ISSN 2689-8268 | Volume 4

American Journal of Surgery and Clinical Case Reports

Case Report Open Access

Anesthesia Management in Patient with Spinal Muscular Atrophy (SMA) Type 2

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Received: 08 May 2022
Accepted: 30 May 2022
Published: 04 Jun 2022
J Short Name: AJSCCR

Citation:

Yasemin Akcaalan, Anesthesia Management in Patient with Spinal Muscular Atrophy (SMA) Type 2. Ame J Surg Clin Case Rep. 2022; 4(16): 1-3

Keywords:

Spinal muscular atrophy (SMA); Anesthesia; Airway management

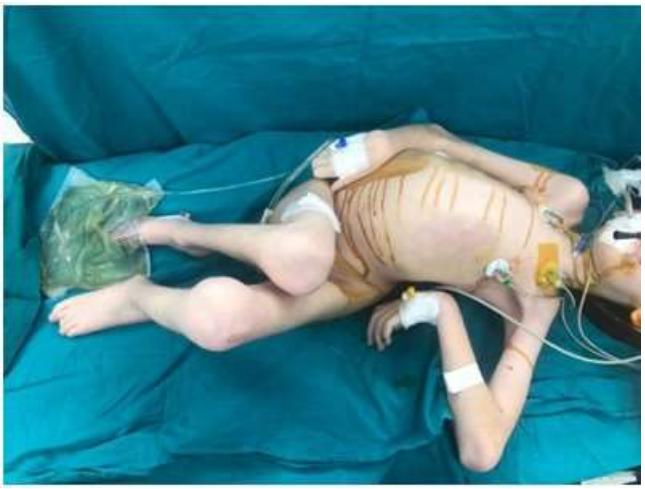
1. Abstact

Spinal muscular atrophy (SMA) is a neuromuscular disease which has varioous clinical progressions. There are difficulties in the management of anesthesia for this disease. The most common anesthetic risks are airway management and respiratory complications. With this case report, we wanted to review the anesthesia management in patients with this disease. Spinal muscular atrophy (SMA) is a neuromuscular disease manifested by weakness and atrophy of the proximal muscles, which occur due to progressive degeneration of the anterior horn cells of the spinal cord. Genetic transition is observed with autosomal recessive inheritance in 98% of patients and autosomal dominant or X-linked inheritance in 2% [1]. It is one of the most common recessive hereditary fatal diseases in children. The incidence of the disease is 1/6,000 to 1/10,000. The disease has various clinical progressions. It progresses from proximal muscle weakness of the lower extremities to dysphagia and respiratory failure. SMA affects proximal muscles more than distal muscles. In severe cases, it results to an increase in hyporeflexia and sensory nerve dysfunction. Sensory nerve function is only impaired in severe cases. The disease does not impair intellectual and cognitive functions [2]. SMA is divided into five subtypes according to its age of occurrence and the severity of muscle weakness. It is further divided into subtypes in terms of anesthetic risks. The early onset of symptoms and their severity increase the risk of complications due to anesthesia. The most common anesthetic risks are caused by lung problems and bulbar dysfunction. In terms of anesthesia management and applications, gastroesophageal reflux, perioperative and postoperative respiratory complications and difficult airway properties, which are more common in patients with SMA Type II, are features that should be considered [1]. In our case, a 14-year-old patient diagnosed with SMA type II who had previously been operated on for scoliosis was operated on for implant removal due to an infected vertebral implant.

2. Case

Our study focused on a 14-year-old, 22-kilogram male patient, with no inbreeding between parents. Despite that he was diagnosed with SMA at the age of one, his other siblings have no history of additional diseases. The patient underwent scoliosis surgery six times. The last operation took place in the external center a year ago. He was subsequently intubated in the postoperative intensive care unit. After having a leakage at the wound site, he has since been admitted to our hospital for the last two months. Our initial contact with the patient showed that his extubated consciousness was open, the spontaneous breathing was relaxed, and he did not need O_2 support. A physical examination found that there was paraplegic lower limb, muscle weakness in the upper limb (muscle strength 2/5), and severe contracture in the lower and upper extremities (Figure 1). The examination also found his Mallampati score at two and the patient's mouth opening to be within normal limits. Cardiology and pulmonology departments were consulted in the preoperative anesthesia evaluation of the patient. While the ECO assessment was normal, recommendations received from pulmonology indicated a severe lung restriction due to spinal deformity in the patient. In the preoperative blood samples, WBC was 4.3/ml, Hb was 10.5g/dL, Hct was 32.1%, Plt was 280^{000} and CRP was 0.041 mg/L. Informed anesthesia consent form was obtained from the patient's family. ECG, oxygen saturation, non-invasive blood pressure and BIS monitoring of the patient were performed on the operating table. The patient underwent general anesthesia. At induction, the doctors administered 2 mg of dormicum, 20 mg of lidocaine, 80 mg of propofol, 25 mcg of fentanyl. No neuromuscular blockers were administered. He was intubated when BIS was

