Anesthesia management in a patient with osteogenesis imperfecta

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ABSTRACT

Osteogenesis imperfecta is a rare autosomal dominant collagen tissue disease that primarily affects the bones. It is a condition that requires to be carefully managed during the preoperative period and when administering anesthesia. In this case report, we aimed to present our experience with supraclavicular nerve block in a patient with osteogenesis imperfecta who underwent intervention for left humeral shaft fracture since it is a relatively simple and safe technique to perform.

Keywords: ultrasonography, osteogenesis imperfecta, supraclavicular nerve block

Osteogenesis imperfecta (OI), is one of the rare diseases with autosomal dominant transition and defective collagen maturation is which primarily affects bones. The main issue is that Type I collagen's structure, which is crucial for bone and other structural strength requirements, has been affected by a genetic mutation (disease) [1, 2]. OI's Clinical signs and symptoms include congenital heart disease, valvular heart disease, thin skin, blue sclera, hearing loss from otosclerosis, hyperthermia, hyperhidrosis, dental structural defects, platelet dysfunction, and cor pulmonale [3]. OI also raises the risk of hemorrhage and in patients with brittle OI, taking a blood pressure reading with a cuff every three to five minutes during receiving anesthesia may result in an iatrogenic fracture [4].

Administering general anesthetic presents a number of challenges, including difficult ventilation, difficult intubation, fractures to the teeth and jaw, risk of cervical injury, and difficulty in positioning. In addition, problems that need to be considered include the possibility of both malignant and non-malignant hyperthermia, respiratory failure caused by kyphoscoliosis, and regulation of cardiac anesthetic settings [5].

In this case report, we aim to discuss the perioperative anesthetic management of an adult patient with osteogenesis imperfecta who had a fracture to the left humeral shaft.

A 24-year-old, 44 kg female patient was taken into surgery due to a fracture to the left humerus shaft. The patient had old fractures in both right radius and left tibia. Surgery for scoliosis had previously performed. Physical examination findings during the preoperative evaluation included kyphoscoliosis, short neck (thyromental distance <6 cm), growth retardation, short stature, deformity, and shortness in both lower and upper extremities. The patient's echocardiogram, lung x-ray, and routine blood tests revealed no additional pathology. The operating room was prepared considering the potential complications in account.For the difficult airway device, arrangements were set up.

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Procedures related to the risk of developing Malignant Hyperthermia (MH), which has been reported to have a higher incidence in comparison to the general population, were additionally noted. The inhalation agent vaporizers were removed from the room. The anesthetic device's carbon dioxide absorber has been changed. Previously unappropiated anesthesia circuit was removed and the device was cleaned with 15 L min-1 fresh gas for 30 minutes.

At this point, the pharmacist provided Dantrolen, which was continually accessible in the operating room. The patient was taken to the operating room once the anesthetic consent was received and the patient was informed of any potential complications.

A continuous infusion of 0.9% NaCl was commenced at a rate of 10 mL/kg/hour after an intravenous 20G cannula was inserted into the contralateral arm. The patient was monitored with standard anesthetic motorization (ECG, blood pressure, pulse oximetry). The initial vital signs were 97 beats per minute, 138/95 mmHg for blood pressure, and 95% for oxygen saturation.The patient was positioned in a supine position and given 0.5 mcg/kg of fentanyl and 0.01-0.1 mg/kg of midazolam intravenously for sedoanalgesia. With povidone iodine, the left shoulder area that intended to be blocked was sanitized. A 22G 100mm long needle (Stimuplex Ultra®, Braun, Melsungen, Germany) was used.Next to the injection site, the ultrasonography probe (GE ML6-15-D Matrix Linear) was positioned 1 cm above the clavicle. The subclavian artery and brachial plexus were visible in the transverse sectional view of the patient when the ultrasonography probe was positioned in the supraclavicular fossa in the coronal oblique plane with the head tilted 45° to the contralateral side. The patient was resting supine at the time (Figure 1). The hypoechoic nodule cluster referred to as the brachial plexus was frequently observed lateral to the hypoechoic subclavian artery, which lay on top of the hyperechoic first rib and pulsed firmly (Figure 2). It allowed for real-time observation of the needle's movement. A nerve stimulator (Stimuplex) was activated after the needle reached the brachial plexus cluster. It initiated at 0.5 mA and increased to a maximum of 1.5 mA to trigger a muscle twitch. Using the in-plane approach, 15 cc of 0.5% bupivacaine and 10 cc of 2% lidocaine combination were administered after the localization of the plexus brachialis was determined. Diffusion across the truncus was observed during the injection of local anesthetic (Figure 3). Sensorial block at the 20th minute, motor block at the 30th minute (Essam scale: 2) (normal muscle strength normal hand gripping, wrist and elbow flexion (3), muscle strength decreased but wrist and elbow flexion protected (2), muscle strength decreased, only elbow flexion protected (1), no muscle strength, wrist and elbow flexion absent (0) was observed) (6). During the 60-minute procedure, no additional analgesics were required.After 30 minutes of postoperative anesthetic care unit monitoring, the patient was transferred to the service with a modified aldrate score of 10, showing no block-related complications.

