CASE REPORT

# Ultrasound-guided and Peripheral Nerve Stimulator-guided Interscalene Brachial Plexus Block in a Patient with Neurofibromatosis during the COVID-19 Pandemic

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## ABSTRACT

An 18-year-old female with neurofibromatosis presented for enucleation biopsy of a slow-growing soft tissue mass on her right anteromedial arm during the COVID-19 pandemic. Ultrasonography was used in the mapping of the brachial plexus and the identification of possible neurofibroma along peripheral nerves. An interscalene brachial plexus nerve block was successfully and safely achieved with ultrasonographic and peripheral nerve stimulator guidance. Ultrasound-guided and nerve stimulator-guided peripheral nerve blocks are safe and viable anesthetic management options for patients with neurofibromatosis, especially in this time of the COVID-19 global pandemic.

*Keywords: ultrasound, neurofibromatosis, von Recklinghausen disease, brachial plexus block, interscalene block, regional block*

## INTRODUCTION

Neurofibromatosis type 1 (NF1) also known as von Recklinghausen's disease or peripheral neurofibromatosis is an autosomal dominant neurologic disorder that has the propensity to form tumors involving ectodermal and mesodermal tissues. It is characterized by hyperpigmented (café au lait) spots, cutaneous and plexiform neurofibromas.[1] It also affects multiple organ systems that impacts the choice of anesthetic technique.[1]

The incorporation of ultrasound guidance in performing nerve blocks has contributed largely to its current popularity. By allowing direct visualization of structures, it has been shown to improve sensory and motor blockade, decrease the minimum effective volume of local anesthetic, decrease the need for intraoperative supplemental anesthetics, decrease the number of attempts, decrease the performance time, and decrease the complication rates.[2,3]

We report a case of an enucleation biopsy of a soft tissue mass over the right anteromedial arm of an 18-year-old female with a history of NF1 complicated by symptomatic cough despite a negative RT-PCR swab test for SARS-CoV-2, performed under ultrasound-guided interscalene nerve block with minimal sedation.

## CASE PRESENTATION

An 18-year-old 55-kg female previously diagnosed with NF1 presented with a slow growing soft tissue mass on the

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**Figure 1.** Soft tissue mass on the right anteromedial arm, with café au lait spots in the axillary area and arm.

anteromedial aspect of her right arm (Figure 1). The patient had no history suggestive of other organ system involvement of NF1. There were no previous anesthetic or surgical experiences. On preoperative assessment, the patient had a history of productive cough with no associated fever, colds, anosmia, ageusia, and a negative SARS-CoV-2 RT-PCR test result.

There were note of multiple café au lait spots over the upper extremities and trunk (Figure 2), positive Tinel's sign, and an unremarkable airway examination. MRI of the arm revealed a 10 cm x 10 cm x 10 cm encapsulated cystic mass with septations, medial to the right biceps brachii muscle, abutting but not enveloping neurovascular structures (Figures 3 and 4). An ultrasound-guided interscalene brachial plexus block combined with minimal sedation was planned for the enucleation biopsy of the large mass proximal to the antecubital fossa. Alternative plans were supraclavicular brachial plexus block with an intercostobrachial block or general anesthesia with provisions for difficult airway scenario.

Standard NPO guidelines were observed. Vital signs upon arrival in the OR were BP 110/70 mmHg, HR 75 bpm, RR 20 cpm, Temp 36.5°C and $SpO_2$ 98% on room air. Prior to nerve blockade, standard monitors (five-lead ECG, pulse oximeter, and noninvasive blood pressure cuff) were applied and supplemental oxygen at 4 lpm via Hudson

**Figure 2.** Multiple café au lait spots around the area of neck and shoulder, with the patient's face mask briefly removed.

