



Use of ultrasound-guided supraclavicular brachial plexus block as an anaesthetic technique in a patient with neurofibromatosis type 1: a case report

Nörofibromatozis tip 1 tanılı hastada anestezi yöntemi olarak ultrason eşliğinde supraklavikuler brakial pleksus blok uygulaması: Olgu sunumu

Emine Aysu ŞALVIZ,¹ Emre Sertaç BİNGÜL,¹ Meltem Savran KARADENİZ,¹ Ömer BERKÖZ,² Erman AK,² Kamil Mehmet TUĞRUL¹

Summary

Neurofibromatosis type 1 is an autosomal dominant condition characterized with cutaneous and/or plexiform neurofibromas and hyperpigmented café-au-lait spots. It affects multiple endocrine and visceral organs and can be associated with several difficulties such as potential airway (ventilation/intubation) problems, abnormal spinal anatomy, and peripheral neurofibromas. Therefore; anaesthesia technique decision becomes more of an issue in terms of avoiding complications, and decreasing morbidity and mortality. In this present case, we describe the utility of ultrasound and successful performance of supraclavicular brachial plexus block for surgery on the arm and forearm in a patient with neurofibromatosis type 1.

Keywords: Neurofibromatosis type I; supraclavicular brachial plexus block; ultrasound.

Özet

Nörofibromatozis tip 1 kütanöz ve/veya pleksiform nörofibromlarla ve hiperpigmente 'café-au-lait' lekeleriyle karakterize otozomal dominant bir hastalıktır. Endokrin ve visseral organları tutmakta, ve potansiyel havayolu problemleri (ventilasyon/entübasyon), anormal spinal anatomi ve periferik nörofibromlar gibi çok sayıda zorlukla ilişkili olabilmektedir. Bu sebeplerle komplikasyonlardan kaçınmak, morbidite ve mortaliteyi düşürmek için anestezi yönteminin seçimi daha da önem kazanmaktadır. Biz, bu olgu sunumu ile kol ve önkol cerrahisi geçirecek olan nörofibromatozis tip 1'li hastada ultrason kullanımının ve başarılı supraklavikuler brakial pleksus blok uygulamasının yararlılığını ifade etmeye çalıştık.

Anahtar sözcükler: Nörofibromatozis tip I; supraklavikuler brakial pleksus blok; ultrason.

Introduction

Neurofibromatosis type 1 (NF-1) (von Recklinghausen disease) is an autosomal dominant, neuroectodermal disease which affects multiple endocrine and visceral organs by localized neural tissue deposition.^[1,2] It is characterized with cutaneous neurofibromas, hyperpigmented café-au-lait spots and plexiform neurofibromas that tend to infiltrate long peripheral nerves as well as its surrounding tissues.^[3]

Looking from the view point of anaesthetists, several

difficulties as potential airway (ventilation/intubation) problems, abnormal spinal anatomy, and peripheral neurofibromas make different methods of anaesthesia difficult for NF-1 patients.^[4] Therefore; pre-operative detailed physical especially neurologic examination, and intracranial, neuroaxial and airway imaging become more of an issue either to decide for anaesthetic technique or to decrease morbidity and mortality. Besides above mentioned reasons, unexpected responses to depolarizing or non-depolarizing drugs during general anaesthesia (GA) and availability of ultrasound (US) make regional techniques

¹Department of Anaesthesiology, İstanbul University İstanbul Faculty of Medicine, İstanbul, Turkey

²Department of Plastic and Reconstructive Surgery, Division of Hand, İstanbul University İstanbul Faculty of Medicine, İstanbul, Turkey

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Correspondence (İletişim): Dr. Emine Aysu Şalvız. İstanbul Üniversitesi İstanbul Tıp Fakültesi, Anesteziyoloji Anabilim Dalı, Cerrahi Monoblok, Giriş Katı, Millet Cad., 34093, İstanbul, Turkey.

Phone (Tel): +90 - 212 - 414 20 20 **e-mail (e-posta):** aysusalviz@gmail.com

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Figure 1. Patient's right arm and forearm with painful neurofibromas, hyperpigmented diffuse café-au-lait spots and progressive weakness.

more preferable for patients with NF-1. However; there is still lack of evidence regarding the success of peripheral nerve blocks (PNBs) and the course of disease after PNBs in these patients.

We describe the utility of US and successful performance of supraclavicular brachial plexus block (SCB) for surgery on the arm and forearm in a patient with NF-1.

