

Anaesthetic Management of a Case of Osteogenesis Imperfecta with Fracture of Femur

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Abstract

Osteogenesis imperfecta (OI) is a rare, genetically inherited syndrome involving connective tissues which leads to fragile bones, short stature, disorders of other organs and body systems. Children with OI are more susceptible for bone fractures. OI poses various anaesthetic challenges like difficult intubation, problems with positioning, tendency to develop malignant hyperthermia, coagulopathy, cardiovascular and pulmonary abnormalities and spinal abnormalities. We report a case of 9 year old male boy with OI type IV, presented with fracture shaft of femur, posted for intra medullary nailing. It was successfully managed under spinal anaesthesia.

Keywords: Osteogenesis Imperfect; Difficult Intubation; Fracture Femur; Spinal Anaesthesia.

Introduction

In a day to day practice Anaesthesiologists frequently come across many congenital diseases and syndromes. Osteogenesis Imperfecta (OI) is one such syndrome. Osteogenesis Imperfecta, also known as Brittle bone disease, Lobstein's disease, Fragilisossium or Vroliks disease is a fibro osseous disorder of the collagen tissue [1,2].

Classically Osteogenesis Imperfecta was divided into two types- the Congenita form which is lethal and the Tarda form which has normal life expectancy. According to SILLENCE classification, the disease has been classified into 4 types. Type I, II, III and IV [3].

The incidence of OI is 6-7 per 100000 people leading to defects in skeletal growth [3]. The disorder poses a challenge to Anaesthesiologists because of multiple organ involvement. Most notably difficult airway, short neck, dental abnormalities, fracture of fragile

bones during positioning, risks of odontoaxial dislocation, tooth fracture, cervical vertebrae fracture, mandibular dislocation during laryngoscopy and intubation can be there. Cardiovascular involvement like Aortic insufficiency, Aortic root dilatation or mitral valve prolapse and pulmonary involvement like restrictive lung disease, severe thoracic kyphoscoliosis, V/Q mismatch can increase the perioperative risks. Lumbar kyphoscoliosis and coagulation disorder may complicate the neuroaxial blockade techniques [4,5].

We report a case of an Osteogenesis Imperfecta type IV with fracture shaft of femur who underwent internal femur fixation under spinal anaesthesia.

Case Report

A 9 year old male child weighing 18 KG with height of 110 cm and a history of fracture of left tibia and

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fibula 2 years back which malunited with conservative treatment and fracture of shaft of right femur 20 days back was posted for internal fixation of right femur fracture. The child was a known case of OI tarda type IV.

On general examination child had short stature, hypermobile joints, mild scoliosis, disproportionate upper limbs and medio-laterally flattened tibia bones with 2 year old fracture of left tibia which united on conservative treatment making an acute angle with two fragments.

On pre anaesthetic evaluation child's heart rate was 90 beats per minute which was regular in rhythm, blood pressure was 110/70 mm of Hg. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy, oedema. Cardiac system showed normal heart sounds. Respiratory system revealed barrel shaped chest with decreased air entry on right side and bilateral rhonchi. Chest X ray revealed thoraco lumbar kyphoscoliosis. ECG and blood investigations viz Complete haemogram, renal function tests and coagulation profile were within normal limits.

On airway assessment child had malampatti scoring of III with acceptable flexion and extension of the neck with adequate mouth opening and normal dentition. Child was advised for Levosalbutamol + Ipratropium bromide nebulisation 4th hourly 12 hours before the surgery. Child was accepted as ASA grade III for the surgery.

In view of risk of odonto axial dislocation, pulmonary complications and normal coagulation profile, we decided to proceed with spinal anaesthesia technique for this case. Patient attenders were informed about the anaesthesia procedure, possible risks, complications and a written consent was taken.