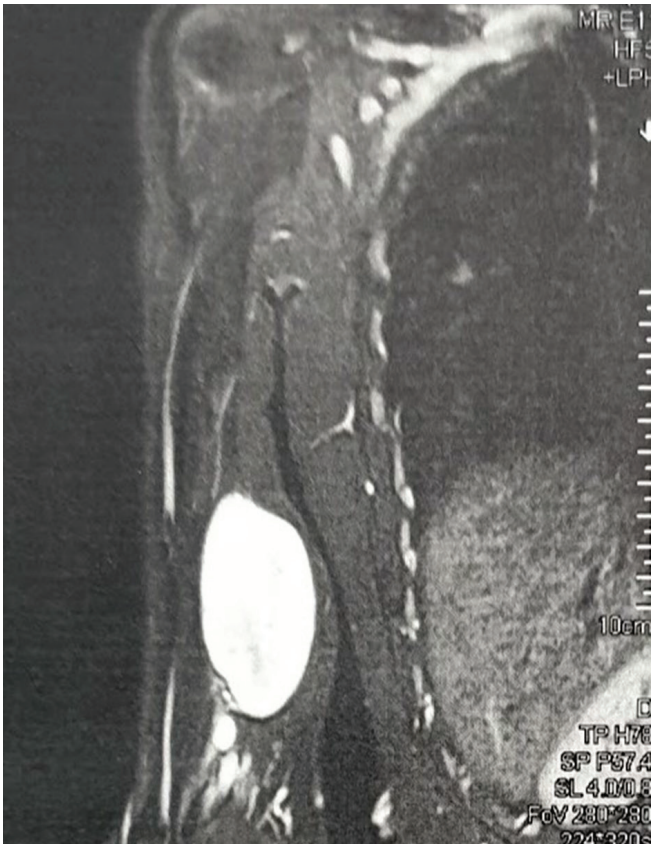

**Figure 3.** Coronal view of MRI showing encapsulated cystic mass medial to the right biceps brachii muscle.

**Figure 4.** Axial view of MRI showing encapsulated cystic mass with septations, medial to the right biceps brachii muscle and not enveloping neurovascular structures.

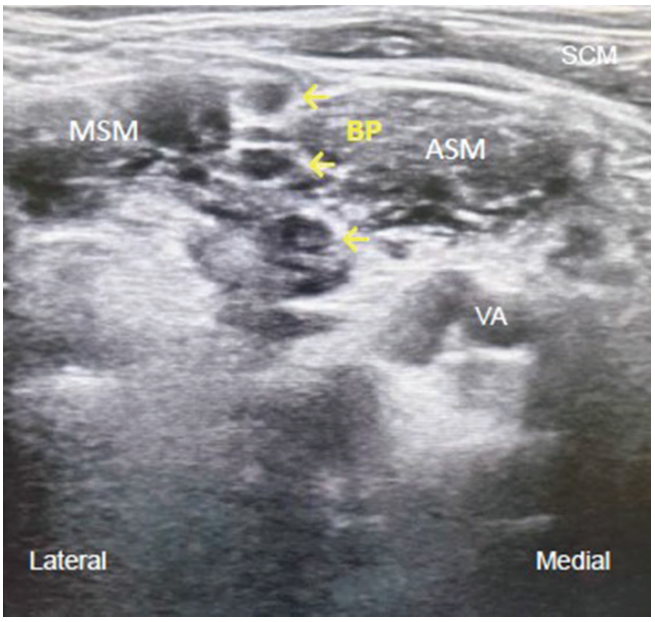


**Figure 5.** Preprocedural scanning showing interscalene brachial plexus trunks *(BP, yellow arrows)*, in the absence of hypoechoic structures with posterior acoustic enhancement *(BP, yellow arrows)*.

