Acta Medica Nicomedia

Cilt: 3 Sayı: 2 Haziran 2020 / Vol: 3 Issue: 2 June 2020
https://dergipark.org.tr/tr/pub/actamednicomedia

Case Report | Olgu Sunumu

SUCCESSFUL ANESTHETIC MANAGEMENT AND RECOVERY WITH SUGAMMADEX IN A PATIENT WITH HUNTINGTON'S CHOREA-A CASE REPORT

HUNTINGTON KORE OLGUSUNDA BAŞARILI ANESTEZİ YÖNETİMİ VE DERLENMEDE SUGAMMADEKS KULLANIMI-OLGU SUNUMU

Meltem Genez[1*], Sule Ozbilgin[2]

[1]SADA Hospital, Department of Anesthesiology and Intensive Care, İzmir, Turkey. [2]Dokuz Eylul University, Faculty of Medicine,Department of Anesthesiology and Intensive Care, İzmir, Turkey.

ABSTRACT

Huntington's chorea (HC) is an autosomal dominant hereditary disease with progressive characteristics affecting the basal ganglia. Patients may encounter complications like increased risk of regurgitation and pulmonary aspiration, prolonged thiopental and succinylcholine response, respiratory difficulties and postoperative shivering advancing to rigid spasms. With a prevalence of 4-10/100.000, there are limited case reports about anesthetic methods for this disease in the literature. In our case report, we aimed to present the anesthetic management of a 50-year old male patient with HC disease. Avoiding the use of anticholinesterase, 100 mg sugammadex was administered, the patient was successfully extubated and recovery was ensured.

Keywords: Huntington disease, anesthetic management, sugammadex

ÖZ

Huntington koresi (HK) otozomal dominant geçişli, ilerleyici özellikte bir hastalıktır ve bazal ganglionların ilerleyici şekilde etkilenmesi ile ortaya çıkar. Hastalarda regürjitasyon ve pulmoner aspirasyon riskinde artma, uzamış tiyopental ve süksinilkolin cevabı, solunum güçlüğü, rijit spazma kadar ilerleyebilen postoperatif titreme gibi komplikasyonlarla karşılaşılabilir. Prevalansı 100000 de 4-10 olan bu hastalıkla ilişkili olarak literatürde bildirilen anestezi yöntemleri sınırlı olguda denenmiştir. Olgu sunumumuzda HK hastalığı olan 50 yaşında erkek hastanın anestezi yönetimini sunmayı amaçladık. Antikolinesteraz kullanımından kaçınmak amacıyla 100 mg sugammadeks verilen hasta başarılı bir şekilde ekstübe edildi ve derlenmesi sağlandı.

Anahtar Kelimeler: Huntington hastalığı, anestezi yönetimi, sugammadeks

[*]**Corresponding author/İletişim kurulacak yazar:** Meltem Genez; SADA Hospital, Esatpaşa, İzmir-Çanakkale Cd., No: 2 D: 1, 35660 Menemen/İzmir, Turkey.
Phone/Telefon: +90 (232) 832 99 00 **e-mail/e-posta:** smyrna_mel@yahoo.com
Submitted/Başvuru: 08.03.2020 • **Accepted/Kabul:** 02.06.2020 • **Published Online/Online Yayın:** 27.06.2020

Introduction

Huntington's chorea (HC) is an autosomal dominant hereditary disease, frequently affecting the nervous system at the basal ganglion level including the caudate nucleus, causing progressive cell degeneration. Huntington's chorea is characterized by a triad of personality changes, dementia and choreiform movements. The abnormal gene responsible for the disease is located on the short arm of the 4th chromosome.[1] The formation mechanism of the disease is associated with neuron destruction in the basal ganglia. Dementia is associated with basal ganglion dysfunction with no disorder in the cortex. Neuron degeneration begins in the striatum and progresses by spreading around it. Due to the autosomal dominant inheritance, it affects men and women equally.

Behavioral changes begin ten years before the occurrence of choreiform movements. Commonly, depression is the earliest symptom. These behavioral changes include abuse, criminal and sexual actions performed thoughtlessly. The earliest choreiform motor movements that occur are commonly the "milk maid sign" involving rhythmic squeezing and relaxing of the hand. The unstoppable repetition of motor movements is frightening for patients. People with Huntington's disease sway while walking and commonly fall. Unintentional motor movements may cause weight loss in people, and even cachexia. Dysphagia occurs with worsening motor symptoms and this situation leads to a tendency toward aspiration pneumonia.[2] Additionally, the reduction in intellectual intelligence is sometimes more frightening than motor symptoms. Dysarthria is very commonly observed.

