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Limb girdle muscle dystrophy and caesarian delivery: Anesthetic management and brief review of literature

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ABSTRACT

Limb girdle muscle dystrophy (LGMD) is group of rare hereditary disorders primarily involving hip and shoulder muscles. Due to scarcity of literature, definite anesthetic management strategy is lacking. We, report the successful management of 28 yr parturient with LGMD for elective caesarian delivery under spinal anesthesia. The anesthetic management is discussed with brief review of literature.

1. Introduction

Limb-girdle muscular dystrophy is a term for a group of diseases that cause weakness and wasting of the muscles in the arms and legs. The muscles most affected are those closest to the body (proximal muscles), specifically the muscles of shoulders, upperarms, pelvic area, and thighs. It is difficult to determine the prevalence of limb-girdle muscular dystrophy because its features vary and overlap with those of other muscle disorders. The prevalence ranges from 1:14,500 to 1:123,000 inhabitants [1]. Some factors such as severity, age at onset, and features of LGMD are varied among patients. During pregnancy symptoms get exacerbated with increased respiratory compromise [2]. Here we present a case of anesthetic management of a woman with autosomal recessive LGMD undergoing elective caesarean section.

2. Case report

A 28 yr old parturient was referred to our hospital with bad obstetric history (BOH). She had her first spontaneous conception 9 years back and spontaneous bleeding per vaginam at 4th month of pregnancy with complete abortion. During her second spontaneous conception, 8 years back, she had spontaneous bleeding per vaginam at 5th month of pregnancy with complete abortion. The third spontaneous conception was 7 years back with IUFD at 7th month of gestation. Fourth spontaneous conception was 3 years back with no fetal cardiac activity at 4th month of gestation. Two years back she had her fifth spontaneous conception. At 5th month of gestation, fetal congenital malformation was detected. At 33 + 6wks of gestation, preterm vaginal delivery was done and a 2.18 kg baby was delivered which had an open meningomyelocele and did not survive. During the present pregnancy she was under regular follow up. She was planned for elective caesarian section in view of transverse lie of fetus.

She was diagnosed with LGMD at age of 5 years with electromyography showing signs of early myopathy and muscle biopsy showing subtle myopathic changes and myeloid degeneration. Creatinine phosphokinase (CPK) level was raised (396 U/L). Her symptoms predominantly affected proximal muscles due to which, she had difficulty in getting up from a sitting posture and had a waddling gait. She had no cranio-bulbar symptoms. On examination there was mild thinning of supraspinatus and infraspinatus muscles although tone was normal. Power in hip was 4/5, infraspinatus 4/5, supraspinatus 4/5, deltoid 4+ respectively. Difficult airway was ruled out. Other preoperative investigations were within normal range. Echocardiography showed no evidence of cardiomyopathy or pulmonary hypertension.

She was planned for caesarian section under spinal anesthesia with NPPV as standby if required. 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 97% on room air. Subarachnoid block (SAB) was given in L3-L4 interspace with 1.8 ml heavy bupivacaine 0.5% with 25 mcg fentanyl. 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. After the surgery, the patient was shifted to recovery room. The

Abbreviations: NPPV, non-invasive positive pressure ventilation; BOH, bad obstetric history; IUFD, intra uterine fetal death

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block receded completely over the next 4 h and on examination, she had no new onset of weakness in her limbs.

3. Discussion

LGMD is a rare genetically inherited disorder. It involves the pelvic or shoulder girdle musculature [3]. Muscle fibers are disintegrated and are replaced with the connective and fiber-fatty tissues which cause myasthenia and atrophy [4].The various forms of limb-girdle muscular dystrophy are caused by mutations in many different genes involved in muscle functioning and maintenance.

Limb-girdle muscular dystrophy is classified based on its inheritance pattern and genetic cause. Type1 is Autosomal Dominant (prevalence 10%) while type2 Autosomal Recessive inherited (prevalence 90%). Various subtypes are identified depending on the gene mutations involved.

The severity, age of onset, and features of limb-girdle muscle dystrophy vary among the many subtypes of this condition and may be inconsistent even within the same family members [4]. Signs and symptoms may first appear at any age and generally worsen with the time [5]. In, our case symptoms appeared at 5 years of age.

Anesthetic management in pregnant patients with LGMD for caesarian delivery has been reported in very few cases in the literature. During pregnancy, there is progression of the disease [6]. Obstetric complications are known to occur in these women especially if they develop severe pelvic girdle muscle weakness and respiratory insufficiency [7]. Elective surgery is recommended as pelvic girdle and trunk muscle weakness are considered risk factors for obstetric interventions [8]. A case of successful vaginal delivery has been reported previously in wheelchair bound multipara patient although she had no respiratory insufficiency [9]. Due to heterogeneity of disease, obstetric management should be individualized [10]. In our case, no trunk muscles were involved but transverse lie of fetus was taken into consideration when planning mode of delivery. Elective caesarian section was planned during normal working hours. In LGDM conduction abnormalities in heart may occur but cardiomyopathies are not consistent feature [7,10]. In severe cases diaphragmatic involvement may lead to respiratory compromise.

In literature there are no trials comparing regional and general anesthesia in these patients. In patients with muscular dystrophies, suxamethonium and to a lesser extent volatile anesthetics may lead to life-threatening complications namely rhabdomyolysis and malignant hyperthermia, and are therefore best avoided [11]. Neuraxial anesthesia may affect respiratory function depending on the extent of motor blockade. Less extensive motor blockade may have minimal effects on ventilatory function [12]. In a study conducted by Conn et al. demonstrated that spinal anesthesia for caesarian section caused a significant reduction in FEV1, FVC and PEFR from pre-spinal levels [13]. Despite this, however, patients were unaware of any deterioration in lung function and moreover, in this study, there was no assessment of arterial oxygenation or carbon-dioxide elimination, the clinical relevance of these changes is uncertain.

General anesthesia has to be considered in patients who are unable to tolerate supine position despite respiratory support, or patients having bulbar muscle involvement [2]. 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. Additionally, we avoided general anesthesia because these patients may be susceptible to malignant hyperthermia. However, the operating room was prepared well beforehand to avoid exposure to volatile anesthetics and succinylcholine. The availability of neuromuscular blockade, nasopharyngeal temperature, bispectral index monitoring, capnography and adequate quantity of dantrolene in the pharmacy were also ensured. Moreover SAB is preferred mode of anesthesia for caesarean section if, there is no contradiction to it.

4. Conclusion

Management of LGMD should be individualized as the symptoms vary among patients. Severe cases are best managed by multidisciplinary team. In this case report we highlight the fact that spinal anesthesia can be safely used to provide anesthesia for caesarian section in patients with LGMD. Further studies are required to establish definite anesthetic management strategies as few references are available for this rare disease.

Conflict of interest

The author declared that there is no conflict of interest.

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Competing interest

Authors have declared that no competing interest exists.

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