PAIN MEDICINE CASE REPORTS

ISSN 2768-5152

Volume 9, Number 6, pp. 323-327

# Role of Peripheral Nerve Blocks for Management of Chronic Pain From Inoperable Neurofibromatosis Type 1-Associated Neurofibromas: A Case Report

Kathryn Kuester, BS[1], Usman Latif, MD[2], Vinicius Tieppo Francio, MD[3], Dan T. Vu, MD[2], Kelsey Gustafson, MD[2], Alaa Abd-Elsayed, MD[4], and Christopher M. Lam, MD[2]

**Background:** Neurofibromatosis (NF) is a genetic condition resulting in tumor formation around neural structures, including peripheral nerves, spinal cord, and brain. Patients afflicted with NF can develop a constellation of symptoms ranging from cutaneous findings to sensory deficits and pain. When pain occurs, it could be associated with compression of neural structures or development of neuropathy itself.

**Case Report:** A 60-year-old woman with a history of NF Type 1 (NF1) presented to our clinic for evaluation of left arm pain. Despite several neuropathic medications and surgical procedures, she continues to have neuropathic arm pain associated with neurofibromas in her brachial plexus. Further surgery was advised against by her surgeons, and she underwent a supraclavicular nerve block with us, with sustained relief of her pain.

**Conclusions:** Here, we present a patient diagnosed with NF1 and neuropathic pain due to an inoperable brachial plexus neurofibroma burden in the left axilla, successfully managed by supraclavicular nerve blocks.

**Key words:** Neuropathic pain, neurofibromatosis, peripheral nerve block, neurofibroma

## BACKGROUND

Neurofibromatosis (NF) is a genetic condition characterized by tumors of the central and peripheral nervous systems. These tumors often cause abnormal growth in the tissue surrounding nerves, leading to pain, loss of function, and disfigurement. The 2 main types of NF are NF Type 1 (NF1) and NF Type 2 (NF2), both of which are autosomal dominant conditions that result in loss-of-function mutations of tumor suppressor proteins (1). Specific chromosomal abnormalities and affected proteins differ between NF1 and NF2, resulting in different clinical manifestations and complications (Tables 1 and 2). The most notable clinical differences between NF1 and NF2 include differences in common symptoms and population prevalence. NF2 is a sporadic disease that characteristically presents as bilateral vestibular schwannomas, resulting in hearing loss, tinnitus, or problems with balance (1). In NF1, the most common clinical manifestations are dermatologic, including characteristic cafe-au-lait macules and subcutaneous and plexiform neurofibromas.

The prevalence of chronic pain in NF1 patients is still unclear, with reported incidences of pain varying from 29% to 70% (2). Pain associated with NF1 can be constant or intermittent in nature and is typically exaggerated with movement or direct contact with painful

From: [1]Kansas City University College of Osteopathic Medicine, Kansas City, MO; [2]The University of Kansas Medical Center, Department of Anesthesiology and Pain Medicine, Kansas City, KS; [3]Washington University School of Medicine, Department of Anesthesiology, St. Louis, MO; [4]University of Wisconsin School of Medicine and Public Health, Department of Anesthesiology, Madison, WI

Corresponding Author: Christopher Lam, MD, E-mail: clam2@kumc.edu
Disclaimer: There was no external funding in the preparation of this manuscript.
Conflict of interest: Each author certifies that he or she, or a member of his or her immediate family, has no commercial association (i.e., consultancies, stock ownership, equity interest, patent/licensing arrangements, etc.) that might pose a conflict of interest in connection with the submitted manuscript.
Patient consent for publication: Consent obtained directly from patient(s).
This case report adheres to CARE Guidelines and the CARE Checklist has been provided to the journal editor.
Accepted: 2025-07-08, Published: 2025-10-31

Table 1. Demographics of neurofibromatosis type 1 vs type 2.

| | NF1 | NF2 |
|---|---|---|
| Incidence | 1 in 3,000 to 5,000 people worldwide | 1 in 25,000 people worldwide |
| Typical Age of Diagnosis | Early childhood | Between 18 and 24 years old |
| Gender | Occurs in men and women equally | Occurs in men and women equally |
| Race | No racial preference | No racial preference |
| Mode of Inheritance | Autosomal dominant | Autosomal dominant |
| Mutated Gene | 17q11.2 | 22q12 |
| Type of Mutation | Loss of function of tumor suppressor protein | Loss of function of tumor suppressor protein |
| Protein Affected | Neurofibromin | Merlin |

Abbreviations: NF1, neurofibromatosis type 1; NF2, neurofibromatosis type 2.

