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Case Report

Epidural anesthesia in a patient with Huntington's chorea

Huntington koresi olan bir hastada epidural anestezi

Mesut Erbaş[1], Burhan Dost[2], Bekir Sami Karapolat[3]

[1]Bünyan State Hospital, Bünyan, Kayseri
[2]Department of Anesthesiology and Reanimation, Faculty of Medicine, Düzce University, Düzce
[3]Department of Thoracic Surgery, Faculty of Medicine, Düzce University, Düzce

Abstract
Huntington's chorea (HC) is a rare, autosomal, dominant hereditary disorder of the nervous system. Symptoms occur in the third and fourth decade of life and manifest as involuntary choreiform movements, ataxia, and progressive mental deterioration. Only a few case reports have been published describing the anesthetic management of patients with HC. Patients are at greater risk for some intraoperative complications (e.g., regurgitation and pulmonary aspiration), poor respiratory function, prolonged response to succinylcholine and thiopental, increased sensitivity to midazolam, and postoperative shivering leading to rigid spasms. All these factors make an ideal anesthesiological management necessary for this group of patients to guarantee an absolute airway protection during all the anesthesia and a fast and safe recovery. To the best of our knowledge, epidural anesthesia in HC patients has not been reported in the literature. Herein, we present our successful experience using epidural anesthesia in a HC patient.
Keywords: Anesthesia; epidural; Huntington's chorea

Özet
Huntington koresi (HK) nadir görülen otozomal dominant geçiş gösteren sinir sistemi bozukluğudur. Semptomları hayatın 3. ve 4. dekatlarında istemsiz koreiform hareketleri, ataksi ve ilerleyici mental bozulma olarak gösterir. HK tanılı olgularda anestezi yönetimi ile ilgili sadece birkaç olgu sunumu yayınlanmıştır. Hastalar bazı intraoperatif komplikasyonlar (regürjitasyon ve pulmoner aspirasyon gibi), zayıf respiratuar fonksiyon, uzamış suksinilkolin ve tiyopental cevabı, midazolama karşı yükselmiş sensitivite, ve rijit spazmlara yol açan titremeler gibi postoperatif komplikasyonlar açısından yüksek risk altındadırlar.Tüm bunlar güvenli geri dönüş ve anestezi sırasında güvenli havayolu sağlamak için ideal anestezi yönetimi gerektirmektedir. Bildiğimiz kadarıyla, literatürlerde HK'li hastalarda epidural anestezi uygulanması bulunmamaktadır. Burada, HK tanılı olgumuzda başarılı epidural anestezi deneyimimizi sunmayı amaçladık.
Anahtar kelimeler: Anestezi; epidural; Huntington koresi

Introduction

Huntington's chorea (HC) is a rare, autosomal, dominant hereditary disorder of the nervous system. Symptoms occur in the third and fourth decades of life and manifest as involuntary choreiform movements, ataxia, and progressive mental deterioration (1). Only a few case reports have been published describing the anesthetic management of patients with HC (2-7).

Patients are at greater risk for some intraoperative complications (e.g., regurgitation and pulmonary aspiration), poor respiratory function, prolonged response to succinylcholine and thiopental, increased sensitivity to midazolam, and postoperative shivering leading to rigid spasms (2-7). All these factors make an 'ideal' anesthesiological management necessary for this group of patients which guarantees an absolute airway protection during the entire anesthesia and provides a fast and safe recovery.

To the best of our knowledge, epidural anesthesia in HC patients has not been reported in the previous literature. A 41 year-old female with HC was scheduled for elective laparoscopic cholecystectomy and epidural anesthesia was performed. Herein, we present our successful experience using epidural anesthesia in a HC patient.

