

Neuraxial and Regional Anesthesia in a Patient with Osteogenesis Imperfecta Undergoing Orthopedic Procedure: A Case Report

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Keywords

Osteogenesis Imperfecta, Anesthesia, Orthopedic surgery.

Introduction

Osteogenesis Imperfecta (OI) is a rare connective tissue disorder characterized by bone dysplasia, deformity, and fragility, affecting 1 in 10,000 to 20,000 live births [1,2]. This condition presents with manifestations such as short stature, craniofacial anomalies, severe scoliosis, hypermobile joints, and bone fractures resulting from minor trauma, often necessitating orthopedic interventions [3]. Additionally, OI is associated with restrictive pulmonary disease, coagulopathy, and a heightened risk of a difficult airway [3]. Managing patients with OI perioperatively poses a significant challenge for anesthesiologists.

This case report aims to provide a comprehensive account of the anesthetic approach employed for a 23-year-old male diagnosed with Osteogenesis Imperfecta Type 3. The patient underwent orthopedic surgery for femur fixation following the exposure of prosthetic material from a previous fracture.

Case Report

The 23-year-old male patient, weighing 25 kg and classified as ASA 2 (American Society of Anesthesiologists status 2), was admitted to the hospital for an elective surgical correction of a femur fracture associated with exposed prosthetic material. Diagnosed with type 3 Osteogenesis Imperfecta (OI), the patient presented chronic pain but was clinically well compensated, asymptomatic, and maintained on alendronate and calcium carbonate at home.

His medical history included multiple fractures and surgical repairs,

with the last procedure performed six years prior to admission. Preoperative lab assessments revealed a low hemoglobin concentration (11.0 g/dL), normal white blood cell count ($3.14 \times 10^9/\mu\text{L}$), low C-reactive protein concentration ($<5 \text{ mg/L}$), and normal renal function. Coagulation evaluation demonstrated an INR of 0.99, partial thromboplastin time of 26.5s/28.9s, and a platelet count of 300×10^3 , with no other abnormalities. Previous cardiac ultrasonography indicated normal cardiac systolic and diastolic function. Upon physical examination, severe scoliosis, extensive deformities in the spine, rib cage, limbs, and a craniofacial malformation with a short neck and restricted mobility were observed (Figure 1).

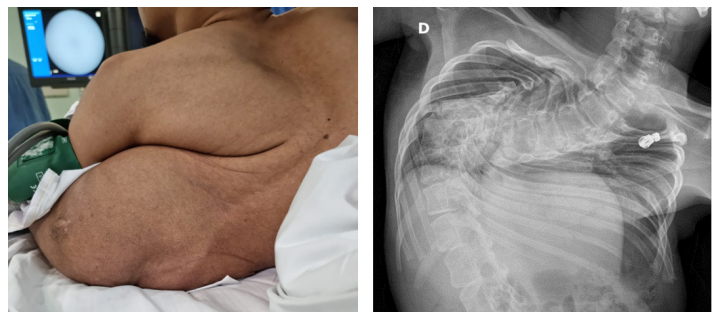


Figure 1: Extensive deformity in the patient with Osteogenesis imperfecta.

In the operating room, the patient underwent monitoring with electrocardiography, pulse oximetry, and non-invasive blood pressure measurement. A careful approach was taken to avoid iatrogenic limb fracture, with a maximum pressure limit set on the monitor at 130 mmHg. Initial vital signs were stable, and an 18-gauge venous access was established in the right upper limb.

Intravenous sedation was initiated with 2 mg of midazolam and 10 mg of ketamine. Following strict aseptic precautions, spinal anesthesia was performed using 10 mg isobaric bupivacaine and 30mcg morphine at the L4-L5 level. A satisfactory blockade was achieved with sensory blockage at T8. The figure 2 depicted the challenge in performing spinal blockade.



Figure 2: Spinal blockade in the patient of the case report.

Throughout the surgery, the patient was carefully positioned, and active heating with a thermal blanket was employed. Dexmedetomidine infusion at 0.4 mcg/kg/h provided adequate sedation on the RASS-3 scale. Oxygen therapy was administered via a low-flow nasal catheter at 2L/min. The surgery, lasting 3 hours and 30 minutes, involved the administration of 1200 ml Ringer's solution and a 284 ml autologous blood transfusion due to increased intraoperative bleeding. Arterial blood gas analysis revealed mild metabolic acidosis.

Figure 3 shows the preoperative and postoperative stages of the surgical correction of an exposed fracture associated with exposed prosthetic material.