Case Report

A 20-year-old female (70 kg, 165 cm), ASA physical status II patient presented to the Department of Plastic Surgery, Division of Hand Surgery with couple of painful neurofibromas, hyperpigmented diffuse café-au-lait spots and progressive weakness on her right arm and forearm (Figure 1). She was scheduled for excision of neurofibromas and localised skin hyperpigmentation.

Besides NF-1, she also had a past medical history of hypothyroidism and diabetes mellitus type 2. Her cranial magnetic resonance imaging demonstrated hamartomatous lesions on the right side of thalamus and pons. In addition, her right arm MRI showed a neurofibroma-like mass with a size of 5.4x3.4 cm next to the vascular structures at the axillary region, and atrophy of flexor muscles on the arm and forearm. At age 9 y, she had neurofibroma excision from right axillary region under GA without any complication. Physical examination revealed normal mouth opening, Mallampati Class 2, mild mental retardation, obesity, multiple diffuse hyperpigmented le-

sions on the skin and flexion disability of the right arm and forearm.

The patient was considered to be at high risk for GA due to her pons lesion and mental retardation. In addition, muscle relaxants were preferred not to be used because of their variable responses. After a discussion with the surgical team, SCB was decided to be performed.

The patient was brought to the operating room and standard monitoring (pulse-oximeter, noninvasive blood pressure and electrocardiogram) was applied. A 20-gauge intravenous (IV) cannula was placed on the left hand of the patient and a free running infusion of 0.9% NaCl was started. Oxygen 2 L/min was applied via mask. Sedation was achieved with IV midazolam 1 mg and a loading infusion of 1 µg/kg/h dexmedetomidine for 10 minutes following by a maintenance infusion of 0.5 µg/kg/h.

With the patient in the supine position and head turned away from the side to be blocked, skin was disinfected. A high frequency linear ultrasound probe (5-13 MHz, GE Healthcare, Wauwatosa, USA) was placed in the transverse plane immediately superior to the clavicle at approximately its midpoint. Subclavian artery was confirmed by Doppler and brachial plexus was visualized anterolaterally as a collection of hypoechoic oval structures (Figure 2a). No neurofibroma was recognised within the sheath. After 2 mL lidocaine 2% infiltration into the skin, a 22-gauge 50 mm insulated needle (Stimuplex A, B Braun, Melsungen, Germany) was inserted lateral to the probe using in-plane

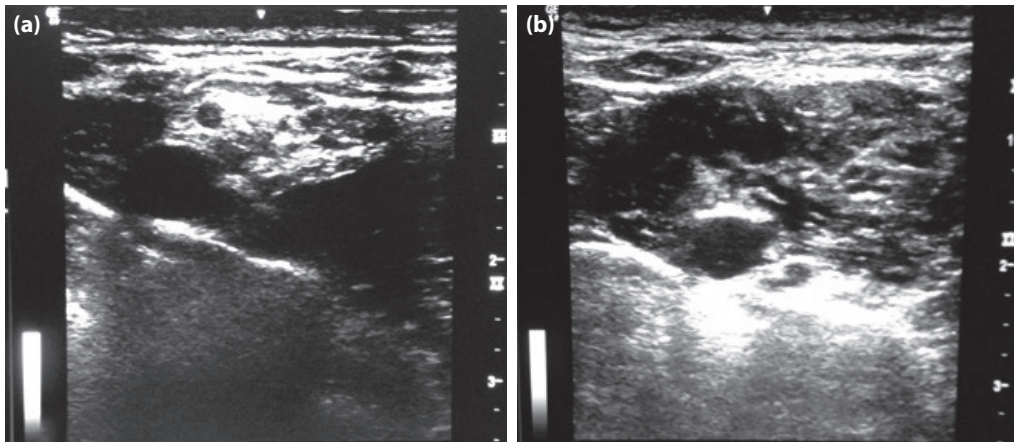


Figure 2. (a) Subclavian artery and anterolaterally identified brachial plexus as a collection of hypoechoic oval structures during supraclavicular brachial plexus block. (b) Observation of LA spread within the brachial plexus sheath.

technique. The correct tip position was confirmed within the brachial plexus sheath by US-guidance and a motor response of the forearm and hand by nerve stimulator (0.5 mA, 0.1 msec). After negative aspiration for blood, the mixture of 10 mL 2% lidocaine and 15 mL 0.5% bupivacaine was injected incrementally in fractionated doses with frequent aspiration. The local anesthetics (LA) spread was observed and SCB was completed (Figure 2b). The patient did not report any paresthesia or discomfort during the procedure.

