Ultrasound-Guided Percutaneous Peripheral Nerve Stimulation for the Treatment of Chronic Intractable Pain Originating From a Lipofibromatous Hamartoma of the Median Nerve

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This case report presents an application of peripheral nerve stimulation to the median nerve to treat a patient with intractable pain due to a lipofibromatous hamartoma of the left upper extremity. Ultra high-frequency ultrasound was used to determine the boundaries of the hamartoma. The patient then underwent an ultrasound-guided implantation of 2 stimulator electrodes distal to the elbow along the median nerve with stimulation coverage achieved at 1.2 and 1.4 mA, respectively. After an uneventful procedure, the pain score immediately decreased from 9 out of 10 to less than 6 on a numeric rating scale. Two weeks after the procedure, the patient reported substantial pain relief, with an average pain level of 5 to 6 out of 10. Twelve months after implantation, the patient maintained significant pain relief, rating her average pain level as a 4 to 6 out of 10. Placement of a percutaneous peripheral nerve stimulator was safe and effective with no adverse events being reported at the 12-month follow-up. *(J Hand Surg Am. 2021;46(3):250.e1-e5.*

Key words Case report, chronic pain, neuropathic pain, percutaneous nerve stimulation, peripheral nerve stimulation.

Online Exclusive

A lipofibromatous hamartoma (LFH) is an uncommon, benign fibrofatty tumor composed of a proliferation of mature adipocytes within the epineurium and perineurium of peripheral nerves, which forms a palpable mass. It affects the median nerve in 66% to 80% of cases, causing pain, sensory, and motor deficits in the affected nerve distribution.[1] Although LFH was first described in the English literature in 1953, this entity and its relation to carpal tunnel syndrome were only accurately described in 1969 by Johnson and Bonfiglio.[2] Until the early 2000s, fewer than 60 cases had been documented in the English literature.[1] Although many new reports have been published since then, the diagnosis remains infrequent, which explains why a lack of consensus exists among the medical community as to what the best care for the condition is.

The outcomes and prognosis of surgical treatment for LFH have ranged from loss of sensory and motor

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function to full return of both sensory and motor function. Numerous factors seem to affect prognosis, including the degree of involvement, the size of tumor burden, the age of the patient, and surgical technique.[3,4]

Over the past 2 decades, electrical neuromodulation techniques have emerged as a viable technical approach in the treatment of medically refractory neuropathic pain. Among the different types of available neuromodulation techniques, percutaneous peripheral nerve stimulation (PNS) is the least invasive, and recently has been gaining momentum in terms of the development of new indications. It may even be particularly effective as a standalone therapy when the pain is localized to a part of a single extremity.[5,6]

Several authors have hypothesized that pain relief from PNS, which the patient feels as a nonpainful or pleasant paresthesia in the painful area, is mediated by orthodromic stimulation of non-nociceptive Aβ fibers present in the free nerve endings of the peripheral nervous system. This stimulation subsequently leads to the activation of the same interneurons that are involved in the processing and transmission of nociceptive information by peripheral Aδ and C nerve fibers in the superficial layers of the dorsal horn of the spinal cord.[6,7]

This case report describes the case of a 50-year-old patient who underwent an ultrasound (US)-guided implantation of a percutaneous PNS along the course of the left median nerve to treat chronic, intractable neuralgic pain in the left upper extremity due to LFH.

CASE DESCRIPTION

A 50-year-old woman presented at our Pain Medicine outpatient clinic for further evaluation of chronic, severe, intractable pain of the left upper extremity after being diagnosed with LFH of the left median nerve during surgical release of the carpal tunnel.

During the first postsurgical follow-up with the surgeon, the patient reported continued pain and weakness in the distal part of the hand and fingers. For this reason, she eventually underwent magnetic resonance imaging 6 months after the surgery, which revealed an area of fibrofatty proliferation expansion of the median nerve. This area, which was at least 8.6 × 2.4 × 1.1 cm, extended from the distal forearm proximal to the field of view through the carpal tunnel and distally beyond the axial field of view, appearing to end at the mid-metacarpal level on the sagittal views.

