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Letter to the Editor

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Anesthetic Approach to Huntington's Chorea

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To the editor,
Huntington's Chorea (HC) is a rare autosomal dominant inherited disease which affects the caudate nucleus and basal ganglia and is characterised with progressive cellular degeneration [1]. A gene mutation on the short arm of the 4th chromosome is responsible for this disease. In HC, an increase in huntingtin production, a mutant protein resulting from the expansion of the CAG repeat in the IT15 gene, results in cell loss and atrophy, especially in the GABAergic striatal mid-spinal output neurons of the caudate, putamen and cortex [1,2]. The prevalence is around 4-6 per 100.000 and it is usually seen between 30-50 years of age. It is characterised by a triad of personality change, dementia and choreiform movements. Personality change starts ten years before choreiform movements with depression usually being the first symptom. While motor symptoms, dysphagia and dysarthria are the leading symptoms, intellectual losses are also frequently observed [3].

In patients with HC who will receive anesthesia or sedation, regurgitation and pulmonary aspiration may pose a risk in the perioperative period due to involvement of pharyngeal muscles. Weakness in respiratory functions and postoperative tremors leading to rigid spasms may cause complications in the postoperative period [3]. There are limited number of case reports in the literature regarding anesthetic management in HC. In this letter, we aimed to share the anesthetic management of a patient with CD and to increase awareness on this issue.

A 71-year-old 50 kg female patient was planned to undergo percutaneous endoscopic gastrostomy (PEG) revision under sedation. The patient's known diseases included Huntington's chorea, dementia and psychosis. Her regular medications were PPI esomeprazole, SSRI escitalopram, antipsychotic olanzapine and statin pitavastatin. On neurological examination before the procedure, the patient was conscious, orientated and coordinated, speech was dysarthric, affection was shallow, thought content was poor, there was no active agitation excitation, pupils were isochoric, there was no cranial nerve abnormality, deep tendon reflexes were vivid, there was no pathological reflex, 4 extremities had at least 4/5 motor strength with in-bed movement, choreiform movements were prominent in the upper extremities, cerebellar tests were incompatible, and oromandibular dyskinesia was present.

Physical examination revealed choreic movements in the upper extremities, swallowing disorder and excessive oral secretion. All laboratory values were within normal limits except anti Hbe and anti Hbc positivity. Increased nodular density on chest radiography and sinus bradycardia on electrocardiogram were detected. In the pre-procedure evaluation by psychiatry and neurology, it was reported that caution should be taken in terms of malignant neuroleptic syndrome and the current treatment was discontinued 24 hours ago. Routine monitoring was performed with pulse oximetry, ECG and noninvasive blood pressure. During the procedure, 5 l/min O2 was given through a nasal cannula. Vital signs and hemodynamic parameters before and during the procedure were within normal limits. Propofol 50 mg was administered for induction and additional doses of propofol were titrated to maintain an SpO2 value >95 during the procedure by monitoring the patient's alertness and vital signs. After successful PEG revision, the patient was transferred to the recovery room and then to the inpatient ward after an uneventful follow-up. Informed consent was obtained from the patient for the publication of the data.

HC is a rare neurodegenerative disease, raising some issues during anesthesia practice. Since patients are frequently treated with antipsychotic, antidepressant, benzodiazepine and antiepileptic drugs, potential interactions between these drugs and the drugs used in anesthesia should be considered. Furthermore, it should be kept in mind that postoperative symptoms such as agitation, chorea and psychosis may be exacerbated if general anesthesia is to be applied (2). Since premedication with metoclopramide and anticholinergics may aggravate choreiform movements, if anticholinergics are required, glycopyrrolate which is a quaternary ammonium that does not cross the blood-brain barrier should be preferred over atropine which is a tertiary amine that can cross the blood-brain barrier [4,5]. These patients are at risk because of prolonged apnea with succinylcholine and thiopental sodium and increased sensitivity to midazolam [6,7]. It has been reported in the literature that short and intermediate lasting nondepolarising agents such as rocuronium and cisatracurium, inhalation agents and total intravenous anesthesia (TIVA) have been used without any problems [2,8]. To avoid the theoretical risks of inhalation agents, such as prolonged shivering and prolonged recovery periods during which the already compromised airway is at increased risk, the use of TIVA seems appropriate as it may provide precise titration during anesthesia, easy induction and a rapid and controlled recovery.

Dysphagia is an important issue to be considered perioperatively in these patients. Regurgitation and aspiration may cause serious morbidity and mortality. Rapid serial intubation and awake fiberoptic intubation should be considered to ensure airway when necessary. When terminating anesthesia, extubation should be planned after airway reflexes are regained to minimise the risk of postoperative aspiration.

We preferred to use propofol in our patient because of its rapid effect and rapid recovery. In addition, we avoided the use of drugs that may worsen the patient's symptoms. Because the patient had secretions and dysphagia, we ensured that the depth of anesthesia was at a level where we could control the airway and secretions.

In this letter, we aimed to review the main goals of anesthetic management of patients with HC, in the light of this rare and infrequently occurring case. Safe anesthesia can be achieved by considering these points in cases with HC.

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