Case Report

Successful management of a rare case of osteogenesis imperfecta using TIVA

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Abstract

Osteogenesis Imperfecta, a rare inherited disorder of connective tissue, is characterised by brittle bones and susceptibility to fractures. Anaesthetic implications range from simply positioning the patient on the operating room table to management of rare occurrences such as malignant hyperthermia and basilar invagination. Commonly encountered complications include a difficult airway, intraoperative bleeding due to platelet dysfunction, respiratory compromise due to skeletal deformity and congenital cardiac anomalies. We report a case of 8 year old child of osteogenesis imperfecta posted for femur nailing; managed successfully using Total Intravenous Anaesthesia (TIVA). Proper preparation and preoperative assessment is important, as well as the choice of anesthetic technique. The perioperative management of this patient is discussed and the literature reviewed.

Keywords: Osteogenesis Imperfecta, TIVA, malignant hyperthermia.

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INTRODUCTION

Osteogenesis imperfecta (OI), also known as brittle bone disease, is a hereditary disease of connective tissue with mutations in type 1 collagen COL1A1 or COL1A2. Inheritance follows an autosomal dominant pattern, sporadic mosaics and recessive forms are also described. The prevalence is 1:60,000 to 1:20,000 life births[1]. The clinical spectrum represents a continum ranging from perinatal lethality (type II) to nearly asymptomatic individuals (Type I) with occasional fractures and normal stature. OI type III is the most severe form in children surviving the neonatal period and leads to extreme short stature. Patients with mild to moderate bone deformities and variable short stature are classified as OI type IV. The pathological fractures may result with little or no trauma. Early hearing loss, blue sclera, bone deformities, scoliosis, hypermobile joints, fragile skin, reduced muscle tone, mitral valve prolapse, restrictive lung disease and platelet dysfunction are the other problems.[2] Anaesthetic challenges encountered in such cases of OI are difficult intubation[2], difficult central neuraxial blockade due to scoliosis or other bony deformities, restriction in using lot of anaesthetic drugs due to tendency to develop malignant hyperthermia[3] and problems with positioning of patient due to brittle bones.

CASE REPORT

An eight year old girl, known case of Osteogenesis Imperfecta type 1, came to our hospital for pathological fracture femur on the right side. She had past history of recurrent fractures post minute trauma which were managed conservatively. She had blue sclera (Figure 1), brittle teeth (Figure 2), bowed lower limbs (Figure 3), gum bleed and abrasive teeth marks on the lips

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Figure 1:


Figure 2:


Figure 3:

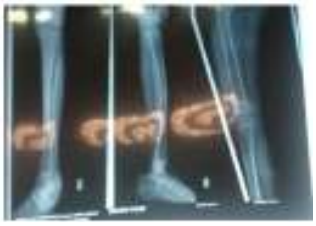
Figure 4:

Routine haematological, biochemical and coagulation tests were normal except for serum alkaline phosphatase and serum phosphorous which were raised (244 U/L and 6.6mg/dl respectively). Her echocardiography was normal. Chest X-ray was done to rule out any rib fractures. Lower limb X-ray showed severe osteopenia, bowing of tibia (Figure 4), fracture right femur and green stick left fibula fracture. High-risk consent and consent for post-operative ventilation was obtained. The inhalational agent vapourisers were removed from the workstation and the circuit was flushed with oxygen to remove any traces of the same. The patient was carefully positioned supine and adequate padding was done. Difficult intubation trolley was kept ready. The child was premedicated with intravenous (iv) midazolam 0.03 mg/kg. Anaesthesia induction was done with iv propofol 2 mg/kg and iv atracurium 0.5 mg/kg. Direct laryngoscopy was performed using size 3 McINTOSH blade, and a 5.5-mm cuffed endotracheal tube (ETT) was inserted. After confirmation of tube placement, anaesthesia was maintained with oxygen, nitrous oxide, propofol infusion (6-10mg/kg/hr) and intermittent atracurium. Inj paracetamol 15mg/kg was administered intravenously for analgesia. Intraoperative monitoring included a pulse oximeter, ECG, manual sphygmomanometer, temperature monitor and capnometer in view of the risk of malignant hyperthermia. The surgery lasted for 120 min and was uneventful. Post-surgery, neuromuscular blockade was reversed and the patient extubated. The patient was shifted to the ICU for observation. Postoperative period was uneventful and patient was subsequently discharged on the seventh postoperative day.

