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Case Report

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Successful application of regional anesthesia for elective cesarean section in a patient with myotonia dystrophica complicated by placenta previa and placenta accreta.

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Abstract

The anesthetic management of a patient with myotonia dystrophica and placenta previa with accreta, presenting for elective cesarean section at 35 weeks of gestation is presented. The onset of muscle weakness had been delayed and presented during the second trimester of pregnancy. The pregnancy was further complicated by findings of restrictive lung disease and anemia. After delivery of the baby, the patient required a sub total hysterectomy due to failure of the placenta to separate. This case highlights the successful application of regional anesthesia in a complicated myotonia dyotrophica parturient with relative contraindication for GA.

Keywords: Myotonia dystrophica, pregnancy, placenta previa, placenta accreta, restrictive lung disease, anemia, regional anesthesia.

CASE REPORT

A 34-year-old mother in her second pregnancy developed symptoms of bilateral lower extremity weakness at 18 weeks of gestation. Her first child had been delivered by caesarean section due to prolonged labour. Difficulty in climbing stairs and rising up from a squatting position were presenting symptoms. She had an impaired ability to cough, but did not have dysphagia or dysarthria. Motor strength was rated as 3/5 in both lower extremities and 5/5 in the upper limbs. Muscle tone, tendon reflexes and sensation were normal. Electromyographic studies indicated a diagnosis of myotonia dystrophica (MD). Her room air oxygen saturation was noted to be 94% and further pulmonary testing demonstrated moderate to severe restrictive lung disease. An ultrasound scan at 35 weeks of gestation revealed a major degree placenta previa, with a high probability of placenta accreta. Electromyographic studies were repeated and reconfirmed the original diagnosis. The patient's hemoglobin was 8.5g/dl. Significantly,

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her four-year-old child was experiencing significant motor development delays. There was no clear family history of MD.

At a multidisciplinary team meeting, it was decided to transfuse the patient and proceed to an elective lower segment cesarean section (LSCS), with combined spinal epidural (CSE) anesthesia. Post transfusion hemoglobin was 10g/dl. Routine ASA monitors were applied and supplemental oxygen by facemask at 4L/min was commenced in the operating room, which resulted in an oxygen saturation of more than 96% throughout surgery. The epidural was performed with an 18 gauge Touhy needle and using a 29 gauge Sprotte needle via the epidural needle, 15mg of hyperbaric 0.5% bupivacaine was administered spinally. An epidural catheter placement was uncomplicated. A surface patient warmer (BearHugger, company) was applied and 15 degree left uterine displacement with a wedge was achieved. Sensory block to the 5th thoracic dermatome was confirmed and a LSCS was performed with a Pfannensteil incision. A 1.9kg baby was delivered, but soon after delivery the baby required intubation and ventilatory support. The surgery was complicated by the inability to extract a morbidly adherent placenta and the decision was made to proceed to a sub total abdominal hysterectomy (AH). Estimated blood loss was 1200ml. Two units of blood and 2000 ml of crystalloids were infused through a fluid warmer (brand, model, company). Tranexamic Acid 1g was administered IV and 5 bolus doses of ephedrine 5mg IV were necessary to maintain blood pressure during the procedure. At the conclusion of surgery, she was transferred to a High Dependency Unit. Postoperative analgesia was provided with a low dose epidural infusion, diclofenac sodium and paracetamol. Regular chest physiotherapy and breathing exercises were continued. The patient had an uneventful postoperative course and was discharged on postoperative day 5. Screening of her siblings for MD was arranged.

DISCUSSION

Myotonia dystrophica (MD) is a generalized muscle disease with an autosomal dominant inheritance pattern. Two types of MD are described [1]. In MD type 1, a defect in chromosome 19 results in a mutation in the gene that codes for myotonic dystrophy protein kinase (DMPK). In DM type 2, a defect in chromosome 3 results in a mutation in the cellular nucleic-acid binding protein (CNBP), also called zinc finger protein 9 (ZNF9)[1]. MD usually presents in young adulthood and is rare before the age of 5. It is characterized by muscle weakness, delayed relaxation or myotonia, ptosis and frontal baldness in males [1]. Weakness of the muscles of the pharynx, mastication, neck and limbs may occur and determine presentation. Multi-system involvement may extend to the respiratory, cardiac, endocrine and central nervous systems [1,2,3]. The presence of documented pulmonary compromise in patients with MD is associated with increased risk of respiratory complications, longer ICU and hospital stays [5]. In females, uterine muscle function may be abnormal leading to dysfunctional labour and postpartum hemorrhages [3]. Clinical diagnosis can be confirmed with electromyography studies. Diagnostic features include predominantly distal myotonic discharges, myopathic motor units and early recruitment [4]. Myotonic discharges are less prominent in MD type 2. Muscle biopsy provides definitive diagnosis [4].

