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Case Study

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ANAESTHETIC MANAGEMENT OF A PATIENT WITH HUNTINGTON'S CHOREA - A CASE REPORT

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ABSTRACT

Huntington's disease is a rare dominantly inherited progressive autosomal disease affecting the basal ganglia. The disease manifests later in life as progressive mental deterioration causing personality change and involuntary choreiform movements. The most ominous motor symptom, dysphagia, hinders nutritional intake and places the patient at an increased risk of aspiration. The real challenge for an anaesthetist in the management of a patient with Huntington's chorea lies in treating the frail elderly uncooperative patient, treating the patients suffering from malnutrition and dealing with increased risk of aspiration and exaggerated response to Succinylcholine and Thiopentone in these patients. We report the anesthetic management of a 62-year-old female patient with Huntington's chorea admitted for an elective orthopedic procedure.

KEYWORDS: Huntington's disease; Basal ganglia; Involuntary choreiform movements; Autosomal; Personality change; GABA.

INTRODUCTION

Huntington disease (HD), a neurodegenerative autosomal dominant disorder, with a worldwide prevalence of 2.7 per 100,000. It is a progressive disease characterized by involuntary choreatic movements with cognitive and behavioral disturbances. The mechanism of the disease process is premature destruction of neurons in the basal ganglia, primarily causing the degeneration of neurons in the putamen, caudate as well as the cerebral cortex.[1] The abnormal gene responsible is located in chromosome 4.[2] The disease occurs as a result of cytosine, adenine, and guanine (CAG) trinucleotide repeats on the short arm of chromosome 4p16.3 in the Huntington (HTT) gene. There is complete penetrance and variable expression of the disease. Since it is an autosomally transmitted disease, men and women are equally affected. The onset of the disease is usually relatively late, at about the age of 30-45 years, and symptoms that begin before the age of 20 (juvenile onset of the disease) occurs in 10% of affected persons.[3,4] Death typically occurs 10-30 years after the onset.[4] The disease is characterized by wide spectrum of symptoms classically defined as occurring in three domains: motor, cognitive, and psychiatric. Late in the disease, dysphagia due to bulbar dysfunction becomes a symptom with high morbid impact, as aspiration is a common occurrence, and cachexia together with aspiration pneumonia is a common cause of death.[5] The anaesthetic management of a patient with Huntington's chorea poses a lot of challenges particularly because these patients are at increased risk of pulmonary aspiration. Moreover, prolonged apnea, intense shivering and generalized tonic spasm, altered response to the administration of barbiturates and succinylcholine, and increased sensitivity to midazolam are considered high risk perioperative complications.[3,6] The primary goal for the anaesthetic management of these patients is to provide airway protection and a safe and rapid recovery.

CASE REPORT

A 62-year-old female, weighing approximately 50 kgs who had been diagnosed with Huntington's chorea at the 42 years-of-age on some unknown medication was admitted for Open reduction and internal fixation (ORIF)with plates and screws for fracture distal one-third shaft of humerus right side with radial nerve palsy. Family history of chorea was present in 2 siblings. Her surgical history included tubectomy 30 years back. On physical examination, the patient displayed gross choreiform movements in all the limbs. Her laboratory values were all within normal limits. The patient's chest radiograph and electrocardiogram (ECG) was also noncontributory. The patient had no history of cardiovascular, pulmonary, hepatic, renal, endocrine, or

gastrointestinal tract disease. Since the patient had gross abnormal choreiform movements, general anesthesia was chosen as the procedure of choice in order to ensure an amicable surgical field. The patient was premedicated with alprazolam 0.25 mg and ranitidine 150 mg orally. The patient was preoxygenated and routine monitors such as pulse oximetry, ECG, noninvasive blood pressure and end-tidal carbon dioxide were placed. A mixture of oxygen and 3% sevoflurane was administered. As the patient became sedated, intravenous access was obtained. To complete the induction, inj. glycopyrrolate 0.2 mg, inj. propofol 100 mg and inj. atracurium 25 mg were administered. Tracheal intubation was performed with 7.5 mm ID cuffed endotracheal tube. Inj. Fentanyl 100 mcg and inj. ondansetron 4 mg were supplemented. Anesthesia was maintained with Oxygen and nitrous oxide (33: 66), Isoflurane 1% and intermittent boluses of inj. fentanyl. The case proceeded uneventfully and at the end of surgery, which lasted for 2 hours, patient was extubated uneventfully with reversal using inj. neostigmine 2.5 mg and glycopyrrolate 0.5 mg intravenously. Recovery and immediate postoperative period was uneventful. The patient was transferred to a general ward and discharged from the hospital after 3 uneventful days.

DISCUSSION

Huntington's disease is a rare neurodegenerative disorder that may have implications for the anesthesiologist. This condition results in increased production of a mutant protein, Huntington that leads to cell loss and atrophy, mainly of GABAergic striatal medium spinal output neurons of the caudate, putamen, and cortex.[7] Psychotropic medications such as antipsychotics, antidepressants, benzodiazepines, and antiepileptics are frequently used for symptom management of this curable condition.[7] So, an anesthesiologist should be aware of potential interactions of these medications with frequently used anesthetics drugs. There are numerous anesthetic considerations for the patient with Huntington's disease. Premedication with metoclopramide is not done because of reports of an exacerbation of the choreiform movements.[8] Anticholinergics should also be avoided with this group, since there is a relative balance between dopamine and acetylcholine in the striatum, and anticholinergics may further increase the choreiform movements.[9] If an anticholinergic must be given, glycopyrrolate, a quaternary ammonium that cannot cross the blood-brain barrier, is preferred over atropine, a tertiary amine that can cross the blood-brain barrier.

