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Case Report
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CASE REPORT

A Complex Clinical Scenario: Perioperative Management of Osteogenesis Imperfecta in Gynaecological Surgery

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ABSTRACT

Osteogenesis imperfecta (OI) presents significant challenges in perioperative management due to fragile bones, airway difficulties, and the potential for other systemic complications. We report a case of a 45-year-old female with OI undergoing a total abdominal hysterectomy. Perioperative complications included difficult positioning, airway management challenges, opioid sensitivity, and postoperative nausea and vomiting (PONV). The patient required critical care admission for postoperative care due to aspiration and oxygen dependency. This case highlights the need for meticulous preoperative planning and a multidisciplinary approach in managing OI patients undergoing major surgery.

Keywords: Osteogenesis imperfecta, Fragile bones, Airway difficulties

INTRODUCTION

Osteogenesis imperfecta (OI) is a rare genetic disorder characterized by varying degrees of skeletal and bone abnormalities. The most common characteristic of osteogenesis imperfecta (OI) is an alteration in type 1 collagen. The revised Nosology and Classification of Genetic Skeletal Disorders categorizes five clinical forms of osteogenesis imperfecta (OI): non-deforming with persistently blue sclera (OI type I), perinatally lethal (OI type II), progressively deforming (OI type III), moderate (OI type IV), and calcification of the interosseous membranes and/or hypertrophic callus.(2)

OI is a multisystem disorder, and the clinical presentation depends on the type of OI.(1.3) The diverse clinical presentation will require different types of surgeries throughout life. Orthopaedic surgeries are the most common type of procedure due to their increased susceptibility to fractures. In addition to skeletal problems, the perioperative period may also be complicated by extraskeletal manifestations.

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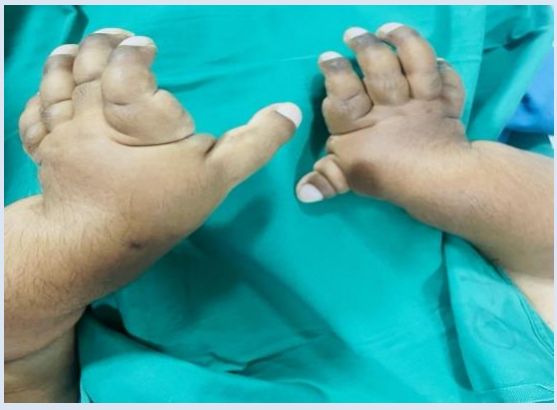
Anaesthetic management in OI patients is complex due to issues such as difficult airways, respiratory problems, and positioning issues. This case report highlights the perioperative challenges and management strategies in managing an OI patient undergoing a total abdominal hysterectomy. These challenges include a difficult airway with failed intubation, issues with positioning and surgical approach, opioid sensitivity, and postoperative complications.

CASE REPORT

A 35-year-old female patient, who was diagnosed with type III OI, presented for an elective total abdominal hysterectomy. The surgery was planned at the request of the patient to enhance her quality of life. At birth, she was diagnosed with OI and, with time, developed various skeletal abnormalities, including malformed fingers and short forearms. (Figure 1).

Figure 1: Maldeveloped fingers with short forearm

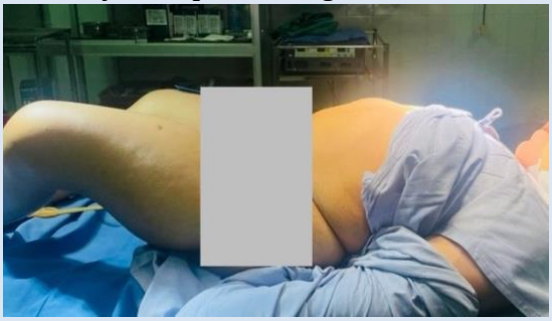


She had a short stature with a height of 112 cm and fixed flexion anomalies both at the hips and knees. Her mouth opening was limited to two finger breadths, accompanied by a fixed cervical spine and limited neck extension. There were no chest wall deformities or cardiac or endocrine abnormalities. The patient had undergone lower-limb orthopaedic procedures under regional anaesthesia and two episodes of general anaesthesia with a laryngeal mask airway without any problems. There were no episodes of PONV or delayed recovery despite using opioids.

Given the smaller size of the abdominal cavity and the fixed flexion deformity of the lower limbs, an open surgical procedure with a midline incision was planned. The patient refused awake fibre optic or awake videolaryngoscopy even after a detailed discussion. Therefore, airway management was planned using general anaesthesia, short-acting muscle relaxants, and a videolaryngoscope.

