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Case Report

Anesthetic Management of a Parturient with Spinal Muscle Atrophy for Cesarean Section: A Case Report

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ABSTRACT

Spinal muscular atrophy is a rare genetic neuromuscular disease characterized by loss of anterior horn cells of spinal cord and brain stem nuclei, resulting in progressive muscle weakness. Anesthesia for these patients is risky because of the risk of worsening of muscle weakness and consequent postoperative respiratory complications. We report anesthesia management in a nulliparous parturient with type 3 disease who underwent urgent cesarean delivery due to progressive decline of amniotic fluid index.

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Spinal muscular atrophy (SMA) is a rare autosomal recessive neuromuscular disease characterized by loss of anterior horn cells of spinal cord and brain stem nuclei, resulting in progressive muscle weakness. Four types of SMA are defined; in which lifespan as well as onset time, signs and symptoms significantly vary. Type 1 is fatal and patients merely survive the second year of their life. Impairment of motor development during infancy (6-18 months) is the early manifestation of SMA type 2 which may result in disability. These patients may survive to their thirties and death occurs due to respiratory problems [1]. SMA type 3 and 4 are more benign and progress more gradually. It might present during childhood and early adulthood and normal life expectancy in both types is expected. Therefore, many patients will survive to the fertile age and become pregnant. Women with SMA type 3 show considerable variability in their symptoms; as some might need wheelchair assistance since childhood while others can ambulate independently. SMA type 4 is the mildest form [1]. However, there is very little information about the management of anesthesia in SMA cases. We report anesthetic management in a SMA pregnant woman who underwent urgent cesarean delivery.

Case Report

A 27-year-old nulliparous parturient presented for urgent cesarean section at 37 weeks of gestational age because of ongoing reduction of amniotic fluid index. She was a known case of SMA type 3 diagnosed through muscle biopsy two years ago. Early manifestations were weakness in shoulder girdle, followed by inability to maintain neck in upright position. She also complained of occasional difficulty in swallowing. After becoming pregnant, the patient experienced progressive weakness and dyspnea, not affecting her daily activities and lower limb tonicity and mobility. Her pregnancy was also complicated with gestational diabetes mellitus controlled by oral agents. Preoperative pulmonary function test (PFT) revealed a moderate restrictive lung disease. Mouth opening was normal with Mallampati 2 and an active herpetic lesion was seen on her lower lip. She had a mild to moderate neck weakness.

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Two medium-bore IV line were placed and crystalloid infusion started. We monitored ECG, noninvasive blood pressure, heart rate and SpO2, all revealing normal values. Then, patient was seated with assistance to perform spinal anesthesia. Mild kyphoscoliosis was detected on examination although not seemingly to result in technical difficulties. We used a 25-gauge Quincke needle through parasagittal approach in L4-L5 lumbar space. First attempt was successful and 10 mg hyperbaric bupivacaine 0.5% (Bupivacaine MYLAM SANT PRIEST-FRANCE) in combination with 25µg fentanyl was injected intrathecally. Patient felt immediate warming in her lower limbs. After 10 minutes, pinprick test revealed both sided sensory block up to T12 dermatome. We then slightly changed the patients position to Trendelenburg. 5 minutes later, she couldn't raise her right leg (Broomage score 3) and sensory block spread to T4 dermatomes in the right side. However, she still was able to move her left knee and foot. So we tilted the patient leftward. After 5 minutes, following cephalad spread of sensory block in the left side, the surgery was begun. We administered 10 mg ketamine with the beginning of incision to improve patient's comfort and two additional 10 mg boluses were also given during surgery. Hemodynamic was stable throughout and she had no respiratory problem.

After successful delivery of an apparently healthy Baby boy (Apgar score 9 out of 10), obstetricians faced with uterine atony, which forced us to take some measures other than oxytocin infusion to resolve atony including administration of 400 mg sublingual misoprostol, followed by 250 mg intramuscular prostaglandin F2a and 500 mg intravenous tranexamic acid. With regard to preoperative Hb 11 g/dl of the patient, 800 ml estimated blood loss was compensated with 3 liters of crystalloids. For postoperative pain management diclofenac suppository administered and patient was admitted to intensive care unit. Postoperative course was uneventful and she was discharged on the third day after surgery.

Discussion

We performed spinal anesthesia to preserve patient's respiratory function, prevent muscle relaxant side effects and avoid spread of herpetic lesion to the lower respiratory tract. In our experience, a small intrathecal dose of fentanyl would not cause respiratory depression but could reduce the total dose of intrathecal bupivacaine. Our challenge was slow progression of sensory blockade, which made us to use positional changes to achieve a desirable block level within 20 minutes. For patient comfort and additional analgesia, we also used small boluses of intravenous ketamine. Delays in obtaining a sufficient level of sensory and motor blocks might be due to lumbar spine deformity and lower concentration of administered bupivacaine.

In a review article by Abati and Corti [1], pregnancy outcomes of SMA parturients was investigated. They frequently reported some co-morbidities, namely: joint contractures, scoliosis and restrictive lung disease. However, there is not much data available about anesthetic technique of choice in these patients.

Anesthesia may be associated with serious risks in SMA patients, mainly depending on degree of muscle weakness and respiratory function and therefore should be evaluated preoperatively. In general anesthesia method, underlying restrictive lung disease, increased sensitivity to nondepolarizing muscle relaxants, succinylcholine-induced rhabdomyolysis and hyperkalemia and likelihood of difficult intubation can complicate anesthesia. Anesthesia induced muscle weakness may prevent early tracheal extubation and cause mechanical ventilation of the lungs after surgery [2-3]. Airway management is challenging in SMA patients because of probable abnormal anatomy of the face and contractures in masticatory muscles, as a result of bulbar weakness, cervical deformities and contractures [3]. In general, it is best to use ultra-short-acting anesthetic drugs in these patients. Anesthesiologists may prefer regional over general anesthesia. However, regional anesthesia has also its own challenges. Spinal deformities and previous corrective back surgeries are common in SMA patients, facing anesthesiologists with technical difficulties and unpredicted spread of local anesthetics as well as the possibility of block failures [4]. There is also concern about the worsening of muscle weakness with neuraxial anesthesia. Epidural anesthesia can be done in SMA, provided that severe deformity would not be an obstacle. Combined spinal epidural anesthesia has the benefit of using smaller and safer doses of intrathecal drugs [5]. The awake fiberoptic intubation with dexmedetomidine without muscle relaxants [6], general anesthesia with the use of propofol and alfentanil without muscle relaxant [7] and transversus abdominis plane block along with ilioinguinal/iliohypogastric block [8] has also been described in SMA patients. Respiratory depression and residual muscle weakness are postoperative concerns that mandate intensive care unit admission for close observation.

Natural childbirth and labor analgesia are also reported in mild form of SMA; though, because the second stage of labor requires adequate muscle strength, the use of assistive devices may be necessary. Furthermore, obstetricians should be aware of the possible need for emergency surgery [9].

Conclusion

Spinal anesthesia with lower than usual doses of bupivacaine plus fentanyl was safe for cesarean section

in the SMA parturient. Additionally, delayed onset and insufficient level of block can be managed with appropriate positioning and low dose intravenous ketamine.

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