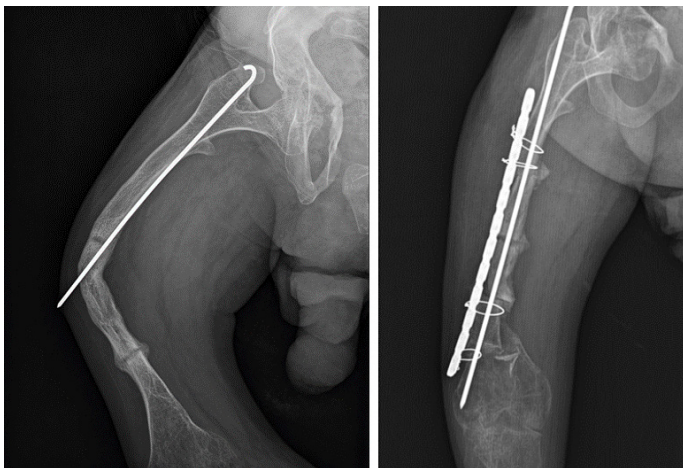


Figure 3: Preoperative and postoperative x-radiographs of the patient

showing the results of the surgical procedure.

Finishing the surgical intervention, an ultrasound-guided femoral nerve block with 15 ml of 0.5% ropivacaine was performed for optimized pain control. Symptomatic drugs, including 1.2 g dipyrene, 4 mg ondansetron, 4 mg dexamethasone, and 25 mg ketorolac, were administered (Figure 3).

The patient woke up at the end of the procedure and was transferred to the intensive care unit (ICU) for monitoring. He remained awake, hemodynamically stable, and complaint-free. Discharged to the infirmary the next day, the patient stayed in the hospital for two more days with effective pain control, without the need for opioid usage.

Discussion

In the case of a patient with Osteogenesis Imperfecta (OI), a meticulous pre-anesthetic assessment and detailed anesthetic planning are paramount, given the systemic involvement commonly associated with this disorder. The selection of an appropriate anesthetic technique necessitates a thorough understanding of the complexities inherent in OI and given the limited evidence in the literature—mostly comprising case reports—it becomes imperative to carefully evaluate the risks and benefits associated with various available anesthesia techniques [4].

Surgical considerations, particularly regarding positioning on the operating table, hold significant importance due to the inherent bone fragility observed in OI patients. Platelet function impairment further underscores the need for anticipating potential fluid replacement and blood transfusion requirements. Additionally, the presence of anatomical airway abnormalities and compromised ventilatory control presents a challenge, reinforcing the rationale for avoiding general anesthesia whenever possible [5]. Historically, the use of regional and neuraxial analgesia in patients with skeletal dysplasia and fragile bones has been perceived as relatively contraindicated due to concerns related to positioning, block placement, spinal cord injury in those with spinal deformity, and reported bleeding diathesis incidences [6]. However, evolving insights, as exemplified in this case, suggest that with a nuanced consideration of patient-specific factors—such as the severity of skeletal involvement and coagulation status—regional anesthesia can be successfully employed in select cases. Strategies to prevent iatrogenic fractures, meticulous positioning, and vigilant monitoring were integral to ensuring the patient's safety. Dexmedetomidine sedation played a pivotal role, particularly given the anticipated challenges associated with airway management in OI patients [7,8].

The intricacies of pain management in individuals with OI arise from the recurrent nature of fractures, frequent surgical interventions, and the potential for repeated exposures to opioids. Chronic pain is a common experience due to bone fragility and deformities, necessitating a nuanced and individualized approach

to pain management. Emphasizing multimodal analgesia, including regional techniques and non-opioid adjuncts, is crucial. Regional nerve blocks offer effective perioperative analgesia with minimal adverse effects, facilitating early rehabilitation exercises and contributing to the swift recovery of patients [8].

The success of patient management in individuals with OI underscores the pivotal role played by multidisciplinary collaboration, bringing together the expertise of anesthesiologists, orthopedic surgeons, and intensivists. This highlights the significance of well-prepared and cohesive teams in navigating the complexities of OI, ensuring comprehensive and effective care for these patients [6].

Conclusion

In conclusion, this report underscores the imperative of a meticulous and tailored anesthetic approach for patients with Osteogenesis Imperfecta Type 3 undergoing orthopedic interventions. Beyond the surgical procedure itself, the journey towards sustained well-being and complete healing necessitates vigilant and regular follow-up. The success observed in this case exemplifies the potential for a multidisciplinary approach, emphasizing the collaborative efforts of anesthesiologists, orthopedic surgeons, and other healthcare professionals. By continually refining our strategies and advancing our understanding of OI, we can pave the way for enhanced care, improved outcomes, and an unwavering commitment to the overall health and quality of life of these individuals.

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