Case of limb-girdle muscular dystrophy for total thyroidectomy: Anaesthetic management

INTRODUCTION

Limb-girdle muscular dystrophy (LGMD) refers to a genetically heterogeneous group of muscular dystrophies that present with weakness mainly involving the shoulder and hip girdles.[1] LGMD has a predominantly proximal distribution of weakness. In the early course of the disease the distal, facial and extra-ocular muscles are spared. The adult-onset disease involves both shoulder and pelvic girdles with gradually increasing proximal limb weakness. This leads to restriction of mobility and eventually leads to wheelchair confinement. Anaesthetic complications/implications in these patients are secondary to effects of anaesthetic drugs on myocardial and skeletal muscles. Events such as cardiac arrest, malignant hyperthermia (MH) and delayed recovery from non-depolarising muscle relaxants pose a challenge to the anaesthesiologist.[2] We describe the anaesthetic management of a patient with LGMD posted for total thyroidectomy for follicular neoplasm of thyroid, under regional anaesthesia.

CASE REPORT

A 40-year-old female patient presented with a history of swelling in the anterior aspect of neck since 1 year and progressive dysphagia for 2 months. She had autosomal dominant LGMD since 16 years. Her children were diagnosed with the same condition. Patient was afebrile with pulse rate of 110 bpm(regular) and blood pressure, 140/80 mm of Hg. Local examination of neck revealed a swelling 3 cm × 3 cm in size which moved with deglutition and was firm in consistency. Cardiorespiratory system examination was normal. Neurological examination revealed quadriparesis with hypotonia of all 4 limbs; the power in the extremities was shoulder-2/5, elbow-3/5, wrist-4/5, hip-2/5, knee-2/5 and ankle-2/5. Deep tendon reflexes were absent and superficial reflexes were normal. Airway assessment showed Mallampati Class-I. Haemoglobin was 9.8 g/dL; platelet count, renal, liver and thyroid profile were normal. Electrocardiogram showed tachycardia and sinus rhythm; echocardiogram was normal.The patient was counselled and consent obtained. Routine nil per oral instructions were advised and the patient was premedicated with tablet alprazolam 0.25 mg, tablet pantoprazole 40 mg and tablet ondansetron 4 mg orally.

The operating room was prepared according to the malignant hyperthermia protocol.[2] We planned for thyroid block with a backup of total intravenous anaesthesia. A bilateral superficial cervical plexus block and bilateral superior laryngeal nerve block was planned for surface analgesia and sensory blockade of the thyroid gland and surrounding structures.

The standard monitors were connected to patient and baseline vitals recorded (pulse-100 bpm, blood pressure-140/80 mm of Hg, respiratory rate-18/min, temperature-37.1°C and SpO_2-100% at room air). Eighteen gauge intravenous line was secured, Ringer lactate infusion started and injection midazolam 1 mg intravenously was administered. Under aseptic precautions, bilateral superficial cervical plexus block and bilateral superior laryngeal nerve block were performed with local anaesthetic mixture of 10 mL of 0.5% bupivacaine and 10 mL of 2% lignocaine (10 mL on each side). After confirming adequate sensory blockade, sedation was maintained with propofol infusion at the rate of 10 mL/h and injection. Fentanyl 50 μg intravenous and oxygen was supplemented through face mask and vitals were maintained throughout the procedure. Duration of surgery was 120 min. Post-operative period was uneventful.

DISCUSSION

Anaesthesia in patients with neuromuscular diseases is a great concern for anaesthesiologists.[3] LGMD are a heterogeneous group of genetically determined progressive disorders of skeletal muscles with both autosomal dominant and recessive inheritance. The prevalence ranges from 1:14,500 to 1:123,000 inhabitants.[2] They are characterised by proximal muscular dystrophy, elevated creatine kinase and associated cardiorespiratory problems.[1,4]

The anaesthetic considerations of LGMD are similar to other muscular dystrophies. However, perioperative complications are not proportional to the severity of the disease and occur even in mildly affected patients which need careful pre-operative evaluation and consultation.[2,4]

