

ANAESTHETIC MANAGEMENT OF SPINAL MUSCULAR ATROPHY FOR LAPAROSCOPIC CHOLECYSTECTOMY

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Abstract

We report the anaesthetic management of a female patient with Spinal Muscular Atrophy (SMA) presented for laparoscopic cholecystectomy. In order to avoid prolonged recovery, we chose to use total intravenous anaesthesia (TIVA) with propofol and remifentanyl. No neuromuscular blocking agent was used. TIVA provided satisfactory anaesthesia and conditions for surgical intervention. The intra- and post-operative course was completely uneventful. We consider that TIVA without neuromuscular blocking agents could be a good anaesthetic choice for patients with SMA when general anaesthesia is required for laparoscopic surgical procedures.

Introduction

Spinal Muscular Atrophies (SMAs) comprise a group of neuromuscular disorders due to degeneration of the anterior horn cells. Published data referring to adult patients with SMA receiving general anaesthesia are rare¹⁻⁴.

Case report

We report a case of a 35-year old female patient (170 cm, 94 kg) with SMA III who presented for laparoscopic cholecystectomy due to acute cholecystitis. The patient was diagnosed with SMA at the age of six and was unable to walk for the last 2 years. The diagnosis was based on electromyography (EMG) (decreased electrical activity in lower limbs) and DNA study (Polymerase Chain Reaction (PCR) technique revealed lack of axons 7 and 8 in her survival motor neuron gene).

Preoperative physical examination and laboratory tests were normal, except for a moderate leucocytosis. Her muscle tone and reflexes, especially in the lower limbs, were remarkably reduced. Her muscular strength was estimated as 2-3/5 normal for upper limbs and 1/5 normal for lower limbs.

Perioperative monitoring included electrocardiography, invasive arterial blood pressure, pulse oxymetry, end-tidal concentration of CO₂ (ETCO₂) and Bispectral Index Score (BIS). Anaesthesia was induced with propofol 120 mg IV and remifentanyl 2.5 µg/min in continuous IV infusion. No neuromuscular blocking drug was administered. Direct laryngoscopy revealed a Cormack-Lehane view grade 1. BIS score at that time was 34. The patient's trachea was intubated with a size 7.5 cuffed tube. Anaesthesia was maintained with continuous IV infusion of propofol 400-600 mg/hr and remifentanyl 3.5-5.5µg/min. Intermittent Positive Pressure Ventilation with 50% oxygen in N₂O was used. During pneumoperitoneum (up to 15 mmHg) Peak Inspiratory Pressure (PIP) was increased from 17 cm H₂O to 21 cm H₂O, respiratory system compliance was reduced from 49 ml/cm H₂O to 27 ml/cm H₂O, and ETCO₂ was between 35 and 40 mmHg. The procedure lasted 70 minutes. TIVA was discontinued and patient was extubated within 10 minutes. She was transferred to the Postanaesthetic Care Unit and back to ward 1 hour later. Postoperative analgesia was achieved with morphine 2 mg IV, parecoxib IV 40 mg /12 hours and paracetamol-codeine suppositories (1000 + 30) mg/6 hours. Her postoperative course was uncomplicated. She was discharged on the 3rd postoperative day.

Discussion

SMA demonstrates autosomal recessive inheritance affecting 1 in 100000 births⁵. The gene responsible was mapped to region q13 of chromosome 5⁶. SMA diagnosis is based on muscle biopsy and direct gene analysis. Decreased EMG activity is also a diagnostic feature^{5,7}. The International SMA Consortium has established the following classification⁷: In type I (Werdnig-Hoffman Disease) symptoms begin before six months of age and survival is less than 2 years. In type II symptoms begin between 7 and 18 months of life. Patients cannot stand unsupported and have a short life span. Type III (Kugelberg-Welander Disease) represents 8% of SMA cases. This milder type affects children past the age of 18 months. Survival is usually normal. Patients are able to stand and often walk unaided, but they eventually end up in a wheelchair. They often suffer from respiratory tract infections.

Published information regarding anaesthetic management of patients with SMA is limited. Anaesthetic problems include a possible

hypersensitivity to non-depolarizing neuromuscular blocking agents, succinylcholine-induced hyperkalaemia, respiratory complications, and need for postoperative mechanical ventilation because of muscle weakness and general anaesthetics' prolonged action¹⁻⁴. In order to avoid all these possible problems we used TIVA with short acting agents. We avoided neuromuscular blocking drugs for their unpredictably prolonged action. Pneumoperitoneum had no major effect on PIP or lung mechanics. Ventilation and operating conditions were satisfactory. Patient's recovery was rapid and satisfactory.

There are no reports about the anaesthetic management of patients with SMA undergoing laparoscopic procedures. According to the experience gained from this case, we suggest that TIVA without neuromuscular blocking agents may be a good anaesthetic choice for patients with SMA when general anaesthesia is needed for laparoscopic surgical procedures.

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