ACU Sağlık Bil Derg 2020; 11(4):738-740
https://doi.org/10.31067/0.2020.326

OLGU SUNUMU / CASE REPORT

Anesthetic Management of an Obstetric Patient with Limb-Girdle Muscular Dystrophy: Case Report and Review of the Literature

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ABSTRACT

Limb-girdle muscular dystrophy is a rare hereditary disease with dangerous systemic components including difficult airway, muscle weakness, increased risk for pulmonary aspiration, cardiomyopathy, and rhabdomyolysis that make anesthetic management challenging. In this case report, we aimed to discuss the anesthetic management of a 36-year-old pregnant woman with Limb-Girdle Muscular Dystrophy (LGMD) who underwent emergent caesarean section with spinal anesthesia and to review of the anesthetic management of obstetric Limb-Girdle patients in the literature.

Keywords: Limb-girdle muscular dystrophy, spinal anesthesia, caesarean section

LİMB-GİRDLE MUSKÜLER DİSTROFİLİ OBSTETRİK HASTADA ANESTEZİ YÖNETİMİ: VAKA TAKDİMİ VE LİTERATÜR TARAMASI

ÖZET

Limb-girdle musküler distrofi zor havayolu, kas zayıflığı, artmış pulmoner aspirasyon riski, kardiomyopati ve rabdomyoliz gibi tehlikeli sistemik komponentleri olan, genetik geçişli, seyrek görülen bir hastalıktır. Bu vakada, Limb girdle musküler distrofili 36 yaşında bir gebenin spinal anestezi ile acil sezaryeni sırasındaki anestezi yönetimini ve literatürdeki obstetrik Limb-girdle musküler distrofi hastalarını gözden geçirmeyi amaçladık.

Anahtar sözcükler: Limb girdle musküler distrofi, spinal anestezi, sezaryen

Limb-Girdle Muscular Dystrophy (LGMD) which was first introduced in 1954 by Walton and Nattrass is an extremely rare progressive disorder of muscles with an incidence of <1/100,000 in which the pelvic or shoulder girdle musculature is predominantly involved and it might be inherited in an autosomal recessive (AR) (90%) or autosomal dominant (AD) (10%) fashion (1–3).

Progressive muscle weakness; possible difficult airway, pulmonary aspiration risk, dysphagia, cardiomyopathy, epilepsy, psychomotor retardation and most importantly increased rhabdomyolysis and malignant hyperthermia risks make this subgroup of patients' anesthetic management challenging (4).

In this case report, we aimed to discuss the anesthetic management of a parturient with LGMD who underwent emergent caesarean section under spinal anesthesia and to review of the anesthetic management of obstetric Limb-Girdle patients.

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Received : May 13, 2019
Revised : May 13, 2019
Accepted : June 19, 2019

Case report

a 36-year-old primigravida woman at 37 weeks of gestation was scheduled for caesarean section in an emergency setting in consequence of the decrease in fetal heart rate. She was diagnosed as LGMD type 2B which is a kind of dysferlinopathies with muscle biopsy ten years ago.

Physical examination shows no abnormalities except micrognathia, mild lumbar lordosis, and restricted mobility of the cervical spine. Mallampati score was 3. Last muscle examination showed a muscle strength of 2/5 for cervical flexors, 4+/5 for deltoid muscles, 4/5 for biceps and triceps muscle, 3-/5 for iliopsoas muscle, 2/5 for gluteus maximus, 2/5 for quadriceps femoris, 3-/5 for femoral abductors, and 4+/5 for both hamstring muscle group and lower-extremity distal muscle group. There was no significant abnormality in preoperative laboratory tests. The initial heart rate was 122/min and systolic blood pressure was 140/84 mmHg while pulse oximetry revealed an oxygen saturation of 98%.

The operating room was prepared by flushing the anesthesia machine with oxygen, changing the components including soda-lime, breathing system. Difficult airway preparation including various blades of Miller and Macintosh type, laryngeal mask airways, tracheal tubes with stilettos, gum-elastic bougie, fiberoptic bronchoscope, and a tracheostomy set were kept ready.

