Spinal Anesthesia in a 21-Year-Old Woman With Osteogenesis Imperfecta: A Case Report

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Abstract

Introduction: The choice of anesthetic technique in patients with osteogenesis imperfecta (OI) can be a challenge for anesthesiologists and must be adjusted based on specific circumstances and abnormalities. We report our anesthetic technique in an OI patient to emphasize this point, and to frame a discussion of this subject. According to the accessible data, we report one of the rare OI patients who have undergone spinal anesthesia for emergency surgery.

Case Presentation: The patient was a 21-year-old female with OI type IV, who was admitted due to severe anal pain. The surgeon decided to perform an emergency reduction of a prolapsed rectum, with late permanent fixation. Considering the circumstances, we decided to perform spinal anesthesia for the operation, which lasted for 30 minutes. The patient underwent rectal prolapse reduction via rectosigmoidoscopy, of up to 25 cm of rectum. She was transferred to the post-anesthesia care unit after her vital signs were stabilized. After gaining lower extremity strength, she was transferred to the surgery ward. The next day, she was discharged from the hospital.

Conclusions: Although most authors believe that general anesthesia following fiberoptic intubation is the preferred method for OI patients, it is likely that spinal anesthesia is acceptable in such patients. Although it is technically difficult, the procedure can be performed by expert anesthesiologists.

Keywords: Osteogenesis Imperfecta, Anesthesiology, Spinal Anesthesia

1. Introduction

Osteogenesis imperfecta (OI) is a rare autosomal-dominant connective tissue disorder characterized by multiple bone fractures, blue sclera, growth retardation, and cardiac involvement. OI presents multiple challenges for anesthesiologists, including difficult airway management, hyperthermia, coagulopathy, spinal abnormalities, and respiratory dysfunction (1-3). Therefore, choosing an anesthetic technique can be a challenge and must be adjusted based on the specific circumstances and abnormalities of each patient; a careful evaluation and risk assessment can optimize the anesthetic management (4-6). The spine is one of the most common sites of bone involvement and deformity in OI patients. Hypotonia, poor bone solidity and firmness, and frequent vertebral compression fractures usually lead to the development of multiple spinal deformities in these patients; this is more likely to happen in more severe types of OI (7). Such abnormalities convince most anesthesiologist to avoid spinal anesthesia in these patients, particularly in emergency situations. We report our anesthetic technique in a case of OI to emphasize this point and to frame a discussion of this subject. According to the accessible data, our reported case is one of the rare OI patients who has undergone spinal anesthesia for emergency surgery.

2. Case Presentation

The patient was a 21-year-old Caucasian woman with a known diagnosis of OI type IV. Her height was 112 cm and weight was 60 kg. She was admitted to the emergency department due to severe anal pain. On physical examination, there was a protruding anal mass and a diagnosis of rectal prolapse was obvious, so the patient was immediately transferred to the operating room (OR). Her past medical history revealed that she had undergone surgery for lower extremity fractures twice under general anesthesia, but there was no available documentation. At the time of her entry to the OR, she was in a semi-sitting position and was agitated due to severe pain caused by the rectal prolapse. The patient's vital signs were a heart rate of 160 beats/min, blood pressure of 110/60 mmHg, and a 94% O₂ saturation. On the head and neck examination, she was edentulous and had a Mallampati score of IV, a thyromental distance of 4 cm, and a 2-finger mouth opening. Neck
movement and head extension were in normal range, and she had thoracic kyphosis of the vertebral column. She had consumed a full solid meal 3 hours earlier. Because of the emergent situation, we had no pulmonary function tests. Her laboratory data showed a hemoglobin of 11 mg/dl, a platelet count of 198,000/cc, and a normal coagulation profile. The surgeon decided to perform an emergency reduction of the prolapsed rectum, with late permanent fixation. Considering the circumstances, we decided to perform spinal anesthesia. After establishment of electrocardiogram monitoring, pulse oximetry, and manual noninvasive blood pressure monitoring, the patient was hydrated with 600 cc of normal saline. A difficult-airway kit was prepared, and spinal anesthesia was performed with the patient in the sitting position, via the L2-L3 space. Two milliliters of bupivacaine 0.5% were injected into the subarachnoid space with a 25-gauge Quincke needle. As the pain diminished, the patient’s position was changed to supine, at which time her heart rate was 100 beats/min and blood pressure was 90/55 mmHg. Due to the hypotension, 10 mg of ephedrine was injected in two divided doses, and her blood pressure increased to 110/80 mmHg. The anesthesia level was fixed at T8. During the surgery, her hemodynamic state was stable, without any dramatic changes. The operation lasted for 30 minutes with the patient in the lithotomy position, and her rectal prolapse was reduced via recto-sigmoidoscopy of up to 25 cm of rectum, which showed normal mucosa. She was transferred to the postanesthesia care unit when her vital signs were a pulse of 90 beats/min, blood pressure of 100/80 mmHg, and 95% oxygen saturation. After gaining lower extremity strength, she was transferred to the surgery ward. The next day, she was discharged from the hospital, and on her one-week follow-up, she had no complaints.