A multi-disciplinary approach is mandatory for the safe management of patient with MD. Chest physiotherapy and breathing exercises are crucial in the pre-habilitation of patients prior to surgery (7).

General anesthesia (GA) is the favored option in the setting of major degree placenta previa with accreta. GA, when compared to regional anesthesia (RA), is less likely to compound the hypotension that is associated with obstetric hemorrhage. Thiopentone, propofol and etomidate have all been safely used for induction of GA in patients with MD [1]. However, the use of succinyl choline for rapid sequence induction (RSI) may result in severe masseter spasm in patients with MD and leads to difficulty with endotracheal intubation [1,8]. Secondly, the resulting exaggerated muscle depolarization may also lead to life-threatening hyperkalemia and cardiac arrest [1,8]. Non-depolarizing muscle relaxants are a safe alternative to facilitate intubation in patients with MD in many surgical settings. However, in an obstetric patient at term, their use for intubation poses a serious dilemma, due to the increased risk

of pulmonary aspiration. The avoidance of these risks associated with GA was the main reasons that predicated our decision to choose CSE in this patient.

Impairment of the protective airway reflexes and compromise of ventilatory reserve are the major considerations for the anesthesiologist when managing a patient with MD. The fact that our patients had impaired coughing ability and the pulmonary function test finding of moderate to severe restrictive lung disease, also contributed to our choice of CSE as the preferred option [6,8]. However, when obtaining informed consent, the possibility that GA might be necessary, was explained to the patient. We were successful in providing satisfactory RA for the LSCS and the caesarean hysterectomy that followed, in the setting of a placenta accreta. Hence, we were able avoid many agents used for GA, that can interact with neuro muscular function and impair central ventilatory drive.

Hypothermia is a factor known to increase muscle weakness in patients with MD [1,8]. We were diligent in maintaining body temperature with the use of surface warmer and a fluid warmer.

The placement of an epidural catheter enabled excellent postoperative analgesia, without the use of narcotic medications. Patients with systemic muscle disorders are at increased risk from the central respiratory depression that is a constant risk with narcotic administration [1,8].

CONCLUSION

Regional anesthesia was successfully used to manage this complicated MD parturient with major degree placenta previa and placenta accreta. She required a caesarian hysterectomy due to failure of the placenta to separate. In exceptional circumstances, regional anesthesia should be considered where general anesthesia would otherwise be the norm.

Author declaration

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Authors and contributions

WMASBW: Collection of information, planning writing the Manuscript
KG: Responsible for clinical care of the patient.
MP: Discussion of the medical management of the patient
LdeS: Proof reading the manuscript

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Ethical approval and consent to participate

Written informed consent was obtained from the patient

Competing interests

The Authors declare that they do not have competing interest

REFERENCES

1. Ferschl M, Moxley R, Day WJ, et al. Practical suggestions for the anaesthetic management of a myotonic dystrophy patient. Myptonic dystrophy foundation care and cure. Available from: https://www.myotonic.org/sites/default/files/MDF_Long Form_AnesGuidelines_01C.pdf
2. Azar I. The Response of Patients with Neuromuscular Disorders to Muscle Relaxants: A Review. Anesthesiology. 1984;61:173-187. doi: 10.1097/00000542-198408000-00011
3. Sarnat HB, O'connor T, Byrne PA. Clinical Effects of Myotonic Dystrophy on Pregnancy and the Neonate. Arch. Neurol. 1976; 33(7): 459-65.doi: 10.1001/archneur.1976.00500070001001
4. Thornton CA. Myotonic dystrophy. Neurologic clinics. 2014; 32 (3): 715-719. doi: 10.1016/j.ncl.2014.04.011
5. Saito H, Hatakeyama K, Konno H, et al. Impact of pulmonary rehabilitation on postoperative complications in patients with lung cancer and chronic obstructive pulmonary disease. Thorac Cancer. 2017;8(5):451-460. doi.org/10.1111/1759-7714.12466
6. Hopkins AN, Alshaeri T, Akst SA, et al. Neurologic disease with pregnancy and considerations for the obstetric anesthesiologist. Seminars in Perinatology. 2014;38(6):359-369. doi.org/10.1053/j.semperi.2014.07.004.
7. Mathieu J, Allard P, et al: Anaesthetic and surgical complications in 219 cases of myotonic dystrophy.

Neurology. 1997; 49:1646-1650. doi.org/10.1212/wnl.49.6.1646

8. Mangla C, Bais K, Yarmush J. Myotonic Dystrophy and Anesthetic Challenges: A Case Report and Review. Case Reports in Anesthesiology. 2019;(9). doi.org/10.1155/2019/428230