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Anesthetic Management of a Pregnant Woman with Limb-girdle Muscular Dystrophy in the Caesarean Section: A Case Study

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

In a study, we present the case of a 33-year-old obese patient with limb-girdle muscular dystrophy (LGMD) admitted for cesarean section. The patient was successfully managed by emergency caesarean section under spinal anesthesia. On physical examination after the operation, the force of the upper and lower extremity muscles were estimated to be 4/5 and 3/5, respectively. In this case, instead of epidural anesthesia we used spinal anesthesia, which is a safe technique recommended for emergency cesarean section in pregnant LGMD patients.

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1. INTRODUCTION

Limb-girdle muscular dystrophy (LGMD) or Erb's dystrophy is an autosomal recessive disorder with varied phenotypic presentations and characterized by genetically heterogeneous group of muscular dystrophies [1]. It is an uncommon form of muscular dystrophy, which involves the proximal muscles, specifically the muscles of the shoulders, upper arm, pelvic area, and thighs. Indeed, the incidence rate of LGMD is less than 0.000001.

In LGMD, myasthenia and atrophy are caused by the disintegration of muscle fibers and their replacement with the connective and fiber-fatty tissues [2]. LGMD takes diverse forms from the first to the fourth decades of life. The severe form of the disorder presents in the early stages of life, while patients with late-onset disorder have a better prognosis. However, LGMD symptoms may appear at any age and gradually worsen with time [3].

Some factors such as severity, age at onset, and features of LGMD are varied among patients. Increasing respiratory compromise may be observed in patients with aggravated LGMD, which ordinarily occurs in pregnant women [4]. LGMD symptoms exacerbate during pregnancy in the vast majority of women suffering from this disorder [5]. In this study, we present the case of anesthetic management of a woman with autosomal recessive LGMD undergoing emergency caesarean section.

2. CASE STUDY

A 33-year-old (gravid: 2; gestational age: 38 weeks) woman was referred to our hospital for parturition after having contractions. The patient had a previous history of cesarean section. The gynecologist ordered an emergency cesarean section after physical examination (height: 160 cm, weight: 85 kg, body mass index [BMI]: 33.2 kg/m^2).

The patient had a 13-year history of LGMD. This autosomal recessive disorder was a result of consanguineous marriage. She had two brothers and four sisters, among whom two other sisters were involved with the disease, as well. In one of her sisters, the lower limbs (both legs) were predominantly affected, while in the other sister the legs, hands, and eyes were involved. Our patient had a history of cesarean section under general anesthesia five years before this study.

In the first month of pregnancy, the patient was referred for blood and urine tests by a gynecologist. According to the blood test, thyroid-stimulating hormone (TSH), as well as hematological and biochemical indicators were normal. The results of the blood test in the second month of pregnancy showed serum glutamic-oxaloacetic transaminase (SGOT), serum glutamic-pyruvic transaminase (SGPT), and layered double hydroxide (LDH) levels were higher than the normal range.

The prenatal screening tests in the first and second trimesters showed no abnormalities in the fetus. In addition, the patient was evaluated in terms of other internal diseases, based on which no disease was detected. Besides, echocardiography showed no evidence of cardiomyopathy or pulmonary hypertension, while the left ventricular function was good. In the third month of pregnancy, the forces of the upper and the lower limbs were 4/5 and 3/5, respectively (BMI: 139 kg/m^2).

Bleeding, watery vaginal discharge, and renal or abdominal pain were not observed within 22 weeks + 4 days. Moreover, the patient did not have any signs of toxicity, frequent urination, and dysuria. The patient reported genital itching and green vaginal discharge. At 22 weeks + 4 days, blood pressure was 100/60 mmHg and fetal heart rate was 136 beats per minute. At 36 weeks + 4 days, blood pressure was 110/50 mmHg and fetal heart rate was 168 beats per minute. Furthermore, she presented with bleeding and watery vaginal discharge. In the sixth month of pregnancy, LDH level decreased to 691 U/l continually to the seventh month. The obtained results of other tests showed that SGOT, SGPT, LDH, creatine phosphokinase (CPK), along with other hematological and biochemical indicators were normal.

A significant proximal muscle weakness in all the limbs, especially the lower limbs, was observed in the preoperative evaluation. However, she was never wheelchair-bound. During hospital stay, the lower limbs, which were extended to hips and hands, were involved. Thus, she had difficulty getting up from the sitting position. After consultation with the treating gynecologist,

emergency caesarean delivery was selected as the best management practice. Hemoglobin level was evaluated before the operation and the anesthesiologist due to the high risk of postoperative pulmonary complications advised spinal anesthesia during general anesthesia. Dietary and medicinal intakes, as well as the details of surgery were explained to the patient before the operation. When the patient was transferred to the operation room, her heart rate, blood pressure, and pulse oximetry read were 92/min, 130/70 mmHg, 98%, respectively.

