

ROLE OF PERIPHERAL NERVE BLOCKS FOR MANAGEMENT OF CHRONIC PAIN FROM INOPERABLE NEUROFIBROMATOSIS TYPE 1-ASSOCIATED NEUROFIBROMAS

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BACKGROUND

- Neurofibromatosis (NF) is an autosomal dominant genetic condition characterized by tumors of the central and peripheral nervous systems, often leading to pain and loss of function.
- Type 1 NF is more common than Type 2, which typically manifests as vestibular schwannoma. Dermatologic manifestations such as café-au-lait spots and cutaneous, subcutaneous, and plexiform neurofibromas are characteristic of Type 1 NF. (**Table 1**)
- Neurofibromas can compress nerves of the CNS and PNS, leading to chronic pain in patients with NF.
- Over 50% of patients with NF1 report significant pain.
- Current treatment of neurofibromatosis includes chemotherapy or radiation therapy, surgical intervention, and agents targeting the Ras signaling pathway.
- Here, we present the case of a patient with neurofibromatosis with extensive inoperable disease burden in the left brachial plexus successfully managed with successive supraclavicular brachial plexus blocks and selumetinib.

CASE

- Our case involves a 60-year-old female with relevant past medical history of hypertension, rheumatoid arthritis, and neurofibromatosis type I that was diagnosed in her 40s.
- She has had a long-standing history of recurrent plexiform neurofibromas, previously requiring multiple surgical excisions.
- Approximately one year prior to presentation, she developed progressive left arm weakness, paresthesias, moderate to severe pain, and decreased grip strength.
- Imaging evaluation revealed soft tissue nodules compatible with neurofibromas within the left axilla compressing elements of her brachial plexus.
- Magnetic resonance imaging (MRI) of her left shoulder redemonstrated these soft tissue masses, with the largest measuring 3.7 by 2.8 centimeters. Some of these masses were noted to track adjacent to some portions of the brachial plexus along the lower trunk. (**Figure 1**)

CASE

- She was evaluated by a general, orthopedic, and neurological surgeon, along with a radiation oncologist, who all determined that the risk of potential nerve injury and resultant disability outweighed any potential benefit of intervention. She was then referred to our clinic.
- After evaluation and informed consent, we performed a left-sided ultrasound-guided brachial plexus nerve block via the supraclavicular approach with 0.5% bupivacaine and triamcinolone.
- 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 brachial plexus block three months after her initial nerve block with reproducible effects.

DISCUSSION AND CONCLUSIONS

- Current treatment for NF1-related pain include over-the-counter medications, prescription pain medication, and size reduction/surgical removal of plexiform neurofibromas.
- Many patients with NF-1 develop plexiform neurofibromas, often causing pain and/or disfigurement.
- In many cases, surgery is not recommended due to size, vascularization, and infiltration of the neurofibroma.
- In our case, we successfully performed successive supraclavicular brachial plexus blocks for pain management in the setting of extensive plexiform neurofibromas of the brachial plexus that were not amenable to resection.
- Our case highlights the importance of multidisciplinary management of NF1-related pain and symptoms with the utilization of selumetinib in conjunction with successive peripheral nerve blocks.

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	NF1	NF2
Most common tumor types	Cutaneous neurofibromas (frequent) Subcutaneous neurofibromas Plexiform neurofibromas Iris hamartomas Optic gliomas Pheochromocytoma Cerebral gliomas	Vestibular schwannomas (95%) Schwannomas of other cranial nerves, spinal roots, or peripheral nerves) Gliomas Multiple meningiomas (50-60%) Ependymomas (90%)
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	Café-au-lait macules Skinfold freckling Learning disability Scoliosis Arterial stenosis Epilepsy Macrocephaly Short stature (10 th to 25 th percentile) Vascular Disease	Hearing loss Balance problems Flesh colored skin flaps Muscle wasting Mononeuropathy or multiple cranial neuropathies Visual impairment

