

Case of Calcaneus Fracture with Anderson-Fabry Disease; Anesthetized by Ultrasound Guided Popliteal Block

Muhammet Ahmet Karakaya^{1*}, Belitsu Salgın¹, Kamil Darçın¹, İlker Eren²,
Ömür Erçelen¹

¹ Department of Anesthesiology and Reanimation, Faculty of Medicine, Koc University, İstanbul, Turkey

² Department of Orthopedics and Traumatology, Faculty of Medicine, Koc University, İstanbul, Turkey

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*Corresponding Author

Muhammet Ahmet Karakaya

Koc University Hospital

Zeytinburnu/İstanbul, Turkey

Phone: +905332375730

E-mail: akarakaya@kuh.ku.edu.tr

ORCID:<http://orcid.org/0000-0001-8026-4783>

Abstract: Anderson-Fabry disease is a rare hereditary disease. It affects the entire body. Globotriacylceramide accumulates especially in the kidneys, myocardium, pancreas and lungs. All this accumulation can increase the risks of general anesthesia. Therefore, regional anesthesia methods can be used. We aimed to share with our anesthesia experience in a patient with Anderson-Fabry disease who was operated due to calcaneus fracture with ultrasound-guided popliteal block and sedation. © 2020 NTMS.

Keywords: Anderson-Fabry Disease, Nerve Blockade, Calcaneus fracture.

1. Introduction

Anderson-Fabry disease (FD) is a multisystemic, X-linked, hereditary disease that presents with vascular, endothelial and smooth muscle lysosomal glycosphingolipid accumulation caused by α -galactosidase A deficiency (in women) and absence (in men). It affects the whole body and causes progressive multiorgan dysfunction in childhood and early adulthood (1-3). Globotriacylceramide accumulates specifically in kidneys, the myocardium, pancreas and lungs. Among its many symptoms, the fact that it affects renal and respiratory functions and peripheral nervous system and causes cerebrovascular disease to hold clinical importance (2). There are only a few case reports on operations performed under general anesthesia in FD patients. In this report, we aim to share our anesthesia experience in a patient with FD undergoing operation for a calcaneal fracture surgery with a sciatic block and sedoanalgesia.

2. Material and Methods

2.1. Case

25 years-old male patient with FD and widespread angiokeratoma (Figure 1) presents with a calcaneus fracture following a fall-from-height. His previous medical history is remarkable for chronic neuropathic pain and myofascial pain with dizziness, day-time fatigue and tendency to over-sleep. He did not suffer from any heart failure symptoms and his echocardiography did not show any signs of heart failure also. His chest X-Ray was unremarkable. His diuresis was sufficient with BUN: 6 mg/dL, serum creatinine: 0,54 mg/dL and Urea: 12 mg/dL. However, his urine analysis revealed protein: 30 mg/dL and RBC: 62/ μ L. No pathology was detected after urological assessment. The patient was using agalsidaz alfa 0.2 mg/kg enzyme replacement therapy once every two weeks (Replagal®; Shire Human Genetic Therapies, Inc) and had no other regular medications.

As premedication, he was given 2 mg midazolam. Sciatic nerve was visualized 5 cm above the popliteal fossa with an ultrasound (USG) device (Acuson Freestyle®, Siemens Medical Solutions USA, Inc.) linear probe (5-13 MHz) in prone position. Peripheral block was applied in in-plane method with a USG compatible 50 mm, 22 gauge, short-bevel stimulating needle (Stimuplex D 50® ; B. Braun, Melsungen AG, Germany) with a 20 mL local anesthetic combination consisting of 200 mg lidocain (Aritmal® %2, Osel Inc.) and 50 mg bupivacaine (Marcaine®, Astrazeneca Inc.). (Figure 2). Blockage level was confirmed after 15 minutes with the pin-prick test and the patient was sedated with 50 µg fentanyl (Talinat®, Vem Ilac Inc.) and 40 mg propofol (Propofol® %2, Fresenius Inc.) for the operation to proceed afterwards. 50 µg fentanyl was added in the 70. minute of the operation. The operation lasted approximately 150 minutes. For post-operative analgesia management, the patient was prescribed intravenous patient-controlled analgesia (PCA) with a 10 µg/mL fentanyl concentration, 10 µg/h infusion rate, 15 µg bolus dose and 15-minute lock period. The effect of the motor block at the postoperative 6th hour and the effect of the sensory block at the 7th hour disappeared. The patient defined pain at the end of post-operative 8th hour; however, his visual analogue score (VAS) remained below 3 throughout. In total, in the first 24 hours, 406 µg fentanyl was given. His fentanyl infusion was stopped after 24 hours, and he received 180 µg and 30 µg in the second and third 24 hours, respectively.



