

Anaesthetic Management in a Patient with osteogenesis imperfecta

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Abstract

Osteogenesis Imperfecta (OI) (Brittle Bone disease) is a rare, autosomal inherited disorder due to a defect in collagen I synthesis leading to skeletal deformities with a characteristic tendency to fracture bones easily, and ocular, otologic, cutaneous and dental abnormalities. Anaesthetic management of such patient is a real challenge to an Anaesthesiologist due to multiple skeleton deformities, risk of odonto axial dislocation during laryngoscopy and intubation and difficulty in achievement of regional anaesthesia due to spastic posture. Again there is an unusual response to muscle relaxants and chances of malignant hyperthermia intraoperative. A case of 11 year old patient with osteogenesis imperfecta with cerebral palsy and mental retardation was posted for bone shortening and K-wire fixation managed successfully under epidural anaesthesia after proper pre operative assessment and workup

Keywords: Osteogenesis Imperfecta, Laryngoscopy, Succinylcholine, Tracheal Intubation.

1. Introduction

Osteogenesis imperfecta (OI) (Brittle Bone disease) is a rare, autosomal inherited disorder due to a defect in collagen I synthesis leading to skeletal deformities with a characteristic tendency to fracture bones easily, and ocular, otologic, cutaneous and dental abnormalities [1]. It may cause various anesthetic complications due to the difficulty in airway management, existence of spinal deformity, respiratory disorders, cardiac anomalies, thrombocyte function disorder, risk of hyperthermia, bacillary invagination, bone deformities and metabolic disorders [2]. OI cases generally require orthopedic surgery due to bone fracture. Even though a direct relationship between OI and malignant hyperthermia has not been proven, malignant hyperthermia symptom and findings have been observed in the OI case under general anesthesia [3]. Due to this we use regional anaesthesia (Epidural anaesthesia) for lower limb surgery.

Anesthetic considerations in this case were recurrent fractures and hyper-mobile joints, platelet dysfunction, skeletal deformities like kypho-scoliosis, shortening, pigeon chest. Patient associated with cardiovascular, neurological anomalies, restrictive lung disease due to skeletal deformities and chances of malignant hyperthermia. We report here anaesthetic management of patient with osteogenesis imperfecta with cerebral palsy and mental retardation managed successfully under regional anaesthesia.

2. Case Report

After obtaining parent's written informed high risk consent, surgery was planned for the case of an 11 year-old male child weighing 11kg with osteogenesis Imperfecta with cerebral palsy with mental retardation. Compound fractures in bilateral femur with exposed necrosed part of the bone observed in the physical examination (Figure 1). In this case lower limb surgeries i.e. bone shortening and K-wire fixation was planned.



Figure 1: Compound fractures in bilateral femur

A detailed preanesthetic evaluation was done. Associated congenital cardiac anomaly was ruled out. ECG and 2D-ECHO reveals no cardiac anomaly. X-ray of right and left upper limb of AP view shows multiple fractures and mal-united old fractures. And X-ray chest of PA view showed scoliotic spine due to this patient was unable to lie flat on bed.

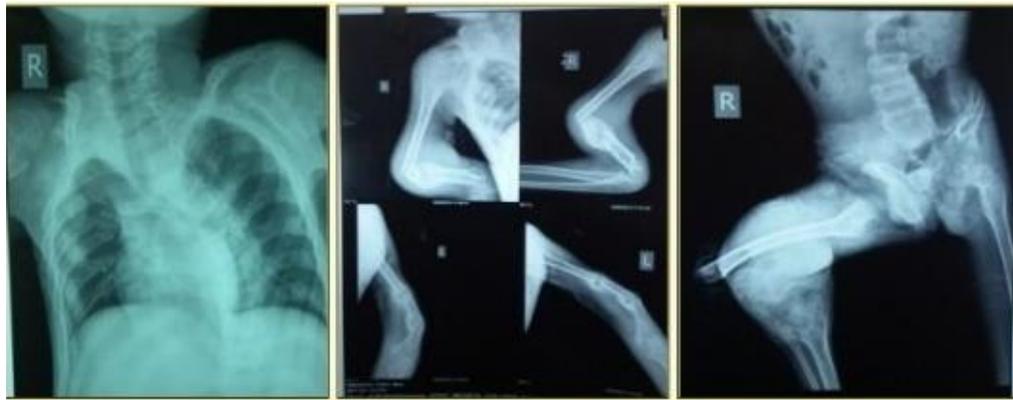


Figure 2: X-Ray chest PA view revealing scoliotic spine and x ray upper and lower limb revealing multiple bone fractures

On general examination patient was drowsy not but responding to deep painful stimulus. Airway cannot be assessed as patient was incooperative. Pulse rate was regular (140/min), low volume and equal on both sides. Patient had Pectus carinatum (pigeon chest), air entry was equal on both sides with bilateral coarse crepitations. Oxygen saturation on room air was 96 %.



