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ANESTHETIC MANAGEMENT USING TIVA IN A PATIENT WITH LIMB-GIRDLE MUSCULAR DYSTROPHY: A CASE REPORT

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Abstract
Limb-Girdle Muscular Dystrophy (LGMD) is a group of progressive neuromuscular disorders characterized by proximal muscle weakness, affecting mobility and respiratory function. The perioperative management of LGMD patients presents significant challenges due to their susceptibility to respiratory compromise, cardiac involvement, and adverse reactions to certain anesthetic agents. We report the case of a 34-year-old male diagnosed with LGMD in 2013, who presented with a right- sided neck swelling requiring surgical incision and drainage under general anesthesia. The patient had no prior cardiac or respiratory compromise but exhibited severe lower limb weakness (0/5) and moderate upper limb weakness (4/5). A total intravenous anesthesia (TIVA) approach was employed to avoid the risk of malignant hyperthermia associated with volatile anesthetics.The patient underwent successful intubation using a CMAC video laryngoscope, and anesthesia was maintained with oxygen and air without inhalational agents. This case highlights the importance of individualized anesthetic management in LGMD patients, emphasizing preoperative evaluation, avoiding high-risk agents, and careful airway management. With a multidisciplinary approach and perioperative vigilance, safe anesthetic care can be achieved, minimizing complications and ensuring optimal surgical outcomes.

INTRODUCTION

Limb-Girdle Muscular Dystrophy (LGMD) is a genetically heterogeneous group of disorders characterized by progressive weakness and wasting of the proximal muscles, particularly those around the hips and shoulders (1). First delineated as a distinct clinical entity by Walton and Nattrass in 1954, LGMD has since been recognized for its significant genetic and clinical variability (2).

The classification of LGMD has evolved over time. Initially, subtypes were categorized based on inheritance patterns: autosomal dominant (LGMD1) and autosomal recessive (LGMD2). However, a 2017 workshop by the European Neuromuscular Centre proposed a refined classification system, emphasizing genetic and molecular characteristics to better reflect the underlying pathophysiology (3).

Clinically, LGMD presents with symmetric muscle weakness, predominantly affecting the pelvic and shoulder girdles. Patients often experience difficulties in activities such as rising from a seated position, climbing stairs, or lifting objects overhead (4). The age of onset and disease progression can vary widely, even among individuals with the same genetic subtype. Some patients may remain ambulatory into late adulthood, while others might require mobility aids within a few years of symptom onset (5). Advancements in genetic testing have enhanced the accuracy of LGMD diagnosis, enabling the identification of specific mutations responsible for various subtypes. This molecular approach not only aids in diagnosis but also informs prognosis and potential therapeutic strategies (6).

Despite these advancements, challenges remain in the management of LGMD. Currently, there is no cure, and treatment focuses on symptomatic management, including physical therapy to maintain mobility and prevent contractures (3). Ongoing research into gene therapy and molecular treatments offers hope for future disease-modifying interventions (3,7).

In this case report, we discuss the anesthetic management in a 34-year-old male patient with a known diagnosis of LGMD, who presented with a neck mass requiring surgical intervention.

Case Report

34-year-old male, diagnosed with LGMD in 2013 following a muscle biopsy prompted by progressive lower limb

weakness, presented with a progressively enlarging swelling on the right side of the neck. One month before presentation, the patient had sustained minor trauma to the neck region. An initial CT scan at that time revealed no abnormal swelling. Over the subsequent weeks, the swelling became increasingly tender and warm, without discharge or overlying skin breakdown.

As baseline, the patient exhibited complete loss of lower limb muscle function (power 0/5) and moderate weakness in both upper limbs (power 4/5), but without significant visible muscle atrophy in either limb. He remained ambulatory only with wheelchair support and was on regular physiotherapy. There was no prior history of respiratory compromise, cardiac symptoms, or significant comorbidities until the current illness.

On examination, the patient was afebrile, with stable Spo2 on room air -97%, Hr- 80/min , Bp – 130/88mmhg. he right cervical swelling measured approximately 8*7 cm, was tender, warm, and firm, without fluctuance or regional lymphadenopathy. No tracheal deviation or airway compromise was observed.

During hospital evaluation, fasting and postprandial capillary blood glucose levels were elevated, and the patient was newly diagnosed with diabetes mellitus. Endocrinology consultation was sought, and insulin therapy initiated with subcutaneous short-acting human insulin (Actrapid) titrated according to capillary blood glucose monitoring, along with long-acting insulin glargine (Lantus) 15 units at bedtime.