Child was shifted and positioned on the OT table carefully to prevent the new fractures from occurring. Difficult airway cart including stylet, different sizes of face masks and ETT, all sizes of laryngoscopic blades and LMA, tracheostomy set were kept ready. An 18G and 20G cannulas were inserted to peripheral veins on either fore forearms. Monitors attached, baseline heart rate 98/min, SPO₂ -92%, respiratory rate 18/min were noted down. Blood pressure measured manually 110/60 and ECG leads connected. Child was preloaded with 250ml of ringers lactate. Sub arachnoid block was given in the sitting position in L₃₋₄ space with 25G Quinke needle. 1.5ml of 0.5% Bupivacaine was mixed with 0.25ml of Fentanyl(12.5mg) to achieve the desired level. The level of block was up to T-10. Patient was positioned cautiously on OT table with all pressure points

padded and monitored throughout the surgery. Duration of the surgery was 1hour 30 minutes. Child was haemodynamically stable throughout the procedure except for one episode of hypotension which was managed with 5mg of ephedrine i.v. recovery was uneventful.



Fig. 1 & 2: External appearance of child with marked bony deformities



Fig. 3 & 4: X-ray appearance child with Osteogenesis imperfecta



Fig. 5: X- ray of femur after nailing

Discussion

Osteogenesis Imperfecta is a rare inherited disease of the connective tissue. It affects 6-7 of 100000 people and usually present with blue sclera, recurrent fracture of bones, hypermobile joints, kyphoscoliosis, short stature, barrel shaped chest and coagulation disorders [3,5].

Anaesthetic management is influenced by co existing orthopaedic deformities, vulnerability of fractures during simple positioning, platelet dysfunction, coagulation abnormalities, difficult intubation, cardiovascular abnormalities and associated pulmonary pathology [6,7].

Shifting and positioning of patients with OI is the initial and most important step. Utmost care should be taken while shifting the patient to OT table as the fragile bones are susceptible for fractures. Malde et al (1993) have reported fracture of shaft of femur in a patient with OI which occurred during shifting the patient to recovery room [8]. Geetha Bhandari et al (2008) reported shoulder dislocation in a patient with OI during positioning the patient on OT table [9]. We took a meticulous care while shifting the patient to OT table to prevent any fractures or dislocations.

Since OI patients manifests with abnormal skeletal growth, defective teeth and high risk of odonto axial dislocation, cervical vertebra fractures a difficult intubation should always be anticipated. Associated kyphoscoliosis may decrease the vital capacity and chest wall compliance with resulting arterial hypoxaemia due to V/Q mismatch. Bergstrom in 1977 and Rampton et al in 1984 have reported several cases of Malignant hyperthermia in OI patients [10,11]. So the use of halothane and succinylcholine should be avoided. Succinylcholine can also cause fasciculation induced fractures. All these factors can complicate the outcome when patient is selected for general anaesthesia.

There have been several successful case reports of surgery being conducted under general anaesthesia. Karabiyik et al (2002) recommended TIVA along with intubating LMA, while Malde et al (1993) used balanced anaesthesia in a case of OI with gross deformity of pelvis for abdominal hysterectomy^{4,8}. Sachin et al (2004) observed a significant degree of movement between 1st and 2nd cervical vertebra during intubation [12].

Automated blood pressure cuff may be hazardous as the over inflation of the cuff may cause fracture of the arm [9]. We measured BP using manual sphygmomanometer throughout the surgery.

Increased bleeding may occur despite the normal coagulation profile. It may be because of capillary fragility, decreased levels of factor VIII and deficient platelet aggregation [13]. In our case though the coagulation profile was normal and blood loss was not significant, we had reserved one packed cells and one FFP before surgery to manage the unpredictable blood loss.

Wherever possible, regional anaesthesia probably looks superior to general anaesthesia provided coagulation profile is within normal limit as it avoids the risks associated with endotracheal intubation and general anaesthesia.

We opted spinal anaesthetic procedure for our patient since he had to undergo lower limb surgery and lumbar spine had only mild degree of scoliosis and normal coagulation profile.

Conclusion

Patients with OI pose a significant challenge to the anaesthesiologist because of difficult airway, complications associated with general anaesthesia and difficult spinal anaesthesia techniques. A detailed pre-operative work up by taking history, examination and assessment of airway and spine is important to improve the outcome in these patients. Regional anaesthesia appears to be safe than general anaesthesia and should be preferred wherever possible and permissible.

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Conflict of Interest: Nil

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