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CASE REPORT

Pre-emptive awake airway management under dexmedetomidine sedation in a parturient with spinal muscular atrophy type-2

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ABSTRACT
Historically, pregnancy in females with spinal muscular atrophy was contraindicated due to the great risk to the parturient, but with improved management and increased survival more patients are becoming pregnant.
We describe the management of a pregnant patient with spinal muscular atrophy type-2, who had severe restrictive lung disease, extensive spinal fusion that precluded neuraxial anesthesia, and chronic respiratory failure on nocturnal Bilevel Positive Airway Pressure. Airway management was further complicated by limited mouth opening and cervical spine ankylosis.

Keywords: Airway; Dexmedetomidine; Spinal muscular atrophy

Introduction

Spinal muscular atrophy (SMA) represents a heterogeneous spectrum of disease with progressive degeneration of motor neurons in the anterior horn of the spinal cord. Although there are X-linked and autosomal dominant forms, it is second only in frequency to cystic fibrosis in the group of autosomal recessive diseases; with a prevalence of 1:6000 to 1:11000 newborns.[1,2] It is the most common genetic cause of infant mortality. Severity correlates inversely with *SMN2* gene copy number.[2] Five variants of SMA have been characterized, delineated by age of onset, age of death, and motor milestones (Fig. 1).[2,3]

The dominant feature of SMA is proximal muscle weakness, with the lower limbs more frequently affected than the upper. Disease progression may involve intercostal and accessory muscles of respiration, leading to respiratory failure. Joint hypermobility or contracture, scoliosis, dislocation, and temporomandibular joint ankylosis may occur. Scoliosis and chest wall dysfunction result in restrictive lung disease, further complicating pulmonary reserve.

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Case report

The patient was a 32-year-old African-American female with SMA type-2. She was 152 cm tall, weighed 56.7 kg, resulting in a body mass index (BMI) of 24.4 kg/m^2. She had undergone extensive posterior fusion of her thoracolumbar spine and was wheelchair-dependent, also having left hip subluxation/dislocation. She had progressive difficulty with mouth opening. Respiratory function had been stable on nebulizer treatments and nocturnal Bilevel Positive Airway Pressure (BiPAP) since her teens, and recent pulmonary studies revealed a severe restrictive defect without obstruction.

Despite being counseled about the implications of pregnancy for her SMA, she wanted to conceive and her implanted contraception had been removed, with pregnancy resulting seven months later. Her trismus had progressed such that she had difficulty eating, which resulted in significant weight loss. Management of her temporomandibular joint dysfunction was planned but deferred due to her pregnancy. She presented to the obstetric clinic at eight weeks of estimated gestational age (EGA), but transport problems delayed her attendance at the anesthetic clinic until 19 weeks of EGA. A multidisciplinary team involving obstetrics, anesthesia, oral-maxillofacial surgery, pulmonary medicine, and otolaryngology was employed.

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SMA Type	Physical Symptoms	Age at Diagnosis	Life Expectancy
0	Severe weakness, hypotonia	Birth	Less than 6 months
1 Werdnig-Hoffman	Unable to sit without assistance	Birth to 6 months	Less than 2 years
2 Infantile	Unable to stand or ambulate; orthopedic complications common	Before 18 months	More than 2 years
3 Kugelberg-Welander	Mild; ambulatory without orthopedic or respiratory complications	Adolescence/Adulthood	Unaltered
4	Mildest form; similar to Type 3 in presentation	Adolescence/Adulthood	Unaltered

Fig. 1 Current classification of Spinal Muscular Atrophy (SMA)

It was clear that previous spinal surgery rendered a neuraxial technique impossible, muscle weakness precluded vaginal delivery, and cesarean delivery under general anesthesia would be rendered difficult due to problems with airway management caused by her limited mouth opening (<1 cm) and cervical ankylosis. Endoscopic evaluation of her airway suggested that even under optimal conditions, fiberoptic intubation would be extremely difficult.

Though any surgical intervention at this time risked the possibility of premature delivery, the potential for deterioration in her respiratory status as pregnancy progressed prompted the decision to perform a mature stoma tracheostomy. It was considered that the advantages over a traditional tracheostomy were that it would be a secure conduit, without risk of airway loss or false passage, and that nocturnal positive pressure ventilation could be provided through it, with reversal after delivery. However, anesthetic management was still challenging due to the lack of pulmonary reserve and the risk of apnea with a difficult airway.

Surgery was performed at 24 weeks of EGA. An awake technique with sedation had been chosen because the patient also experienced claustrophobia and panic attacks. Two doses of antenatal corticosteroids were administered prior to surgery for fetal lung protection. Pre-operative fetal heart tones were obtained. She had been told that emergency cesarean delivery would only be performed in the setting of severe maternal or fetal compromise.

Due to her small body habitus, oxygen at 2 L/min was supplied by a standard pediatric anesthesia facemask that was strapped to the face. The patient was positioned on the operating table in a semi-supine position, with the back of the table elevated approximately 30°. Dexmedetomidine was chosen since it does not typically cause apnea and permits maintenance of spontaneous respiration. The recommended dose is a 1 μg/kg bolus; we slowly administered 0.5 μg/kg to assess the effect and to allow for the detection of bradycardia. An infusion was started, slowly titrating up to 1.2 μg/kg/h. Immediately prior to commencement of surgical stimulation, the other half of the dexmedetomidine bolus was given. Fentanyl boluses (25 μg) were given as needed to a total of 100 μg. The surgeon injected 10 mL of 1% lidocaine with 1:100 000 epinephrine into the anterior neck, and after incising the skin created "I" shaped flaps of the trachea which were then sutured to the skin to create a mature stoma. The patient tolerated the procedure well and slept throughout, waking comfortably at its conclusion. A cuffed tracheostomy cannula was placed through the stoma to provide oxygen at 6 L/min, allowing rest while under observation in the surgical intensive care unit (ICU). She was able to transition to a tracheostomy collar that same night. Since postoperative fetal heart rate monitoring showed periods of absent variability, continuous monitoring was used while in the ICU.

On the first postoperative day the cuffed tracheostomy cannula was changed to a stomal stent to allow mouth-breathing and phonation. By the second postoperative day, the fetal heart tracings had normalized and the biophysical profile was reassuring. She used the stomal stent during the day and slept with high-flow nasal cannula oxygen at night. Three days postoperatively she transferred to a general ward and she was discharged home 12 days after her procedure.

Six days after discharge (27 + 1 weeks of EGA) she returned to the obstetric emergency department complaining of progressive upper extremity weakness and

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