Case Report

Anesthesia in a Patient with Huntington's Chorea

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Abstract

Huntington's chorea is a hereditary disorder characterized by continuous involuntary movements. Symptoms usually occur between the ages of 30 and 45 years and include choreatic movements, progressive mental deterioration, and ataxia. Various anesthetic techniques have been tried in these patients, both regional as well as general anesthesia. These patients are of special concern to anesthesiologists with respect to preoperative assessment and securing intravenous access due to their continuous movements. Such patients have to be managed with respect to providing airway protection and ensuring rapid and safe recovery. We report a case of Huntington's chorea who was anesthetized successfully for removal of foreign body cricopharynx without any deleterious postoperative outcome.

Key words: Cricopharynx, foreign body, general anesthesia, Huntington's chorea

Introduction

Huntington's disease is a neurodegenerative genetic disorder that affects muscle co-ordination and leads to cognitive decline and psychiatric problems. It is the most common genetic cause of abnormal involuntary movements called chorea and so the disease is called Huntington's chorea. These patients have continuous jerky, random, and uncontrollable movements, and so when they are posted for any surgical procedure, they are of special concern to anesthesiologists. Due to the continuous movements, their preoperative assessment becomes difficult. History taking is difficult as these patients tend to forget past things due to some psychiatric component of the disease. Similarly, obtaining some investigations such as electrocardiogram becomes difficult due to continuous movements.

These patients are at higher risk of intraoperative complications such as pulmonary aspiration, prolonged response to succinylcholine and thiopental, and increased sensitivity to midazolam.

General anesthesia with newer anesthetic agents such as propofol and sevoflurane were tried in our patient with rapid and safe recovery.

Case Report

A 42-year-old female, diagnosed to be suffering from Huntington' chorea 5 years back, was brought to the operation theater for emergency removal of foreign body cricopharynx. The patient was diagnosed as a case of chorea, but the details of the previous records were not available with the relatives. This patient was on some drugs for her disease, but had stopped them 1 year back. As she did not have her previous records, the exact treatment which she was receiving could not be assessed.

She had a history of ingestion of mutton piece 4 h back, and following which she had a feeling of something stuck in her throat and was painful. She had difficulty in deglutition even for her saliva. After examination, she was diagnosed as a case of ingestion of foreign body cricopharynx and was posted for emergency removal of the foreign body.

On physical examination, she had continuous involuntary movements of her head, hands, and legs. She did not have any respiratory difficulty. Her motor power was normal and there was no neurological deficit. There was no history suggestive of rheumatic fever. Rest of the history and physical examination was not significant. Her laboratory values were

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within normal limits and electrocardiogram could not be taken due to continuous movements.

After obtaining an informed consent, the patient was shifted to the operation theater. As she had continuous movements of her hands, intravenous access was difficult. Hence, she was made unconscious with 100% oxygen and sevoflurane using a face mask and intravenous access with 20 gauge cannula was secured in her left forearm. Routine monitors such as pulse-oximeter, twelve lead electrocardiogram, and noninvasive blood pressure monitors were attached to the patient. Her electrocardiogram was normal.

She was premedicated with intravenous injections of ranitidine 50 mg, ondansetron 4 mg, and glycopyrrolate 0.2 mg. Then, she was induced with injection propofol 100 mg intravenously. Immediately after induction, a continuous cricoid pressure was applied and injection succinylcholine 80 mg was administered intravenously to facilitate endotracheal intubation with cuffed endotracheal tube of 7.5 mm internal diameter. Endotracheal tube was connected to Bain's circuit and anesthesia was maintained with oxygen and nitrous oxide at a ratio of 30:70. The patient's spontaneous respiratory attempts were regained 5 min after administration of succinylcholine. Further relaxation was then maintained with injections atracurium 25 mg and sevoflurane.

Rigid esophagoscopy was done and the foreign body was removed. The procedure lasted for 15–20 min. After ensuring adequate respiratory attempts of the patient, she was administered injection neostigmine 2 mg and glycopyrrolate 0.4 mg. She was extubated once adequate spontaneous respiration and consciousness were regained. After extubation, her involuntary movements re-appeared in the recovery room. She was observed for signs of upper respiratory tract obstruction.

