Spinal anesthesia during cesarean section in a patient with severe osteogenesis imperfecta

− A case report −

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Obstetric anesthesia in a parturient with severe osteogenesis imperfecta is challenging in many aspects, particularly concerning maternal pathophysiological problems and the technical difficulties of anesthesia. Here, we report a case of successful spinal anesthesia, instead of general or epidural anesthesia, during a cesarean delivery in a patient with severe osteogenesis imperfecta. (Korean J Anesthesiol 2009; 57: 662∼5)

Key Words: Cesarean delivery, Osteogenesis imperfecta, Spinal anesthesia.

Osteogenesis imperfecta is a rare genetic disorder of collagen type I synthesis characterized by bony fragility. There are several types of this disorder, from mild (type I) to severe (type IV). Many anesthetic reports postulated that anesthesia of parturient with mild type was not different from that of normal obstetric patient. But a parturient with severe type of osteogenesis imperfecta was reported one case of successful epidural anesthesia and failed spinal anesthesia.

This report presents management of a parturient with severe osteogenesis imperfecta undergoing cesarean delivery under spinal anesthesia.

CASE REPORT

A 28-year-old parturient with type IV osteogenesis imperfecta at 36 week gestation was admitted for cesarean delivery. In the past medical history, she suffered fracture sixty times with several orthopedic operations under general anesthesia, and received medical treatment for bronchial asthma 2 years ago. Physical examination demonstrated severe scoliosis and deformities of all extremities by multiple fractures. She was 116 cm tall, and her body weight was 41 kg with 10 kg of weight gain during pregnancy. She showed an airway of Mallampati class II [1], 3 fingerbreadths thyromental distance, fragile den-
Laboratory findings of hematology and coagulation profile (bleeding time, prothrombin time, activated partial thromboplastin time) were normal. Chest and L-spine roentgenogram showed severe scoliosis with enlarged heart (Fig. 1, 2), but pulmonary function test was normal.

We scheduled regional anesthesia under general anesthesia preparation. Fiberoptic bronchoscope and laryngeal mask airway were prepared. The patient was admitted to operating room without premedication.

To avoid fracture, the patient was carefully positioned supine on the operating table with sponge and cotton pads under both arms and legs.

After peripheral intravenous lines were assured with two large bore (18 G) needles, the hydration of 500 ml colloid solution (PENTASPAN®, JEIL, Korea) and 500 ml lactated Ringer’s solution was done before anesthesia. Direct radial artery blood pressure, electrocardiogram, pulse oximetry, temperature were monitored. All monitored vital signs were within normal range.

Oxygen was supplied to the patient via face mask at the rate of 5 L/min. The patient was positioned in the left lateral decubitus position. Lower thoracic spinous process was palpated but not those of other areas. We scheduled to try lumbar spinal anesthesia and then lower thoracic epidural anesthesia. After two attempts with 25 G pencil point needle (Pencan®, B/Braun, Germany), CSF was freely flowing. 8 mg of 0.5% hyperbaric bupivacaine (Marcaine®, Astra-Zeneca, Sweden) was intrathecally injected for spinal anesthesia.

To avoid hypotension, continuous ephedrine-phenylephrine infusion (1.2 g of phenylephrine and 96 mg of ephedrine was mixed with normal saline to make 40 ml of total volume) was started at the rate of 20 ml/hr [2].

Anesthetic level evaluated by coldness with alcohol sponge was T10 after 1 minute and T8 after 3 minute. 5 minute after spinal anesthetic injection, blood pressure decreased to 90/45 mmHg, and heart rate 135 beats/min. Phenylephrine 100 μg was intravenously given to treat hypotension and tachycardia. Blood pressure elevated to 120/60 mmHg and heart rate decreased to 35 beats/min. Atrovent 0.5 mg was given to treat bradycardia. The anesthetic level was T4 at that time.

Cesarean delivery was performed via lower midline incision, and 2,230 g male was delivered. Apgar score was 8 at 1 minute, 9 at 5 minute. After delivery, 20 units of pitocin mixed in 1 L of Hartmann’s solution and 3 mg of midazolam were given. Total anesthetic duration was 100 minutes and surgical duration 60 minutes. Patient was transferred to post anesthetic care unit without transfusion or any other medication, and discharged without complication.