The prevalence of the disease in western countries is 4-6 per 100,000. There are 6000 patients with HC diagnosis in England, with 20,000 known to be at risk.[1] The age of incidence typically varies from 30-50 years.[2] Additionally, 10% of those affected have the possibility of juvenile (younger than 20 years) symptoms. Death occurs 10-30 years after the beginning of the initial symptoms.

In this case report, we wanted to share anesthetic management of HC disease which is very difficult to manage and unordinary.

Case Report

A 50-year old, ASA (American Society of Anesthesiology) II male patient with a weight of 47 kg and BMI of 14.50, was operated for amputation of third toes on foot. History included 8-years since HC diagnosis, smoking habit and septoplasty operation before the disease. Before the operation, the patient's laboratory values and radiological investigation showed no abnormal findings. Current disease treatment was 100 mg Quetiapine, 15 mg Olanzapine, 5 mg Haloperidol and 2 mg Trihexyphenidyl. With widespread choreiform movements in four extremities, cooperation could not be made with the patient due to dysarthria. After a 20-gauge branula was placed in the left hand for venous access, the patient was administered 50 mg Ranitidine, 150 mg Propofol and 50 mg Rocuronium (1mg/kg) and rapid sequence intubation was performed. We didn't use any pre-medication. To avoid aspiration during ventilation, head raised position was given to the operating table and cricoid pressure was applied. After inflating the cuff, cricoid pressure was stopped. Anesthesia maintenance continued with 50% N_2O/O_2 and 2% sevoflurane. The operation was finished in 40 minutes. No hemodynamic changes were observed during the operation. For analgesia, 50 mg meperidine fluid was added. At the end of the operation, to avoid anticholinesterase use, rocuronium bromide was antagonized with 100 mg (2mg/kg) sugammadex. When spontaneous respiration movements were observed, the patient was extubated. With postoperative shivering, the patient was administered an additional dose of 25 mg meperidine. Monitored in the recovery room after operation, the patient did not develop complications in the postoperative period.

This case report was carried out with the informed consent of the family of the patient.

Discussion

Huntington's chorea patients are at high risk of intraoperative complications like regurgitation and pulmonary aspiration linked to involvement of pharyngeal muscles. The study by Gilli et al. found agents with low dissolution coefficient like isoflurane and sevoflurane were more reliable for HC patients as they ensure rapid induction and rapid awakening.[2] As aspiration is most often seen during intubation, rapid-series intubation should be performed. For this, 8% sevoflurane and rocuronium could be used along with cricoid pressure. Choreiform movements may be severe enough to prevent venous access in patients. In this situation, following ventilation with sevoflurane, intravenous cannulation can be performed.[3] In our case, firstly, we placed branula, followingly we enjected 150 mg propofol and 50 mg rocuronium accompanied by cricoid pressure with the operating table in head-up position and then rapid-series intubation was completed.

The use of metoclopramide in premedication is shown to increase the severity of choreiform movements. The use of anticholinergic agents disrupts the dopamine, acetylcholine balance in the striatum causing more severe movements. If anticholinergic use is necessary, glycopyrrolate, an agent which does not pass the blood-brain barrier, should be used. Meperidine is relatively contraindicated due to its atropine-like structure.[3] We used meperidine twice in our case. First of all we used 50 mg meperidine for analgesia. Secondly we administered low dose (25 mg) for post-operative shivering. Consequently, we didn't experience any side effects with meperidine.

Anesthetic agents like succinylcholine and thiopental sodium may cause prolonged response while there may

be increased sensitivity to midazolam and anticholinergic agents. The 1982 study by Balonleil showed that thiopental use caused extended apnea of 1 hour and above in patients.[4] Additionally, Stoelting and Miller reported thiopental at doses of 3-5 mg/kg did not affect apnea duration in HC patients.[5] There is no prolonged apnea associated with propofol use in patients with HC diagnosis. As a result, we used 2-3 mg/kg dose of propofol for our patient and did not encounter any side effects.

