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Anesthetic Management of a Patient with Spinal Muscular Atrophy Type III Undergoing Emergent Caesarean Section: A Case Report

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Abstract

In this case report, we describe the anesthetic management for a 36-year-old G2P0010 at 36 weeks gestation with Spinal Muscular Atrophy Type III who underwent an emergent caesarean section due to fetal footling breech position. The patient is a wheelchair-bound quadriplegic with kyphoscoliosis and a lack of cough reflex who required nasal continuous noninvasive ventilatory support (CNVS) for chronic hypercapnic respiratory failure. Surgery was done under general anesthesia due to its emergent nature, and the patient was successfully extubated and transitioned to nasal CNVS in the operating room at the end of the case. Postoperative care was provided in the medical intensive care unit for three days without complication and the patient was discharged home uneventfully.

Keywords

Spinal Muscular Atrophy, General Anesthesia, Cesarean Section, Obstetric Anesthesia

1. Introduction

Spinal muscular atrophy (SMA) is an autosomal recessive neuromuscular disease characterized by the degeneration of spinal cord anterior horn cells and lower brainstem motor nuclei, causing progressive symmetrical muscular weakness, hypotonia, and bulbar symptoms. Only lower motor neurons are involved, meaning that sensory function and cognition remain intact. The effects of motor neurons of cranial nerves III, IV, and VI, and sacral motor neurons are also not affected, leading to preserved eye movement and continence [1]. It is the second most

common autosomal recessive disease in humans with an incidence ranging from 4 to 10 per 100,000 live births [2].

SMA is caused by mutations in the survival motor neuron 1 gene (SMN1) on chromosome 5q13.2 or bi-allelic mutations resulting in a deficiency or proapoptotic form of the SMN1 protein, which normally protects motor neurons by inhibiting apoptosis and may also play a role in motor neuron mRNA synthesis [3] [4]. SMN protein is also integral for maintaining the integrity of the neuromuscular junction and maturity of acetylcholine receptors**.** The survival motor neuron 2 gene (SMN2), which lies telomeric of SMN1, can compensate for SMN1 protein synthesis. Thus, SMA displays a range of symptom severity, generally correlated inversely with SMN2 copy number and directly with the level of SMN protein [5]. Infants with Type 0 SMA generally only have one copy of SMN2, type 1 typically has 2 - 3 copies, type 2 has three copies, type 3 has 2 - 4, and type 4 has 4 - 8 copies [6].

The spectrum of SMA is generally divided by age of onset and degree of motor impairment. Typically, the later age of onset correlates with better prognosis. The most common causes of death from SMA include respiratory insufficiency and pneumonia due to aspiration. Type 0 SMA, which affects <1% of all SMA cases, features a prenatal onset with very severe disease severity. In the prenatal period, pregnant mothers might notice decreased or absent fetal movements. At birth, there is severe hypotonia, muscle weakness, areflexia, congenital heart defects, and joint contractures. Patients have a life expectancy of less than 6 months due to death from respiratory failure [7]. Type 1 SMA, otherwise known as Werdnig-Hoffmann disease, is the most prevalent type of SMA, representing about 55% of all SMA cases. Symptoms start after birth and up to six months of life with severe symmetric flaccid paralysis causing "floppy baby syndrome" or a frog leg posture. Respiratory muscle weakness increases slowly, often manifesting with paradoxical breathing due to intercostal muscles being affected more so than the diaphragm. These issues can also lead to death from respiratory failure. There is severe bulbar palsy, atrophy and fasciculations of the tongue and fingers, weak cry and cough, and the inability to swallow. Deep tendon reflexes can be diminished or absent and hip dislocations can occur recurrently and life expectancy is less than 2 years of age [7]. Type 2 SMA has an intermediate level of symptom severity and represents about 30% of total SMA cases (8). Symptoms are typically intermediate in nature and start about at 7 - 18 months old. Key symptoms include hand tremors and difficulty feeding due to bulbar weakness leading to poor weight gain. Musculoskeletal issues include scoliosis, kyphosis, and hip dislocation. Patients often never reach the developmental milestone of standing without support and have a life expectancy of 30 years. Type III SMA, otherwise known as Kugelberg-Welander disease, is another type of mild SMA, with an onset of symptoms beginning at greater than 18 months. Because onset often happens in childhood, it can also be referred to as juvenile SMA. It represents up to 30% of SMA cases and has differing levels of muscle weakness [8]. Type IV SMA, also known as the adult SMA type, manifests with symptoms at ages 10 - 30. This is the mildest form of

SMA with variable levels of muscle weakness, often able to reach developmental milestones, and accounts for less than 1% of cases [9].

In this paper, we will discuss the anesthetic management of an emergent cesarean section of a patient with Type III SMA and hope to inform future anesthetic practice for patients with this condition.

2. Case Presentation

Our patient was a 36-year-old G2P001 woman at 36 weeks and 6 days gestation who was brought into the hospital by EMS for active labor. On exam, the fetus was in breech presentation with a fetal foot protruding from the birth canal. Subsequently, the patient was sent to the OR for an emergent Cesarean section.

