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Osteogenesis imperfecta in a dwarf male with SOL: A head on challenge taken up by anaesthesiologists

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Abstract

Osteogenesis imperfecta (OI) is a rare genetically inherited disorder of connective tissue, resulting in anatomic and physiological abnormalities which pose unusual challenges for the anaesthesiologists during any form of anaesthesia. We present the anaesthetic management of a 48-year-old dwarf male with OI, who underwent craniotomy and excision of a skull-based meningioma SOL with midline shift at our institute. After a successful surgery under general anaesthesia, patient was shifted to ICU on mechanical ventilator for post-op care.

Keywords: Osteogenesis Imperfecta, craniotomy, meningioma, dwarf patient

Introduction

Osteogenesis imperfecta (also known as brittle bone disease) is a rare genetically inherited disorder of connective tissue caused by defect or deficiency in collagen formation and structure. OI is caused by mutations of collagen type 1- COLIA 1 and COLIA 2 genes.

Silence classification of OI is based on genetic, radiographic and clinical findings ^[1]. The defect in skeletal growth is a result of lack of normal ossification of endochondrial bone resulting in increased fragility of bones. These patients usually have history of recurrent fracture of bones; present with hypermobile limbs, progressive hearing loss, muscle weakness and other associated skeletal deformities like kyphoscoliosis, short neck and pigeon chest. The patient's cardiovascular status should be evaluated especially mitral and aortic valve function for congenital and valvular heart disease and for cystic degeneration of proximal aorta. The anaesthetic considerations include difficult airway and risk of odontaxial dislocation, cervical vertebra, mandible and teeth fractures during laryngoscopy and intubation ^[2].

We are sharing our experience of the anaesthetic challenges faced by us in a dwarf male diagnosed with Osteogenesis imperfecta, who presented to the hospital with a skull-based meningioma SOL and midline shift. The patient underwent a craniotomy and excision of the SOL during which the challenges encountered included complex airway management, respiratory compromise secondary to skeletal deformity, dwarfism and the risk of fractures from positioning.

Case Report

In the pre-anaesthetic check-up clinic, a 48-year-old male presented with complaints of generalised tonic clonic seizures (GTCS) associated with frothing. He was a known case of Osteogenesis imperfecta with characteristic features of short stature, brittle bones, hypermobile joints and history of recurrent fractures of long bones for which he had been operated 4 times under general anaesthesia. Patient is a known case of hypertension and diabetes mellitus for which he had been taking medications regularly. Patient denies history of any cardiac disease or any family history of OI.

On general examination he was short statured (143 cm / 4 feet and 8 inches), 81kg and with a BMI of 39.6 kg/m² (obese class II), afebrile and with normal sclera (Fig 1). Respiratory system revealed barrel shaped chest with bilateral basal crept. Airway examination revealed a short neck with limited neck extension, one-fingerbreadth thyromental distance, adequate mouth opening, Mallampati class 3 and poor dentition.



Fig 1: General Built of Patient

Routine haematological investigations were normal. ECG showed left ventricular hypertrophy (LVH) pattern, echo showed an ejection fraction of 59% and diastolic dysfunction grade (DDG) I.

X-ray chest (AP) was suggestive of apparent cardiomegaly, multiple dorsal vertebral flattening and clear lung fields. X-ray cervical spine (lateral) was suggestive of degenerative changes in C4-C7 vertebrae. Pulmonary function tests (PFT) was suggestive of mild obstructive airway disease with significant reversibility post bronchodilator. CT brain with contrast was suggestive of a large hyper dense intensely enhancing lesion in right frontal lobe measuring 5.6 x 4.7 x 6.5 cm which is causing a mass effect on adjoining parenchyma and a midline shift of 7.5mm to the left is seen (Fig 2).

