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Case Report
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Anesthetic Management of a Patient with Huntington's Disease (HD): A Case report

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Abstract

Huntington disease (HD) is a rare autosomal dominant disease affecting the central nervous system (CNS) that is attributed with distinct anesthetic challenges and implications. We present a patient with HD who underwent general anesthesia for two different orthopedic surgeries using, respectively, total intravenous anesthesia (TIVA) and inhalational anesthesia; neuromuscular blockade reversal was obtained utilizing, respectively, traditional agents and Sugammadex. There have been no prospective studies on anesthetic management of patients with HD, and clinicians rely solely on case reports or prior experience in assessing general anesthetic safety in these patients. This case report presents different methods in the anesthetic management of the same patient with HD.

Introduction

Huntington disease (HD) is an autosomal dominant disease caused by cytosine-adenine-guanine (CAG) trinucleotide repeat in the Huntingtin gene on chromosome 4p. The protein, Huntingtin, is present throughout the body but symptomatology is primarily due to central nervous system (CNS) dysfunction. Pathological destruction of neurons in the basal ganglia, particularly in the putamen and caudate nucleus, is evident [1]. Symptoms manifest in the form of progressively worsening cognitive deterioration, involuntary choreiform movements, psychiatric disturbances, and dysphagia [2]. Death typically occurs 10-30 years from onset of disease and is generally due to aspiration complications [3]. Age of onset is generally between ages 30-45 but is dependent on the number of trinucleotide repeats found in the gene. Greater than 40 repeats demonstrates full penetrance with earlier onset, while less than 28 repeats yield no symptoms. Anticipation is commonly seen between generations with HD. It is associated with people of caucasian ancestry due to its prevalence of approximately 5.7 per 100,000 in North America, and 0.4 per 100,000 in Asia and 2.7 per 100,000 worldwide [1].

The anesthetic management of patients with HD has been complicated by pharyngeal muscle involvement, prolonged apnea, generalized tonic spasm, postoperative fever, prolonged recovery, increased sensitivity to benzodiazepines, hypertonia, and postoperative shivering [4]. The optimal anesthetic management of these patients has yet to be established due to the increased risk of aspiration, although multiple different agents have been successfully used for induction, muscle relaxation, and maintenance of anesthesia. In this report, we present our intraoperative anesthetic management of the same patient with HD who underwent two orthopedic operations.

Case Report

A 63-year-old, caucasian female weighing 55.3 kilograms with PMH of HD, seizures, dementia, anxiety, depression, hyperlipidemia, and adult failure to thrive presented for open reduction internal fixation (ORIF) of a right proximal humerus fracture due to a fall from her bed. Physical examination did not demonstrate any choreiform movements, but she did have difficulty communicating. Her home medications included Amantadine, Buspirone, Donepezil, Fluoxetine, Levetiracetam, Lorazepam, Quetiapine, and Risperidone. She was brought to the OR, and, after pre-oxygenation, a modified rapid sequence induction was performed with 100 mcg of Fentanyl, 50 mg of Lidocaine, 60 mg of Propofol, and 50 mg of Rocuronium. The patient was intubated with a Miller 2 blade with a grade 1 view. Anesthesia was maintained with TIVA using Propofol and intermittent boluses of Fentanyl. She received Dexamethasone, IV Acetaminophen, and

Ketorolac. Neuromuscular blockade was reversed with 100 mg of Sugammadex and extubation was without issue. She tolerated the procedure well without complications.

Five days later, the patient presented to the emergency department with dizziness, weakness, and a hemoglobin of 5.9 g/dL with a hematoma over her surgical site. She was brought to the operating room for a revision of ORIF and hardware removal of the right proximal humerus and was taken care of by a different anesthesia team. After pre-oxygenation the patient received 100 mg of Propofol, 50 mg of Lidocaine, 100 mcg of Fentanyl, and 30 mg of Rocuronium, but was found to have an infiltrated midline. A second IV was placed and 100 mg Propofol, 50 mg of Fentanyl, and 20 mg of Rocuronium were administered. Bag mask was easy without an airway device. She was intubated with a Mac 3 blade with a grade 2 view. Anesthesia was maintained with Sevoflurane throughout the case. At the end of the case, she received Ondansetron for PONV prophylaxis. Neuromuscular blockade was reversed with 3 mg of Neostigmine and 0.4 mg of Glycopyrrolate. She was again extubated without issues. She tolerated this procedure well without complications.

