



Anesthesia Management in Rare Case: Osteogenesis Imperfecta

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Authors' contributions

This work was carried out in collaboration between all authors. Authors AK, NKÖ, ASK, GÇ and AAO managed the anesthesia period of patient during the operation. Authors AK, NKÖ, ASK, GÇ, AAO, KI and BK designed the case report. Authors AK, NKÖ, ASK, GÇ, AAO, KI and BK wrote the first draft of the manuscript. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Osteogenesis imperfecta is a rare genetic disorder and a collagen tissue disease for which preoperative preparation and intra-operative anesthesia management must be performed with great care on patients.

An operation was planned for a 5-year old female patient with Type I osteogenesis imperfecta due to a right femoral fracture. Her medical history showed that she had been operated due to a left femoral fracture 5 months ago under sevoflurane inhalation anesthesia without any complications. On her physical examination, she was observed to be a short child with growth deficiency, kyphoscoliosis, and bone and shape deformities on her extremities. Her modified Mallampati score was III and neck extension was limited. Preoperative echocardiography, complete blood cell count, coagulation profile, and biochemical values were found in normal limits. She was taken into the

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operation theatre and monitored. Pressure points were supported by silicon pads. Vascular access could not be established at first because of her agitation, and then it was achieved after the patient's anesthesia induction was performed with sevoflurane. Her neck was kept stable and laryngeal mask was placed in the mouth in the first intervention. Anesthesia was provided through 50% O₂ + 50% air and 2% sevoflurane. At the end of the operation that took 90 min, the patient was extubated uneventfully, and taken to the recovery room.

Main anesthetic problems in patients with osteogenesis imperfecta are the difficulties in maintaining the airway management and malignant hyperthermia. We used sevoflurane both at the induction and at the maintenance of anaesthesia due to the difficult vascular access of the patient, and we did not encounter any problems.

Inhalation anesthesia such as sevoflurane as well as TIVA could be used for the anesthesia for the patients with osteogenesis imperfecta. Great care must be given because of difficult airway in such patients, and necessary precautions must be taken. Laryngeal mask airway could be preferred in order to secure the airway and avoid traumatic complications.

Keywords: Osteogenesis imperfecta; malignant hyperthermia; airway management; sevoflurane; laryngeal mask airway.

1. INTRODUCTION

Osteogenesis imperfecta (OI), which is clinically characterized by blue sclera, bone fragility, kyphoscoliosis, fragile skin, hearing loss, bleeding diathesis, dental and cardiac abnormalities, is a rare autosomal inherited disease of the connective tissue [1,2]. It is observed approximately one in 30,000 live births [3] and is caused by mutations of type I collagen genes [4]. There are four main types of OI described in literature. Type I is the most common form and is characterized by blue sclera, hyperextensible joints, dentinogenesis imperfecta and variable bone fragility. Type II manifests as early as in utero or at birth. Type III patients usually die during childhood or adolescence period because of cardiopulmonary complications. Type IV OI is similar to type I, with the exception of blue sclera, audiological and dental abnormalities. More recently, other forms have been identified [2,5].

These patients often need orthopedic surgery due to the bone fractures. Therefore, they frequently require anesthesia. Patients with OI may also increase the risk of malignant hyperthermia, bleeding diathesis due to platelet dysfunction, respiratory dysfunction due to secondary to thoracic skeletal deformity, congenital heart defects such as aortic regurgitation and mitral valve prolapse, difficult airway, injuries during positioning and intubation [6,7]. Therefore, in this group of patients, the anesthetic management should be carefully implemented. In order to prevent the development of intraoperative malignant hyperthermia, many reports have recommended anesthetic management using total intravenous

anesthesia (TIVA) [1,7,8]. However, in a view of anticipated difficult airway situations or difficult vascular access especially in pediatric patients, it states that inhalational agents such as sevoflurane may also be preferred [2].

In this case report, we present the use of sevoflurane for induction and maintenance of anesthesia and laryngeal mask airway (LMA), uneventfully, in a patient with OI who needed surgery because of right femur fracture.

2. CASE

A 5-year-old girl was scheduled to orthopedic surgery due to right femur fracture. In preoperative physical examination, the patient had revealed short length, severe growth retardation, bone deformities on lower and upper extremities, kyphoscoliosis, micrognathia, limitation of head and neck movements and Mallampati III score. She had no hearing loss and congenital heart disease, but had blue sclera.

She had previously left femur fracture surgery 5 months ago under sevoflurane anesthesia without any complications. There was no abnormality in the complete blood count, coagulation profile and biochemical analysis. Preoperative echocardiography was normal.

The patient was taken to the operating room without premedication. She was monitored with 5 lead electrocardiography, noninvasive blood pressure, pulse oximeter (SpO₂), FiO₂ and EtCO₂, esophageal core temperature and rectal temperature. To prevent the development of

intraoperative malignant hyperthermia; dantrolene sodium, sodium bicarbonate and cold IV solutions were available in the operating room. We avoided the use of agents which could trigger malignant hyperthermia such as succinylcholine, halothane, enflurane and anticholinergics. After inhalational induction with sevoflurane and dry air in oxygen, a peripheral intravenous line was placed. After the stomach contents were removed via a nasogastric tube, we cautiously inserted a size 2 laryngeal mask airway (LMA) (ProSeal LMA, laryngeal Mask Company, Herley on Thames, UK) having her head in neutral position to avoid the damage of the lower jaw or teeth. We inserted a thermistor temperature probe into the esophagus via an LMA drain tube and monitored the esophageal temperature. The patient was positioned carefully on the operating table and pressure points was supported by soft peds. Anesthesia was maintained with 2% sevoflurane and 50% dry air in oxygen. Intraoperatively, temperatures were within a range from 36°C to less than 37°C. Surgery was uneventful in about 90 min and we did not observe excessive blood loss. The patient was stable with a heart rate of approximately 130 bpm. At the end of the surgery, tachycardia was observed 150 bpm as maximum, but other parameters (SpO₂, ETCo₂) remained normal. We did not find any increase of temperature and significant hemodynamic changes. Once spontaneous respiration and protective airway reflexes was confirmed, then the LMA was removed. Postoperative analgesia was provided by 15 mg/kg paracetamol, 15 minutes before the end of the surgery. Recovery was uneventful. In postoperative period, there was no abnormality in serum hematological parameters.