Figure 3: Child with pectus carinatum (pigeon chest)

Routine blood investigations were within normal limits, apart from 9.3 gm% hemoglobin. Preoperative work-up for coagulation profile was done to rule out bleeding disorders. Adequate blood and blood products were arranged to overcome haemorrhage during surgery considering the present condition, we decided to go for procedure under epidural anaesthesia.

In the operation theatre, patient was properly placed with cotton gauze padding of all bony pressure points and intravenous line was secured with 22 G cannula. After that multipara monitoring device SpO₂, ECG, NIBP, temperature probe were applied to the patient and baseline parameters were recorded. Patients were induced with Oxygen + Sevoflurane (2-4%) using paediatric breathing circuit. Patient was put on left lateral position and epidural was performed at L3-L4 level under all aseptic precaution with a19G Touhy's needle. Epidural space was identified at a depth of 1.5 cm from skin and confirmed by LOR method. However epidural catheter was inserted 5 cm from skin and epidural anaesthesia was given with 10 ml of 0.25% Bupivacaine along with 10ug of inj. fentanyl which achieved T₁₀ level of block. After induction surgical procedure i.e. bone shortening and bilateral K-wire fixation of compound fractures of femur was done. IJBR (2016) 7 (04)

Intraoperatively vital parameters were monitored and it remained stable. No rescue analgesic medication was required. Total duration of surgery was 40 minutes.

Postoperative recovery was uneventful and patient was discharged 6th postoperative day.

3. Discussion

Osteogenesis imperfecta is a connective tissue disorder that affects a lot of individuals. There are different classifications that range in their severity. OI seen in two forms 1) Osteogenesis Imperfecta Congenita-usually death occur in utero and 2) Osteogenesis Imperfecta Tarda-manifest in childhood or adolescence. Patients with this disorder suffer from brittle bones that may lead to frequent recurrent fractures. Special care must be used in caring for these individuals to avoid injuries. Certain complications arise when performing medical and anesthetic procedures on these patients depending on the severity of their condition [4].

The general anesthesia was used in several reports of patients with OI who experienced a perioperative hypermetabolic state with fever. This hyperthermia seems not to be of the malignant type. Either increased cellular energy metabolism or central nervous temperature dysregulation are discussed as possible causes. Although one case series describes normal caffeine halothane contracture test (CHCT) results in patients with OI and reported malignant hyperthermia (MH). However there is one convincing report of a patient with OI and MH who underwent general anesthesia. In summary evidence of an association between OI and MH is weak. Due to this single case and small case series report of successful anesthetic treatment of patients with OI with neuroaxial anesthetic procedures (spinal anesthesia, epidural anesthesia and caudal nerve block). One has to keep in mind that the full implications of coagulopathy have not been delineated, and due to growth retardation the epidural dosage should be reduced and adapted accordingly [5].

During general anaesthesia there is always risk of odonto axial dislocation while performing laryngoscopy and intubation.

The present case showed Osteogenesis Imperfecta type I with cerebral palsy, mental retardation, multiple fractures with bilateral femur. Patient had bilateral crepitations suggestive of underlying lung infection. Use of succinylcholine for tracheal intubation may result in malignant hyperthermia and occurrence of bone fractures due to fasciculations. So to avoid complication of intubation, muscle relaxants, and respiratory complication general anaesthesia was avoided. Preoperative assessment and plan of anaesthesia made our job easy to provide safe anaesthesia. It was a lower limb surgery and hence planned under regional anaesthesia (Epidural anaesthesia) which resulted in the uneventful and successful outcome of the patient.

4. Conclusion

In conclusion, we think that preoperative evaluation, appropriate preparation and plan of anaesthesia were a prerequisite in the anesthetic management of OI patients with severe anesthetic problems. Also in osteogenesis imperfect, special attention is required to rule out associated anomalies, bleeding disorders and anticipation of difficult airway. An extra gentle care is must during positioning and transfer of these patients.

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