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CASE REPORT

Anesthetic Management of a Pregnant Patient with Limb-girdle Pattern of Muscular Dystrophy for Caesarian Section: A Case Report

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ABSTRACT

Muscular dystrophies and myopathies are inherited disorders that affect the neuromuscular junction.Limb-girdle muscular dystrophy (LGMD) is a rare neuromuscular condition that includes a variety of disorders with heterogeneous causes, as well as mutations within the same gene that can result in different phenotypic expressions. As the natural course of the disease progresses, the risk of surgery increases concomitant with the increased comorbid conditions associated with the later phase of the disease. Notably, perioperative complications do not correlate directly with the severity of the disease, as they can manifest even in patients with mild symptoms. Therefore, it is essential for patients to receive thorough preoperative consultations and evaluations.

Factors such as the severity of the condition, age at onset, and specific characteristics of LGMD can differ significantly among individuals.Pregnant women with LGMD are particularly susceptible to obstetric complications, especially if they experience significant weakness in the pelvic girdle muscles and respiratory insufficiency. Symptoms may worsen during pregnancy, leading to increased respiratory compromise. Additionally, the progression of the disease is observed during this period.

The various anaesthetic challenges presented by a patient with LGMD complicate the anaesthetic management. However, with thorough pre-operative evaluation and a judicious selection of anesthesia and anaesthetic agents, it can be managed safely.Here we discuss a case of pregnant patient diagnosed with LGMD scheduled for elective caesarean section under spinal anesthesia.

KEYWORDS

• Limb girdle muscular dystrophy • Pregnancy • Spinal anesthesia

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INTRODUCTION

Muscular dystrophy refers to a group of hereditary diseases with progressive muscle weakness with preserved reflexes and sensation, each with unique phenotypic and genetic features. Progressive skelet al. muscle weakness is characteristic of these disorders, and it frequently leads to respiratory failure[1].

Limb-girdle muscular dystrophy (LGMD) is a rare type of neuromuscular disorder which encompasses agroup of disorders with heterogeneous causes and Mutations within the same gene which may lead to different phenotypes. Proximal muscle (shoulder and pelvicgirdle) weakness is the characteristic feature of this group of diseaseswith varying degrees of pulmonary and cardiac manifestations[2]. In most cases,there is a sparing of facial and bulbar muscles[3].

We report the successful management of 34 year parturient with limb girdle pattern of muscular dystrophy for elective cesarean delivery under spinal anesthesia.

CASE REPORT

37 years old $G_4P_2L_1A_2$ withgestational age of 36 weeks weighing 88kg (BMI-34) who belongs to ASA PS 3 with history of PIH on tab labet al.ol 100mg BD, bronchial asthma not on regular inhaler therapy and history of limb girdle Pattern of muscular dystrophy admitted for elective LSCS. Patient was born of consanguineous marriage and was diagnosed with limb girdle Pattern of muscular dystrophy at the age of 20 years during 3rd trimester of her first pregnancy. There was no family history of LGMD. Symptoms started as difficulty in releasing bilateral hand grip for which she consulted the obstetrician and taken calcium tablets. She underwent a term normal vaginal delivery. After her first delivery symptoms got increased as difficulty in getting up from squatting position, climbing up stairs and painless buckling of both knees. Weakness was predominantly involved the proximal muscles of bilateral lower limbs started on tab prednisolone 40mg per day and tab azathioprine 150mg OD and stopped by her own 5 years back. Weakness was progressive and now presented with difficulty in getting up from a sitting posture and had a waddling gait. She had no cranio-bulbar symptoms. Her EMG showed myopathic process, muscle biopsy showed dystrophy with rimmed vacuoles, CPK was moderately raised and her genetic testing showed pathogenic variant suggestive of LGMD.

Her Clinical examination revealed wasting of bilateral lower limbs and Power of upper limbs - 5/5 and lower limb-hip flexors and extensors - 4/5, trunk muscles were weak, deep tendon reflex-intact, plantar reflex-flexor response B/L. Sensory nervous system & cranial nerves examination were within normal limits.

The airway examination showed no findings predictive of the difficult airway and her spine was normal. Her vitals were within normal limits and room air oxygen saturation of 93%.

Her blood counts, blood chemistry, urinalysis, CPK and Chest X-ray were normal. ECG showed sinus rhythm and echocardiography showed normal cardiac chamber and valve morphology with a left ventricular ejection fraction of 67%.