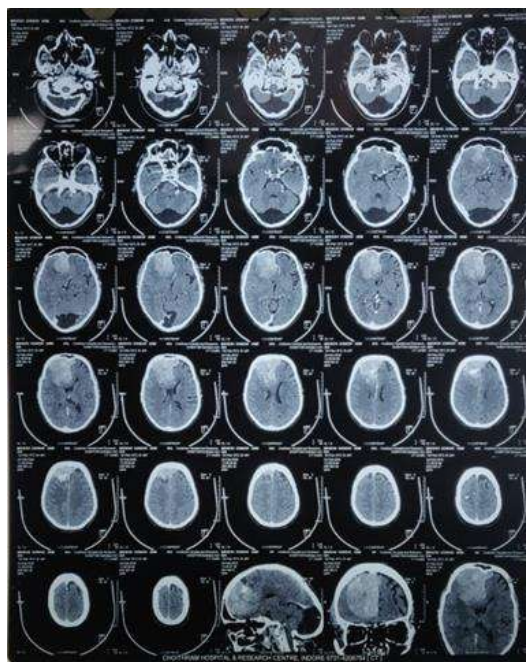


Fig 2: CECT Brain Film

Patient was labelled for surgery as ASA grade III. In the operating room, monitors were then applied (ECG, pulse oximeter). Blood pressure was measured manually once. Under USG guidance an 8F 4 lumen central line catheter was inserted in the right internal jugular vein (IJV). An arterial line was secured in the right radial artery for continuous BP monitoring. Difficult airway cart was kept ready.

Patient was placed in ramp position to ease ventilation and intubation (Fig 3). Patient was premedicated and preoxygenated with 100 % O₂ for five minutes. Anaesthesia was induced with propofol 200 mg and ability to mask ventilation was assessed before administering vecuronium 8 mg. The patient remained hemodynamically stable throughout the intubation procedure. For analgesia fentanyl infusion was started at a rate of 50mcg/hr.



Fig 3: RAMP Position for Induction

Anaesthesia was maintained using N₂O:O₂ in 2:1 ratio with sevoflurane to maintain a MAC of 1. Muscle relaxation was achieved by starting vecuronium infusion at a rate of 3mg/hr. Two units of blood were given to compensate for excessive bleeding.

The patient remained hemodynamically and vitally stable during the 10.5-hour surgery before being shifted to the ICU on mechanical ventilator. Patient was successfully intubated on post-operative day 2 without any complications and discharged on day 10.

Discussion

Osteogenesis imperfecta (OI) is a hereditary disease characterized by bone fragility and short stature^[3].

The anaesthetic considerations for patients with OI include:^[2]

(1) Preoperative

- Obtain detailed medical history to determine the type and severity of the patients' disease.
- Obtain echocardiography to evaluate cardiac anatomy and function if indicated.
- Accurately assess the airway to determine difficulty of intubation.
- Devise the Anaesthetic plan and alternative plans.
- Confirm blood type and screen.

(2) Intraoperative

- Transport and position patient with care.
- Consider arterial cannulation in place of blood pressure cuff to avoid bone fractures and bruising.
- Avoid succinylcholine use if feasible for patient clinical management.
- Be vigilant of hemodynamic and ventilation changes.

- e) Be cognizant of the risk for hyperthermia and malignant hyperthermia.

(3) Postoperative

- Ensure adequate oxygenation and ventilation.
- Monitor for postoperative haemorrhage.
- Pain control: patients with OI may have chronic bone pain that is not related to surgical site.

When case history shows symptoms of congenital heart defect or malformation of the thoracic vessels a preoperatively echocardiography should be performed ^[4].

T Sitsopoulos, *et al* in 2008 reported a 46-year-old male with a known family history of OI type I presented with progressive gait disturbances and diminished muscle strength. Brain MRI scans revealed an infiltrative intracranial mass occupying both front parietal lobes. The patient underwent surgical intervention. The histological diagnosis was an atypical (Grade II) meningioma. The bony parts demonstrated a mixture of osseous defects due to OI and infiltration by the tumor. At one-year follow up the patient's muscle power partially returned while repeat MRI scans were negative for tumor recurrence.

Jasveer Singh, *et al.* in 2017 reported a case of OI Tarda with difficult airway who underwent general anaesthesia uneventfully for internal fixation (nailing) of fracture shaft femur where endotracheal intubation (ETI) was executed with a Glide Scope™ (Verathon, Bothell, WA, USA) video-laryngoscope and a novel 180° upside down technique was used.

Conclusion

Patients with Osteogenesis imperfecta pose significant challenges to even the most trained anaesthesiologists.³ Such cases reemphasize the importance for an open discussion addressing the patient's goal of care and the surgical and anaesthetic concerns prior to a procedure.⁴ Hence, in our opinion, thorough and scrupulous workup of the patient pre-operatively will have a favourable outcome of anaesthesia in the hands of a perfectly vigilant anaesthetist.

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