İletişim/Correspondence to: Burhan Dost, Department of Anesthesiology and Reanimation, Faculty of Medicine, Düzce University, 81620 Konuralp, Düzce, TURKEY
Tel: +90 532 7042493 *burhandost@hotmail.com*

Case Report

A 41-year old female, 75 kg of weight with diagnosis age of 31 years HC was admitted to the hospital with stomachache, and was scheduled for elective surgery for cholecystectomy. The patient was on antipsychotic treatment (haloperidol; 5 mg/daily). Her father died at the age of 50 for the same disease and her elder sister (44-year old) was diagnosed as HC 3 years ago as well. Results of a preoperative and standard laboratory tests were unremarkable, except for elevated total bilirubin (1.68 mg/dl, normal range 0-1 mg/dl), amilase (271 U/l, normal range: 0-100 U/l) and lipase (112 U/l, normal range: 0-60 U/l) levels. Her physical examination was revealed mild dysarthria, mild ataxia, and gross choreiform movements in all extremities. ASA (American Society of Anesthesiologhy) physical status was I.

The patient was pre-medicated with ranitidine 50 mg iv and midazolam 1 mg iv 1 h before surgery. In the operating room after routine electrocardiogram, non-invasive arterial blood pressure, pulse oximetry monitoring, we sedated the patient with iv midazolam (0.5 mg). Before anesthesia the patient's oxygen saturation was 92%, tension arterial was 120/75 mmHg and heart rate was 78 beats/min. The patient was administered oxygen through a face mask at 2 litre/min.

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The patient was in the sitting position. A lumbar epidural catheter was placed at the interspace between the 12nd thoracal and first lumbar vertebrae using a midline approach and a loss of resistance to saline through a 18-gauge Touhy needle. A test dose of 3 ml of 2% lidocaine was administered without appreciable evidence of either intravascular or intrathecal placement of the catheter. Sensory analgesia till the sixth thoracic dermatome was achieved with incremental doses of 0.5% bupivacaine (20 ml) combined with 50 µg fentanyl before surgery commenced. After 20 minutes, surgery was started. Approximately 45 minutes after first bolus, we reinfused 10 ml of the same solution. Throughout this period, electrocardiography was normal, arterial blood pressure was in normal range, SpO_2 was greater than 98%, and the patient had no respiratory discomfort. The operation lasted 2.5 hours. For the postoperative desensibilization, the patient controlled epidural analgesia was performed by mixing morphine 4 mg with 0.125% bupivacaine 100 ml, totally 104 ml, and infusing mixture at the flow rate of 1.5 ml/h. The vital signs in the recovery room were stable. The Visual Analogue Scale (VAS) was maintained at 0 to 1 during the hospitalization. The epidural catheter was removed on the 3rd post-operative day. The patient was discharged on the fifth day after the operation without a specific finding.

Discussion

Huntington's disease is a premature neurodegenerative disease characterized by marked atrophy of the basal ganglia, particularly the caudate nucleus. It is a genetic disease transmitted in an autosomal dominant pattern (10). An ideal anesthesiological management for patients suffering from HC is still not described, but considering the high risk of perioperative complications, various techniques have been introduced by different authors, especially using the most recent anesthesiological agents, both intravenous and inhalation agents with fast kinetics (8). Until recently, a case of successful epidural anesthesia has not been reported. Experience with the management of anesthesia in HC is too limited to propose specific drugs or techniques. Reported anesthesia experience in patients with Huntington's disease is largely anecdotal and consists of a small number of case reports and letters (9).

Our patient was prescribed antipsychotic medications. The anesthesiologist should be aware of potential interactions of this medications with frequently used anesthetic drugs.

Although not observed in our patient, theoretically, general anesthesia could exacerbate psychiatric symptoms resulting in postoperative agitation, chorea, and psychosis. It is probably prudent for patients to continue the use of psychotropic medications until the day of surgery. Increased sensitivity to barbiturates and benzodiazepines among patients with HC has been reported (11).

As a result; HC in appropriate surgical procedures epidural anesthesia may be effective, comfortable, and preferable to general anesthesia, yielding frequent success rates while preventing the possible disadvantages of general anesthesia.

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