OLGU SUNUMU

Prone epidural anesthesia in a patient with Huntington's chorea; Case Report

Huntington Koresi tanılı hastada pron pozisyonda epidural anestezi; Olgu Sunumu

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ÖZET

Huntington koresi (HK); nadir görülen otozomal dominant geçiş gösteren bir sinir sistemi bozukluğudur. Semptomları hayatın 3. ve 4. dekatlarında istemsiz koreiform hareketleri, ataksi ve ilerleyici mental bozulma olarak kendini gösterir. HK prevelansı 4-10/100.000 olarak belirtilmekte ve erkek ve kadın cinsiyet açısından farklılık göstermemektedir. HK tanılı olgularda anestezi yönetimi ile ilgili sadece birkaç olgu sunumu yayınlanmıştır. Hastalar bazı intraoperatif komplikasyonlar, zayıf respiratuvar fonksiyon ve rijit spazmlara yol açan titremeler gibi postoperatif komplikasyonlar açısından yüksek risk altındadırlar. HK tanılı olgularda anestezi yönetimi ile ilgili sadece birkaç olgu sunumu yayınlanmıştır. Burada, HK tanılı olgumuzda prone Jack Knife pozisyonunda uyguladığımız başarılı epidural anestezi deneyimimizi sunmayı amaçladık.

Anahtar kelimeler : Huntington, anestezi

ABSTRACT

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. The prevalence of HC is 4-10/ 100.000 cases with no difference between males and females. Patients have greater risk for some intraoperative complications, poor respiratory function and postoperative shivering leading to rigid spasms Only a few case reports have been published describing the anesthetic management of patients with HC. In this case report; we present our successful experience using epidural anesthesia in prone Jack Knife position in a HC.

Keywords: Huntington, anesthesia

Introduction

Huntington's chorea (HC) is a rare autosomal dominant neurodegenerative syndrome. Symptoms commonly occur in the third and fourth decades and manifest as involuntary choreiform movements, ataxia, and progressive mental deterioration. The prevalence of HC is 4-10/ 100.000 cases with no difference between males and females (1).

Only a few case reports have been published about the anesthetic management of patients with HC. Patients have 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 (2,3,4). Moreover gastric content aspiration, prolonged apnea, intense shivering and generalized tonic spasm, altered response to administration of barbiturates and succinylcholine, increased sensitivity to midozolam and to anticholinergics drugs are considered at high risk as perioperative complications. All these factors make an ''ideal'' anesthesiological management necessary for this group of patients to guarantee an absolute airway protection during all anesthesia also fast and safe recovery, especially in emergency and urgency cases (3,4).

To the best of our knowledge, epidural anesthesia in HC patients has been reported only in a few case reports. But; there are no literature about prone epidural anesthesia in HC patients. In this case report; we present our successful experience of using epidural anesthesia in prone Jack Knife position in a HC.

CASE REPORT

A 67 years old female, 45 kg of weight patient with diagnosis age of 6 years HC was admitted to the Sincan State Hospital with left femur fracture. The patient has not used any medication for HC. Her father died at the age of 58 due to same disease, her brother (51 years old) was diagnosed as HC 9 years ago and her sister (53-year old) was diagnosed as HC 4 years ago as well. Six years ago; the patient has been operated for breast carsinoma at general

anesthesia and she has admitted in intensive care unit for rigid spasmus in postoperative 2 days. ASA (American Society of Anesthesiologhy) physical status was II.

After the informed content taken from patient; she was pre-medicated with ranitidine 50 mg iv and midazolam 1 mg iv 1 hour before surgery. In the operating room after routine electrocardiogram, noninvasive arterial blood pressure, pulse oximetry monitoring, we sedatized the patient with iv midazolam (2 mg). Before anesthesia the patient's oxygen saturation was 93%, tension arterial was 130/70 mmHg and heart rate was 67 beats/min. The patient was administered oxygen through a face mask at 3 litre/min.