Table 1 Comparison Table of Neurofibromatosis Type 1 vs Type 2

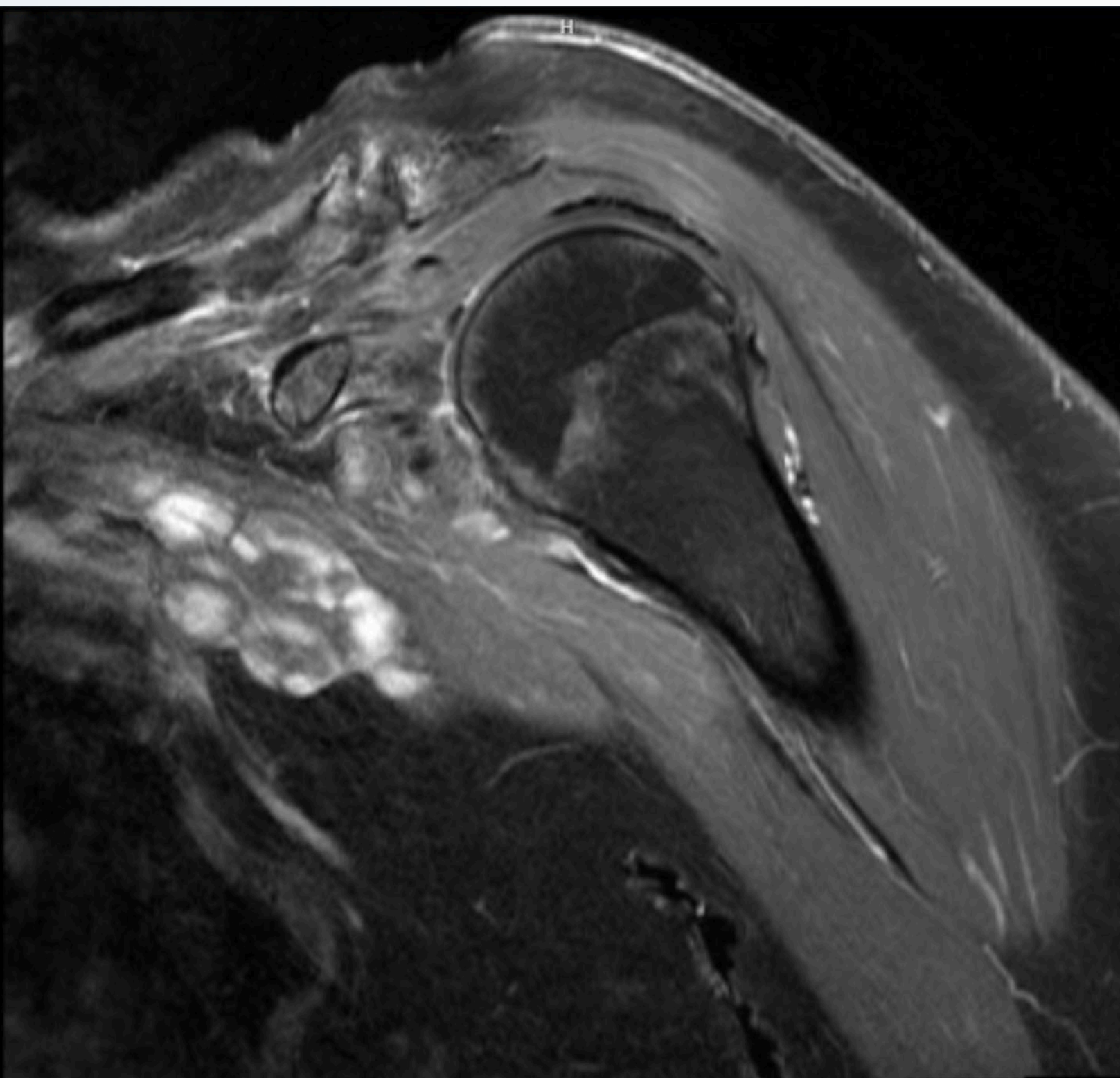


Figure 1. Magnetic resonance imaging of left upper extremity demonstrating neurofibromas compressing brachial plexus