The delivery occurred in the 38th week of gestation (October-November, 2016). The patient underwent electrocardiography, and non-invasive blood pressure (NIBP) and peripheral capillary oxygen saturation (SpO_2) monitors were attached in order to control the patient's condition. Two intravenous catheters (16 gauge) were secured on the patient's hand under aseptic precautions. Intravenous ranitidine (100 mg) and 500 cc normal saline solution were administered to the patient before induction of anesthesia. The patient's back was washed while she was in sitting position. Then, 12.5 mg hyperbaric 0.5% of bupivacaine (AstraZeneca company, Sweden) was injected with 25-gauge Quincke spinal needle at L4 or L5.

During the operation, systolic and diastolic blood pressures were 120/60 and 110/70 mmHg, respectively, while the heart rate was 70-75/min and pulse oximetry read was 98%. Finally, a healthy baby was delivered with the first- and second-minute Apgar scores of 8 and 9, respectively. After parturition, diffuse muscle weakness was noted on physical examination. The patient was transferred to the intensive care unit after the C-section, and then was moved to the ward after 24 h due to normal respiratory status. According to the neurologist, the forces of the upper- and lower-extremity muscles were estimated to be 4/5 and 3/5, respectively.

3. DISCUSSION

LGMD is a rare disorder that may be caused by autosomal recessive inheritance in 90% of the cases. The involved sites in LGMD are the pelvic or shoulder girdle musculature [1]. Moreover, myasthenia and atrophy are caused by the disintegration of muscle fibers and their replacement with the connective and fiber-fatty tissues [2].

Generally, the symptoms of LGMD may appear at any age and gradually deteriorate with time [3]. In our case, the symptoms began at the age of 13. Some factors, such as severity, age at onset, and the features of limb-girdle muscle, are different among patients, even among the same family members [2]. In the present study, two sisters of the patient were differently involved with this disorder.

So far, four pregnant patients with LGMD who were anesthetized during parturition have been reported in the literature. According to the previous studies, the cases with LGMD present with disease progression during pregnancy [3-6]. Since obstetric complications are common in women with severe pelvic girdle muscle weakness and respiratory insufficiency, a multi-disciplinary approach was employed [7]. To avoid any possible complications, caesarian section was conducted at the presence of a team of obstetricians, anesthesiologists, and neurologists. In this case, spinal anesthesia was used due to the high risk of postoperative pulmonary complications during general anesthesia.

To the researcher's best knowledge, no study has yet compared the effects of general and regional anesthesia on LGMD patients. Since segmental epidural analgesia is associated with fewer side effects on the respiratory and cardiovascular systems, as compared to the other methods of anesthesia, spinal analgesia was applied. This technique was used in two other similar studies [3,6]; however, they used spinal and epidural anesthesia in patients with LGMD [4,5].

The harmful effects of spinal anesthesia on the respiratory system in pregnant women were revealed in the previous studies [8,9]. According to a study by Yun et al. [8] epidural anesthesia in women with elective cesarean section was not associated with a significant change in forced vital capacity (FVC), forced expiratory volume (FEV1), peak inspiratory flow rate (PIFR), peak expiratory flow rate (PEFR), or peak inspiratory pressure (PIP). Nonetheless, spinal anesthesia caused a significant reduction in FEV1, FVC, PIFR, and PEFR. As the respiratory function can be affected by an epidural block in muscular dystrophy patients, segmental epidural for general anesthesia induction was administered.

In fact, regional anesthesia is not a suitable option in all pregnant women. It is not recommended for patients who cannot lie in supine position, except for those who are at risk of aspiration and require respiratory support, which was not observed in this case [3]. In our study, general anesthesia was also possible in the absence of any problems.

For pregnant women with LGMD, difficult intubation cart should be available because the majority of the patients with LGMD are obese, which increases the risk of respiratory problem. Besides, in LGMD patients, neuromuscular function, temperature, and end-tidal CO2 monitoring are recommended during general anesthesia. The use of any medications leading to severe contracture should be avoided in LGMD patients since some neuromuscular blocking medications, such as succinylcholine, lead to severe myotonic response of a muscle and makes ventilation and tracheal intubation difficult [10-12].

According to the physical examination, the forces of upper- and lower-extremity muscles were 4/5 and 3/5, respectively, after the operation. Therefore, the patient became wheelchair-bound. A significant proximal muscle weakness in all the limbs, especially the lower limbs, was noted in preoperative evaluation. During hospital stay, the lower limbs, which were extended to the hips and hands, were involved. Consequently, she had difficulty getting up from the sitting position.

4. CONCLUSION

Given the high risk of general anesthesia in pregnant women with LGMD, patient management is of great importance. Therefore, presence of a multi-disciplinary team comprising of obstetricians, neurologists, anesthesiologists, and critical care physicians is critical for patient management. We decided to administer spinal block considering the emergency condition of our patient and the slower onset of action of epidural injection compared to spinal injection. Furthermore, spinal anesthesia is safe for emergency cesarean section in pregnant LGMD patients.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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