Her past medical history was significant for bilateral percutaneous Achilles tenotomy status post clubfoot deformity diagnosed at 12 months of age. The patient was diagnosed with Type III SMA at age 10 due to repeated falls, and uncontrollable lower extremity movements. She denied any known family history of SMA and is dependent on her mother for all activities of daily living. She became wheelchair bound at age 14 following years of progressively worsening weakness and instability and lost motor function of her hands in her early 20's. The patient lost the ability to cough at age 26 requiring a Mechanical Insufflation-Exsufflation device. The patient has a history of recurrent pneumonia and was previously noncompliant with outpatient pulmonary follow-up during the time of pregnancy. At 30 weeks gestation, the patient complained of symptomatic nocturnal dyspnea, and subsequent pulmonary function tests were performed and compared with results from 2015. The results showed that her vital capacity decreased from 1290 mL to 730 mL in the sitting position and maximum insufflation capacity decreased from 2200 mL to 1650 mL. Unassisted peak cough flows were 1.2 L/s, her oxygen saturation ranged from 91% - 93% and her end-tidal pCO_2 was 41 mmHg, so the patient was subsequently started on nocturnal nasal CNVS. However, at 34 weeks the patient complained of daily headaches, fatigue and hypersomnolence. She was then transitioned from nocturnal to continuous nasal CNVS due to worsening respiratory function.

On physical examination, the patient was anxious but communicative verbally and quadriplegic with generalized muscle wasting of upper and lower limbs in acute distress from labor. Musculoskeletal exam showed kyphoscoliosis. Airway exam was significant for Mallampati Class III due to difficult mouth opening. She had a thyromental distance greater than 3 with a mildly hypertrophic neck. Her neck range of motion and mobility was not limited and she had normal dentition. Pulmonary exam on auscultation was positive for fine crackles on the left side. Her cardiovascular exam was unremarkable.

On the day of the procedure, a 20-gauge IV cannula was inserted into the right median cubital vein. After a pelvic exam was performed, the case was deemed emergent due to fetal malpresentation and she was immediately moved to the operating room. Patient endorsed that she had eaten breakfast consisting of juice and bread 2 hours prior to presentation. Due to the urgency of the case and the

potential difficulty with regional anesthesia as a result of kyphoscoliosis and anxiety, general anesthesia with endotracheal tube was planned. An NG tube was placed for decompression of the stomach. Preoxygenation was achieved with a face mask administering 100% oxygen in addition to the patient's own Nasal CNVS device. A modified rapid sequence excluding muscle paralysis with succinylcholine and nondepolarizing neuromuscular blockers was performed to avoid potential hyperkalemia-induced cardiovascular complications and extended paralysis respectively. With fiberoptic scope present as backup, a C-mac video laryngoscope with size D-blade, and a 6.5 Endotracheal tube were chosen for intubation given her mallampati score. Induction was achieved with slow bolus of a total of 150 mg of propofol with cricoid pressure applied and suction on standby to further reduce the risk of aspiration. One attempt was made successfully, however, it was challenging as the patient's airway was edematous with difficult mouth opening. Upon successful intubation anesthesia was maintained with positive pressure ventilation on 100% oxygen and 1.8% Sevoflurane for a total MAC of 0.8. Intraoperative Azithromycin 500 mg and Cefazolin 2 g were administered. Body warmers were applied and set to 43˚C to maintain adequate temperature and to prevent the insensible heat loss from general anesthesia. A live healthy female infant was then delivered. We administered 20 units of pitocin via 1L drip shortly after delivery, given over 30 minutes. 1 g acetaminophen and 15 mg toradol were administered for multimodal pain control and the patient was provided a TAP block for postoperative incisional surgical pain. The patient was extubated and immediately placed on nasal CNVS in assist control ventilation mode with a target tidal volume of 350 mL. She was then transferred to the medical ICU for continuous monitoring postoperatively.

3. Discussion

We report in this case the general anesthetic management for an emergent cesarean section of a patient with Type III SMA who was successfully extubated from endotracheal tube back to nasal CNVS. The management of anesthesia in patients with SMA can be very challenging due to the anesthesia-induced pulmonary complications, muscle weakness, succinylcholine-induced hyperkalemia and the potential hypersensitivity to nondepolarizing muscle relaxant [10]. The literature suggests that pregnancy in women with SMA can be successful despite these potential complications stated above. However, SMA effects on the pulmonary physiology during pregnancy have been underreported and there needs to be greater awareness as special consideration must be given to the respiratory management of these patients.