There have been previous reports that sodium thiopental can cause prolonged apnea. Davies and Blanloeil reported prolonged apnea for a period of 1 hour following induction with sodium thiopental.[10,11] Although excessive induction doses of sodium thiopental were considered as the cause in these reports (greater than 5 mg/kg), we chose to avoid thiopental and instead employed propofol, an agent that has not been implicated in the literature for prolonged apnea in this group. The unpredictability of thiopentone can be circumvented by using induction agents like midazolam and propofol.[10,12]

Gualandi and Bonfanti reported a case of prolonged apnea of 2 hours following the administration of 50 mg succinylcholine.[13] Genetic variation of plasma cholinesterase is a potential cause of prolonged muscle paralysis after receiving succinylcholine.[14] However, in some studies conducted by Brown and Cross[15] and Costarino and Gross[16], succinlycholine did not cause prolonged apnea. Despite this, muscle paralysis after succinlycholine may be prolonged in patients with Huntington's chorea. Therefore, succinylcholine should be used with caution. Short and intermediate duration agents, such as rocuronium and cisatracurium, also appear acceptable for use.[14]

Further, the incidence of postoperative shivering can be minimized by use of isoflurane and sevoflurane in place of halothane. The low solubility coefficients of these agents allow early recovery without postoperative confusion and early return of protective airway reflexes, thus reducing the risk of pulmonary aspiration. However, these inhalational agents carry the theoretical risks of generalized muscle spasms and protracted recovery periods in which the already compromised airway is at increased risk. To avoid these risks associated with inhalational agents, using total intravenous anesthesia (TIVA) can be beneficial in that it allows precise titration of anesthetic, ease of induction, and a rapid and controlled recovery.[6]

Another anesthetic consideration in this patient population centers around the dysphagia that is so common to this group. Dysphagia is probably the most significant motor symptom because it hinders the nutritional intake of those with Huntington's disease and places them at risk for aspiration.[4] Therefore, at the termination of the anesthetic, airway reflexes must be assurred to minimize the risk of postoperative aspiration.

We chose to use sevoflurane for induction (since isoflurane is a modest airway irritant) and then switched to isoflurane for the remainder of the procedure. This allowed for a more rapid recovery and faster return of airway reflexes, thus minimizing the risk of aspiration. We administered an intermediate duration, non-depolarizing neuromuscular blocking drug, atracurium and short acting anaesthetic agents like propofol and fentanyl. Using the above techniques, we could successfully anesthetize a patient with Huntington's chorea without perioperative complications.

REFERENCES

1. Purdon SE, Mohr E, Ilivitsky V, Jones BD. Huntington's disease: pathogenesis, diagnosis and treatment. Journal of Psychiatry and Neuroscience, 1994; 19(5): 359.

2. Gusella JF, Wexler NS, Conneally PM et al. A polymorphic DNA marker genetically linked to Huntington's disease. Nature, 1983; 306(5940): 234-8.
3. Gilli E, Bartoloni A, Fiocca F, Dall'Antonia F, Carluccio S. Anaesthetic management in a case of Huntington's chorea. Minerva anestesiologica, 2006; 72(9): 757-62.
4. Cangemi Jr CF, Miller RJ. Huntington's disease: review and anesthetic case management. Anesthesia progress, 1998; 45(4): 150.
5. Martin JB. Huntington's disease: New approaches to an old problem: The Robert Wartenberg Lecture. Neurology, 1984; 34(8): 1059.
6. Nagele P, Hammerle AF. Sevoflurane and mivacurium in a patient with Huntington's chorea. British journal of anaesthesia, 2000; 85(2): 320-1.
7. Walker FO. Huntington's disease. The Lancet, 2007 Jan 20; 369(9557): 218-28.
8. Patterson JF. Choreiform movement associated with metoclopramide. Southern medical journal, 1986; 79(11): 1465.
9. Stewart JT. Huntington's disease. Am Fam Physician, 1988; 37(5): 105-14.
10. DAVIES DD. Abnormal response to anaesthesia in a case of Huntington's chorea. British Journal of Anaesthesia, 1966; 38(6): 490-3.
11. Blanloeil Y, Bigot A, Dixneuf B. Anaesthesia in Huntington's chorea. Anaesthesia, 1982; 37(6): 695-6.
12. Kaufman MA, Erb T. Propofol for patients with Huntington's chorea? Anaesthesia, 1990; 45(10): 889-90.
13. Gualandi W, Bonfanti G. A case of prolonged apnea in Huntington's chorea. Acta Anaesthesiologica, 1968 Jan 1; 19: Suppl-6.
14. Kivela JE, Sprung J, Southorn PA, Watson JC, Weingarten TN. Anesthetic management of patients with Huntington disease. Anesthesia & Analgesia, 2010; 110(2): 515-23.
15. Browne MG, Cross R. Huntington's chorea. BJA: British Journal of Anaesthesia, 1981; 53(12): 1367.
16. COSTARINO A, GROSS JB. Patients with Huntington's chorea may respond normally to succinylcholine. The Journal of the American Society of Anesthesiologists, 1985; 63(5): 570.