Her Pulmonary function test showed a restrictive pattern and her ABG at room air was normal with a Pao2 of 76.

ANESTHETIC MANAGEMENT

- She was planned for caesarian section as pelvic girdle and trunk muscle weakness are considered risk factors for obstetric interventions and the procedure was planned under spinal anesthesia.
- Operating room was prepared according to malignant hyperthermia protocol. Aspiration prophylaxis was given. Standard monitors were attached. Baseline vitals were within accepted limits. SpO2 was 93% on room air.
- The subarachnoid block was given in L3-L4 interspace with 2 ml of 0.5% hyperbaric bupivacaine. The procedure was started after a block level of T6 was achieved. A healthy baby was delivered with APGAR score of 8/10 at 1 min and 10/10 at 5 min and was handed over to the attending pediatrician. Caesarian section was completed uneventfully with stable intra-operative hemodynamics and SpO2 of 99–100% on oxygen face mask.
- Post operative pain managed with bilateral TAP block and iv paracetamol.

DISCUSSION

Muscular dystrophies are a group of congenital and heterogeneous diseases that have a diverse genetic origin and have a wide range of clinical manifestations. Each of them has significant anaesthetic effects that pose a significant perioperative risk for patients, and it requires accurate knowledge to anticipate and provide specific management in each case. Malignant hyperthermia, cardiac arrhythmias, hyperkalemia, rhabdomyolysis, cardiovascular instability, ventilatory failure, and sudden death can complicate anesthetic management for muscular dystrophy making it particularly difficult[4].

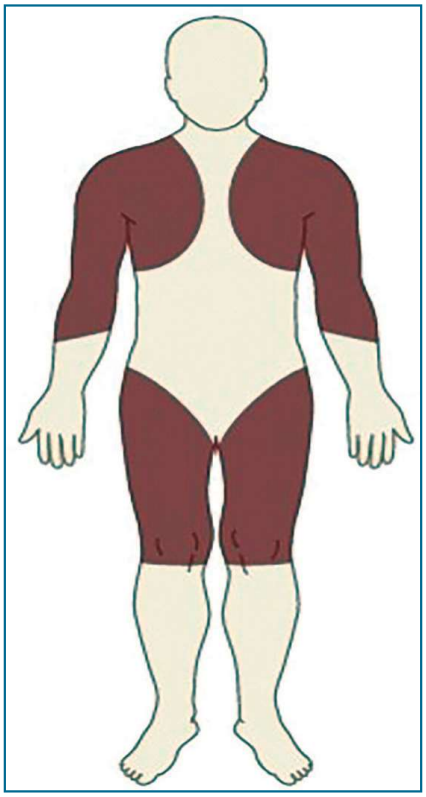

Fig. 1: Distribution of predominant muscle weakness in LGMD

LGMD is a rare neuromuscular disorder that predominantly affects pelvic and shoulder girdle muscles. It is classified as type 1 (Autosomal dominant) and type 2 (Autosomal recessive). The estimated incidence of this disease is 1 to 6.5 in 100,000[5]. The prevalence ranges from 1:14,500 to 1:123,000 inhabitants[6].

The multiple anaesthetic concerns in a patient with Limb-Girdle muscular dystrophy make the anaesthetic management of the patient difficult. Adequate pre-operative assessment and careful choice of anaesthetic agents can ensure safe management.

It is recommended to conduct a thorough pre-surgical evaluation to determine the general condition of each patient, the surgical risk, and to use a multidisciplinary approach to ensure the most effective perioperative care.

An echocardiogram and electrophysiological studies may be required during pre-operative evaluation based on cardiac signs and symptoms.

Systemic involvement can include minor cardiac lesions or significant cardiac lesions like dilated cardiomyopathy, and malignant arrhythmias.The majority of patients with muscular dystrophy exhibit some cardiac abnormalities, but only ten percent of them are clinically significant.In 10% to 25% of patients, an echocardiogram will reveal mitral valve prolapse[1].

Similarly, pulmonary involvement varies from minimal to severe pulmonary restriction[4]. Preoperative pulmonary assessment is crucial for this reason.

The many subtypes of limb-girdle muscular dystrophy cause variations in severity, age of onset, and features, which can even be inconsistent between family members[7]. Symptoms and signs can start at any age and usually worsen over time[8]

During pregnancy symptoms get exacerbated with increased respiratory compromise[9]

Obstetric complications are known to occur in these women especially if they develop severe pelvic girdle muscle weakness and respiratory insufficiency[10].

