Anesthetic Management in a Patient with Huntington's Chorea: A Case Report

Sezen Kumaş Solak

Department of Anesthesiology and Reanimation, Bağcılar Training and Research Hospital, Istanbul, Turkey

Correspondence: Sezen Kumaş Solak, Bağcılar Training and Research Hospital, Department of Anesthesiology and Reanimation, Istanbul, Turkey

ABSTRACT

Huntington's chorea (HC) is a rare autosomal dominant neurodegenerative disorder characterized by progressive choreiform movements, cognitive decline, and psychiatric disturbances. The perioperative management of patients with HC poses significant challenges, particularly due to pharyngeal muscle involvement, increased aspiration risk, and altered responses to anesthetic agents. We present the case of a 54-year-old male patient with HC who underwent emergency choledochotomy under total intravenous anesthesia (TIVA) with awake fiberoptic intubation. The use of propofol and remifentanil-based TIVA, combined with low-dose rocuronium and bispectral index (BIS) monitoring, allowed for safe anesthetic management without perioperative complications. This case highlights the importance of careful preoperative planning, appropriate agent selection, and vigilant intraoperative monitoring in patients with HC.

Keywords: Huntington's chorea; anesthesia; awake fiberoptic intubation; total intravenous anesthesia; aspiration

INTRODUCTION

Huntington's chorea (HC) is an autosomal dominant hereditary disorder characterized by progressive neurodegeneration, primarily affecting the basal ganglia and the caudate nucleus. The clinical triad of the disease consists of personality changes, dementia, and choreiform movements. The causative mutation is located on the short arm of chromosome 4, and due to its dominant inheritance pattern, both sexes are affected equally [1,2].

The prevalence of HC in Western countries is estimated between 4 and 6 per 100,000 individuals. Symptom onset typically occurs between the ages of 30 and 50, although juvenile forms accounting for approximately 10% of cases may manifest before the age of 20.

Behavioral symptoms, including depression, frequently precede motor findings by up to a decade. Death usually occurs 10 to 30 years after disease onset [3].

From an anesthesiological perspective, HC presents unique perioperative risks. Pharyngeal muscle dysfunction significantly increases the risk of pulmonary aspiration. Additionally, HC patients may exhibit altered or unpredictable responses to several anesthetic and adjuvant agents, including succinylcholine, thiopental sodium, midazolam, anticholinergics, and meperidine [1,4]. Careful agent selection and meticulous intraoperative monitoring are therefore essential. Herein, we report the successful anesthetic management of a patient with HC undergoing emergency abdominal surgery.

CASE REPORT

A 54-year-old male patient (weight: 65 kg, American Society of Anesthesiologists physical status II) with a known diagnosis of HC was scheduled for emergency choledochotomy. On physical examination, choreiform movements of the lower extremities, intellectual disability, and excessive salivation were observed. A family history was significant; the patient's mother had exhibited similar symptoms, although she had never received a formal diagnosis. Written informed consent was obtained from the patient prior to the procedure.

Preoperative laboratory evaluation revealed elevated white blood cell count and C-reactive protein levels, consistent with an acute inflammatory process. Chest radiography showed no pulmonary pathology, and the 12-lead electrocardiogram demonstrated normal sinus rhythm. The patient's current medications included tetrabenazine 75 mg/day, paroxetine 40 mg/day, and olanzapine 10 mg/day.

Given the high aspiration risk secondary to pharyngeal dysfunction, awake fiberoptic intubation was planned. The patient was not premedicated, as metoclopramide was avoided due to its potential to exacerbate choreiform movements, and anticholinergic agents were withheld to prevent further disruption of the dopamine-acetylcholine balance in the striatum. Topical anesthesia was achieved with 10% lidocaine spray applied to the base of the tongue and the pharyngeal wall. A 7.0 mm cuffed endotracheal tube was successfully placed into the trachea via a fiberoptic bronchoscope (LF-TP, Olympus, Japan) under awake conditions.

Standard intraoperative monitoring included pulse oximetry, electrocardiography, non-invasive blood pressure measurement, and continuous end-tidal CO_2 monitoring. A bispectral index (BIS; BISquatro™, Aspect Medical Systems, USA) sensor was applied to monitor

anesthetic depth, targeting a BIS value between 40 and 60. Neuromuscular function was assessed using train-of-four (TOF) stimulation of the ulnar nerve with an electrical stimulator (Relaxograph®, Datex, Finland).

