.[4] Consequently, a multidisciplinary evaluation involving anaesthesiologists, neurologists, cardiologists, and respiratory physicians is recommended before surgery.

Cardiac assessment is particularly important because conduction abnormalities and cardiomyopathy are common and may influence anaesthetic management.[5] Airway management can also be challenging due to

restricted neck movement, reduced mandibular mobility, upper airway muscle weakness, and decreased functional residual capacity. Therefore, difficult airway equipment should always be readily available. In addition, pharyngeal muscle weakness and delayed gastric emptying increase aspiration risk, warranting routine aspiration prophylaxis.[7]

Maintenance of normothermia is essential, as hypothermia and shivering may precipitate myotonic contractions.[6] Continuous temperature monitoring and active warming measures should therefore be incorporated into intraoperative care.

Patients with muscular dystrophy may exhibit variable sensitivity to non-depolarizing neuromuscular blocking agents. In our patient, atracurium was administered only for tracheal intubation, and anaesthesia was subsequently maintained with total intravenous anaesthesia (TIVA) using propofol, fentanyl, and dexmedetomidine, avoiding volatile agents throughout the procedure.[5]

Previous reports have also demonstrated the safety and efficacy of TIVA-based techniques in minimizing perioperative complications in this patient population. Deshpande et al.[5] reported successful anaesthetic management using a combination of propofol and dexmedetomidine along with an ultrasound-guided erector spinae plane block in a 31-year-old patient with Duchenne muscular dystrophy undergoing percutaneous nephrolithotomy.[5]

Extubation should be performed only after complete recovery of neuromuscular function and adequate spontaneous ventilation. Because delayed respiratory depression and cardiac complications may occur, postoperative observation in a high-dependency or intensive care setting is advisable.[7]

Baseline and postoperative serum creatine kinase levels were not measured; therefore, subclinical muscle injury or rhabdomyolysis could not be objectively excluded.

CONCLUSION

Muscular dystrophy poses significant anaesthetic challenges owing to multisystem involvement and the risk of severe reactions to trigger agents. Careful preoperative assessment, avoidance of succinylcholine and volatile anaesthetics, meticulous airway preparation, temperature management, and close postoperative monitoring are essential for safe perioperative care. Our case demonstrates that a TIVA-based anaesthetic technique using propofol, fentanyl, and dexmedetomidine can provide safe and effective anaesthesia in patients with muscular dystrophy undergoing surgery.

AUTHORS' CONTRIBUTIONS

Concept and design: UB, SK

Literature review: SK, SS

Compilation of manuscript: SK, SS

Manuscript editing and proofreading: SS, UB

DECLARATION OF CONFLICTS OF INTEREST

The authors declare no potential conflicts of interest relevant to this article.

DECLARATION OF PATIENT'S CONSENT

Written informed consent was obtained from the patient and his family for publication of this case.

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