For example, normal physiologic changes occur in pregnancy in order to cope with the increased oxygen demand and worsening of restrictive lung disease, including an elevation in minute ventilation that is achieved by increasing respiratory muscle drive [11]. However, this physiologic adaptation may be impaired in patients with advanced SMA due to weakened accessory muscle function. Skeletal deformities such as scoliosis distort abdominal anatomy and serve to further

impair pulmonary function. Thus, a patient with SMA may be ill-equipped to adapt to the pulmonary changes that occur during pregnancy. In our case, the patient presented with worsening respiratory status at 30 weeks of gestation when she was started on nocturnal CNVS. By 34 weeks of gestation, the patient required CNVS throughout the day. It is not uncommon for patients with SMA to present to the operating room with a Noninvasive Positive Pressure Ventilation (NIPPV) device due to their predisposition for hypoventilation and severe restrictive lung disease. Ideally, these patients should undergo respiratory status assessment and monitoring before and during pregnancy with a pulmonary medicine specialist in order to prevent adverse outcomes. In this case, preoperative respiratory monitoring did not occur due to loss of follow-up until the patient developed symptoms of worsening pulmonary function requiring the non-invasive ventilatory support device. Thus, the patient's inability to cope with high oxygen demand increased her risk for preterm labor and fetal distress.

Intra-operatively, regional anesthesia should be highly considered as an anesthetic plan for patients with SMA undergoing labor in order to decrease the risk of postoperative hypoventilation that occurs following general anesthesia [12]. Wilson *et al.* and Maruotti *et al.* both describe cases of successful cesarean sections in patients with SMA, done with epidural and spinal anesthesia, respectively [13] [14]. However, due to severe scoliosis, vertebral abnormalities, and previous spine surgical interventions, neuraxial anesthesia can be difficult and often impossible to achieve. Additionally, the local anesthetic spread in patients with SMA can be altered due to spine abnormalities, increasing the risk of a higher-than-desired block and therefore worsening respiratory function intraoperatively. For our case, our team felt strongly about pursuing general anesthesia from the beginning of the case due to the history of quadriplegia and kyphoscoliosis which would make neuraxial anesthesia difficult to obtain taking into consideration the acuteness of presentation, in addition to securing an airway on this patient with a lack of a cough reflex. There are a select number of cases discussed in literature with similar circumstances and outcomes that support this decision. For example, Habib *et al.* described a case in which a wheelchair-bound 23-year-old parturient with SMA type II underwent a cesarean section under general anesthesia [15]. Although our case presented different challenges comparatively, including a worse baseline respiratory function and an edematous airway, it was similar in that general anesthesia without the use of nondepolarizing muscle relaxants was opted over regional anesthesia due to the anatomical challenges presented and risk of worsening respiratory function. The patient in his case ended up remaining stable both during and after the surgery and was extubated uneventfully, representing a successful case of general anesthesia.

Notably, during the induction of our patient, we elected to avoid all neuromuscular blockers given her history of recurrent respiratory infections and lack of cough reflex. Neuromuscular blockade with succinylcholine is well known to be contraindicated in patients with neuromuscular disorders due to increased risk of fatal arrhythmia [16]. Additionally, blockades with non-depolarizing neuromuscular

blockers, while not contraindicated, are known to frequently cause prolonged weakness in patients with SMA. This prolonged weakness predisposes patients to increased risk of postoperative respiratory infection [10], thus informing our decision.

Although it was decided against in our case, there are cases reported in literature describing the successful use of neuromuscular blockers in patients with this condition. For example, McLoughlin *et al.* details a case of a 24-year-old woman with type III SMA who underwent an elective cesarean section under general anesthesia and was induced with the help of rocuronium [17]. In this case, reversal was attempted with neostigmine 40 minutes after administration and was successful, with the patient being extubated uneventfully. Notably, the patient had an intact cough reflex and underwent immediate chest physiotherapy afterwards, which are both variables that were not present in our case and were critical in their recovery from general anesthesia. Additionally, when considering their choice of reversal agent, it should be noted that this case report, was published prior to the widespread adoption of sugammadex. Current publications suggest reversal with sugammadex results in quick and effective restoration of neuromuscular function in non-pregnant patients with SMA [9]. A 2011 case report from Vilela et al details the anesthetic management of a patient with SMA type III undergoing a procedure to close an atrial septal defect, and in this case, rocuronium was used and successfully fully reversed with sugammadex in 69 seconds with no adverse effects [18]. Although this was one of the few reported uses of sugammadex in a patient with SMA, this was done in a male undergoing a cardiac procedure, rather than a parturient undergoing a cesarean section. Therefore, with further studies, we must consider the possibility that sugammadex can make the reversal of neuromuscular blockades even more safe and effective in parturients with this condition.

4. Conclusion

In summary, we report the anesthetic management of an emergent cesarean section of a patient with Type III SMA complicated by kyphoscoliosis and lack of cough reflex. Our case was conducted under general anesthesia without the use of neuromuscular blockade due to the aforementioned challenges, however, there is literature supporting the use of regional anesthesia as well as general anesthesia with nondepolarizing muscular agents. Given the discussed variables that contribute to the maintenance of such a patient's respiratory status, every anesthetic plan must be individualized. There are few studies on the anesthetic management of parturients with this condition, so we hope to inform future practice with the discussion of our case.

Ethics

Verbal informed consent was obtained from the patient for publication of this case report.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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