Table 2. Comparison table of neurofibromatosis type 1 vs type 2.

| | NF1 | NF2 |
|---|---|---|
| Most Common Tumor Types | Cutaneous neurofibromas (frequent) | Vestibular schwannomas (95%) |
| | Subcutaneous neurofibromas | Schwannomas of other cranial nerves, spinal roots, or peripheral nerves |
| | Plexiform neurofibromas | Gliomas |
| | Iris hamartomas | Multiple meningiomas (50% to 60%) |
| | Optic gliomas | Ependymomas (90%) |
| | Pheochromocytoma | |
| | Cerebral gliomas | |
| Affected Nerves | Cranial nerves, spinal nerves, and peripheral nerves | Cranial nerves, spinal nerves, and peripheral nerves |
| Affected Organ Systems | Nervous system, skin, eyes, bone | Nervous system and skin |
| Clinical Presentation | Cafe-au-lait macules | Hearing loss |
| | Skinfold freckling | Balance problems |
| | Learning disability | Flesh-colored skin flaps |
| | Scoliosis | Muscle wasting |
| | Arterial stenosis | Mononeuropathy or multiple cranial neuropathies |
| | Epilepsy | Visual impairment |
| | Macrocephaly | |
| | Short stature (10th to 25th percentile) | |
| | Vascular disease | |

Abbreviations: NF1, neurofibromatosis type 1; NF2, neurofibromatosis type 2.

neurofibromas (3). Current treatment of NF includes chemotherapy, radiation therapy, surgical intervention, and agents targeting the Ras signaling pathway (4). Opioid pain medication has been shown to have little to no effectiveness for the management of the associated pain (2,5). Buono et al (5) conducted a review of 255 patients diagnosed with NF1. Fifty-five percent of these patients reported being prescribed opioid medication in their lifetime, and 85% of those patients reported opioid treatment for pain management had "little to no effectiveness" (5). Due to the recurrent and progressive nature of plexiform neurofibromas, patients require a variety of options to treat their pain and manage the underlying disease.

Patients with NF1 may exhibit multiorgan involvement, affecting the patient's response to anesthesia or posing challenges in airway management (6). As a result, multiple studies (6-8) in the literature establish the safety and efficacy of local anesthetics for perioperative purposes in patients with NF1. However, few reports demonstrate successful use of peripheral nerve blocks for pain alleviation in patients with pain due to plexiform neurofibromas. We report the case of a patient with NF1 who presented with chronic arm pain secondary to an extensive inoperable disease burden in the left brachial plexus that is managed with successive supraclavicular nerve blocks and selumetinib.

## Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

## CASE PRESENTATION

Our case involves a 60-year-old woman with a relevant past medical history of hypertension, rheumatoid arthritis, and NF1 that was diagnosed in her forties. She

has had a long-standing history of recurrent plexiform neurofibromas. The patient has previously required multiple surgical excisions of a soft tissue mass surrounding her left rib and soft tissue masses on her left chest wall via thoracotomy.

Approximately one year before presentation, the patient began to develop left arm weakness. This progressively worsened over that period, and she developed paresthesias, moderate to severe pain, and decreased grip strength. At the time of presentation, she reported constant left upper extremity pain, rating it at an 8 to 10 in intensity by Numeric Rating Scale scoring, worsening with movement. This pain was limiting the function of her left arm and was negatively affecting her sleep. It was associated with numbness, paresthesias, and decreased grip strength. She was taking acetaminophen, oral diclofenac, gabapentin (600 mg in the morning and 300 mg at night), nortriptyline, and tizanidine to manage symptoms associated with her pain and rheumatoid arthritis.

Her neurological exam noted decreased pinprick sensation to her left lateral forearm, left thumb, and second through fourth fingers. She had mild weakness involving the left triceps muscle and severe weakness of the left abductor digiti minimi. The patient had 4/5 strength in her finger extensors bilaterally and 4+ to 5/5 in her first dorsal interosseous muscle and abductor pollicis brevis bilaterally. Her upper extremity reflexes, including her left biceps and triceps reflexes, were equal.

A positron emission tomography and computed tomography scan revealed several soft tissue nodules within the left axilla and lateral chest subcutaneous tissue with mild tracer uptake, compatible with her known neurofibromas. Further evaluation with magnetic resonance imaging of her left shoulder redemonstrated these soft tissue masses, with the largest measuring 3.7 cm by 2.8 cm (Fig. 1). Some of these masses were noted to track adjacent to some portions of the brachial plexus along the lower trunk.