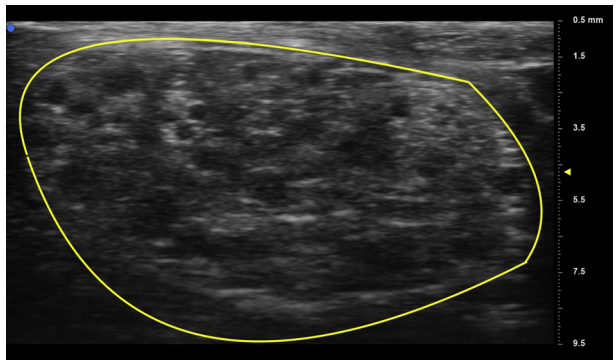

FIGURE 1: Transverse ultra high-frequency US image (at 50 MHz) of the left median nerve shows proliferation of mature adipocytes within the epineurium and the perineurium of the nerve. Yellow circle, boundaries of the LFH to the unaffected median nerve.

At presentation, the patient reported maintenance of the symptoms she felt before the surgery: left upper extremity pain that she rated as an 8 to 9 out of 10 on the numeric rating scale (NRS), described as sharp, shooting, squeezing, and throbbing. She also described worsening during nighttime and with using her left hand, as well as loss of sensation and weakness in the distal part of the hand and fingers, which she felt was greater in the palmar aspect of her thumb, index, and middle fingers. The physical examination was notable for a nontender, well-healed, midline proximal palmar scar. In addition, the patient presented with a positive Tinel sign along the midline volar distal forearm, from approximately 4 cm proximal to the carpal tunnel and extending throughout the carpal tunnel area. There was no intrinsic muscle or thenar atrophy The patient demonstrated diminished pinprick sensation in the volar aspect of the thumb, index, and middle fingers, with normal proprioceptive testing. At this point, the symptoms were so severe that the patient inquired about a possible amputation of her hand.

Using ultra high-frequency US (at 50 MHz), the boundaries between the LFH and the unaffected nerve were determined (Figs. 1, 2) and a US-guided block of the median nerve was performed. This proved to be successful in ameliorating the pain in the forearm, hand, and fingers. After the success of the block was established, the patient underwent a US-guided implantation of a percutaneous PNS (Bioness Stimrouter; Bioness, Valencia, CA) (Fig. 3) parallel and in close proximity to the left median nerve. Two stimulator electrodes were implanted

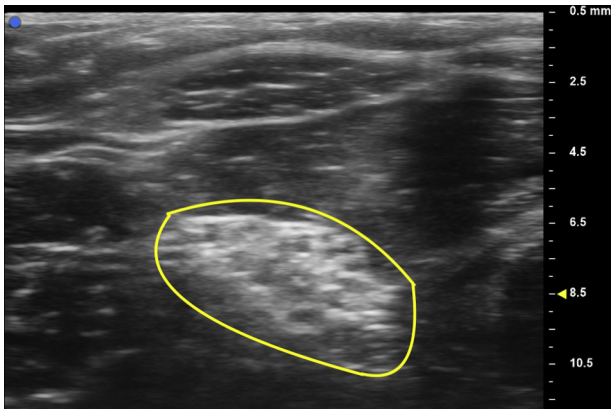

FIGURE 2: Transverse ultra high-frequency US image (at 50 MHz) of the normal-appearing median nerve proximal to the LFH. Yellow circle, boundaries of the median nerve.

longitudinally just distal to the elbow but proximal to the LFH with excellent stimulation coverage of the nerve (as indicated by an area of paresthesia that fully overlapped with the patient's area of pain and muscle weakness) achieved at 1.2 and 1.4 mA, respectively (Figs. 4, 5).

Although stimulation coverage of the median nerve typically only requires 1 stimulator electrode, in this instance, the authors opted to implant 1 electrode on the palmar side and 1 electrode on the dorsal side of the nerve. This deviation from the standard technique was decided during the implantation procedure, after a single electrode proved to be insufficient to cover all painful areas. This decision was made taking 2 particular points into special consideration: first, the much larger than normal diameter/area of the patient's median nerve; and second, the nature of the hamartoma tissue itself and its possible different conductivity compared with healthy human nerve tissue.

After the procedure, the pain score immediately decreased from 8 to 9 out of 10 to less than 6 on the NRS. Two weeks after the procedure, the patient reported continued pain relief, with an average pain level of 5 to 6 out of 10, located mainly in the distal part of the hand and the volar aspect of the index and middle fingers. At this point, the patient described that the area of pain was fully overlapped by an area of paresthesia when the device was turned on and for 2 to 3 hours after she had turned it off. In addition, with the device turned on, the patient reported having a stronger grip, being able to grab and carry small objects in her hand. By this time, the patient was intermittently using the device for periods of 8 hours.