DISCUSSION

Type I OI as in our case is characterized by bony fractures during childhood that can result from mild trauma. It is the mildest and most common form of the disease. Affected individuals have a nearly normal stature, little bone deformity, and characteristically blue sclera. Frequent fractures begin to occur with ambulation during childhood and osteoporosis is present throughout life.[4] There have been several successful case reports of conductance of surgery under general anaesthesia as well as under regional anaesthesia in patients of osteogenesis imperfecta.[2,3,5-7,9] Balanced general anaesthesia with meticulous attention to use of neuromuscular blocking agents, inhalational agents, airway management, positioning of the patient and acute pain management can be used in patients of OI when general anaesthesia is considered for proposed surgical procedure or if there is relative contraindication to regional anaesthesia[5]. General anesthetic management should be carefully implemented in consideration of the potential risks of malignant hyperthermia (MH). Many reports have recommended anesthetic management using TIVA rather than inhalation anesthetics, which contribute to temperature elevation.[3,6] Karabiyik *et al*[3] have recommended TIVA along with Intubating Laryngeal Mask Airway (ILMA) to manage elective case, while Malde *et al*[7] have successfully used balanced general anaesthesia in a case of osteogenesis imperfecta with gross deformity of pelvis for abdominal hysterectomy. Patient should be moved carefully from the stretcher to the operating room table as it can result in a fracture. Proper padding of the pressure points, avoiding overextension during positioning and use of a molding mattress to avoid joint dislocation is important[4].Temperature monitoring and end tidal CO2 is mandatory due to susceptibility to hyperthermia. Pressure applied during insertion of an intravenous catheter and pressure from an automated blood pressure cuff or tourniquet can cause fractures[4], hence, we used manual sphygmomanometer. Perioperative hypermetabolic state with fever is common in patients with osteogenesis imperfecta. It may be the result of either abnormal central nervous center temperature regulation or abnormal cellular energy metabolism. The later can be due to increase in basal creatine kinase, elevated pyrophosphate concentration or an elevated serum thyroxine level[8].We avoided inhalational agents and depolarizing muscle relaxant like succinycholine as they may trigger malignant hyperthermia in these patients. Contraction induced fractures after adminstration of suxamethonium have been reported. Also lethal hyperkalemic responses may occur in immobilized patients. Similarly anticholinergics may produce hyperthermia and should be avoided if possible[3]. Overextension of the cervical spine can lead to odonto-axial dislocation or fracture and must be avoided[9]. A short neck, a protruding mandible, large

tongue and the presence of a pigeon chest can make visualization of the glottis difficult and should be anticipated. Jaw fracture and easily chipped or dislodged teeth is common[4].Intubation with a pediatric fibreoptic bronchoscope is ideal [4] in such cases, but due to its unavailability in our institute and no visible anatomical difficulty; patient was intubated after gentle direct laryngoscopy. Laryngeal mask airway (LMA) may not be able to protect the airway against reflux that is commonly seen in these patient. Though our patient had no coagulopathies; bleeding and clotting abnormalities are well documented in these patients. Platelet counts and standard coagulation tests should be done along with platelet function test and factor VIII activity if there is any past history of hemorrhagic diasthesis[10]. Associated kyphoscoliosis along with pectus carinatum may decrease vital capacity, chest wall compliance with resulting arterial hypoxemia due to ventilation perfusion mismatch and this can lead to increased risk under GA. Preoperative spirometric tests and blood gas analyses may help. High FiO2 or PEEP may be required intraoperatively. However, our patient had no thoracic deformity, so there was no increased pulmonary risk under general anaesthesia. Preoperatively echocardiography was done to rule out congenital heart defect or malformation of the thoracic vessels as incidence of aortic dissection, left ventricular rupture and valve incompetence is high in these patients. In severely affected patients there is a risk of a basilary invagination and an atlanto- occipital dislocation: Therefore a x-ray of the cervical spine might be useful especially when the operation requires a complex positioning of the patient. Abnormal anatomy of extremities may pose difficulty for regional anaesthesia. Central neuraxial blockade can be given safely if the coagulation profile is normal. Epidural is safe but dosage needs to be reduced due to growth retardation. Bharadwaj *et al* discussed successful outcome of spinal anaesthesia in a patient of OI posted for femur fixation[11]. Although regional anaesthesia avoids need for tracheal intubation; it may be difficult in cases of associated kyphoscoliosis. In our patient, we avoided central neuraxial block to prevent risk of further fractures associated with positioning during regional anesthesia. Although general anaesthesia is not ideal in all cases of OI; thorough preoperative evaluation and prompt and meticulous perioperative management leads to successful outcome after general anaesthesia using TIVA.

CONCLUSION

A Osteogenesis Imperfecta patient poses many challenges to the anaesthetist. A proper pre operative assessment, optimization of cardiopulmonary status and proper materials for aggressive management of any complications will prevent poor performance and maximize the patient's opportunity for a favorable outcome.

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