

Infraclavicular Block and Anesthesia Management in Two Cases with Osteogenesis Imperfecta

Osteogenesis İmperfektali İki Olguda İnfraklavikuler Blok ve Anestezi Yönetimi

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ABSTRACT

Osteogenesis imperfecta, is a rare autosomal hereditary connective tissue disease. Extremely fragile bones, other organ and body system disorders are prominent in such cases. There are few studies in the literature related to anesthesia for patients with Osteogenesis Imperfecta. In terms of anesthesia, cases with osteogenesis imperfecta have risks such as difficult airway and malignant hyperthermia. Therefore, they require a careful anesthesia management. Although, in literature general and epidural anesthesia applications on patients with osteogenesis imperfecta have been reported, peripheral block usage is very rare. In this article, we aim to present infraclavicular block experiment, a method which we prefer due to its convenience and safety, in two cases with osteogenesis imperfecta.

Key Words: Anesthesia management, infraclavicular block, Osteogenesis imperfecta

ÖZET

Osteogenesis imperfekta, nadir görülen otozomal kalıtmı bir bağ dokusu hastalıdır. Bu olgularda son derece kırılğan kemik, diđer organ ve vücut sistemlerinin bozuklukları ön plandadır. Osteogenesis imperfekta'lı hastalarda anestezi ile ilgili literatürde az sayıda çalışma vardır. Osteogenesis imperfektalı olgular anesteziik açıdan zor hava yolu ve malign hipertermi gibi riskler taşımaktadır. Bu nedenle dikkatli bir anestezi yönetimi gerektirirler. Literatürde Osteogenesis imperfekta'lı hastalarda genel ve epidural anestezi uygulamaları bildirilmiş olmasına rağmen, periferik blok kullanımı çok seyrekdir. Bu yazıda Osteogenesis imperfekta'lı iki olguda uygulanması kolay ve güvenli bir yöntem olduđu için tercih ettiğimiz infraklavikuler blok deneyimimizi sunmayı amaçladık.

Anahtar Kelimeler: Anestezi yönetimi, infraklavikuler blok, Osteogenesis imperfekta

Introduction

Osteogenesis imperfecta (OI) is a rare autosomal hereditary connective tissue disease. In live birth, the incidence ratio is reported as approximately 1:20,000 (1). Extremely fragile bones, other organ and body system disorders are prominent in such cases. There are few studies in the literature related to anesthesia for patients with OI. These reports are often written to emphasize the risk of iatrogenic trauma, coagulopathy risk and malignant hyperthermia (2).

Peripheral blocks are some of the safest methods among anesthesia methods. Therefore, in cases of risky general anesthesia, it is a proper method of anesthesia. With the use of ultrasound (US) in peripheral blocks, the drug doses have been decreased, and it has become possible to make blocks by displaying the vascular structures (3).

Although, in literature general and epidural anesthesia applications on patients with OI have been reported, peripheral block usage is very rare. In this article, we aimed to present infraclavicular block experiment, a method which we prefer due to its convenience and safety, in a case with osteogenesis imperfecta.

Case Reports

Case 1: A male patient at the age of 14 who was intended to be operated due to implant removal from left arm elbow was assessed by anesthesia clinic. The patient history informed of 20 previous fractures, none of which was subjected to surgical procedure while about 1.5 years ago an implant was implemented to the patient due to left forearm elbow fracture. It was informed that during this process, general anesthesia was

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preferred and no problems were faced. The patient's routine laboratory tests, physical examination, electrocardiography and PA lung X-ray were normal. To remove the implant, instead of general anesthesia, peripheral block implementation to the patient was planned. Patient and relatives were given sufficient information about peripheral blocks and their confirmation was obtained. The patient was monitored with standard anesthetic motorization (pulse, blood pressure, pulse oximetry). After placing the patient in supine position, 2 mg midazolam, 50 mg ketamine and sedoanalgesia were intravenously applied before the operation. The block area on the left shoulder was disinfected with povidone-iodine. 20 cc local anesthetic with %0.375 bupivacaine concentration was prepared. 22G 100mm in length (Stimuplex Ultra®, Braun, Melsungen, Germany) needle was used. Ultrasonography (Esaote® MyLab 5, Florence, Italy) probe was positioned 1 cm below the clavicle right next to the injection area. With the help of the probe, subclavian artery and brachial plexus cords were spotted and needle was referred to the posterior of the subclavian artery with in-plane technique. With US guided lateral sagittal infraclavicular approach, 14 ml of the local anesthetics mixture was injected to the posterior cord of plexus, while 3 ml local anesthetic was injected to anterior and superior cords by practicing intermittent aspiration and observing drug distribution. The distribution around the cord was observed via US during local anesthetic implementation. No complications such as; vein puncture, pneumothorax, and local anesthesia toxicity were observed. The surgical anesthesia occurred within 45 minutes and no additional analgesic was required during the 45 minutes operation. The operation was successfully completed and the patient was sent to the service.