The postoperative period was uneventful and there were no complications. Patient was transferred from recovery to the ward after regaining of reflexes and discharged from the hospital after 3 days.

Discussion

Chorea is the most visible feature of Huntington's disease, a genetic disorder affecting brain. It is caused by an autosomal dominant mutation in either of an individual's two copies of a gene called Huntington.[1] Degeneration through the cortex and abnormalities of dopaminergic substantia nigra pars compacta occur.[2] There occurs widespread dysregulation of glutamatergic and dopaminergic signaling system.

Chorea is initially exhibited as general restlessness, small unintentional movements, lack of co-ordination, or slow saccadic eye movements. These symptoms may progress to continuous involuntary movements, difficulty in chewing and swallowing, and cognitive dysfunction. Cachexia and aspiration are the most common causes of death in these patients.

These patients have various anesthetic concerns. Preanesthetic assessment becomes difficult in these patients as they tend to forget things due to psychiatric component of the disease. In addition, obtaining basic investigations such as electrocardiogram is difficult due to continuous movements. These patients are on psychotropic medications such as haloperidol and fluphenazine for management of their symptoms. Anesthesiologists should be aware of the interaction of these medicines with anesthetic drugs.

These patients have bulbar muscle dysfunction, so there is a risk of aspiration. The risk of aspiration was more in our patient as we had planned for general anesthesia. Furthermore, there was a risk of dislodgment of foreign body after administering general anesthesia, but since the foreign body was at the level of cricopharynx, which is the narrowest part, the risk of dislodgment was less.

The use of pro-motility drugs to decrease the risk of aspiration should be avoided as these drugs can aggravate chorea symptoms in these patients.[3] Hence, we preferred ondansetron for premedication. Similarly, anti-cholinergics should be avoided in these patients as there is relative imbalance between dopamine and acetylcholine in the striatum, and anti-cholinergics may further increase the choreiform movements.[4] Glycopyrrolate is preferred over atropine for premedication as it does not cross blood–brain barrier, so we used it for premedicating our patients. Meperidine is also avoided in these patients as it has anticholinergic properties such as atropine. As few studies have reported prolonged effect of benzodiazepines, we avoided midazolam in our patient.

In these patients, various anesthetic techniques have been used, both regional as well as general. Use of Total intravenous anaesthesia (TIVA) has been found to be a good option in these patients. Volatile anesthetic agents viz. halothane[5] and isoflurane[6] have been safely used in these patients. We preferred to use sevoflurane in our patient, which gave us a dual advantage for securing intravenous access as well as for maintenance of anesthesia and had a property of rapid recovery.

These patients show increased sensitivity to barbiturates[7] and hence we preferred propofol for induction, which also has advantages such as short duration of action and rapid recovery. Prolonged response to thiopental sodium in these above-mentioned cases could be attributed to larger doses (7.5 mg/kg) given in such diseased and debilitated patients. In contrast, few cases had normal response to both thiopental sodium and propofol.

Though previous studies had shown prolonged effects from succinylcholine, we did not find such findings in our patient. Rather our patient, being an emergency case, we were planning for rapid sequence intubation and so succinylcholine was used to facilitate intubation. Our patient did not show prolonged effect of succinylcholine and had spontaneous respiratory attempts just 5 min after administration of succinylcholine. In one case, prolonged action of succinylcholine was found, and most probably,

it might be because of abnormal plasma cholinesterase and not related to Huntington's chorea.[6] Alternatively, rocuronium could have been also used for rapid sequence induction, but our institute did not have this drug and so we opted for succinylcholine. In addition, the use of nondepolarizing muscle relaxants such as atracurium in our case did not have any prolonged muscle relaxant action which shows that nondepolarizing muscle relaxants can be safely used in these patients. Few studies have proved safety of mivacurium in these patients.

Finally to conclude, patients with Huntington's chorea can be safely managed with the use of newer induction agents such as propofol and volatile agents such as sevoflurane with rapid recovery and no adverse postoperative outcome.

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Conflicts of interest

There are no conflicts of interest.

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