The patient was subsequently evaluated by a general surgeon, an orthopedic surgeon, and a neurosurgeon for possible surgical excision of these neurofibromas, causing her symptoms. Ultimately, she was advised against surgical intervention as the risk of potential nerve injury and resultant disability outweighed the potential benefit. The patient was also evaluated by a radiation oncologist for possible radiation therapy; unfortunately, she was not a candidate for radiation therapy given the risk of malignant transformation of her neurofibromas in the setting of her relatively young age. She was recommended by her medical oncologist to start treatment with selumetinib, a mitogen-activated protein kinase (MEK) inhibitor associated with neurofibroma-size reduction and possible symptomatic improvement, and was referred to our clinic in the interim.

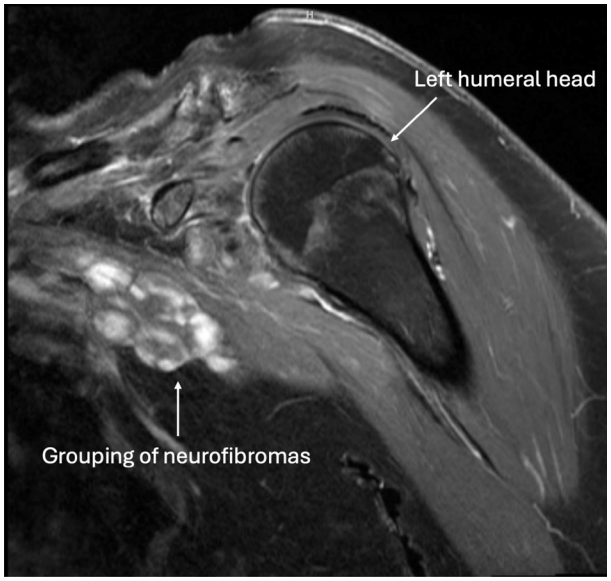


Fig. 1. Magnetic resonance imaging of the left upper extremity demonstrates neurofibromas compressing the brachial plexus.

The patient was seen in our clinic, and after a thorough explanation of the risks, benefits, and alternative treatments, informed consent was obtained for a supraclavicular nerve block. Under ultrasonic guidance, a left-sided supraclavicular nerve block was performed with 8 mL of 0.5% bupivacaine and 80 mg of triamcinolone (Fig. 2). At follow-up a month later, the patient reported symptomatic improvement after the nerve block with a 75% improvement in her pain, improved sleep, and increased function of her left upper extremity. Given this significant durable improvement, she opted to undergo a repeat supraclavicular nerve block 3 months after her initial nerve block, which was completed without issue with reproducible effects.

## DISCUSSION

Between 20% and 50% of patients with NF1 develop plexiform neurofibromas, often causing pain and/or disfigurement (6). One option for these patients

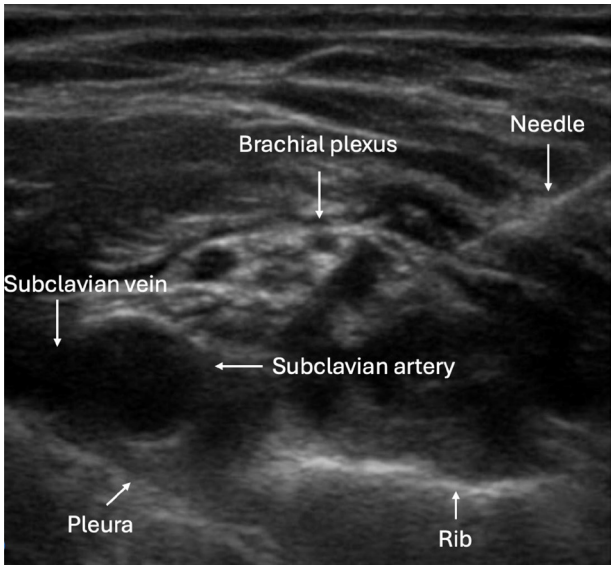


Fig. 2. Ultrasound-guided peripheral nerve block. Left-sided brachial plexus trunks are visualized.

is surgical resection, but in some instances, surgery is not recommended due to size, vascularization, and infiltration of the neurofibroma (2). Prior literature is scarce regarding the use of peripheral nerve blocks for pain relief in patients with neurofibromatosis (9). One previous case report conducted by Jevotovsky et al (9) describes a patient with a history of NF1 and extensive plexiform neurofibromas who presented with several years of buttock pain radiating into the left lower extremity. The patient was treated with perineural steroid injection (lidocaine and dexamethasone) at the level of the inferior gemellus with complete pain relief for up to one month following the procedure and no reported complications (9). Similar to this patient, the location of the nerve block was chosen to treat the nerves cranial to the area of the lesion to provide analgesia, as the pain was thought to be associated with the growth of the lesion itself into the nerve bundle.