In the operating room, ECG, pulse oximeter and blood pressure were monitored and infusion of 1000 ml isotonic solution from a venous line was initiated. Spinal anesthesia was performed in the sitting position, using a 27 Gauge spinal needle (Braun Spinocan) at the L3-L4 spinal space. After visualizing clear cerebrospinal fluid; hyperbaric bupivacaine 2.0 ml was injected intrathecally. Then the patient was re-positioned. In 10 minutes, adequate motor and sensory block were obtained at the T4 dermatome. Moderate hypotension (systolic blood pressure 78 mmHg) occurred during surgery and was controlled by administration of 10 mg ephedrine. Except for this temporary hypotension, the spinal block was well tolerated by the patient. A live female 2290 g baby with APGAR scores of 8–9 was delivered. 10 units of oxytocin and 1 g cefazolin were administered intravenously following the delivery.

The operation time was 38 minutes and the patient was transferred to the ward afterwards. SaO2 and ECG were monitored continuously in the patient's room for 6 hours to detect any abnormalities. Intramuscular diclofenac was used for post-operative pain. 2 hours later, the Bromage scale indicated no remaining block. The patient was discharged on the day after surgery without any complications.

Discussion

LGMD is a heterogeneous group of diseases characterized by progressive weakness and atrophy in the pelvic and shoulder girdle muscle groups (5). The clinical course of LGMD might be variable, symptoms may begin in the late 40's or early stage of life. Late-onset diseases like this patient who was diagnosed at 26 years old show a milder course.

The systematic components of LGMD make anesthetic management of the disease challenging and because of the limited published data, there is still no consensus on the anesthetic management. The optimum approach seems to be individualized to the anesthetic approach due to the variable clinical manifestations.

The management of obstetric LGMD patients is more complicated due to the possible progression of symptoms including severe pelvic girdle muscle weakness and respiratory insufficiency (6). The PubMed search revealed four cases of caesarean section in women with LGMD and only one was performed in an emergency setting (Table 1). In these case reports, combined spinal-epidural anesthesia

Table 1. Anesthetic management of Caesarean section (C/S) in LGMD patients Anesthetic management of Caesarean section (C/S) in LGMD patients

Author	*Case*	*Age*	*Weeks*	*Technique*
Allen T. 2005 (2)	Emergency C/S	28 y	34 wk	CSE (1.8 ml Bupivacaine 0.5% + fentanyl 20 mg)
Ranjan RV. 2005 (6)	Elective C/S	27 y	38 wk + 2 d	Epidural anesthesia (12 ml 0.5% Bupivacaine)
Yilmaz R. 2019 (8)	Elective C/S	27 y	–?	CSE (6.25 mg Bupivacaine + 15.5 μ fentanyl)
Pash M, 1996 (9)	Elective C/S	19 y	37 wk	Epidural anesthesia (15 ml lidocain 1:200000 epinephrine)

was preferred for the management of caeserean section. In our case, labor was initiated and fetal heart rate was decreased and time for epidural anesthesia was insufficient due to the status of the fetus, we preferred low-dose spinal anesthesia. However, spinal anesthesia did well and therefore epidural catheter was not used either for anesthesia nor postoperative analgesia. In addition to spinal anesthesia with low doses, we did not administer any opioids as patients with LGMD are vulnerable to respiration problems resulting from respiratory muscle dysfunction.

Data from prospective trials demonstrate that lowering the spinal dose improves maternal hemodynamic stability. Doses of intrathecal bupivacaine between 5 and 7 mg are sufficient to provide effective anesthesia. If general anesthesia is preferred in patients with LGMD, life-threatening complications including rhabdomyolysis and malignant hyperthermia should be taken into consideration. Neuraxial anesthesia not only provides avoidance from these complications, but also prevents the development of pulmonary complications which might occur with intubation and invasive ventilation. Moreover, less extensive motor blockade may have minimal effects on pulmonary gas exchange with arterial oxygenation and carbon dioxide elimination being well maintained during most spinal and epidural anesthesia (7).

Anesthesia for patients with severe muscular dystrophy can be accomplished through a careful review of the patient's respiratory status, the anticipation of cardiac dysfunction, and careful selection of anaesthetic agents based on the potentially increased risk of malignant hyperthermia and their residual effects on respiratory function (8–9). Although low dose spinal anesthesia proved successful in our case, more severe cases of LGMD are best managed by a multidisciplinary team including obstetricians, neurologists, anesthesiologists, and critical care physicians.

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