Gualandi and Bonfanti reported prolonged response linked to reduced plasma cholinesterase activity in HC patients. The researchers showed that apnea duration was two hours following administration of 50 mg succinylcholine.[6] In contrast, the case report by Costarino et al. reported that prolonged apnea was not observed following 0.6 mg/kg succinylcholine use for rapid-series intubation.[7] Cangemi and Miller used low-dose morphine and desflurane and successfully completed anesthesia induction without requiring the use of muscle relaxant agents.[3] Additionally, rocuronium use was common as a muscle relaxant agent for rapid-series induction in the relevant articles.

Sevoflurane or isoflurane may be chosen for anesthesia maintenance. Desflurane use should be avoided as it is an irritant.[3] We preferred sevoflurane in our case, and we didn't experience any problem. Gilli and Kang used remifentanil and propofol infusion in HC cases with total intravenous anesthesia (TIVA).[2,8] Ensuring intraoperative normothermia reduced postoperative shivering and reduced the risk of developing generalized tonic spasms. Some studies have shown TIVA use reduces the risk of postoperative tonic spasms compared with inhalation agents.

As the use of atropine should be avoided for antagonization of neuromuscular blockage, glycopyrrolate which does not pass the blood-brain barrier should be chosen instead.[3] Kang et al. used 20 mg prydostigmine and 0.4 mg glycopyrrolate for antagonization.[8] Glycopyrrolate does not pass the blood-brain barrier as it is a synthetic quaternary ammonium compound. Atropine passes the blood-brain barrier due to tertiary amine structure.[3] As glycopyrrolate is not found in Turkey, we found it appropriate to use the cyclodextrin molecule of sugammadex to avoid using atropine in our patient. Moreover, sugammadex appears to reverse neuromuscular blockade more rapidly than neostigmine.[9] In the literature, there are limited numbers of case reports about the use of sugammadex for HC cases. Publications have stated that the use of sugammadex for HC patients is reliable and may be chosen as superior to neostigmine.[10]

In conclusion, dysphagia is the most important motor symptom in HC patients. Aspiration of food linked to motor function disorder of pharyngeal muscles increases the risk of aspiration during anesthesia. Rapid-series induction should be performed to reduce this risk, with cricoid pressure applied or awake fiberoptic intubation performed in patients who are cooperative. The appropriate agents should be chosen for anesthesia maintenance with precautions taken against complications that may occur in the postoperative period. To avoid use of anticholinergic agents like atropine, sugammadex may be an appropriate choice for antagonization of neuromuscular blockage.

Compliance with Ethical Standards

Permission was granted for this report.

Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Author Contribution

MG: Concept; MG: Design; SO: Supervision; MG, SO: Literature research.

References

1. Tollis D. Who was ... Huntington? *Nurs Times*. 1995;91(27):45.
2. Gilli E, Bartoloni A, Fiocca F, Dall'Antonia F, Carluccio S. Anaesthetic management in a case of Huntington's chorea. *Minerva Anestesiol*. 2006;72:757-762.
3. Cangemi CF Jr, Miller RJ. Huntington's disease: review and anesthetic case management. *Anesth Prog*. 1998;45:150-153.
4. Blanloeil Y, Bigot A, Dixneuf B. Anaesthesia in Huntington's chorea. *Anaesthesia*. 1982;37:695-696. doi:10.1111/j.1365-2044.1982.tb01287.x
5. Stoelting RK, Miller RD. *Basics of Anesthesia*. 3rd Edition. New York: Churchill Livingstone Inc.; 1994.
6. Gualandi W, Bonfanti G. A case of prolonged apnea in Huntington's chorea. *Acta Anaesthesiol (Padova)*. 1968:19(suppl 6):235-238.
7. Costarino A, Gross JB. Patients with Huntington's chorea may respond normally to succinylcholine. *Anesthesiology*. 1985;63:570. doi:10.1097/00000542-198511000-00032
8. Kang JM, Chung JY, Han JH, et al. Anesthetic management of a patient with Huntington's chorea- A case report. *Korean J Anesthesiol*. 2013;64(3):262-264. doi:10.4097/kjae.2013.64.3.262
9. Blobner M, Eriksson L, Scholz J, et al. Sugammadex (2 mg/kg) significantly faster reverses shallow rocuronium-induced neuromuscular blockade compared with Neostigmine (50 microgram/kg). *Eur J Anaesthesiol*. 2007;24(suppl 39):125-126. doi:10.1097/00003643-200706001-00465
10. Uluer MS, Sargin M, Ozmen S. High dose Suggammadex administiration in a case of Huntington chorea. *Med Science*. 2016;5(1):301-305. doi:10.5455/medscience.2015.04.8313