*ASM – anterior scalene muscle; MSM – middle scalene muscle; SCM – sternocleidomastoid; VA – vertebral artery.*

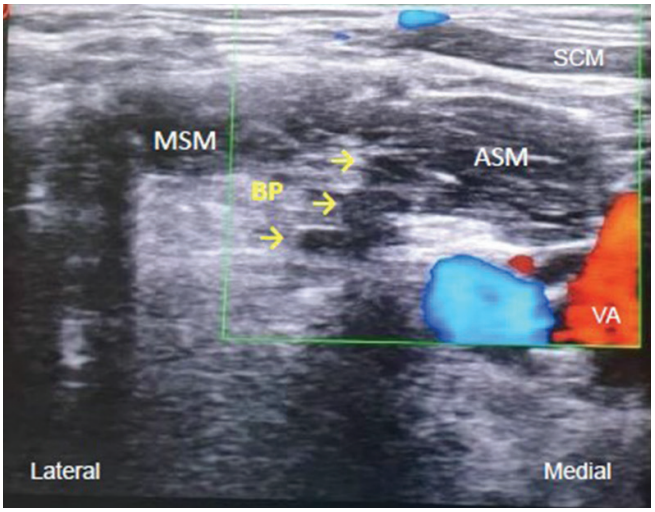


**Figure 6.** Color flow imaging showing vascular structures and interscalene brachial plexus trunks *(BP, yellow arrows)*.

*ASM – anterior scalene muscle; MSM – middle scalene muscle; SCM – sternocleidomastoid; VA – vertebral artery.*

mask was administered. Minimal sedation was achieved with midazolam 1 mg and fentanyl 25 mcg after completion of sign in and the regional block preprocedural checklist.

The patient was positioned supine, with the head turned to the contralateral side, and an elevated back rest at 45 degree angle. A systematic preprocedural sonographic survey in the cephalad and caudad orientation was then performed to plan a suitable approach for the nerve block. The ultrasound transducer (linear, 8-14 Hz, Wisonic Navi) was placed transversely on the right side of the neck, approximately three centimeters superior to the clavicle. Once the carotid artery has been identified, the transducer was moved laterally across the neck, to identify the anterior and middle scalene muscles, with the trunks of the brachial plexus located between them (Figure 5). Color Doppler was also used to identify vascular structures (Figure 6).

Under an aseptic technique, the skin was infiltrated with 2 mL of 2% lidocaine, and a 50 mm 22-gauge SonoPlex STIM needle (Pajunk) was utilized. A real-time view of the SonoPlex STIM needle tip, carotid artery, anterior and middle scalene muscles, and the brachial plexus was maintained at all times during the block (Figure 7). The brachial plexus in the ultrasound-guided interscalene approach was further confirmed through the use of nerve stimulation (0.5 mA, 0.1 msec, 2 Hz) with a motor response of the right deltoid and triceps muscles accepted as successful localization of the brachial plexus. A total of 17 mL of 0.5% ropivacaine was injected with intermittent negative aspiration for blood. The local anesthetic spread circumferentially around the trunks of the brachial plexus, as noted on ultrasound evaluation, with the patient not reporting any paresthesia or discomfort during the procedure (Figure 8).

On examination, the patient had complete sensory loss to sharp pain and cold temperature, and heaviness of her right arm within ten minutes of the procedure. Intraoperatively, no additional sedation was provided. No response to surgical incision was elicited and she remained hemodynamically stable for the duration of the three-hour surgery, wherein a well-circumscribed soft tissue mass was excised completely with adequate margins. There was minimal blood loss. Intravenous ondansetron 4 mg was given 30 minutes prior to the completion of surgery. The patient was awake and oriented at the Post Anesthesia Care Unit (PACU) with residual sensory loss and motor weakness.

**Figure 7.** Transducer placement and needle insertion to obtain the desired ultrasound-image for an interscalene brachial plexus nerve block.