The anaesthetic management is quite challenging in patients with LGMD. Regional anaesthesia is preferred over general anaesthesia, whenever it is feasible. In regional anesthesia, the risk of rhabdomyolysis, malignant hyperthermia, post-operative respiratory compromise due to opioid sensitivity, and residual muscle paralysis is omitted.

Myotoxic effects of local anesthetics, particularly in peripheral blocks, are a concern, but the neuraxial block has been used without any complications[3].

Anaesthesia management during pregnancy is more challenging than administering

anaesthesia to non-pregnant females because of the numerous endocrinal, systemic, and physiological changes. Delivering safe anaesthesia services require a thorough knowledge and understanding of these facts. The advantages of regional anesthesia for maternal and fet al. outcomes outweigh those of general anesthesia[11].

The respiratory function may be affected by neuraxial anesthesia depending on the extent of motor blockage. Less extensive motor blockade may have minimal effects on ventilatory function[12]

General anesthesia has to be considered in patients who are unable to tolerate supine position despite respiratory support, or patients having bulbar muscle involvement.[9] Our patient had weakness only in proximal muscles and no cardio-respiratory involvement. Hence, we planned to give spinal anesthesia to our patient with NPPV if required.

CONCLUSION

- To warrant safe anesthesia for surgery in muscular dystrophy patients, a comprehensive preoperative evaluation, advanced intraoperative monitoring, thoughtful choice of anesthetic technique and pain management strategy, as well as with cooperation of a multidisciplinary team is mandatory.
- In this case report, we emphasize the safety of spinal anesthesia in performing caesarean sections in patients with muscular dystrophy.

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REFERENCES

1. Miller's Anesthesia International Edition, 2 Vol - 9780323612630 | Elsevier Health. Available from: https://www.mea.elsevierhealth.com/millers-anesthesia-international-edition-2-volume-set-9780323612630.html
2. Kocum A., Sener M., Caliskan E., Aribogan A. Anesthetic management for a child with unknown type of limb-girdle muscular dystrophy. Pediatr Int Off J Jpn Pediatr Soc. 2010 Feb; 52(1): e37-38.
3. Shakya B.M., Shrestha S. Anaesthetic Management of Patient with Limb Girdle Muscular Dystrophy Scheduled for Laparoscopic Cholecystectomy. Nepal J Med Sci. 2023 Jan 31; 8(1): 57–60.
4. Echeverry-Marín P.C., Bustamante-Vega Á.M. Anesthetic implications of muscular dystrophies. Colomb J Anesthesiol. 2018 Sep; 46(3): 228.
5. Iyadurai S.J.P., Kissel J.T. The Limb-Girdle Muscular Dystrophies and the Dystrophinopathies. Contin Minneap Minn. 2016 Dec; 22 (6, Muscle and Neuromuscular Junction Disorders): 1954–77.
6. Baldeón-Chávez E.J., Ortiz-Gómez J.R., Díez-Sánchez N, Cerdán-Rodríguez G. General anaesthesia in two patients with Limb-Girdle muscular dystrophy. Anaesthesiol Rescue Med. 2013 Dec 22; 7(4): 397–400.
7. Zatz M., de Paula F., Starling A., Vainzof M. The 10 autosomal recessive limb-girdle muscular dystrophies. Neuromuscul Disord NMD. 2003 Sep; 13(7–8): 532–44.
8. Ranjan R.V., Ramachandran T.R., Manikandan S., John R. Limb-girdle muscular dystrophy with obesity for elective cesarean section: Anesthetic management and brief review of the literature. Anesth Essays Res. 2015; 9(1): 127–9.
9. 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 Oct 1; 16(4): 370–4.
10. Chestnut's Obstetric Anesthesia: Principles and Practice - 5th Edition | Elsevier Shop [Internet]. [cited 2024 Mar 11]. Available from: https://shop.elsevier.com/books/chestnuts-obstetric-anesthesia-principles-and-practice/chestnut/978-1-4557-4866-2
11. Iddrisu M, Khan ZH. Anesthesia for cesarean delivery: general or regional anesthesia—a systematic review. Ain-Shams J Anesthesiol. 2021 Jan 6; 13(1): 1.
12. Duggan M., Kavanagh B.P. Pulmonary atelectasis: A pathogenic perioperative entity. Anesthesiology. 2005 Apr; 102(4): 838–54.