At the 6-month follow-up consultation, the patient had continued pain relief and reported no adverse

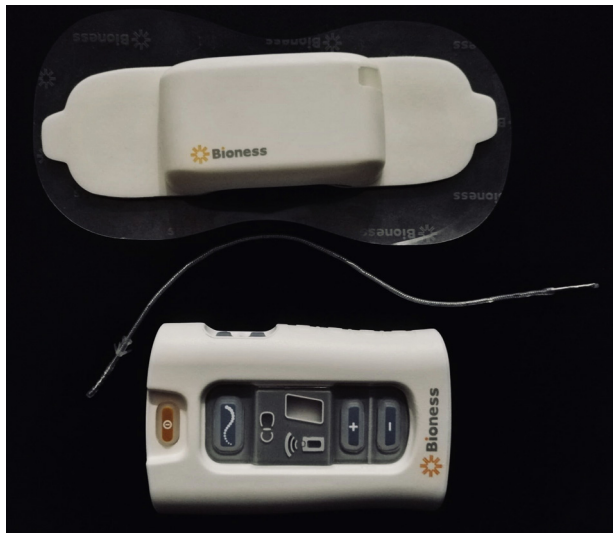

FIGURE 3: Image of a percutaneous PNS showcases the 3 components of the system: the external pulse transmitter (EPT) and electrode patch, the implanted lead, and the patient programmer. **Top** The EPT on top of the electrode patch. Whenever the patient wishes to trigger stimulation for pain relief, she places the electrode patch on her forearm's skin directly overlying the trajectory of the implanted lead. The EPT is then attached to the electrode patch, delivering neuromuscular electrical field stimulation through the electrode patch to the implanted lead. **Middle** One of the leads that was implanted parallel to the patient's left median nerve. **Bottom** The patient programmer, which the patient and the medical staff utilize to adjust the stimulation parameters after implantation.

events of any kind with the implanted PNS. During the months leading to this follow-up consultation, the patient had been using the PNS system on an intermittent basis, with periods of active stimulation ranging between 4 and 6 hours. At this time, the

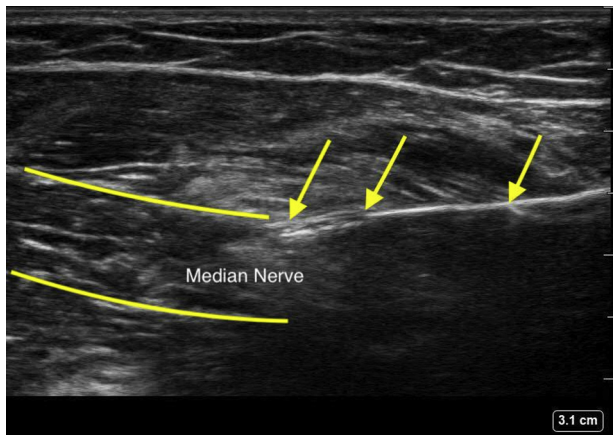

FIGURE 4: Longitudinal ultra high-frequency US image (at 50 MHz) of the stimulator electrode implanted in close proximity to the median nerve just distal to the elbow. Yellow arrows, Lead.