Following confirmation of correct endotracheal tube placement, anesthesia was induced with an intravenous bolus of 2% propofol, remifentanil target-controlled infusion (TCI) at an effect-site concentration of 4 ng/mL, and rocuronium 0.5 mg/kg. An arterial line was inserted via the radial artery for continuous invasive blood pressure monitoring. Throughout the procedure, propofol and remifentanil concentrations were titrated according to BIS values and mean arterial pressure. No additional neuromuscular blocking agents were required, as TOF monitoring revealed complete absence of muscle contractions following the initial rocuronium dose. Core temperature was maintained within normal limits throughout the surgery to minimize the risk of postoperative shivering and tonic spasms.

The surgical procedure lasted 190 minutes. At the conclusion of surgery, ondansetron 4 mg was administered intravenously for antiemetic prophylaxis. Remifentanil and propofol infusions were then discontinued. The patient recovered spontaneous respiration and consciousness within 6 minutes, and the endotracheal tube was removed without difficulty. The postoperative course was uneventful; no nausea, vomiting, shivering, or neurological complications were observed. The patient was transferred to the general surgery ward from the post-anesthesia care unit.

DISCUSSION

This case illustrates the successful perioperative management of a patient with HC undergoing emergency abdominal surgery. The management strategy was guided by an appreciation of the disease-specific pharmacological vulnerabilities and the primary concern of preventing pulmonary aspiration.

HC patients are at heightened risk for aspiration pneumonia due to progressive involvement of the pharyngeal musculature, as well as dysarthria and dysphagia. Awake fiberoptic intubation is widely recognized as the technique of choice for securing the airway in patients at high aspiration risk [1,3]. In the present case, this approach allowed for controlled airway management without compromising the patient's protective reflexes.

The pharmacological considerations in HC are numerous. The use of metoclopramide as a premedication agent is contraindicated, as its dopamine antagonist properties can exacerbate

choreiform movements. Similarly, anticholinergic agents disrupt the striatal dopamine-acetylcholine equilibrium, intensifying involuntary motor activity; if an anticholinergic is deemed necessary, glycopyrrolate, which does not cross the blood-brain barrier, is the preferred agent. Meperidine is relatively contraindicated due to its atropine-like structure, although isolated case reports have described its use without adverse effects [1].

Sodium thiopental has been associated with prolonged apnea in HC patients, particularly at doses exceeding 5 mg/kg [4]. Although subsequent reports have not consistently reproduced this finding at lower doses [5,6,7,8,9], propofol is generally preferred given its more predictable pharmacokinetic profile and the absence of reported prolonged apnea in this patient population. Succinylcholine may also elicit prolonged responses in HC and is best avoided. Increased sensitivity to midazolam has also been documented.

Total intravenous anesthesia (TIVA) with propofol and remifentanil is an established approach in HC [3]. Remifentanil is particularly advantageous due to its metabolism by non-specific plasma and tissue esterases, independent of hepatic function, enabling high-dose infusion without drug accumulation and facilitating rapid, predictable recovery. In the present case, TIVA permitted precise titration of anesthetic depth as guided by BIS monitoring, rapid emergence, and a smooth postoperative course.

Rocuronium, a non-depolarizing neuromuscular blocking agent of intermediate duration, was used at a reduced dose (0.5 mg/kg) and was not required to be re-administered, as evidenced by TOF monitoring. This approach minimized the risk of residual neuromuscular blockade in the postoperative period. Intraoperative normothermia was maintained to reduce the incidence of postoperative shivering, which in HC patients may trigger generalized tonic spasms.

In conclusion, the combination of awake fiberoptic intubation and propofol-remifentanil TIVA, guided by BIS and TOF monitoring, proved to be a safe and effective anesthetic strategy in this patient with HC. Avoidance of known precipitants of adverse effects, alongside careful intraoperative monitoring, is essential to minimize perioperative morbidity in this challenging patient population.

CONCLUSION

Anesthetic management of patients with Huntington's chorea requires meticulous preoperative planning and individualized drug selection. Awake fiberoptic intubation is an effective and safe technique for airway management in patients with elevated aspiration risk. TIVA with propofol

and remifentanil, combined with low-dose rocuronium and multimodal intraoperative monitoring (BIS and TOF), provides a reliable approach associated with favorable perioperative outcomes. Avoidance of potentially harmful agents such as metoclopramide, succinylcholine, high-dose thiopental, and anticholinergics is strongly recommended.

AUTHOR CONTRIBUTIONS

Concept and design: S.K.S. Data collection: S.K.S. Data analysis and interpretation: S.K.S. Manuscript writing: S.K.S. Critical revision: S.K.S. Final approval: S.K.S.

CONFLICT OF INTEREST

The author declares no conflict of interest.

ETHICS STATEMENT

Written informed consent was obtained from the patient for publication of this case report.

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