To Block or Not to Block: Role of Ultrasonography in Guiding an Anesthetic Plan for a Patient With Charcot-Marie-Tooth Disease

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A 31-year-old man scheduled for a fifth metatarsal head resection secondary to osteomyelitis presented to the preoperative holding area for placement of an ultrasound-guided popliteal nerve block as part of a multimodal pain management plan. During the preoperative evaluation, a medical history of Charcot-Marie-Tooth disease was noted. The patient had decreased range of motion and neuropathy in both lower extremities and required an assistive device when ambulating. Before placement of the block, a pre-procedure scan of the popliteal fossa revealed abnormal sonoanatomy of the distal sciatic nerve as well as the proximal tibial and common peroneal nerve branches. The surgeon was consulted regarding the ultrasonography findings, and the proposed block was abandoned. A field block proximal to the surgical site was performed under monitored anesthesia care, with an understanding that the case would convert to general anesthesia using a laryngeal mask airway if the procedure was not tolerated. The surgery was performed as planned without any difficulties, and the patient was transferred to the postanesthesia care unit. The postoperative course was uneventful, and the patient was discharged home.

Keywords: Charcot-Marie-Tooth, double-crush phenomenon, peripheral neuropathy, regional anesthesia, ultrasound.

The integration of ultrasonography (US) into the specialty of anesthesia continues to expand. The benefits of ultrasound guidance when performing peripheral nerve blocks are well documented,[1] and more recently authors have demonstrated the benefits of incorporating US for the placement of neuraxial blocks and perioperative point-of-care testing.[2,3] The ability to see key anatomic structures beneath the skin in real time provides practitioners with information to make clinical decisions in the patient's best interest.

As the patient population continues to age, the number of associated comorbidities will surely increase. Charcot-Marie-Tooth (CMT) disease is the most common inherited neuropathy and one of the most common inherited diseases in humans.[4] Although several variations of CMT exist, all are characterized by distal muscle atrophy, weakness, and sensory loss with reduced muscle tendon reflexes.[5] In recent years, US has emerged as a modality to assess changes in peripheral nerve structure. There are numerous anesthetic implications in the care of patients with CMT, such as increased sensitivity to nondepolarizing neuromuscular agents and potential hyperkalemia with succinylcholine administration[6]; thus, determining the best anesthetic plan can be difficult. Currently, there is not a clear consensus regarding the performance of peripheral nerve blocks in patients with preexisting neuropathy. It is theorized that these patients are at increased risk of a "double crush" phenomenon, where a second minor insult to an already compromised nerve can lead to permanent injury. Although multiple case reports[7-9] have demonstrated successful regional anesthetics in patients with CMT, current evidence is considered too sparse to allow for comprehensive recommendations regarding the best anesthetic approach.[10] In the specific case reported here, the lack of available information regarding the patient's medical history; unexpected abnormal sonoanatomy of the distal sciatic, tibial, and peroneal nerves; and preexisting neuropathy contributed to the decision not to perform the peripheral nerve block.

Case Summary

The anesthesia service was consulted to see a 31-year-old man scheduled for a left fifth metatarsal head resection secondary to osteomyelitis. The proposed plan was to place a popliteal nerve block as part of a multimodal pain management plan and to perform the case using monitored anesthesia care(MAC). During the preoperative evaluation, a medical history of CMT disease was noted. Physical assessment revealed moderate bilateral lower extremity neuropathy and weakness, as well as extensive inverted, plantar-flexed position of the feet that caused difficulty with ambulation and occasionally confined the patient to a wheelchair. The patient was treated for this disease at an outside facility, and access to those medical records was not available on the day of the procedure.