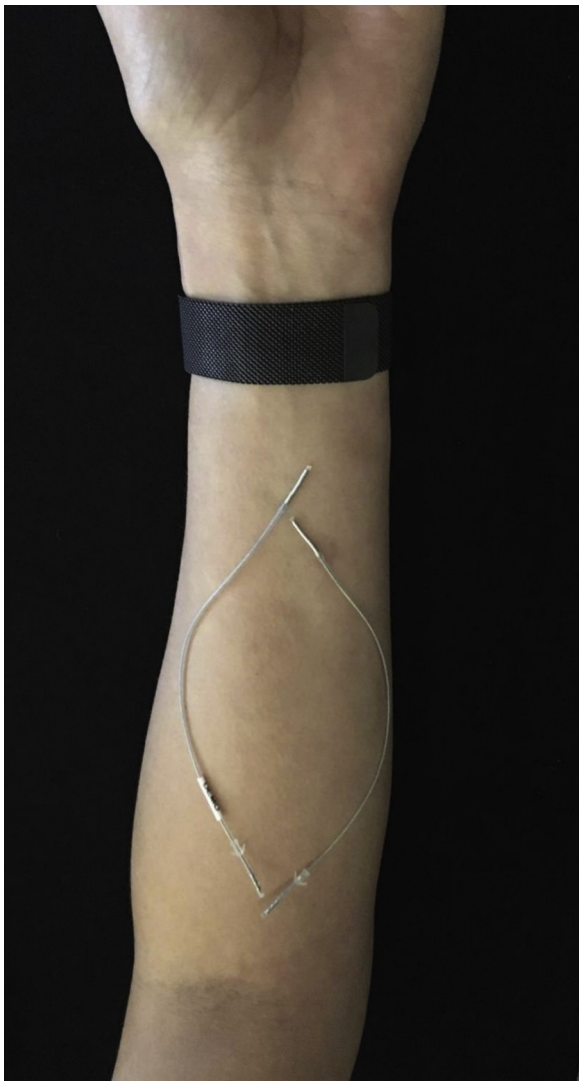

FIGURE 5: PNS placed over a volunteer's forearm skin shows the trajectory of the leads implanted in the patient.

physical examination was notable for a return of normal pinprick sensation in the volar aspect of the thumb, with continued diminished pinprick sensation in the volar aspect of the index and middle fingers, and normal proprioceptive testing. Given her tolerance of the device, as well as the benefit of the treatment up to that point, a determination was made to follow her on a 6-monthly basis to ascertain the long-term potential benefit of the PNS system. At the 12-month follow-up consultation, the patient maintained significant pain relief, rating her average pain level as a 4 to 6 out of 10 on the NRS, and reported no adverse events associated with the use of the device.

DISCUSSION

Pain modulation in the peripheral nervous system is mainly controlled by the nociceptive system. Primary nociceptive neurons in the periphery contain free nerve endings (Aδ and C fibers) that respond to noxious stimuli or tissue injury (eg, thermal or chemical). These stimuli originate nociceptive signals that travel into the spinal cord where they synapse with second-order neurons in the gray matter of the dorsal horn. Some of these second-order neurons contain axons that ascend the spinal cord and project to the brainstem or thalamocortical system where the conscious pain response is generated.[6,7] Another way nociceptors can mediate pain signaling is by the release of neuropeptides (eg, substance P, calcitonin gene–related peptide) at the terminal end of peripheral nerve fibers, leading to an increased inflammatory response, also known as neurogenic inflammation, and causing further local changes that magnify the pain response (eg, vasodilation, plasma extravasation, attraction of macrophages, degranulation of mast cells).[6]

The original explanation for the mechanism of action of PNS, based on the gate control theory by Wall and Melzack (1965),[8] postulates that orthodromic stimulation of non-nociceptive Aβ nerve fibers results in the activation of the interneurons of the superficial layers of the dorsal horn of the spinal cord, the same interneurons that are involved in the processing and transmission of nociceptive information by peripheral Aδ and C nerve fibers. This nonpainful stimulation provided by PNS inhibits these interneurons, therefore decreasing or interrupting the transmission of pain signals.[7] Furthermore, some studies have suggested that PNS may also directly change the excitability of peripheral nerve fibers, increasing the threshold for nociceptive stimulation to occur.[6,7,9] It is possible that this direct peripheral inhibition happens through an alteration in the local concentrations of biochemical mediators that enhance the pain response. By changing the local concentrations of neurotransmitters and endorphins, it is possible that PNS directly inhibits some of the mechanisms responsible for peripheral neurogenic inflammation.[6,7,9]

In this case, a 50-year-old woman underwent a successful implantation of a percutaneous PNS along the left median nerve to treat chronic, severe, intractable left upper extremity pain. This suggests that, even in cases of severe neuropathic pain, percutaneous PNS might prove to be a successful treatment option.

To date, mainly 2 different therapeutic strategies have been employed for the treatment of LFH of the median nerve: surgical decompression of the carpal tunnel and neurolysis. However, given the rarity of the condition and the associated lack of published research in the literature examining its treatment,

controversy still exists regarding the most suitable approach to the problem. In addition, the varying outcomes and prognosis reported in the few series described in the literature, ranging from variable loss of sensory and/or motor function to full return of both motor and sensory function, are testament to the challenging nature of providing lasting pain relief for this condition.[2,3,10]

This case report suggests that, for patients experiencing chronic, severe, intractable neuralgic pain in the upper extremity due to an LFH of the median nerve, implantation of a percutaneous PNS in close proximity to the nerve may provide continued and significant pain relief where other options have failed.

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