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Spinal Anesthesia Management in Central Core Disease: A Case Report

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Abstract: Central core disease is a rare neuromuscular disorder associated with leg weakness. It is a familial disease with autosomal dominant inheritance. Central core disease has been reported to be associated with malignant hyperthermia. A 25-year-old woman with central core disease was scheduled to be operated for lumbar disc hernia at L4-5 and L5-S1 interspaces. Oral dantrolene was administered prophylactically. Spinal anesthesia was performed with a 25 G Whitacre spinal needle at the L3-4 interspace. No complication was observed during the intraoperative and postoperative periods. We present a case with central core disease who was operated under spinal anesthesia with oral prophylactic dantrolene.

Key Words: Central core disease, spinal anesthesia

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Santral Kor Hastalığı Olan Olgudaki Spinal Anestezi Yaklaşımı

Giriş: Santral kor hastalığı bacaklarda güçsüzlük ile seyreden nadir görülen bir nöromüsküler hastalıktır. Otozomal dominant geçişlidir. Santral kor hastalığının malign hipertermi ile ilişkili olabileceği rapor edilmiştir. 25 yaşında santral kor hastalığı olan kadın hastanın L4-5 ve L5-S1 disk nedeniyle operasyonu planlandı. Oral dantrolen profilaktik olarak verildi. L3-4 aralığından spinal anestezi 25 G Whitacre iğne ile yapıldı. İntraoperatif ve postoperatif dönemde komplikasyon gözlenmedi. Biz profilaktik oral dantrolen verilip spinal anestezi uyguladığımız santral kor hastalığı olan olguyu sunduk.

Anahtar Sözcükler: Santral kor hastalığı, spinal anestezi

Introduction

Central core disease (CCD) is a rare neuromuscular disorder that is most often characterized by weakness in the legs, but occasionally affects other muscle groups as well. CCD is not a progressive disease. It is familial with autosomal dominant inheritance [Chromosome 19 q13.1; Gene Locus RyR1 (Ryanodine receptor-1 gene]. A mutation of the RyR1 results in two clinical phenotypes - CCD and malignant hyperthermia (MH) (1). CCD has been reported to be associated with MH in a small proportion of patients (1-3). MH is an inherited condition that causes muscle rigidity and uncontrollable fever in response to the administration of certain anesthetics. One of the most common anesthetics that can cause this reaction is the combination of halogenated inhalation agents and succinylcholine. Surgery is possible for patients with CCD, but it requires extra precautions. For the CCD patient, safe drugs are barbiturate, benzodiazepines, droperidol, ketamine, local anesthetics (lidocaine, bupivacaine), nitrous oxide, and propofol. The halothane-caffeine contracture test remains the gold standard. Prophylactic dantrolene administration is controversial. There are some studies indicating that dantrolene therapy is effective in these cases (2,4,5). We present a case with CCD who was operated under spinal anesthesia and was administered prophylactic oral dantrolene.

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Case Report

A 25-year-old woman with CCD was scheduled to be operated for lumbar disc hernia at L4-5 and L5-S1 interspaces. In infancy, she acquired head control at the normal time, sat independently at 10 months and began to walk at 12 months. But she had difficulty in squatting down, running, jumping, climbing up the stairs and turning from left to right in bed. She was diagnosed as CCD at 12 years of age as a result of a muscle biopsy taken from the vastus lateralis. Oxidative stains showed uniform fiber typing and abundant central cores.

The patient presented with low back pain, and numbness and weakness in her left leg. She preferred regional anesthesia. We planned spinal anesthesia for the operation. Muscle biopsy and contracture test were planned, but the patient refused. Oral dantrolene (4 mg kg^{-1} day^{-1}) was administered prophylactically for three days. For premedication, oral diazepam (5 mg) was given on the evening before surgery, and diazepam (5 mg) and famodine (40 mg) were given orally two hours before the operation.

The anesthesia machine was flushed with oxygen (10 L min^{-1}) for 10 minutes. Before the operation, vaporizers and soda lime were removed. General anesthesia equipment, iced solutions and non-dissolved dantrolene were ready for use. Monitoring included ECG, SpO_2, rectal temperature probe, invasive blood pressure (placed from radial artery with local anesthesia) and central venous pressure.