3. DISCUSSION

The relationship between development of temperature elevation and osteogenesis imperfecta has been stated in several reports [4,9,10]. In osteogenesis patients, the mechanism of temperature elevation under general anesthesia is associated with malignant hyperthermia (MH) and /or non-malignant hyperthermia. Hypermetabolic state with a temperature elevation due to the increase of the thyroid hormones is accused to cause non-malignant hyperthermia [8] and it is stated that, compared to malignant hyperthermia, non-malignant hyperthermia is self-limiting, muscle rigidity is not observed and normocarbica is protected [2]. Porsbog et al. stated that [11] metabolic acidosis and malignant hyperthermia

had developed following general anaesthesia using barbiturate, fentanyl, pancuronium and nitrous oxide in a patient with osteogenesis imperfecta. It is also recommended that MH triggering drugs such as volatile anesthetics, succinylcholine, anticholinergic agents and ondansetron should be avoided [12]. We also avoided to use of this agents.

In the literature, it is highly discussed that which is the safest anesthesia for OI patients. In order to prevent the development of intraoperative malignant hyperthermia, many authors have recommended anesthetic management using TIVA [13,14]. Furdere et al. [15] investigated the influence of TIVA and enflurane anaesthesia on the intra and postoperative body temperature in OI and they showed that body temperature was significantly lower when TIVA used and they also recommended TIVA with propofol. However, TIVA is still controversial in pediatric patients because of intraoperative awareness [16] and propofol infusion syndrome [17]. Several case reports confirmed the association of serious side effects such as metabolic acidosis with increased serum lactate, rhabdomyolysis, bradyarrhythmia with myocardial failure resistant to treatment with the long term use of propofol in children. Because of this, long term infusion of propofol has been discontinued in most paediatric patients [17]. Killc et al. described [17] severe metabolic acidosis in a 7 year old boy with OI after a 2.5 h propofol infusion. It is stated that the option of sevoflurane anesthesia is reasonable to perform with paediatric patients when a difficult airway or difficult vascular access is anticipated [2]. Santos et al. [18] observed body temperature was not rise and hemodynamic stability was provided under sevoflurane anesthesia in OI patients. We used both induction and maintenance of sevoflurane and we did not observe any temperature elevations during intra and postoperative period.

The mutations of type I collagens result as extreme bone fragility, joint laxity which may lead to bone fractures. Excessively fragile bones could cause perioperative morbidity. Cervical or mandibular fracture may occur with tracheal intubation during the laryngoscopy due to excessive extension of the neck [19]. Over inflation of the cuff during non-invasive blood pressure measurement may also cause bone fractures. Oliverio [20] described a humerus fracture with intraoperative use of non-invasive blood pressure cuff. Fragile and brittle teeth can easily be damaged or may be missing in patients

with dentinogenesis imperfecta during the laryngoscopy [2]. Due to the brittleness of bone, some authors suggested avoiding the use of succinylcholine because of fasciculations may result in bone fractures [21]. However, this concern has never been clinically documented and succinylcholine has been previously used in these patients uneventfully. Bojanic et al. [4] used succinylcholine uneventfully in 19 patients of 34 procedures without any complications. Movement of the patient should be done with extreme care and all pressure points should be supported with use of soft pads while positioning, transporting and placement of the patient on the operation table during the perioperative period [1]. In our patient, we did not identify perioperative fractures or other type of positioning injuries.

Restrictive lung disease due to kyphoscoliosis, thoracic deformities such as pectus carinatum or pectus excavatum and cardiac anomalies such as mitral valve prolapse or aortic regurgitation is usually observed [22]. Preoperative clinical examination, chest radiograph and echocardiography can be helpful in detecting these abnormalities. Excessive perioperative haemorrhage due to thrombocyte dysfunction may also be a problem with these patients and it has been described to be successfully treated with desmopressin [2]. Regional anesthesia may be acceptable in selected patients as it avoids need for tracheal intubation. But before regional anaesthesia, coagulation profile should be screened in terms of increase in bleeding time despite normal platelet count [22]. Successful epidural and spinal blocks have been reported in obstetrical practice [23-25].

Difficult airway management because of megaloccephaly, macroglossia, short neck, mandibular malformation, limited mobility of cervical spine and tooth deformities in OI patients have been reported previously [4,18,19]. In the literature, airway management with the LMA is preferred to prevent complications including mandibular or cervical fracture and dental injury related to tracheal intubation [8]. In addition, airway management with LMA can prevent the possible movement related bone fractures and complications due to soft extubation [1]. Successful intubation using fiberoptic bronchoscope have also been reported [6].

4. CONCLUSION

Because of the bone fractures during the tracheal intubation and movement of the patients, difficult airway and risk of malignant hyperthermia, anesthetic management is performed with great care on patients with OI. Sevoflurane, alternates TIVA, can be preferred especially when a difficult airway or difficult vascular access is anticipated in pediatric patients. The airway management using LMA is suitable for patients with OI.

CONSENT

As per international standard or university standard written patient consent has been collected and preserved by the authors.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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