Case Report

Managing Anesthesia in a Patient of Osteogenesis Imperfecta: Practical Tips and Review of Literature

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ABSTRACT

Osteogenesis Imperfecta (OI) also known as “brittle bone disease” is an autosomal dominant disorder of the connective tissue associated with abnormalities of type 1 collagen leading to skeletal deformities with a characteristic tendency to fracture bones easily, and ocular, otologic, cutaneous and dental abnormalities. We hereby share an experience of anesthetic management of a 30-year-old man with OI with dislocation of cervical spine (C2 - C3) for correction, to emphasize the need for a detailed pre-anesthetic evaluation and preparation.

KEY WORDS: connective tissue disorders, difficult airway, fibreoptic intubation

INTRODUCTION

Airway management in a case of osteogenesis imperfecta (OI) patient coming for surgery is a major challenge for an anesthesiologist[1,2]. This is attributed to a short neck, large tongue, prominent occiput, fragile mandible and cervical spine and predisposition to odonto-axial dislocation, cervical vertebra and teeth fractures and mucosal bruising during laryngoscopy and intubation[3]. Fibreoptic intubation is an ideal technique in such situations, but if unavailable, an intubating laryngeal mask airway (ILMA) has also been recommended as it facilitates intubation with minimal neck movement. Patients with OI often undergo surgery, most frequently orthopedic. There are a number of important issues relating to the anesthetic management of these patients which are summarized in this article.

CASE REPORT

A 30-year-old male patient came with chief complaints of pain in the neck and stiffness since one year associated with tingling and numbness in both upper limbs. He had a history of frequent skeletal fractures for which he had under gone emergency closed reduction twice under general anesthesia, uneventfully.

There was no history of dyspnea on exertion, paroxysmal nocturnal dyspnea, chest pain, palpitation or syncope. There was no history of bleeding gums, GI bleed or easy bruising. On pre-anesthetic evaluation, he weighed 42 kilograms with height of five feet (60 inches). He had a characteristic blue sclera. The skeletal deformities included bilateral lower limb deformity in the form of genu valgus (knock knee), femur and tibia bow-shaped, bilateral dislocation of hip since childhood, with restricted hip abduction and extension and a waddling gait. He had no obvious kyphoscoliosis or any other vertebral abnormality, but there was increased lumbar lordosis.

Respiratory system revealed prominent ribs with restricted chest movements. Airway assessment revealed a short neck with restricted movements and pain. His dentition was firm and airway was Mallampati class- II. Spine was easily palpable and there was no associated spina-bifida. A chest X-ray showed crowding of rib with slanting and bilateral patchy opacities. A cardiology consult was done for the cardiac dysrrhythmia and a 2D-Echocardiography was done to rule out any valvular heart disease.

A MRI reported large posterior disc herniation, compression at C2 - C3 level, segmental hypertrophy, calcification and ossification over C2 - C3 level. His

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pulmonary function tests showed severe obstructive lung defect, with mild response to bronchodilator. Pre-operatively, the patient was started on incentive chest physiotherapy with breathing exercises. Pre-operative arterial blood gases revealed carbon dioxide retention with normoxia. After arranging adequate blood and blood products, he was accepted for surgery as ASA class II.

In view of the anticipated difficult airway and the concern to attenuate cardio-vascular response and minimize neck manipulation, it was planned to secure the airway with endotracheal intubation following IV induction. Monitoring included 5-lead ECG, NIBP, heart rate, pulse oximetry, skin temperature and end tidal CO₂ (EtCO₂). Anesthesia was initiated with premedication in the form of Injection (Inj) glycopyrrolate 0.2 mg, Inj. ondansetron 4 mg and Inj. hydrocortisone 100 mg. After pre-oxygenation with 100% oxygen, induction was done using Inj. Propofol (2 mg/kg). After checking for ventilation, Inj. rocuronium 0.6 mg/kg IV was given. Laryngoscopy was done after 60 sec with a Macintosh number 3 blade but the vocal cords could not be visualized. Only tip of the epiglottis could be seen and the view was graded as Cormack-Lehane grade 3. Intubation was attempted blindly with a No. 36 cuffed armoured flexometallic tube but was not successful. Another attempt was done with a No. 8 Portex cuffed endotracheal tube with a stilette in situ but was unsuccessful.

