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Case Report

Anesthetic Management of a Pregnant Patient with Spinal Muscular Atrophy for Cesarean Section: Case Report

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Abstract

Spinal muscle atrophy (SMA) is a group of rare inherited neuromuscular disorders characterized by selective degeneration of anterior horn cells of the spinal cord which leads to proximal muscle weakness and paralysis. We describe the anesthetic management of a multigravida diagnosed with type 3 SMA for a repeat cesarean section. She had worsening symptoms of progressive lower motor weakness during her second trimester with preserved pulmonary function. Combined spinal epidural anesthesia using an intrathecal local anesthetic with opioids was used to maintain the mechanics of pulmonary function.

Depending on disease symptomalogy, both general and regional anesthesia can be performed safely.

Keywords: Spinal muscular atrophy; Anesthesia; Pregnancy; Cesarean section; Case report.

Introduction

Spinal Muscle Atrophy (SMA) is a group of inherited neuromuscular disorder characterized by selective degeneration of anterior horn cell motor neurons in the spinal cord leading to proximal muscle weakness and paralysis. The disease can cause respiratory dysfunction and in 20% of cases cranial nerves are involved, leading to disabilities like dysphagia [1]. As there are very few reports of the anesthetic management of a parturient with SMA, we are adding to the body of knowledge of this rare disorder. Patient gave written consent for publication of the case report.

Description of the Case

A 28-year-old African-American woman with type 3 SMA presented for elective repeat cesarean section. She had been a healthy child who achieved her milestones until the age of five when she began to have difficulty climbing stairs; she later developed a wide base gait. Because of worsening limb girdle weakness, she had a muscle biopsy at age of 8 which resulted in a diagnosis of type 3 SMA.

The patient had had two prior surgeries. She had a cholecystectomy under general anesthesia at age 14. Then, for her first pregnancy at age 20, she was given an epidural for labor analgesia. Because of fetal distress, the plan was changed to a C-section under spinal anesthesia. No perioperative complications were documented in either procedure.

During the second trimester of her more recent pregnancy, her extremities became weaker and she had fallen multiple times. Electromyography revealed evidence of neutrophilic changes consistent with her diagnosis. She was scheduled for a C-section at 37 weeks. The preoperative assessment was unremarkable except for her girdle weakness. There was no respiratory muscle weakness. She was obese (BMI of 35) and had a Mallampati score of 3. Combined spinal epidural technique was chosen for her anesthesia; 1.4 mL heavy bupivacaine was injected into the intrathecal space with 15 mcg fentanyl and 250 mcg morphine for postoperative analgesia. The baby was born with Apgar scores of 9 and 10 at 1min and 5 min. She had an uneventful postoperative course was discharged after three days. She was later followed up by neurology and PFT suggested a normal study.

Discussion

SMA is a rare autosomal recessive inherited disorder with variable presentation depending on the severity of the disease. There are four types of SMA that are categorized by the severity, age of on set, and prognosis (Table 1) [2].

In addition to motor weakness, patients can present with severe kyphosis, pulmonary dysfunction, and cranial nerve involvement. Our patient, who had survived into adulthood with no scoliosis or respiratory dysfunction, had type 3.

	Age of onset	Maximum function achieved	Prognosis
Type 0 (very severe)	Neonate with prenatal sign	Never sits	If untreated no survival after one month of birth
Type 1 (severe)	0-6 months	Never sits	If untreated life expectancy <2 years
Type 2 (intermediate)	7–18 months	Sits but never stands	Survival into adulthood
Type 3 (mild)	>18 months	Stands and walks	Survival into adulthood
Type 4 (adult)	10–30 years	Stands and walk	Survival into adulthood

Table 1: Classification of spinal muscle atrophy [2].

In addition to motor weakness, patients can present with severe kyphosis, pulmonary dysfunction, and cranial nerve involvement. Our patient, who had survived into adulthood with no scoliosis or respiratory dysfunction, had type 3.

The literature on the anesthetic management of these patients is limited. Both regional and general anesthesia has been used with favorable outcomes. Our patient had three procedures with three different anesthetic technique and had a safe perioperative course.

Because of the rarity of SMA, there is limited worldwide experience of anesthesia for C-section. There is higher incidence of premature labor and C-section. Muscle weakness may worsen during second trimester, with possible improvement after delivery. Loss of respiratory function and mobility are the most important risk factors during pregnancy.

Patients should be warned of the deterioration of respiratory functions and be monitored with PFTs to assess for restrictive disease.

During general anesthesia, bulbar weakness and cervical spine changes alter the intubation anatomy unfavorably. Succinylcholine is associated with the risk of life-threatening hyperkalemia and rhabdomyolysis. Rocuronium may lead to prolonged residual weakness due to increased sensitivity as a result of impaired production of choline acetyltransferase [3]. An alternate technique to avoid muscle relaxant will be rapid sequence induction with propofol with alfentanil [1]. Use of spinal and epidural anesthesia has been reported (Table 2) [4-6].

We chose to do a combined spinal epidural because we anticipated a longer procedure as this was a repeat C-section. Moreover, our patient had tolerated regional anesthesia previously, did not have scoliosis or prior back surgery, and, although she had muscle atrophy, there was no change in her respiratory status. We used 1.4 mL heavy bupivacaine 0.75% with a block to last the entire procedure. A long-acting intrathecal opioid was used for postoperative analgesia.

General anesthesia • Muscle relaxants • Succinylcholine: risk of life threatening hyperkalemia and rhabdomyolysis. • Non-depolarizing relaxants- increased sensitivity due to impaired production of choline acetyltransferase. • Bulbar weakness and cervical spine changes alter the intubation anatomy – anticipate difficult airway • Delayed extubation and prolonged mechanical ventilation. • Pulmonary complications. • Risk of aspiration – pharyngeal muscle weakness.
Epidural Anesthesia • Slow controlled block. • Technical difficulties – disease process – kyphoscoliosis and prior instrumentation. • Risk of Dural puncture, patchy block, inadequate anesthesia and block failure.
Spinal Anesthesia • Technical difficulties – consider Ultrasound. • Positioning concern due to kyphosis. • Potential risk of exacerbation of chronic neurological condition. • Acute restrictive respiratory deficit.

Table 2: Special consideration: various anesthetic techniques.

There is a possibility that the patient's neurological condition might deteriorate spontaneously in the perioperative period, quite unrelated to anesthesia. Our patient had no neurologic deficit in either the immediate postoperative period nor after 4 months.

Conclusion

Understanding the pathophysiology of the disease and the implications during pregnancy are important for safe anesthetic and postpartum care. The risks and benefits of different anesthetic techniques should be considered and be tailored to the variable clinical presentation.

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