Case 2: 13 years old, 28 kg male patient was scheduled for operation due to left humerus fracture. The patient had old fractures in the right femur and the left tibia. In the preoperative physical examination, short stature, growth retardation, short neck (thyromental distance <6cm), kyphoscoliosis and disfigurement and shortness in both sub and upper extremities. Difficult airway in the patient was not considered of because of the general anesthesia due to previous fractures. No additional pathology in patient's routines, in the lung film and echocardiography was detected. Patient's relatives were informed that instead of general anesthesia, there was the opportunity to implement anesthesia

only on the left arm. Approval of patient's relatives for implementation of infravascular block was obtained.

Vascular access was implemented to the patient with 20G granules on the right arm from the antecubital area and 0.9% NaCl infusion started at a rate of 10 mL kg/h. The patient was premedicated with 1 mg of midazolam and was taken to the operation room. The patient was monitored with standard anesthetic motorization (pulse, blood pressure, pulse oximetry). After placing the patient in supine position, 1 mg midazolam was intravenously applied and sedoanalgesia with 50 mcg fentanyl was applied. The left shoulder area where the block was to be made was disinfected with povidone-iodine. 10 mL local anesthetic with %0.375 bupivacaine concentration was prepared. 22G 100mm in length (Stimuplex Ultra®, Braun, Melsungen, Germany) needle was used. Ultrasonography (Esaote® MyLab 5, Florence, Italy) probe was positioned 1 cm below the clavicle right next to the injection area. With the help of the probe, subclavian artery and brachial plexus cords were spotted and needle was referred to the posterior of the subclavian artery with in-plane technique. With US guided lateral sagittal infraclavicular approach, 6 mL of the local anesthetics mixture was injected to the posterior cord of plexus, while 2 mL local anesthetic was injected to anterior and superior cords by practicing intermittent aspiration and observing drug distribution. The distribution around the cord was observed via US during local anesthetic implementation. No complications such as; vein puncture, pneumothorax, and local anesthesia toxicity was observed. The surgical anesthesia occurred within 17 minutes and no additional analgesic was required during the 60 minutes operation. The operation was successfully completed and the patient was sent to the service.

Discussion

OI is a disease that collocates with bone deformities, deafness, blue sclera, macrocephaly, kyphoscoliosis, anemia, thrombocyte dysfunction, and metabolic disorders, cardiac and pulmonary disorders (1). In OI cases, extremely fragile bones, other organ and body system disorders are prominent.

These patients need surgical intervention and anesthesia practice from birth onwards for various reasons, especially bone fractures (4,5). However, it was reported that fractures could even occur

during the non-invasive blood pressure measurement due to the over-inflation of the arm cuff. Kyphoscoliosis or neck movement limited situations could cause difficult airway and cause teeth injuries in the perioperative period. But in literature, many examples regarding successful intubation and use of the laryngeal mask are also available (6,7).

Although OI is a rare disease, it requires a careful anesthesia management due to risks such as difficult airway and malignant hyperthermia (4,8). We aimed minimizing possible risks during intubation by preferring infraclavicular block in our cases. Because peripheral blocks are preferred more often than general anesthesia as they are easier to implement, safer and less risky compared to general anesthesia (3).

There is no specified completely secure anesthetic technique for IO cases as it is a rare disease seen in a heterogeneous population. However, sevoflurane induction and TIVA have been proposed particularly for cases who are anticipated to have difficult airway and difficult vascular access (2,9).

In management of general anesthesia of patients with OI, the attention should be given to malignant hyperthermia risk and mandibular bone fractures due to tracheal. We believe that peripheral block can be preferred as a method which is easier to implement, safer and less risky compared to general anesthesia (3).

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