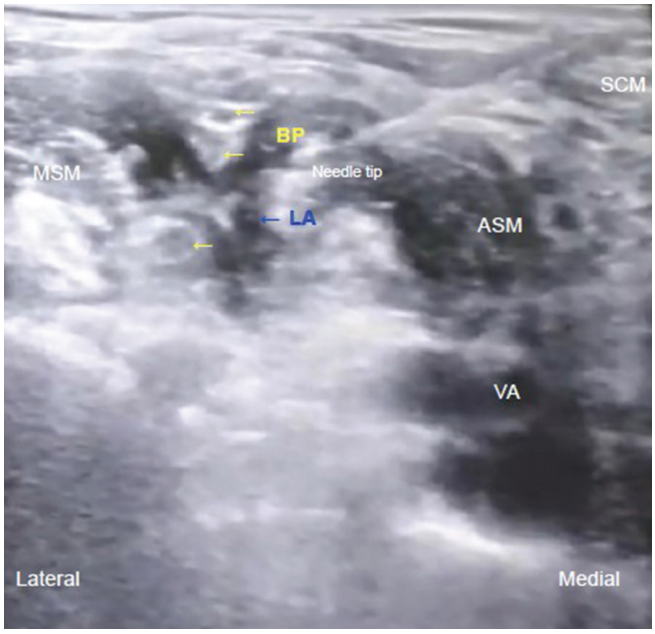


**Figure 8.** Actual needle placement in the interscalene groove with dispersion of local anesthetic *(LA, blue arrows)* surrounding the brachial plexus trunks *(BP, yellow arrows)*.

*ASM – anterior scalene muscle; MSM – middle scalene muscle; SCM – sternocleidomastoid; VA – vertebral artery.*

On post-operative visit, the patient reported return of baseline motor and sensory functions, eight and twelve hours following nerve block, respectively. Pain score at rest was pain numeric rating scale score of 1-2/10, which was adequately addressed with intravenous paracetamol and ketorolac.There were no reported redness, hematoma, or tenderness at the site of the regional block. The patient was discharged on the first postoperative day.

## DISCUSSION

Neurofibromatosis type 1 (NF1) has a birth incidence of 1 in 2,500-3,300 and a prevalence of 1 in 5,000. It is caused by an NF1 gene mutation.[1] This gene encodes a protein called neurofibromin which is an important negative regulator of Ras, a proto-oncogene that serves as a signaling molecule in cell growth and differentiation.[4] Reduced levels of neurofibromin due to NF1 gene mutations explains the development of tumors observed in patients with NF1.

NF1 has three clinically and histologically distinct subsets: cutaneous, nodular and plexiform. Cutaneous neurofibromas are benign tumors found within the dermis and occur in more than 95% of patients. Nodular neurofibromas arise in peripheral nerves and can become very large. Plexiform neurofibromas, the hallmark lesion of NF1, affect long portions of peripheral nerves and can infiltrate the nerve and surrounding tissue.[1] Apart from possible mechanical compression of surrounding structures and gross disfiguration, plexiform neurofibromas can rarely transform into malignant nerve sheath tumors.[1]

As NF1 affects ectodermal and mesodermal tissues, there are varying degrees of multiple organ system involvement posing several anesthetic considerations during surgical procedures. Neurofibromas in the oral cavity can lead to airway obstruction after anesthesia induction or failure of fiberoptic intubation eventually requiring emergency tracheostomy.[5,6] Vertebral deformities, like scoliosis and kyphosis, or neurofibromas in the spinal cord may make neuraxial anesthetic techniques technically difficult but it is not a contraindication and has been successfully performed.[7,8] Other considerations include undiagnosed neurofibroma in the respiratory and gastrointestinal systems, hypertension, pheochromocytoma or renal artery stenosis.[1] There are also reports of increased sensitivity to neuromuscular blockers among patients with neurofibromatosis resulting in prolonged episodes of apnea of unexplained mechanism. However, a large retrospective study concluded a normal response among NF1 patients.[9]

Several authors have reported the successful use of peripheral nerve blocks for patients with neurofibromatosis.

