Spinal Anesthesia of a Patient with Central Core Disease for Caesarean Section: A Case Report

Se Jin Kim[1], Yeon Ji Roh[2], So Yeon Cho[3], Jangho Bae[4], Seongsik Kang[5, *]

[1,2,3,4,5]Department of Anesthesiology and Pain Medicine, Kangwon National University Hospital
Chuncheon, South Korea

[*]*Corresponding author's email: sskang [AT] kangwon.ac.kr*

ABSTRACT ---- ***Central core disease is an autosomal dominant congenital myopathy. It typically manifests as muscle weakness and developmental delay. Central core disease is also associated with malignant hyperthermia which can be developed by volatile agent or succinylcholine. Here, we are reporting a case of a 34-year-old primigravida with central core disease who underwent an emergency cesarean section under spinal anesthesia without complications.***

Keywords ---- Central core disease, Congenital myopathy, General anesthesia, Malignant hyperthermia, Spinal anesthesia

1. INTRODUCTION

Central core disease (CCD), first described in 1956 by Shy and Magee, is a dominantly inherited congenital myopathy [1]. It usually appears in childhood in the form of muscle weakness (developmental delay) and skeletal deformities (scoliosis, congenital dislocation of the hip, short stature and abnormal gait) [2]. Although CDD is a rare disease, many patients need surgical correction at some time, and anesthesiologists may come into contact with a relatively high number of these patients.

It has long been known that Central core disease is associated with malignant hyperthermia [3]. Therefore, it is important to exactly evaluate the patient before the surgery and make a thorough anesthetic plan.

2. CASE

A 34-year-old primigravida was taken to the emergency room due to ruptured membrane, and she had been diagnosed with central core disease confirmed by muscle biopsy at Gangnam Severance Hospital in 2009. In fact, she was supposed to undergo elective Cesarean section two days after she visited the emergency room. The patient was short of stature (1.50m) but had no history of scoliosis. She showed a mild weakness of limb muscles and abnormal gait. She had been having a hard time jumping and climbing stairs since she was young. ECG and Chest X-ray were normal without any evidence of arrhythmia or cardiomyopathy.

Therefore, a Cesarean section at 38 weeks of gestation was performed under spinal anesthesia. A 18-gauge peripheral intravenous cannula was inserted, and 25-gauge needle was used for spinal anesthesia.

The patient was placed in the lateral decubitus position with the right side up, and 0.5% bupivacaine 9mg with fentanyl 15mcg were injected into the intrathecal space at L4-5 with midline approach. After the injection, the level of anesthesia was checked by using alcohol swabs, and sensory block level was reached to T4.

The cesarean section was performed uneventfully with total blood loss of 400 ml. The induction-to-incision interval was 10 minutes, and the incision-to-delivery time was 8 minutes. The newborn baby did not have breathing problems or require any drugs. His Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. The umbilical cord arterial pH was 7.4 with a base excess of -1. He weighed 3.14kg. Duratocin, which is an oxytocic agent and cause uterine contractions, was given to prevent uterine atony and postpartum hemorrhage. Surgery lasted 60 minutes, and the patient was carried to the post anesthetic care unit without any problem.

The newborn baby had no problem with muscle tone except he had a single umbilical artery. However, he had been taken care of at Neonatal Intensive Care Unit (NICU) to rule out the possibility of hypotonia. During the stay, he showed no sign of hypotonia and no other remarkable findings were observed in brain sonography and abdominal sonography. As there were no specific problems, mother and baby were discharged from the hospital three days after the birth.

3. DISCUSSION

Muscle weakness (developmental delay) and skeletal deformities (scoliosis, congenital dislocation of the hip, short stature and abnormal gait) are key characteristics of Central core disease, and it usually appears in childhood [2]. The patient in this case had a delay in walking when she was young as well as difficulties in running or climbing stairs, which finally led to the diagnosis of CCD in 2009.

Dysregulation of calcium signals due to defects of the skeletal muscle sarcoplasmic reticulum calcium release channel (ryanodine receptor; RyR1) is a causative factor in several congenital muscle disorders including malignant hyperthermia, central core disease [4]. CCD is a typical congenital myopathy, but in this case, only our patient was diagnosed with CCD while her siblings (old sister, young brother) were not.

It has long been known that Central core disease is associated with malignant hyperthermia [3]. Although the patient did not experience any complication after anesthesia, she had a high risk of developing malignant hyperthermia. By performing spinal anesthesia, the avoidance of general anesthesia helped in preventing a common complication.

It is well known that Bupivacaine, one of amide local anesthetic drugs, is a safe agent to use in patients susceptible to malignant hyperthermia [5]. If a regional technique can be carried out successfully, intubation and ventilation are not necessary. It also excluded the possibility of developing malignant hyperthermia in this case. Even though our patient showed no retrognathism or high-arched palate from physical examination, it is significant to keep in mind that intubation should be conducted carefully when dealing with CCD patients who have facial dysmorphism. Since succinylcholine may cause malignant hyperthermia during intubation, rocuronium should be considered as an alternative when preparing an anesthetic plan. As a result, spinal anesthesia with bupivacaine was the best option for the patient, as she did not have a spinal deformity.

Propofol is a safe anesthetic agent in terms of malignant hyperthermia [6]. If a general anesthesia is required, total intravenous anesthesia can be an alternative. Administering propofol with a combination of remifentanil by continuous infusion has been reported to be secure for the induction of anesthesia during cesarean section [7]. After anesthetic machine without vaporizers is flushed with oxygen for two hours, anesthesia can be maintained with propofol and remifentanil by target control infusion [8].

4. CONCLUSION

Even though our patient was aware of her congenital disease, CCD, more caution should be taken when it comes to dealing with undiagnosed patients. Any patient with an ill-defined myopathy or non-specific features of muscle disease can develop malignant hyperthermia. Therefore, meticulous anesthetic premedication through precise history taking and physical examination is important for anesthesiologists before everything else. For thorough anesthetic plan, the possibility of malignant hyperthermia should be considered as well.

5. REFRENCES

1. Shy GM, Magee KR. A new congenital non-progressive myopathy. Brain 1956; 79: 610–2.

2. Ronald D. Miller. Miller`s anesthesia 8th edition Volume 1 1130

3. Denborough MA, Dennet X, Anderson RM. Central-core disease and malignant hyperpyrexia. British Medical Journal 1973; 1: 272–3.

4. Treves S, Jungbluth H, Muntoni F, Zorzato F. Congenital muscle disorders with cores: the ryanodine receptor calcium channel paradigm. Curr Opin Pharmacol 2008; 8: 319–26

5. A K Brownell, R T Paasuke. Use of local anesthetics in malignant hyperthermia. CMAJ. 1986 May 1; 134(9): 993–994.

6. A J McKenzie, K G Couchman, N Pollock. Propofol is a 'safe' anaesthetic agent in malignant hyperthermia susceptible patients. Anaesth Intensive Care. 1992 May;20(2):165-8. doi: 10.1177/0310057X9202000208

7. Liguo HuJianhui, PanShantang, ZhangJinjing, YuKeqiang, HeShuhua, ShuRuiting Wang. Propofol in combination with

remifentanil for cesarean section:Placental transfer and effect on mothers and newborns at differentinduction to delivery intervals. Taiwanese Journal of Obstetrics and Gynecology Volume 56, Issue 4, August 2017, Pages 521-526

8. A.P. Georgiou, J. Gatward. Emergency anaesthesia in central core disease. BJA CORRESPONDENCE VOLUME 100, ISSUE 4, P567, APRIL 01, 2008