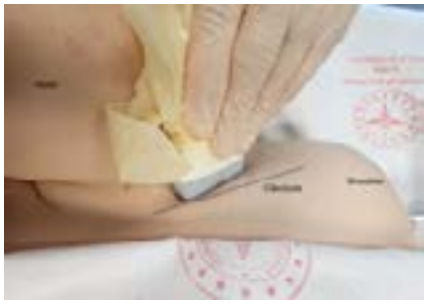

Figure 1. Patient resting at the supine position

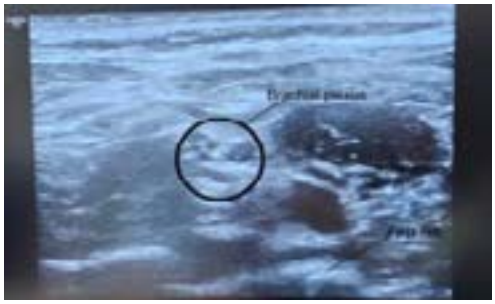

Figure 2. The hypoechoic nodule cluster referred to as the brachial plexus

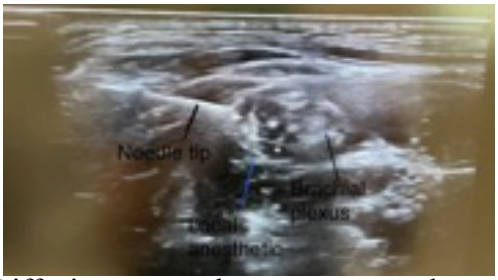

Figure 3. Diffusion across the truncus was observed during the injection of local anesthetic

The ultrasound image of the supraclavicular brachial plexus at the level of the first rib.

DISCUSSION

OI is also called "brittle bone disease" since the fractures can occur following a slight or no trauma. According to a previous study, OI affects approximately 6-7 persons out of every 100,000 and is present in about 1 in 20,000 infants [7].The disorder is caused by a defect in type I collagen synthesis, secondary to a gene mutation. Type I collagen is the principal type in bone, skin, sclera, dentine, and other tissues; therefore, lesions in these areas are more visible [8].

In addition, recurrent pharyngeal collapse and even apnea may occur during sleep due to abnormalities in pharyngeal anatomy, upper airway dilatation and ventilatory regulations [9]. Consequently, managing anesthesia during surgery in individuals with these diseases presents serious difficulties. Patients with OI need to be carefully managed under general anesthesia to minimize the risk of malignant hyperthermia and mandibular bone fractures during tracheal intubation.

During the perioperative period, kyphoscoliosis-related issues or neck movement restrictions may result in difficult airway management and dental injuries. However, the literature also has a large number of cases of successful laryngeal mask use and intubation [10, 11].

The development of MH is definitely the most feared complication, apart from the difficult airway. The syndrome known as MH develops from a hereditary deficiency in the skeletal muscles and is characterized by the emergence of a hypermetabolic reaction following exposure to a trigger, such as depolarizing neuromuscular blockers or volatile anesthetics. Increased carbon dioxide, hyperthermia, tachypnea, tachycardia, muscle rigidity, acidosis, hyperkalemia, and rhabdomyolysis are among the symptoms. This condition develops rapidly and has a high death rate; early diagnosis and treatment are crucial. Remember that cardiac symptoms related to hyperkalemia may also appear [12]. A standard clinical rating score was developed by Laranch and colleagues in 1994, which can be used in the diagnostic approach and is accepted by the international consensus. According to this scoring system, the probability of MH development can be estimated/calculated proportionally [13]. Removal of the triggering substance should be the first action taken once there is clinical doubt.Symptomatic treatment should be applied afterwards.Dantrolene sodium is the only medication that known to stop the MH cascade. After the introduction of the drug, the mortality rate due to MH decreased from 80% to 10%.Though the exact mechanism of action of this drug is unknown, it is believed to be related to ryanodine receptor inhibition-induced blockage of calcium release from the sarcoplasmic reticulum [14]. Total intravenous anesthesia (TIVA) administration is an optimal anesthetic maintenance technique that may be applied to patients at risk for MH. As they are easier to use and less hazardous than general anesthesia, peripheral blocks are utilized more often than general anesthesia [15].

OI is a severe disease with a number of complications. For patients who need surgery, it's important to assess the severity of the state of affairs, practice a comprehensive preoperative evaluation, and create a personalized anesthetic management strategy. Anesthesia safety and smoothness are enhanced by early identification of risk factors and optimizing the preoperative health status of patients with OI.

In our case, we aimed to minimize the risks that may occur during intubation by preferring supraclavicular block.

We believe that peripheral block can be used as an easier, safer and less hazardous method compared to general anesthesia.

Conflict of Interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Authors' Contribution

Study Conception: EU, EÇ; Study Design: EU, EÇ; Literature Review: EU, EÇ; Critical Review: EU, EÇ; Data Collection and/or Processing: EU, EÇ; Analysis and/or Data Interpretation: EU, EÇ; Manuscript preparing: EU, EÇ.

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