Another attempt was made using a gum elastic bougie (GEB) but failed. A senior anesthesiologist was called for help and in the meantime patient was ventilated with 100% oxygen. A fiberoptic bronchoscope is an ideal intubating aid in such a situation, but it was not available in our institute. A LMA could not be passed due to restriction of mouth opening. It was then decided to do an emergency tracheostomy. The tracheostomy tube was checked for correct position and then the cuff was inflated.

Intraoperatively, the patient was maintained on 50:50 oxygen in N₂O with isoflurane. Muscle relaxation was maintained with Inj. rocuronium at 0.3 mg/kg dose. The patient remained hemodynamically stable throughout the procedure. The total volume of intravenous fluid administered was 2500 ml and surgery lasted for 300 min with a blood loss of 250 ml and urine output of 800 ml.

Post-operatively, the patient was shifted to the intensive care unit (ICU) for ventilatory support and weaning. Six hours later, he had a sudden episode of bradycardia and looked extremely distressed. This was soon followed by a cardio-respiratory arrest. Resuscitation was initiated immediately but the patient could not be revived despite continued efforts for over an hour.

**DISCUSSION**

Airway management in an OI patient coming for surgery is a major challenge for an anesthesiologist. This is attributed to a short neck, large tongue, prominent occiput, fragile mandible and cervical spine. Associated basilar invagination, with an upward translocation of the cervical spine distorts the airway anatomy further. These patients frequently have associated congenital neurological and cardiovascular abnormality, the most common being valvular heart diseases in the form of mitral valve prolapse and aortic dissection[4, 5].

There is a greater predisposition to pulmonary disease in patients of OI because of kyphoscoliosis and thoracic cage deformity, and recurrent aspirations requires aggressive preoperative optimization of lung function. There may also be hypoxemia secondary to ventilation-perfusion mismatch. Delay in extubation is anticipated in these patients for the same reason[6].

They are also clinically distinct by the presence of hypermetabolism[7]. Therefore, during anesthesia they may tend to develop malignant hyperthermia, although a direct relationship of OI to malignant hyperthermia is not substantiated[8]. Halothane and succinylcholine should be avoided in these patients; availability of rapid cooling methods is important.

Up to 30% incidence of bleeding diathesis in patients of OI has been reported[9]. Platelet dysfunction is common leading to bleeding disorder and easy bruisability. Increased capillary fragility, decreased levels of factor VIII and deficient collagen induced platelet aggregation has been implicated as causes for bleeding diathesis. An increase in intraoperative bleeding may occur despite normal bleeding times and coagulation values, accounting for the adequate arrangement of blood made in our case.

Patients with OI often undergo surgery, most frequently orthopedic. There are a number of important issues relating to the surgical and anesthetic management of these patients which can be summarized as follows:

1. The ease of fracture of bone and teeth;
2. Increased tendency to bleed secondary to platelet dysfunction and possible vascular disorders;
3. Increased tendency to develop malignant hyperthermia;
4. Difficulty in intubation as many patients may have short neck, large tongue and thoracic deformity; and
5. Gas exchange defects. Repeated respiratory infections are complications of OI
Regional anesthesia is difficult in patient with OI, due to skeletal abnormalities. Karabiyik et al have recommended total Intravenous anesthesia along with intubating LMA to manage elective cases[1]. Anticipating these problems helped us achieve a relatively uneventful intraoperative course in our patient. However, we were unprepared for the sudden and dramatic terminal events that we attribute to either an acute, extensive MI or a massive pulmonary embolism, neither of which could be confirmed since all our efforts were directed towards resuscitating the patient first. A third possibility of acute aortic dissection can also be considered. Later, a review of literature showed two reports of patients with OI without a known cardiac disease presenting with acute aortic dissection[10,11]. In the absence of confirmatory tests or autopsy, our diagnosis remains speculative but we now feel that a preoperative echocardiogram and venous Doppler of the lower limbs (in a bedridden patient like ours) could have helped.

CONCLUSION

In conclusion, we would like to emphasize the need for a detailed pre-anesthetic evaluation and preparation for anesthesia in a patient of OI. Special attention should be paid to exclude associated cardiovascular abnormalities, bleeding disorder, difficult airway or any other co-morbidity. An extra gentle care is essential in handling these patients to prevent the complications which can occur in the perioperative period like, fracture of bones and teeth, odontoaxial dislocation, occurrences of hyperthermia, and excessive bleeding. Proper positioning and adequate padding of all pressure points during surgery and transfer is required.

REFERENCES