Jevotovsky et al (9) reported the use of dexamethasone, a nonparticulate steroid and frequently used adjunct in peripheral nerve blocks. In our case, we used a corticosteroid as a peripheral nerve block adjunct. Triamcinolone, a particulate steroid, was used in conjunction with bupivacaine to prolong the effects of pain relief. Major concerns associated with steroid adjuncts include arteriolar occlusion due to precipitation of noncompatible admixtures (10). Triamcinolone is compatible with bupivacaine and thus diminishes the concern for precipitation (10).

Aside from the treatment with successive supraclavicular nerve blocks, the patient was also receiving chemotherapy treatment to reduce the tumor burden. In April 2020, the US Food and Drug Administration approved selumetinib for use in patients > 2 years with symptomatic plexiform neurofibromas who are not amenable to surgical intervention based on the findings of the Systolic Blood Pressure Intervention Trial. Selumetinib is a highly specific MEK1/2 inhibitor often prescribed for patients with inoperable plexiform neurofibromas. The mechanism of selumetinib is to block the activity of MEKs, halting a critical pathway for cell proliferation and survival (11). This results in reduced volume of plexiform neurofibromas and subsequent pain relief (2).

## CONCLUSIONS

Patients with NF1 and NF2 can develop a diverse spectrum of neurologic symptoms ranging from sensory deficits to neuropathies. When neuropathic pain occurs, treatment may be difficult, particularly in refractory cases where surgeries are not indicated and medications are ineffective. Few case reports have been published describing the utilization of peripheral nerve blocks for pain management in patients with this condition. Fewer studies have detailed the utilization of peripheral nerve blocks in conjunction with MEK1/2 inhibitors for management of neuropathic pain. Here, we illustrate a potential role for peripheral nerve blocks in conjunction with selumetinib for management of neuropathic pain in patients with NF1.

## REFERENCES


1. Ferner RE. Neurofibromatosis 1 and neurofibromatosis 2: A twenty first century perspective. *Lancet Neurol* 2007; 6:340-351.
2. Bellampalli SS, Khanna R. Towards a neurobiological understanding of pain in neurofibromatosis type 1: Mechanisms and implications for treatment. *Pain* 2019; 160:1007-1018.
3. Creange A, Zeller J, Rostaing-Rigattieri S, et al. Neurological complications of neurofibromatosis type 1 in adulthood. *Brain* 1999; 122:473-481.
4. Tamura R. Current understanding of neurofibromatosis type 1, 2, and schwannomatosis. *Int J Mol Sci* 2021; 22:5850.
5. Buono FD, Grau LE, Sprong ME, Morford KL, Johnson KJ, Gutmann DH. Pain symptomology, functional impact, and treatment of people with neurofibromatosis type 1. *J Pain Res* 2019; 12:2555-2561.
6. Das S, Singh N, Sahoo S, Mohanty CR, Goswami V. Ultrasound-Guided ankle block is a suitable alternative to other anesthesia modalities for a patient with neurofibromatosis having an isolated foot injury posted for emergency surgery: A case report. *Perioper Care Oper Room Manag* 2023; 33:100344.
7. Shahid M, Sebastian B. A safe alternative in neurofibromatosis for lower limb surgeries: Combined femoral and sciatic nerve block. *J Clin Diagn Res* 2015; 9:UD03-UD04.
8. Desai A, Carvalho B, Hansen J, Hill J. Ultrasound-Guided popliteal nerve block in a patient with malignant degeneration of neurofibromatosis 1. *Case Rep Anesthesiol* 2012; 2012:753769.
9. Jevotovsky DS, Callahan E, Libby J, Portugal SE. Successful treatment of peripheral nerve sheath tumor-related pain with perineural steroid injection: A case report. *Interv Pain Med* 2024; 3:100394.
10. Nestor CC, Ng C, Sepulveda P, Irwin MG. Pharmacological and clinical implications of local anaesthetic mixtures: A narrative review. *Anaesthesia* 2022; 77:339-350.
11. Galvin R, Watson AL, Largaespada DA, Ratner N, Osum S, Moertel CL. Neurofibromatosis in the era of precision medicine: Development of MEK inhibitors and recent successes with selumetinib. *Curr Oncol Rep* 2021; 23:45.