The patient was taken to the block suite and placed in the supine position with the operative extremity el-

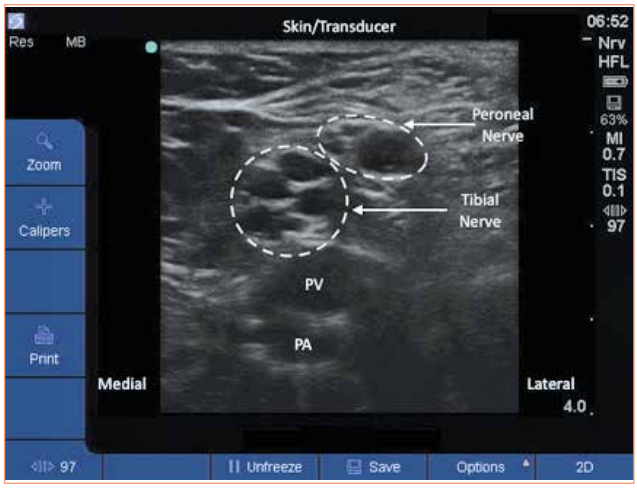

Figure 1. Sonoanatomy of Tibial and Peroneal Nerves in Popliteal Fossa in the Patient with Charcot-Marie-Tooth Disease

Note: See enlarged dark, hypoechoic fascicles of the nerves
Abbreviations: PA, popliteal artery; PV, popliteal vein.

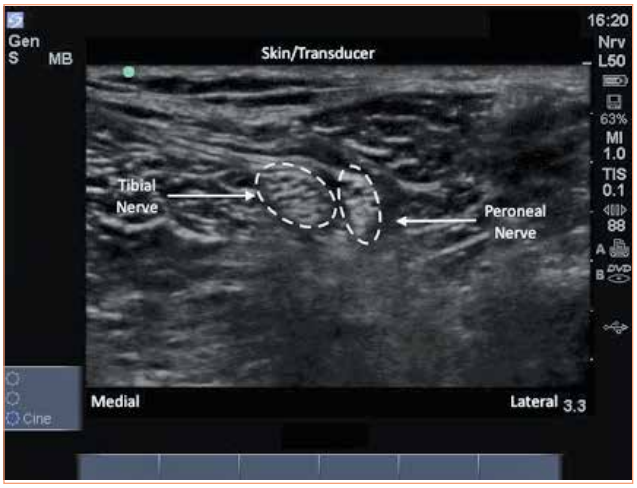

Figure 2. Sonoanatomy of Tibial and Peroneal Nerves in Popliteal Fossa in a Healthy Subject With No Known Disease

Note: Nerves display a honeycomb appearance commonly seen in peripheral nerves on ultrasound.

evated, permitting access to the popliteal fossa. After applying standard monitors, an S-Nerve ultrasound system (Fujifilm SonoSite Inc, Bothell, WA) with a 13-6 MHz HFL38 high-frequency linear array transducer was placed on the popliteal fossa. The popliteal artery and vein were identified. Dorsal to the vessels were multiple hypoechoic circles, initially thought to be collateral, aberrant vascular structures (Figure 1). Color flow Doppler US was applied to the image and confirmed the presence of flow in the artery and vein. Flow was not detected in any of the suspected collateral structures. Light pressure was applied to the fossa, resulting in compression of the vein, but not the artery nor any of the collateral structures. A pre-procedure scan was then performed caudad and cephalad along the popliteal fossa to identify the location of the sciatic nerve and the location at which the tibial and peroneal nerves divided. It was determined that the "collateral" structures were, in fact, enlarged nerve fascicles of the tibial and peroneal nerves.

Given the unexpected abnormal size of the fascicles on US, the surgeon was consulted, and the peripheral nerve block was abandoned. Infiltration of local anesthetic proximal to the surgical site by the surgeon using MAC was agreed on. If the patient was not able to tolerate the surgery, the plan was to convert to general anesthesia using a laryngeal mask airway without a paralytic agent. The case proceeded without incident, and the patient tolerated the procedure well. Following the case he was transferred to the postanesthesia care unit, before returning to the preoperative holding area. He was discharged without complaints of pain and did not require analgesics. On postoperative day 1, in a follow-up phone call, the patient stated that his pain was controlled with rest, ice, and oral medications.

