Successful Perioperative and Anesthetic Management of a Cancer Patient With Myotonic Dystrophy (Steinert’s Disease) Undergoing A Thyroidectomy and Mastectomy

Primary Author: Yury Potylchansky MD
The University of Texas MD Anderson Cancer Center

Co-Authors: Acsa Zavala, MD; Elena Potylchansky, MD; Marc Rozner, MD (deceased); Omar Alnatour, Medical Student; Ravish Kapoor, MD; Shital Vachhani, MD; Ursula Uduak Willaims, MD;

Potylchansky Y, Alnatour O, Kapoor R, Tsai JY, Vachhani S, Williams UU, Zavala AM, Potylchansky E

The University of Texas MD Anderson Cancer Center Department of Anesthesiology and Perioperative Medicine, Houston, TX

Introduction

Myotonic dystrophy (MD) is an autosomal dominant multisystem disease characterized by muscle weakness and wasting, cardiac conduction abnormalities, endocrine dysfunction (thyroid and adrenal), and myotonia. It is amongst the most common forms of muscular dystrophy and has a reported incidence of 1:8000. It begins in early adulthood and is classified into types 1 and 2. Type 1 MD (T1MD), also known as Steinert’s disease, represents 98% of MD cases and involves mutations in the DMPK gene on Chromosome 19q. Type 2 presents with milder symptoms and involves a mutation in the CNBP gene on Chromosome 3q, representing only 2% of patients with T1MD. In both types, these mutations result in variable systematic abnormalities that can prove fatal for patients undergoing surgery if not managed properly. We present a case of the successful perioperative anesthetic management of a cancer patient with T1MD undergoing a segmental mastectomy and thyroidectomy.

Case Report

A 47 year-old woman with T1MD presented for segmental mastectomy with oncoplastic reconstruction and total thyroidectomy with bilateral central neck dissection. Common symptoms of T1MD, such as dyspnea, sleep apnea, dysphagia, dysarthria, dysphonia, aspiration, and cardiac abnormalities were not reported. Prior to surgery, a multidisciplinary approach incorporating various medical specialties including internal medicine, surgery, and anesthesia was employed.

In the holding area, a peripheral intravenous (IV) line was started. After signing the anesthesia consent, the patient was pre-medicated with IV midazolam 0.5 mg and famotidine 20 mg. In the operating room, standard monitors were applied and the patient was induced with IV fentanyl 100 mcg, lidocaine 100 mg, propofol 150 mg, and glycopyrrolate 0.2 mg. Muscle relaxants were avoided. The patient was carefully intubated with a cuffed neural integrity monitor electromyogram endotracheal tube (6.0) using a Miller 2 blade (grade 1 view). An arterial line was placed for blood
pressure monitoring and serial arterial blood gases. A hemodynamic monitor was used to assess additional cardiac parameters. We maintained total intravenous anesthesia (TIVA) with a propofol infusion. Analgesia was achieved using IV acetaminophen 1000mg and infiltration of surgical sites with liposomal bupivacaine. In addition to increasing the room temperature to 24 degrees Centigrade, a lower body forced air warmer, fluid warmer, and heat-loss prevention cap were used to maintain normothermia. The surgery was uneventful and the patient was extubated without complications after meeting extubation criterias. She stayed in overnight recovery and was discharged home the next day.

Discussion

In a retrospective analysis of 219 T1MD patients who underwent surgery under general anesthesia, Matheiu et al. found that most perioperative complications were related to the pulmonary system. It is recommended that pulmonary function tests be performed and in this patient they were within normal limits. T1MD patients have an increased risk of temporomandibular joint dislocation due to bone remodeling caused by altered jaw biomechanics, so extreme caution was taken in manipulating and instrumenting the airway. Patients with T1MD are also very sensitive to the sedating effects of opioids and anxiolytics, which can compromise respiratory drive, so precautions were taken in the administration of such medications. A multimodal approach to analgesia allowed us to minimize opioid administration. Fentanyl was used to alleviate laryngeal response to direct laryngoscopy and subsequently, acetaminophen and local anesthesia with liposomal bupivacaine were used for pain control. One of the most common post-operative complications is delayed-onset apnea, so the patient was admitted and monitored overnight.

Patients with T1MD are at an increased risk of sudden cardiac death due to conduction abnormalities and increased risk of dysrhythmias. Strict fluid management and electrolyte replacement as necessary allowed us to help prevent cardiac complications. Additionally, an esophageal pacer was readily available had an arrhythmia occurred. Succinylcholine was avoided since its effects in T1MD patients are unpredictable and it can lead to hyperkalemic cardiac arrest since these patients have a reduced sodium-potassium pump capacity. Volatile anesthetics can exacerbate cardiomyopathy, so they were avoided as well.

Since hypothermia and shivering can induce a myotonic contracture, the operating room was kept warm with addition to the use of warmed IV fluids, forced-air blankets, and a heat-loss prevention cap. With these measures, normothermia was maintained.

Lidocaine was given prior to propofol to minimize response to direct laryngoscopy and prevent pain-induced myotonic contractures. T1MD patients have a high risk for aspiration due to weakness of pharyngeal muscles and delayed gastric emptying, so famotidine was administered prior to induction. Muscle relaxants were avoided because they can cause a prolonged and unpredictable response in T1MD patients.

Given the multifaceted and variable nature of this disorder, a multi-disciplinary approach was taken for the entirety of this patient’s care. Perioperative planning took place months in advance and it was the combined multidisciplinary efforts that allowed for optimal care of our patient.

References