3. Discussion

There are a few case reports on regional anesthesia in OI, such as that of Munish Garg et al., who reported a caudal epidural technique for the anesthetic management of an OI patient with urinary bladder stones (5). Baranovic et al. reported unilateral spinal anesthesia in an OI patient with a lower leg fracture (8). There was also a case report of an urgent cesarean delivery in a woman with OI using spinal neuraxial block (9). Such reports indicate the importance of this condition, as well as ambiguities in the field. OI presents multiple challenges during anesthesia, including a tendency for bony fractures, difficulty with airway management, a restrictive pulmonary system, bleeding diathesis, and malignant hyperthermia (4, 10). An understanding and a review of these issues, sufficient and efficient preoperative preparation, accurate intraoperative monitoring, and careful postoperative management can minimize such complications.

3.1. Bony Fracture Tendency

This problematic issue is quite important in that it may lead to mandibular, tooth, and cervical spine fractures or dislocations during laryngoscopy and tracheal intubation. Even applying an automated blood pressure cuff can produce fractures. Gentle transferring, proper positioning, and avoidance of over-extension and over-pressure could minimize these injuries (4).

3.2. Airway Management Difficulties

There are several risk factors for difficult airway management in OI patients, such as a miniature mandible, brittle teeth, and an enormous tongue. Fracture or dislocation of the atlantoaxial axis may occur after neck extension or direct laryngoscopy (8, 11). Upward translocation of the cervical spine, called basilar invagination, is one of the rare reported airway difficulties in these patients (12).

3.3.Restrictive Pulmonary System

Spinal deformities, severe kyphoscoliosis, and pectus excavatum could lead to decreased lung capacity and severe respiratory dysfunction (7). Low functional residual capacity may predispose OI patients to early development of hypoxemia following the administration of induction agents. Mucosal drying, barotrauma, aggravated ventilation/perfusion mismatch, and precipitation of laryngeal and bronchial spasms can all occur during invasive positive-pressure ventilation (13, 14). Volatile anesthetic agents, such as halothane, that could decrease the body’s response to hypercapnia and hypoxia, are not recommended. Instead, ketamine not only preserves respiratory drive, but also maintains airway reflexes, and can be considered the preferred intravenous induction agent. Intraoperative administration of morphine and non-steroidal anti-inflammatory drugs is discouraged because of the possibility of inducing histamine release and exacerbating asthma (4, 11).

3.4. Bleeding Diathesis

Increased capillary fragility, decreased platelet retention, decreased Factor VIII formation, decreased platelet aggregation caused by collagen disorders, and decreased secretion of platelet factors can all cause coagulopathy in OI patients. Therefore, evaluation of the coagulation profile is necessary before surgery, even if the platelet count is normal (3, 5). Inability of the small vessels to constrict...
and the presence of clotting abnormalities can cause major intraoperative and postoperative bleeding. Preoperative assessment and treatment with platelets, fresh frozen plasma, and cryoprecipitate can prevent such complications (4). This entity is so problematic that it can cause mucosal bruising and bleeding during direct laryngoscopy.

3.5. Hyperthermia

One of the concerns in OI is the possibility of malignant hyperthermia during general anesthesia, resulting from a hypermetabolic state. Dysfunction of the thermoregulatory system in the central nervous system, or abnormal cellular metabolism and elevated serum thyroxin levels, are among the proposed pathogeneses of hyperthermia in OI patients (15, 16). Fasciculation is known as a predisposing factor for hyperthermia. Non-depolarizing muscle relaxants, except for atracurium, are preferred over succinylcholine, which theoretically has more potency for inducing muscle spasms and fasciculation (10). It was reported that elevated creatine kinase or pyrophosphate are considered predictors of malignant hyperthermia occurring during or after surgery (4).

Considering all of the abovementioned issues, some authors believe that fiberoptic intubation is the preferred method to overcome most complications when general anesthesia is required for OI patients. However, although difficult to perform, regional techniques should be kept in mind because these bypass the problems accompanying general anesthesia (4, 9).

3.6. Conclusion

Although most authors believe that general anesthesia following fiberoptic intubation is the preferred method for OI patients, it is likely that spinal anesthesia would be acceptable in such patients. Although it is technically difficult, the procedure can be performed by expert anesthesiologists.

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Footnote

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References