Rocco and Rosenblatt performed an ultrasound-guided popliteal sciatic nerve block for excision of a painful lesion in a patient's left lateral foot and ankle who requested to have a regional anesthetic.[10] The nerve identified appeared to have hypoechoic structures resembling vessels but it was confirmed through Doppler that these structures were not compressible and did not have evidence of blood flow. These hypoechoic areas may represent neurofibromas in the absence of direct visualization or histopathologic sampling. Desai and colleagues described an ultrasound-guided popliteal sciatic nerve block for open biopsy of left tibia.[11] They noted that clinically silent neurofibromas may involve the spinal cord and nerve roots in up to 40% of patients and emphasized preintervention anatomic mapping via CT and MRI. Shahid and Sebastian, likewise, reported a 66-year-old patient with NF1 who was successfully managed with peripheral nerve stimulator-guided femoral and sciatic nerve block for lower limb wound debridement.[12] Suvar and Mehaffey also reported the use of ultrasound-guided superficial and deep peroneal and tibial nerve blocks in the soft tissue biopsy of the foot in a 27-year-old male with a history of NF1 complicated by multiple cervical spinal fusion surgeries and significant airway issues.[13]

Upper extremity peripheral nerve blocks have likewise been used in the surgical management of NF1 patients. Salviz used an ultrasound-guided supraclavicular nerve block for excision of neurofibroma, while Bagam and his team used a brachial plexus block in a 60-year-old NF1 patients who needed an intramedullary interlocking nail for her humerus.[14,15]

Given this patient's neurofibromatosis throughout her trunk and upper extremities, and the demonstrated success and safety of peripheral nerve blocks in previous reports, a decision to proceed with an upper extremity nerve block was made. A systematic sonographic survey of all suitable block sites in this patient was initially performed prior to obtain valuable information on the morphology of the nerves, vessels, and presence of any peripheral neurofibroma previously described in literature as hypoechoic structures with posterior acoustic enhancement. This initial scanning demonstrated the absence of any perineural lesion, thus, supporting a safe window in performing an interscalene block, while avoiding neurovascular complications. Unlike the cited studies which utilized only either ultrasound or peripheral nerve stimulator guidance, both ultrasound and peripheral nerve stimulator was used in this patient to increase the safety of the performance of the procedure.

Being a COVID-19 referral center, the volume of surgical cases dramatically decreased primarily due to hospital rooms and personnel being redirected to the COVID-19 wards. A similar significant drop in surgical admissions was seen worldwide.[16] The patient presented with productive cough at the time of pre-operative examination which heralded a beginning upper respiratory tract infection (URTI). Pre-COVID-19 pandemic, this would have automatically warranted a deferral. The patient would have been discharged and will be admitted weeks after resolution of URTI. Considering the difficulties in securing an admission, the negative SARS-CoV-2 RT-PCR test result coupled with clear breath sounds were significant determinant factors in proceeding with the surgery.

The provision of regional anesthesia through ultrasound-guided upper extremity nerve block was favored despite COVID-19 being ruled out as it avoided the associated risk from aerosol-generating procedures from an infectious agent aside from SARS-CoV2 and airway manipulation-induced hyperreactive airway issues. The ultrasound-guided interscalene block also reduced the need for sedatives and hypnotics, which is helpful in light of expected anesthetic drug shortages during the COVID-19 pandemic.

## CONCLUSION

Peripheral nerve blocks are viable and safe anesthetic options for patients with neurofibromatosis, especially in this time of the COVID-19 global pandemic. Guidance through ultrasonography allows visualization and avoidance of lesions in patients while increasing the success of nerve identification and blockade. Nerve stimulation was likewise used as another confirmation of proper needle placement. However, there are several potential issues that still require further studies including possible alterations in nerve conduction, local anesthetic spread, onset and duration of nerve blockade. Hence, the decision to perform nerve blocks on this patient population should be individualized for its efficacy and safety.

### Declaration of Patient Consent

The authors certify that they have obtained appropriate patient consent forms granting use of the patient's images and clinical information for publication. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

### Statement of Authorship

KCS participated in the acquisition of data, drafting and revising both the original and final manuscripts, and approved the final submitted case report; NCB participated in the acquisition of data, drafting the original manuscript, and approved the final submitted case report.

### Author Disclosure

Both authors declared no conflicts of interest in preparing this article.

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