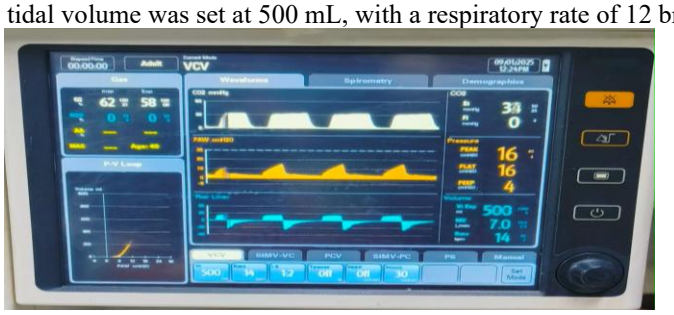
There was no history of systemic hypertension, bronchial asthma, coronary artery disease, seizures, or cerebrovascular disease. The patient denied any previous surgeries.

Preoperative Evaluation and Anesthetic Management

Prior to surgery, the patient underwent pre-anesthetic evaluation, chest X-ray - showed no opacities .The breath-holding time was 18 seconds. The ECHO revealed an ejection fraction (EF) of 58% with normal cardiac function. Airway assessment showed a Mallampati Grade 2 (MMG-2) airway with adequate mouth opening. Restricted neck movement was noted, with pain on extension, while flexion of neck remained unrestricted. On the night before surgery, the patient was administered tablet Anxit (0.25 mg) . The patient was scheduled for incision and drainage of the neck swelling under general anesthesia using a Target-Controlled Infusion (TCI) pump.

Intraoperative Course

On the day of surgery, CBG levels, urine ketones, and serum electrolyte levels were within normal limits. The patient was transferred to the operating theater (OT), where non-invasive blood pressure (NIBP), pulse oximetry, and ECG monitoring were initiated. Venous access was secured with an 18G venflon in the left arm and a 20G venflon in the right arm. The TCI pump was set based on the patient's height (166 cm), weight (70 kg), age (34 years), and BMI (25.2), with an effect-site concentration (Ce) of 1 mcg/mL. The patient was preoxygenated with 100% oxygen, followed by the administration of glycopyrrolate (0.2 mg) and midazolam (1 mg). Fentanyl (100 mcg) was administered through the 20G venflon, and a propofol infusion was initiated via the 18G venflon. Once anesthesia was induced, the patient was ventilated and intubated using CMAC with an endotracheal tube (ETT) size 7.5. The anesthesia was maintained with a propofol infusion along with a flow rate of 2 liters of oxygen and 2 liters of air per minute. Inhalational agents were strictly avoided due to the risk of malignant hyperthermia. The tidal volume was set at 500 mL, with a respiratory rate of 12 breaths per minute.



The surgery proceeded without complications, and the patient remained hemodynamically stable throughout the procedure. Toward the end of surgery, the propofol infusion on the TCI pump was gradually tapered to reduce the effect-site concentration, allowing for a smooth and controlled emergence from anesthesia. Once spontaneous respiratory efforts increased and end-tidal CO_2 and oxygen saturation were within target ranges, suctioning of the oral and oropharyngeal cavity was performed to clear secretions.

Neuromuscular function was preserved throughout (no relaxants used), so extubation criteria were based on clinical signs — including sustained head lift (as tolerated by the patient's baseline muscle strength), purposeful movement , obeying commands , on spontaneous ventilation and adequate tidal volumes (>6 mL/kg). The ETT cuff was deflated, and extubation was performed in a semi-upright position to optimize respiratory mechanics. The patient transitioned smoothly to breathing spontaneously with oxygen supplementation via face mask, maintaining stable hemodynamics and oxygen saturation above 98%. Postoperatively, he was fully awake, reported no awareness of the extubation process, and was pain-free and comfortable in the recovery area.

DISCUSSION

Limb-Girdle Muscular Dystrophy (LGMD) presents unique challenges in perioperative anesthetic management, primarily due to its progressive impact on skeletal, respiratory, and cardiac muscles. The perioperative care of LGMD patients requires meticulous planning to mitigate potential complications and ensure optimal patient outcomes.

Patients with LGMD are at an increased risk for perioperative respiratory complications, including hypoventilation, aspiration, and respiratory failure (8). Preoperative pulmonary function assessment is crucial to evaluate the extent of respiratory muscle involvement, as these patients may have restrictive lung disease that can significantly impact postoperative recovery (9). Studies have shown that individuals with neuromuscular diseases often experience perioperative respiratory complications, necessitating a thorough preoperative evaluation (10). Similarly, cardiac involvement in LGMD can manifest as cardiomyopathy or conduction abnormalities, increasing the risk of perioperative cardiac events (11). Preoperative cardiac evaluation, including electrocardiography (ECG) and echocardiography (ECHO), is recommended to identify potential risks and prevent intraoperative hemodynamic instability (12). A recent study emphasized the importance of assessing cardiac function in neuromuscular disease patients, as undiagnosed conduction defects can significantly increase surgical risks (13).