Block success was assessed as complete sensory loss to pinprick with a paper clip on the skin over the right arm, forearm and hand at 30 minutes and ability to complete surgery with IV sedation using infusion of 0.5 µg/kg/h dexmedetomidine maintaining spontaneous breathing via an oxygen mask. During the surgery, firstly hyperpigmented areas were excised. Secondly, two neurofibromas that originate

from lateral cutaneous nerve of forearm (11.8x4.3 cm) (branch of musculocutaneous nerve) and inferior lateral cutaneous nerve of arm (8x3 cm) (branch of radial nerve) were both excised (Figure 3). Surgery was completed uneventfully in 60 minutes with no pain. Motor and sensorial blockage wore off at 12th and 16th hours; respectively. Post-operative analgesia consisted of IV paracetamol 2x1 g resulted in adequate pain control during the first 48 hours (NRS≤3). She did not experience any surgery- or anaesthesia-related post-operative complications. After obtaining written informed consent from the patient and her parents for publication of this report, the patient was discharged on postoperative day 2.

Discussion

NF-1 patients appear with several potential challenges including potential airway difficulty, abnor-



Figure 3. Patient's right arm and forearm after excision of neurofibromas and localised skin hyperpigmentations.

mal spinal anatomy, and peripheral neurofibromas for GA, RA and PNBs.^[4,5] Besides routine physical examinations, neurologic exams and radiologic imaging assessments as computerized tomography and magnetic resonance imaging must be complete in every case. Possible cranial mass and midline shift, increased intracranial pressure, herniation, spinal neural sheet tumors should be taken into consideration. Then, anaesthetists should choose their anaesthetic technique balancing the benefits and risks of patient-specific procedures.

In this case, we preferred a PNBs technique as SCB to avoid possible post-operative GA-associated problems because of brain stem hamartomas seen on magnetic resonance imaging and preserved the spontaneous ventilation by administering only midazolam and dexmedetomidine for sedation.

The effectiveness of anaesthesia and analgesia accomplished with US-guided PNBs are well described. However; the efficacy and outcomes of PNBs in patients with NF-1 is still limited.^[6,7] Ambardekar et al.^[8] reported that there has been no evidence to show that NF itself is a contraindication to peripheral nerve block techniques. However, perineural LA injection in the presence of NF may have risks as ineffective spread, failed block, hemorrhage into the neurofibroma causing nerve compression, or spreading neurofibromatous cells through the needle's track. The use of US becomes more evident as it is a diagnostic tool for neurofibromas and a guide for interventional procedures. Therefore; in our case, we describe the clinical utility of US on the successful performance of SCB for surgery on the arm and forearm in a patient with NF-1.

Brachial plexus was decided to be blocked at the supraclavicular level either to achieve a sufficient block for surgical area or to provide a tumor-free level by walking away from neurofibromas placed at the axillary region and more distally. During the block performance, we performed a sonographic survey to identify possible lesions, to minimize injury to the neurofibroma and to reduce associated risks. No neurofibroma was recognized as there might not be any within the sheath or it/they might be unnoticed because of having hypoechoic nodular sonographic appearances exactly as brachial plexus trunks/divi-

sions at this level.^[8-12] Beggs described neurofibromas as well-defined solid hypoechoic masses that have faint distal acoustic enhancement.^[9] Reynolds et al.^[13] also reported that these tumors are often hypoechoic, with posterior acoustic enhancement. The Doppler function was used to highlight vasculature structures and US allowed us to insert the needle through the sheath without injuring the hypoechoic structures and see the LA spread. Blockade onset time and duration were definitely normal, it was successful both for anaesthesia and post-operative analgesia, and there was no complication.

Nerve stimulator-guided axillary brachial plexus block^[1] and US-guided sciatic nerve/popliteal block performances were reported previously in NF diagnosed patients,^[6-8] however, to our knowledge, there has been no data about SCB performance.

In conclusion; US-guided SCB can be used as an alternative to accomplish anaesthesia and post-operative analgesia in patients with NF-1. At this point, US guidance provided us success and avoidance of neurofibroma puncture. Nevertheless, it is hard to prove regional anaesthesia techniques' safety and efficacy in all NF cases, we recommend anaesthesia technique preferences to be individualized.

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