Figure 1: Fabry disease and widespread angiokeratoma.

3. Discussion

FD is a hereditary, X-linked, lysosomal storage disease with a multisystemic presentation. Galactosidase alpha (GLA) gene mutation, that codes for α -galactosidase A, results in glycolipid, especially globotriacylceramide (GL-3), accumulation in various cell types, especially in vascular endothelial and smooth muscle cells (1). It causes cellular dysfunction not only in skin, renal, heart, lung and brain cells but also in the gastrointestinal system and the cornea. Injured endothelial cells in large and small vessels and pathologic vascular smooth muscle cells cause early-

onset hypertension, left ventricular hypertrophy, cardiac coronary disease and vertebrobasillar artery signs and symptoms with headache and tendency to over-sleep. Small fiber neuropathy may start early and presents with acroparesthesia of the hands and feet. FD includes a variety of symptoms, like acroparesthesia, cornea verticillata and stomach aches in childhood, angiokeratomes, renal failure, electrocardiography (ECG) anomalies in teenage years, and sleepiness, obstructive sleep apnea, hypertension, cardiomyopathy, kidney failure and strokes in adulthood. The most common first sign towards a diagnosis is the early onset renal failure (3). Given that it is a multisystemic disease, the preoperative evaluation should be individualized and careful. Those with FD are under great risk for surgery because of their renal and respiratory dysfunction, cerebrovascular and cardiovascular disease incidence and their intra-operative management may prove to be challenging. Pre-, intra-, and post-operatively, hypertension, cardiac valvular problems, arrhythmias, cardiac conduction pathologies, obstructive lung disease, hyperalgesia, proteinuria, renal damage and nausea-vomiting and malabsorption constitute the primary base of problems that can arise (3, 4). FD shows a variety of signs and symptoms that change with sex, severity and age that should be monitored and evaluated carefully pre-operatively (3). In addition, extra effort should be made to preserve organ functions in milder cases. In comparison with similar patient demographics without FD, chronic neuropathic and myofascial pain, dizziness, daytime fatigue, neurological signs like excessive sleeping and renal impairment were problems we had to manage with this case. No invasive monitoring was needed in our case; however, this could change with more intense symptoms. Dizziness was considered as a neurological symptom and therefore regional anesthesia was preferred over general anesthesia.

Renal pathologies are among the distinguishing signs of FD and usually are the cause of death between 30-50 years old. Polyuria because of concentrating incapacity might be the first sign and is always followed by rapidly progressing proteinuria (4). Therefore, the proteinuria in our patient was probably a sign of fore coming renal failure.

Spheric concentric ventricular hypertrophy, valvular anomalies, cardiac conduction defects are seen commonly in men with FD over 30 years old. In addition, lung and airway diseases are significantly worse in men and smokers. It is known that many patients with FD have explicit airway obstructions and intrinsic airway diseases (2, 4). This was one of the most important reason of why we chose regional anesthesia in this case.

None of the medications used were contraindications in enzyme replacement therapy. As a result, agalsidase alfa treatment should be prescribed as usual after regional anesthesia.

Using USG guidance for peripheral nerve blocks is a considerably new method in anesthesia but has grown rapidly over the past years. Of its reported benefits, faster initiation of block effect, higher success rates and longer operative and post-operative analgesia management can be counted. Better pain management, quicker return of gastrointestinal motility, lesser opioid use and resulting decreased nausea, decreased hospitalization times, better preservation of respiratory functions and easier involvement in physical therapy made regional anesthesia extremely popular in recent times (5, 6). In addition to there being very few case reports on general anesthesia and FD patients on literature, we did not come across one that has performed a peripheral block. Additionally, there is little information on peripheral blocks and post-operative analgesia. Planning of peri-operative pain management is of top-most importance for the patient. However, there is still no specific protocol to follow and both the patient and the surgeon's individual circumstance and preferences make it challenging to construct one (5, 6).