The selection of anesthetic agents is a critical aspect of perioperative planning in LGMD patients. The use of depolarizing muscle relaxants, such as succinylcholine, is contraindicated due to the risk of hyperkalemia and rhabdomyolysis (14). Non-depolarizing neuromuscular blockers are often preferred, but their effects may be prolonged due to altered pharmacodynamics in LGMD patients (15). Total Intravenous Anesthesia (TIVA) with agents like propofol and remifentanil is frequently recommended, as it minimizes the risk of malignant hyperthermia, a potentially life-threatening complication in patients with neuromuscular disorders (16).

In this case, the patient was specifically assessed in the immediate postoperative period for any recall or awareness during intubation and extubation. This was done through a structured interview once the patient had regained full orientation and stable vital parameters. Questions included whether the patient recalled hearing voices, feeling the endotracheal tube being removed, experiencing choking or gagging, or any sensation of distress during emergence. The patient reported no memory of either intubation or extubation, and no unpleasant sensory experiences or discomfort. This lack of awareness indicates that the depth of anesthesia was appropriately maintained throughout emergence, avoiding accidental intraoperative awareness—a complication that can be psychologically distressing and may lead to post-traumatic stress disorder.

Additionally, the patient had no throat pain, hoarseness, or cough, suggesting atraumatic airway management and reduced postoperative airway irritation. This is particularly beneficial in LGMD patients, as avoiding postoperative airway discomfort and respiratory compromise is crucial given their baseline muscle weakness. Case reports have similarly demonstrated the successful use of TIVA in LGMD patients undergoing orthopedic and general surgeries, highlighting its role in providing stable hemodynamics, adequate amnesia, and smooth recovery without airway-related complications (17)

Airway management is often challenging in LGMD patients due to potential facial and pharyngeal muscle weakness, which may complicate intubation and increase the risk of aspiration (18). Difficult airway management strategies, such as awake fiberoptic intubation, have been reported as effective methods in securing the airway safely while minimizing risks (19). In a recent case report, awake fiberoptic intubation was successfully used in a patient with LGMD, allowing for controlled and secure airway access (20). Regional anesthesia is a viable alternative or adjunct to general anesthesia in LGMD patients, particularly when avoiding the risks associated with general anesthesia is preferable (21). Postoperative care in LGMD patients should focus on close monitoring for respiratory depression, as the effects of anesthetic agents may be prolonged due to the underlying muscle disorder (20). Pain management strategies should balance adequate analgesia with minimization of respiratory depression. Non-opioid analgesics, such as paracetamol and NSAIDs, along with regional anesthesia techniques, are often preferred to reduce opioid requirements. The anesthetic management of LGMD patients requires a multidisciplinary approach, incorporating thorough preoperative assessment, careful selection of anesthetic agents, effective airway management strategies, and vigilant perioperative monitoring. Given the complexity of neuromuscular disorders, an individualized anesthetic plan is essential to minimize risks and optimize surgical outcomes.

CONCLUSION

The anesthetic management of Limb-Girdle Muscular Dystrophy (LGMD) patients presents unique challenges due to the progressive involvement of skeletal, respiratory, and cardiac muscles. This case highlights the importance of thorough preoperative evaluation, including respiratory and cardiac assessments, to identify potential perioperative risks. In patients with neuromuscular disorders, a multidisciplinary approach involving anesthesiologists, neurologists, and physiotherapists is essential for optimizing perioperative care. Total Intravenous Anesthesia (TIVA) offers distinct advantages in the anesthetic management of patients with Limb-Girdle Muscular Dystrophy. By avoiding inhalational agents and depolarizing muscle relaxants, TIVA minimizes the risk of malignant hyperthermia–like reactions, hyperkalemia, and rhabdomyolysis—serious complications to which these patients are predisposed. The use of propofol-based TIVA allows for precise titration of anesthetic depth, maintaining hemodynamic stability and enabling rapid, smooth emergence without residual sedation. In addition, avoiding volatile agents and neuromuscular blockers reduces the likelihood of postoperative respiratory depression, an important consideration in patients with baseline muscle weakness. These benefits make TIVA a safe and effective technique for LGMD patients, supporting optimal surgical outcomes and enhancing postoperative recovery..The careful airway management using C-MAC strategies ensured a secure airway, particularly given the risk of pharyngeal muscle weakness. Intraoperative monitoring and adherence to strict perioperative protocols contributed to a stable surgical course without complications. Postoperatively, vigilant respiratory monitoring is essential in LGMD patients due to the potential for prolonged anesthetic effects and reduced respiratory function. The use of opioid-sparing analgesic strategies, including regional anesthesia and non-opioid medications, can effectively manage pain while minimizing respiratory depression risks. This case highlights the need for individualized anesthetic plans tailored to the specific pathophysiological characteristics of LGMD patients. With careful perioperative planning and anesthetic modifications, surgical interventions can be performed safely, ensuring optimal patient outcomes and reducing complications. Further research and clinical guidelines are necessary to refine anesthetic protocols for patients with neuromuscular disorders and the use TIVA , ensuring the highest standards of perioperative care and patient safety.

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