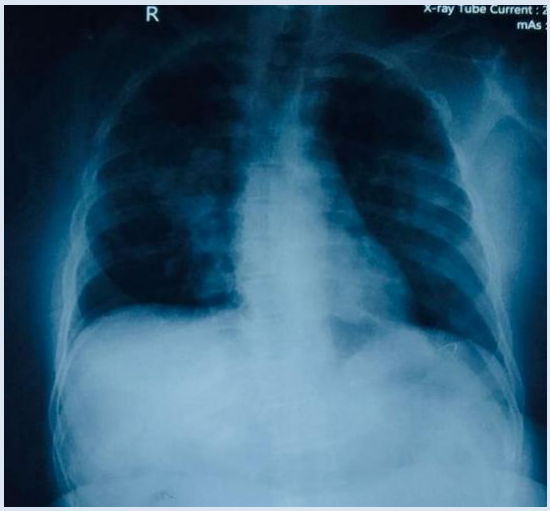
IV suxamethonium was given after induction with sevoflurane and confirmed the possibility of mask ventilation. The repeated attempts at orotracheal intubation with a videolaryngoscope were unsuccessful due to the inability to visualise the vocal cords. The patient was able to ventilate using a size 3 LMA, and it was decided to proceed with the surgery with ventilation via LMA. She received morphine 7mg and IV paracetamol as pain relief methods. The surgical procedure was difficult due to the patient's positioning, owing to the fixed flexion deformity of the legs. (Figure 2) It was further complicated by the limited space inside the pelvic cavity. Surgery lasted for 60 minutes with a blood loss of 75 mL.

Figure 2: Short stature with fixed flexion deformity after positioning



After complete recovery, the LMA was removed; however, during the process, the patient developed desaturation due to pulmonary aspiration. The recovery period was prolonged and lasted for 1 hour. The patient was admitted to the High Dependency Unit for observation, where she required oxygen therapy. Chest X-ray showed diffuse shadowing suggestive of aspiration. (Figure 3) She had several episodes of postoperative nausea and vomiting, which required multimodal therapy. On day 2, the hospital weaned her off the oxygen, and on day 4, she was discharged from the hospital.

Figure 3: Post operative CXR showing bilateral diffuse shadows suggestive of aspiration



DISCUSSION

Challenges in the perioperative management of patients diagnosed with osteogenesis imperfecta (OI) encompass various critical aspects, making surgical procedures challenging for both the anaesthetist and the surgeon.

Individuals with OI may exhibit craniofacial anomalies, micrognathia, and restricted neck mobility, posing challenges in airway management. An anticipated difficult airway necessitates meticulous preoperative assessment, with alternative airway management strategies readily available. The patient declined the safe options for awake fibreoptic intubation, thereby requiring alternative approaches. Despite limitations in neck mobility, the patient was ventilated with an LMA. However, this may have led to pulmonary aspiration, as OI may be associated with gastric dysmotility. If she had underlying pulmonary pathology such as kyphoscoliosis, pulmonary aspiration could have adversely affected her. Therefore, securing the airway in a safe manner is mandatory in patients with OI. Numerous studies have been conducted on patients with OI, specifically focusing on airway management. Studies done on the paediatric population show a low incidence of difficult airway and failed intubation.[4,5] Mohamad et al. describe using supraglottic airway devices in patients with OI to reduce airway-related injuries in this population. They reported 3.22% of difficult airways in this cohort.[6]

Patients with OI have delicate bones that are susceptible to fractures, particularly during positioning, transportation, and the use of automated blood pressure cuffs. Adequate padding of pressure points and gentle handling during surgical procedures and transfers are imperative to prevent fractures. Previous unprovoked fractures in this patient necessitated careful positioning. The presence of fixed flexion deformities in the lower limb made positioning as well as mobilization challenging.

The literature does not describe the incidence of prolonged recovery or the alterations in the pharmacology of anaesthetic medications. Most often, anaesthetic procedures were performed using regional techniques. This method would provide postoperative analgesia,

resulting in a shorter hospital stay. The nature and complexity of the surgery precluded the use of regional techniques in this patient. We observed prolonged recovery in our patient with opioid sensitivity. There is no evidence to support patients with OI having a high incidence of postoperative nausea and vomiting, as seen in our patient. This may be due to the use of opioids in gynaecological surgery, which anyway carries a high incidence of PONV. The presence of a fixed flexion deformity and the patient's reduced axial length complicated the surgical course, leaving the surgeon with inadequate space to reach the pelvic organs. Laparoscopic surgeries may pose challenges in OI patients due to the risk of inadvertent injuries during trocar insertion, insufflation, and manipulation of instruments within the abdominal cavity.[8,9] Farid et al. report successful laparoscopic management of suppurative appendicitis in a dwarf patient with OI. However, in OI patients with fragile tissues and bones, the risk of inadvertent visceral or vascular injuries during laparoscopic procedures may outweigh the potential benefits of minimally invasive surgery. Therefore, this should be considered on an individual basis.

CONCLUSION

This case illustrates the complexities involved in the perioperative management of patients with osteogenesis imperfecta undergoing major surgery. A multidisciplinary approach, including thorough preoperative planning and flexibility in intraoperative and postoperative management, is crucial for optimizing outcomes in these high-risk patients.

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