Dural puncture was performed with a 25 G Whitacre spinal needle at the L3-4 interspace with the patient lying in the lateral position. Hyperbaric bupivacaine 0.5% 7.5 mg and fentanyl 20 µg were injected. She was maintained in the supine position for 30 min and was then turned into the prone position and surgery was commenced. During the pre- and peri-operative periods and the evening of the first post-operative day, the following parameters were recorded: body temperature (BT), heart rate (HR), creatine kinase (CK), blood gases, electrolyte concentrations, and clinical signs of MH (myoglobulinuria, muscle pain, muscle cramps, rigidity, fever, cyanosis, tachycardia, tachypnea). One mg midazolam was injected two times during the operation. No complications occurred during or after surgery. The patient was admitted to the intensive care unit and followed for 24 hours and she was discharged from the hospital five days after the operation.

Discussion

CCD is an autosomal dominant congenital myopathy and accompanies MH. The incidence of CCD is not clearly known. The most often presented symptom of CCD is weakness in legs. Patients have difficulty in jumping or climbing stairs. The findings often begin in the first year of the disorder; however, the weakness is not of a progressive trait (3). Our patient could sit independently when she was 10 months old and was able to walk at 12 months, but she had been experiencing difficulties in sitting and standing and turning from side to side in her bed. She had been presumed to have a congenital hip dysplasia, for which she had been treated for a long period. No information about the anesthesia protocol could be obtained from the patient concerning a reported tonsillectomy, which was conducted without any complications when she was 10 years old.

Due to the high risk of MH existence in patients with CCD, anesthetic management is of particular importance. MH causes muscle rigidity and uncontrollable fever, and most often occurs in cases when halothane + succinylcholine are used. Prophylactic usage of dantrolene is controversial in patients suspected of MH. Flewellen et al. (6) reported that dantrolene had no effect on respiratory functions in healthy adults. Oikkonen et al. (7) suggested that if dantrolene was to be used in patients undergoing spinal or epidural anesthesia, respiratory functions should be monitored closely.

Although dantrolene is used prophylactically via i.v. route, some researchers prefer to administer it orally. Allen (8) had administered the drug to 10 patients suspected of MH orally as 5 mg kg^{-1} day^{-1} at six-hour intervals and applied the last dose four hours before the operation. The operations were concluded without any complication and 6-18 hours post-operatively the dantrolene level in blood was found to be normal. In seven patients, weakness was observed. Bernhardt (9) reported that after three days of 4 mg kg^{-1} day^{-1} dantrolene usage orally in five patients, CK values dropped, but eight days after treatment rose again to the pre-operative values. However, there are instances when it may be used, and oral dantrolene should be kept in mind as an option. Oral dantrolene is much less expensive than the intravenous preparation. It does not carry the risks of thrombophlebitis or tissue necrosis. Intravenous dantrolene comes mixed with mannitol, which can lead to problems in patients with cardiac or renal failure (8). We

used prophylactic oral dantrolene at 4 mg kg^{-1} day^{-1} for three days at six-hour intervals in our patient as well. We observed a mild weakness in the patient. However, no severe clinical status developed that required respiratory support.

Goto et al. (4) administered oral dantrolene to a 26-year-old patient who previously experienced fulminant MH during cesarean operation. Dantrolene was applied for four days as 75 mg day^{-1} and a further 25 mg was administered before the operation. The cesarean was conducted under spinal anesthesia. During the operation, 1-2 mg kg^{-1} i.v. dantrolene was used and no complication occurred. Sariego et al. (10) applied epidural anesthesia for cesarean operation to a patient with muscular dystrophy and encountered no problems. However, they noted that during permanent spinal and epidural anesthesia, the blockage level should be duly controlled. Matsuoka et al. (5) applied epidural catheter in a patient suspected to be MH and administered ropivacaine and reported no complications. Lucy (11), without administering dantrolene prophylaxis, applied a spinal anesthesia with bupivacaine + fentanyl to a patient with a family history of MH. Abballe et al. (12) applied dantrolene prophylaxis to a patient suspected to be MH and conducted a spinal anesthesia with bupivacaine. No complications occurred.

There was no family history in our case. MH did not occur in her previous operation. However, considering the diagnosis of CCD, we considered her at high risk of development of MH and administered prophylactic dantrolene for three days. We conducted the operation with a combination of bupivacaine + fentanyl spinal anesthesia and no complications occurred.

We conclude that prophylactic dantrolene usage provides protection in CCD patients against the high risk of MH and that spinal anesthesia is a proper method of anesthesia in these patients.

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