forty-six. Since the patient had a difficult intubation, the Cormack Lahen score was three. He was intubated with the spiral cuffed number 5 tube with direct laryngoscopy. One percent sevoflurane induction and remifentanyl infusion of 0.1-0.2 mcg/kg/min were used to maintain anesthesia. After the patient was given the left lateral decubitus position, the surgical procedure commenced. While his hemodynamics remained stable throughout the surgery, his BIS was monitored between 42-48. The surgery lasted for 2 hours and 15 minutes. Dexketoprofen 25 mg iv was administered for postoperative analgesia near the end of surgery. By the extubation stage, after all the anesthetics were turned off, the patient received 100% O_2 for respiration. When the BIS value was 70-75, the patient had a tidal volume of 100-120 ml, after which the patient was extubated. After the operation, he was taken to ICU without O_2 support but with $SpO_2$97. The patient was followed up in intensive care for one day postoperative and then taken to the department.



3. Discussion

The most common surgical operations performed in patients with SMA are orthopedic operations such as osteotomy, tendon relaxation and scoliosis surgery. There is no ideal and standard method of anesthesia in these situations. Anesthetic risks vary between the subtypes of SMA and the severity of the disease. These risks include various problems such as respiratory failure, difficulty in airway management, gastroesophageal reflux and associated aspiration, increased need for postoperative intensive care, and restrictions on the use of neuromuscular blockers [3]. In SMA patients, especially those with SMA types I and II, the gradually increasing worsening of respiratory function and dyspnea are usually observed. On the one hand, weakness in the inspiratory muscles can cause respiratory failure. While on the other hand, weakness in the expiratory muscles can cause accumulation of secretions because of insufficient cough and chronic atelectasis [4]. In addition, due to spinal deformities such as kyphoscoliosis, restrictive lung disease may occur, which may have a severe clinical course. As a result, a bad lung with a reduced respiratory capacity may be encountered. Such patients need to be monitored in the postoperative intensive care unit. Even after minor surgical procedures, prolonged hospitalization should be recommended [5].

Airway control and difficult intubation are the main anesthetic problems in SMA patients. Both mask ventilation and endotracheal intubation difficulties can be encountered due to scoliosis-related deformities and the patient's joint contractures. The decrease in mouth opening may be parallel to the increase in age or may be secondary to ankylosis of the mandibular joint. Existing studies have found that 30% of SMA type II young patients have difficulty opening their mouth. This rate may increase to 80% in later ages [6]. In patients who are considered difficult to intubate, laryngeal mask, fiberoptic intubation, retrograde intubation and/or intubation with a video laryngoscope should be kept in mind as alternative options. Caution should be exercised during the use of LMA in terms of gastroesophageal reflux and aspiration, which are especially common in SMA type I and II patients [1,2]. The most important and noteworthy things to consider in terms of anesthetic drugs used during general anesthesia in SMA patients are non-depolarizing neuromuscular blocking agents and opioids. A more accurate approach would be to follow up by monitoring TOF in patients using muscle relaxants. Opioids have only been used perioperatively in SMA patients. Opioids should be used in a titrating, monitoring and follow-up manner. When used for postoperative analgesia, they can cause respiratory depression due to a limited respiratory reserve in patients. Thus short-acting opioids will be a more suitable option for intraoperative use [1].

4. Results

There is no ideal anesthesia management in SMA patients. The most common anesthetic risks are airway management and respiratory complications. With these in mind, we successfully performed both induction and maintenance of anesthesia to the patient. This was done with a minimum number of opioids without muscle relaxants. Although sevoflurane has a central muscle relaxant effect, we believe that the combination of 1% sevoflurane and remifentanyl can be used reliably both for intraoperative anesthesia management and postoperative recovery.

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