Regional anaesthesia should be performed whenever possible as general anaesthesia in LGMD needs careful monitoring, due to the high incidence of fatal complications. In LGMD, cardiac muscle and conducting pathways are affected and the sudden appearance of a Wenckebach type of block can occur especially during change in patient's position. In the absence of cardiomyopathy, propofol and thiopentone can be safely used as induction agents.[2,3] Respiratory compromise may occur early due to severe diaphragmatic involvement resulting in hypoventilation. Hence, sedative-hypnotics and opioids should be used judiciously.[2,5] In patients with muscular dystrophies, suxamethonium and to a lesser extent volatile anaesthetics should be avoided as they may lead to life-threatening complications such as rhabdomyolysis and MH.[6] These patients are sensitive to non-depolarising agents and it is recommended to use titrated doses of rocuronium and atracurium under neuromuscular monitoring. We avoided inhalational, depolarising and non-depolarising muscle relaxants (being triggering agents in MH and also with risk of respiratory insufficiency). Local anaesthesia is also beneficial for post-operative analgesia.

Sugammadex is an alternate option for cholinesterase inhibitors for steroid neuromuscular blockade reversal. These patients may have decreased laryngeal reflexes, prolonged gastric emptying time and masseter spasm with high risk of aspiration and hence should receive aspiration prophylaxis. It is important to be prepared for a difficult airway, especially in patients with potential airway problems.[2]

Thyroid surgery is routinely performed under general anaesthesia but cervical epidural anaesthesia or regional anaesthesia can be used. A bilateral superficial cervical plexus block with superior laryngeal nerve block can be used for total thyroidectomy. The bilateral deep cervical plexus block is technically more difficult to perform and is associated with phrenic nerve block which may lead to respiratory compromise. Thyroidectomy under local anaesthesia is generally supplemented with midazolam, fentanyl and/or propofol to provide conscious sedation. The choice of local anaesthetic is based on anaesthesiologist's preference and anticipated duration of surgery. The studies reviewed indicate that thyroidectomy under regional anaesthesia is associated with fewer complications.[7]

LGMD patients undergoing non thyroid procedures can also be managed under regional anaesthesia; spinal and epidural anaesthesia have been successfully used for caesarian section.[2,4,7]

LGMD patients are confined to bed and are more prone for the development of deep vein thrombosis which can be prevented by use of compression stockings or sequential compression devices intraoperatively and low molecular weight heparin prophylaxis.[8]

CONCLUSION

Anaesthesia in patients with neuromuscular diseases is a great concern for anaesthesiologists as there is a risk of delayed recovery from the neuromuscular blockade and fatal complications. Total thyroidectomy in a patient with LGMD can be performed with good outcome using bilateral superficial cervical plexus and superior laryngeal nerve blocks.

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Conflicts of interest

There are no conflicts of interest.

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REFERENCES

1. Mathews KD, Moore SA. Limb-girdle muscular dystrophy. Curr Neurol Neurosci Rep 2003;3:78-85.
2. Baldeón-Chávez EJ, Ortiz-Gómez JR, Díez-Sánchez N, Cerdán-Rodríguez G. General anaesthesia in two patients with limb-girdle muscular dystrophy. Anaesthesiol Rescue Med 2013;7:397-400.
3. Klingler W, Lehmann-Horn F, Jurkat-Rott K. Complications of anaesthesia in neuromuscular disorders. Neuromuscul Disord 2005;15:195-206.
4. Kulkarni PM, Takalkar MU, Kelkar YP. Limb girdle muscle dystrophy: Anaesthetic management in a patient for caesarean section. Int J Curr Med Appl Sci 2015;6:141-2.
5. Allen T, Maguire S. Anaesthetic management of a woman with autosomal recessive limb-girdle muscular dystrophy for emergency caesarean section. Int J Obstet Anesth

2007;16:370- 4.
6. Stevens RD. Neuromuscular disorders and anesthesia. Curr Opin Anaesthesiol 2001;14:693-8.
7. Arora N, Dhar P, Fahey TJ 3rd. Seminars: Local and regional anesthesia for thyroid surgery. J Surg Oncol 2006;94:708-13.
8. Narani KK. Deep vein thrombosis and pulmonary embolism – Prevention, management, and anaesthetic considerations. Indian J Anaesth 2010;54:8-17.

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