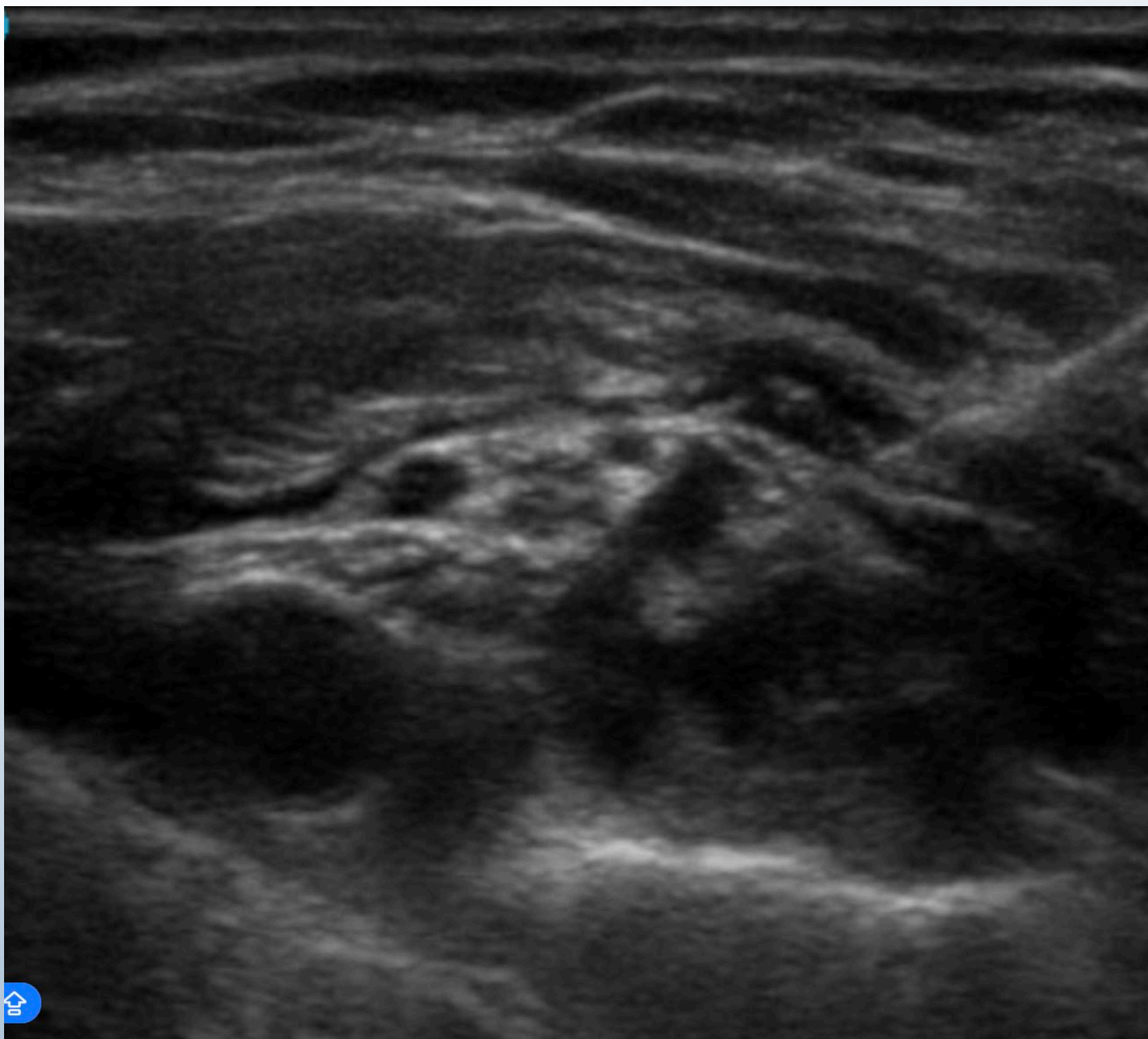


Figure 2. Ultrasound-guided nerve block of brachial plexus via supraclavicular approach