We preferred opioids instead of non-steroid anti-inflammatory drugs in post-operative analgesia to prevent renal damage. Recently, intravenous lidocaine has been used successfully to manage pain-episodes in FD patients (1). Therefore, we chose lidocaine to be the half of our local anesthesia in the peripheral block. Lastly, opioids are shown as effective in pain episode' management; however, it should be kept in mind that chronic use of opioids may result in constipation, addiction and drowsiness (3, 4). Additionally, if renal functions are compromised, wanted and unwanted effects of opioids may be prolonged and stressed. Accumulation of morphine and its metabolites may cause post-operative delayed respiratory depression. Fentanyl was chosen since it's metabolized in liver and has no toxic metabolite and is therefore safe to use in renal damage (7).

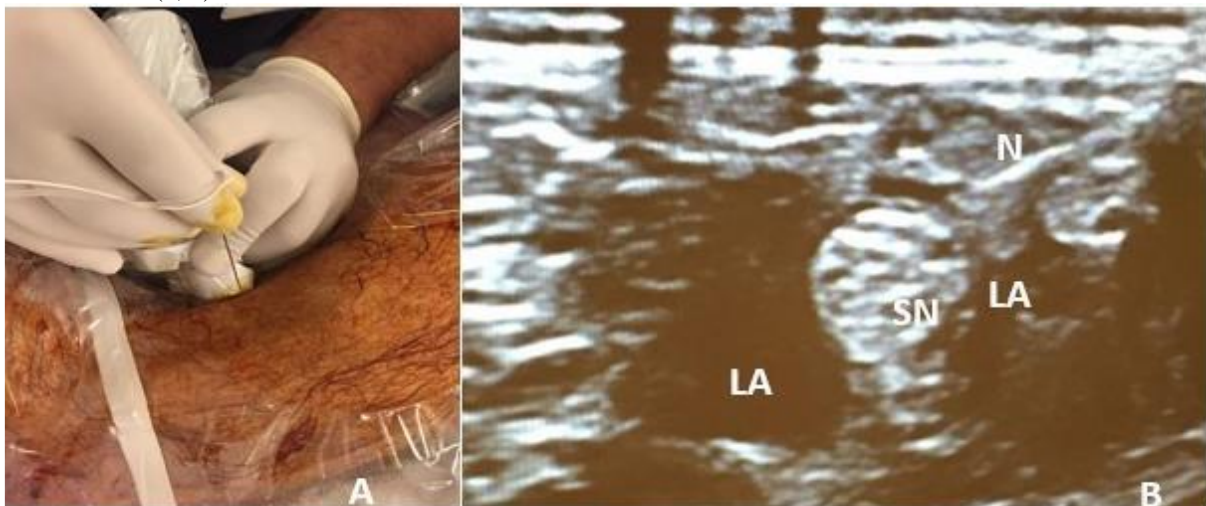


Figure 2: A. Sciatic Nerve block, USG guided in in-plane method. B. Sciatic Nerve USG view. LA: Local anesthetic, SN: Sciatic Nerve, N: Needle.

4. Conclusions

In conclusion, Anderson-Fabry disease' preoperative evaluation should focus on heart, brain, lung and renal organ damage assessment. Extra caution should be taken to preserve these organ functions. Our clinical experience showed peripheral blocks can be used in FD patients. We think careful post-operative pain management is additionally important in these patients who are inclined for chronic pains. We believe regional anesthesia practices are extra advantageous in these favored patients whom airway management is unpredictable.

Conflict of interest statement

No conflict of interest.

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Authors' ORCID

Muhammet Ahmet Karakaya
<http://orcid.org/0000-0001-8026-4783>
Belitsu Salgın
<http://orcid.org/0000-0003-3134-3447>
Kamil Darçın
<http://orcid.org/0000-0002-0004-8392>
İlker Eren
<http://orcid.org/0000-0003-2965-7690>
Ömür Erçelen
<http://orcid.org/0000-0002-5508-1077>



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