Discussion

HD is a rare disease, there have been no prospective studies on anesthetic management, and we must solely rely on case reports to assess general anesthetic safety in these patients. There are numerous anesthetic considerations that must be considered. Many HD patients are on antipsychotics, antidepressants, antiepileptics, and benzodiazepines for management of psychiatric disorders and chorea. Theoretically, there may be inhibition or induction in the cytochrome P450 system leading to changes in the medication(s) half-life. Anesthesiologists should be aware of these medications and their potential to interact with anesthetic agents.

Previous cases have reports of prolonged apnea after administration of 50 mg of Succinylcholine, but the patient was found to have a low plasma cholinesterase level. There have been conflicting population studies that show a genetic connection between HD and a rare plasma cholinesterase fluoride-resistant allele. But in a report of the literature, there have been multiple incidences of Succinylcholine use without apnea [4]. In both of our cases, muscle relaxation was asked for by the surgical team and Rocuronium was used. Reversal of muscle relaxation was treated by Sugammadex in the first case. In the second operation, our patient did receive Glycopyrrolate without any deleterious effects in post-operative recovery. There have been case reports of Metoclopramide and Anticholinergic medications worsening choreiform movements [3]. It is preferred to utilize a quaternary amine, Glycopyrrolate, to reverse muscular blockade because it does not cross the blood-brain-barrier.

Prior studies report uses of 5mg/kg of Sodium Thiopental causing prolonged apnea over 1 hour. Although this dose seems aggressive, as most HD patients are debilitated [3], Sodium Thiopental is no longer in use as an induction agent. For both our cases induction was performed with Fentanyl, Lidocaine, and Propofol without complications. In the first case, a decision was made to perform an RSI, which is generally used for patients with HD, as late symptoms include dysphagia, which places HD patients at an increased risk of aspiration. The most common cause of death in this patient population is aspiration pneumonia. For the second case the decision was made for an RSI, but an infiltrated IV necessitated bag mask ventilation until a second IV could be placed. In both operations, ventilation throughout the case was normal and no signs of aspiration were noticed. There have been case reports of HD patients requiring awake fiberoptic intubation [5].

Maintenance of anesthesia has been safely described using inhalation agents and total intravenous anesthesia. Kang et al. [5] theorize that a TIVA with Propofol and Remifentanil would be ideal for HD patients due to ease of titration and rapid recovery without lingering effects potentially leading to airway compromise. In this patient, both inhalational with Sevoflurane and TIVA with Propofol were used without complications. Both cases used a Bispectral Index for titration of anesthesia. Postoperative delirium can be increased in HD patients after general anesthesia [4]. Our patient did not experience any alterations that affected her post-operative care in either case.

Successful spinal anesthesia has been reported in the literature twice. Due to the location of the operation this was not considered, and although regional anesthesia was potentially an option, it was not utilized [6,7].

There have been many case reports of patients with HD showing successful anesthetic management, although it seems none have shown two different anesthetic regimes on the same patient in such short succession, as in our case report. The perfect anesthetic is likely never going to be found and will be at the discretion of the provider for each individual patient. Further studies should be performed to help elucidate the best anesthetic choices for these patients.

Author Contributions

David McEnerney D.O.: This author assisted in the anesthetic management of the patient and drafting of manuscript.

Mohamad El Churafa D.O.: This author assisted in study drafting of manuscript.

Giuseppe Giuratrabocchetta M.D.: This author assisted in the anesthetic management of the patient and drafting of manuscript.

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