**DISCUSSION**

Osteogenesis imperfecta is a rare hereditary connective tissue disorder and primarily involves ossification of the endochondral bone, which causes multiple fractures due to bony fragility. Depending on the inheritance pattern of osteogenesis imper-
fecta, the disease can vary in severity.

Type I is most common, mild form with minimal deformity, Type II (perinatal lethal osteogenesis imperfecta) is incompatible with life, and Types III and IV are severe, more debilitating forms of the disease resulting in short stature and kyphoscoliosis.

Osteogenesis imperfecta also frequently affects sites including tendons, ligaments, skin, sclera, teeth, and middle and inner ear, so results in hyperthermia, hyperhidrosis, blue sclerae, conductive hearing loss, dental abnormalities, platelet dysfunction, cor pulmonale, congenital heart disease, valvular heart disease, joint laxity, and thin skin [3-5].

Osteogenesis imperfecta superimposed on the preexisting physiologic changes of pregnancy presents unique anesthetic challenges. In severe forms of the disease, the bone abnormalities, metabolic disorders, and cardiac and pulmonary insufficiency can significantly increase maternal morbidity. Additionally, many of these patients require cesarean delivery because of a contracted maternal pelvis, cephalopelvic disproportion, or a fetus with osteogenesis imperfecta.

There were several reports regarding anesthetic management of a patient with osteogenesis imperfecta undergoing cesarean section [6-9]. Although epidural or general anesthesia was done in most reports about cesarean delivery in patients with mild to moderate osteogenesis imperfecta, spinal anesthesia was successfully carried out for cesarean section in a patient with severe form of the disease in this case.

A spinal anesthetic technique can be done safely and effectively as illustrated by this case and provide more relaxed muscle tone than epidural anesthesia. By using lumbar spinal anesthesia, we were able to avoid tracheal intubation, its inherent risk of aspiration, and mandibular and teeth injury. Because of increased metabolic rate and hyperthermia associated with osteogenesis imperfecta, general anesthesia was not chosen as well [10,11]. Although the patient had history of successful general anesthesia, the risk of malignant hyperthermia could not be completely excluded. In addition, because of the patient’s anatomical distortion, the operator expected delayed time from induction to delivery, therefore we preferred regional anesthesia over general anesthesia. And the patient also desired to be awake during the surgery to meet her newborn infant. In preparation for failure of spinal anesthesia and high spinal anesthesia, fiberoptic bronchoscope and laryngeal mask airway (LMA) must be ready.

Technical difficulty with block, inability to tolerate the supine position when awake, and preexisting coagulopathy from platelet abnormalities may preclude regional anesthesia in patients with osteogenesis imperfecta. Fortunately, there were no problems in patient’s positioning and no preexisting coagulopathy in this case.

In the spinal anesthesia, because of repeated puncture, kyphoscoliosis coupled with short stature can predispose these patients to inadvertent post-dural puncture headache (PDPH), and may make it difficult to predict the level of any block produced by a given dose of local anesthetic.

To avoid PDPH and the likelihood of respiratory insufficiency developing from an unintended inadequate high block, we used pencil point spinal needle and 8 mg of 0.5% hyperbaric bupivacaine as small dose for cesarean section. Even though minimal dose of bupivacaine was used, it was difficult for us to predict the block level.

For inadequate block whether too high or too low, mask or LMA ventilation were considered. To avoid hypotension, continuous ephedrine-phenylephrine mixed infusion was prepared. This case presented a very severe form of osteogenesis imperfecta in a pregnant woman for whom cesarean delivery was successfully done under spinal anesthesia.

It is important to understand and consider the advantages and complications associated with spinal anesthesia compared to general or epidural anesthesia in this patient population.

We suggest that it is also helpful to use a multidisciplinary approach in planning a technique not only appropriate for the level of expertise of the anesthesiologist and obstetrician but also suited to the severity of physiologic and anatomic abnormalities of the individual patient.

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