The patient was in the sitting position. A lumbar epidural catheter was not placed at the interspace between the 3 th and 4 th lumbar vertebrae using a midline approach and a loss of resistance to saline through a 18-gauge Touhy needle. After she has been positioned laterally and epidural cateter was not placed again because of involuntary choreiform movements. The patient didn't accept general anesthesia. After that; the patient was in prone Jack Knife position. A lumbar epidural catheter was placed at the interspace between the 4 th and 5 th lumbar vertebrae using a midline approach and a loss of resistance to saline through a 18- gauge Touhy needle. The patient had no paresthesias. A closed -end catheter (0.9 mm) with three lateral orifices was inserted 5,5 cm into the epidural space. There was no blood or cerebrospinal fluid (CSF) leak through the needle or the catheter. 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 8 th thoracic dermatome was achieved with incremental doses of 0.5% bupivacaine (8 mL) combined 2 mL saline before surgery commenced. After 20 minutes, surgery was started. Approximately 75 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, SpO2 was greater than 94%, and the patient had no respiratory discomfort. The operation lasted 2.5 hours. Total fluid infusion during surgery consisted of 3000 mL of crystalloids and 500 mL of colloids. Estimated blood loss was 1000 mL and urine output was 500 mL. We replaced 1 Ü erythrocyte suspension. In postoperative term; we accepted the patient to Intensive Care Unit for haemodynamic and respiratory monitoring for 24 hours. The postoperative course was uneventful; in particular no episode of nausea, vomiting or shivering were observed. The patient was discharged from hospital on postoperative day 3.

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Discussion

Huntington's disease is a premature neurodegenerative disorder which is characterized by marked atrophy of the basal ganglia, particularly the caudate nucleus. An ideal anesthesiological management for patient suffering from Huntington's Chorea is stil not described.

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 (4,5,6). Until recently, in patients with Huntington's disease a case of successful epidural anesthesia in prone Jack Knife position has not been reported.

The prone position for epidural catheterisation is employed when epidural nerve block is used in chronic pain management; fluoroscopy is usually required. Position the patient prone on the procedure table with a pillow under the waist to decrease the lumbar lordosis (7).

In this case report; the patient didn't accept general anesthesia and we could not place the epidural catheter in epidural space on sitting and lateral positions; because of involuntary choreiform movements. In necessary; we have attempted to placement epidural catheter in prone position.

As a result; epidural anesthesia may be effective, comfortable, and preferable to general anesthesia in HC. Patients with Huntington Chorea is very difficult to place epidural catheter in sitting and lateral position because of involuntary coreiform movements.In this case we placed the epidural catheter in prone Jack Knife easily and this technique can be the first choice.

Conclusion:

Huntington's disease is a premature neurodegenerative disease. An ideal anesthesiological management for patient suffering from Huntington's Chorea is stil not described. Epidural anesthesia may be effective, comfortable, and preferable to general anesthesia in HC.

References

1. White T, Neustein S. Monitored anesthesia care for a patient with advanced Huntington's chorea. Middle East J Anesthesiol. 2013;22(2):185-186.
2. Sriganesh K, Saini J. Exacerbation of Involuntary Movements After Propofol Anesthesia in a Patient With Huntington Disease. J Neurosurgi Anesthesiol 2013; 25(2): 212.
3. Nagele P, Hammerle AF. Sevoflurane and mivacurium in a patient with Huntington's chorea. Br J Anaesth 2000; 85(2):320-321.
4. Gıllı E, Bartoloni A, Fiacco F, Dall'antonia F. Anaesthetic management in a case of Huntington's Chorea; Case Report. Minerva Anestesiol 2006;72(9):756-762.
5. Kivela JE, Sprung J, Southorn PA, Watson JC, Weingarten TN. Anesthetic management of patients with Huntington disease. Anesth Analg 2010;110(2):515-523.
6. Erbaş M, Dost B, Karapolat B. Epidural anesthesia in a patient with Huntington's chorea. Gaziantep Med J 2012;18(2): 113-114.
7. Chawla J, Raghavendra M, Lovato L. Epidural Nerve Block. 2013: http://emedicine.medscape.com/article/149646-overview