Discussion

Charcot-Marie-Tooth disease is the most common inherited neuropathy, and with an estimated prevalence of one in 2,500, one of the most common inherited diseases in humans.[4] It commonly results in a progressive distal to proximal weakness with atrophy and sensory deficits starting in the feet and legs. The patient usually presents with complaints of lower extremity weakness, foot drop, and deformity, although other neurologic symptoms, such as optic atrophy, ataxia, and spasticity may also be seen. Assessment of these individuals demonstrates distal neuropathy and motor weakness, associated with muscular atrophy and absence of deep tendon reflexes. Charcot-Marie tooth disease has been classified into types 1 to 7, based on different genotypes and phenotypes.[4] Type 1 (demyelinating) is one of the most commonly encountered types of CMT.[4] Diagnosis has traditionally been based on electrodiagnostic findings of either nerve conduction slowing (representing demyelinating disease), or decreased compound muscle action potential amplitudes. Other clinical findings may be present as well.[4]

Over the past several years, US has emerged as a means to assess changes in peripheral nerve structure. Reports involving nerve entrapment syndromes, as well as hereditary, immune-mediated infectious and axonal peripheral neuropathies have been reported. Ultrasound imaging of peripheral nerves was first described by Fornage,[11] in which their features corresponded to their macroscopic and microscopic anatomy. The appearance of nerve tissue on US varies with the level at which the nerves are imaged. In cross-section, nerves that are centrally located, such as the cervical nerve roots and brachial plexus, appear more hypoechoic (dark) because of larger proportion of fascicles and reduced amount of connective tissue.[5] Conversely,

more distal peripheral nerves, such as the tibial and peroneal nerves in the popliteal fossa, demonstrate a bright honeycomb appearance, made by the dense fibrous hyperechoic connective tissue that surrounds small bundles of hypoechoic nerve fascicles[12] (Figure 2).

Heinemeyer and Reimers[13] were the first to describe the US findings of nerves in patients with CMT in 1999. They concluded that nerve diameter and echogenicity did not vary significantly compared with healthy subjects; however, this was partly attributed to the lack of higher resolution US that is currently available today.[5] Several authors recently conducted US assessments of patients with a diagnosis of CMT and found consistent enlargement of fascicles in both upper and lower extremity nerves.[14,15] Noto and colleagues[14] compared the cross-sectional area (CSA) of the median, sural, and auricular nerves and the C6 nerve root of patients with CMT to controls. They noted the CSA of all nerves and the nerve root were increased in CMT type 1 compared with controls. The authors also noted that with regard to the median nerve, the CSA correlates to disease severity and peripheral nerve function.[14] Goedee and colleagues[15] compared the US characteristics of the median, ulnar, peroneal, and posterior nerves and the brachial plexus between patients with CMT and hereditary neuropathy with pressure palsies. Both groups demonstrated nerve enlargement but without increased vascularization. However, the subjects with hereditary neuropathy with pressure palsies tended to have larger CSA at areas of entrapment, such as the carpal tunnel, whereas patients with CMT had larger fascicles at areas proximal to entrapment. Additionally, both groups were assessed for symptom duration and muscle strength according to the Medical Research Council (MRC) grading system. The authors found no significant correlation between fascicle size and the overall MRC score, nerve size, or number of nerves.[15]

The performance of regional anesthesia in patients with preexisting peripheral neuropathy remains debatable. It is thought that patients with preexisting neurologic disease are at increased risk of permanent nerve injury through a "double crush" phenomenon, in which a preexisting neurologic compromise is more susceptible to denervation when exposed to a second minor insult, as may occur during the perioperative period.[10] The American Society of Regional Anesthesia (ASRA) Practice Advisory on Neurologic Complications states "clinical evidence is too sparse to allow for definitive recommendations other than if a regional technique is chosen; extra precautions should be taken to minimize other surgical or anesthetic risk factors."[10] Patients with CMT present a particular problem to anesthesia providers as they have an increased sensitivity to certain nondepolarizing neuromuscular medications. Additionally, in severe cases, the administration of succinylcholine can result in hyperkalemia.[6] The incorporation of US into regional anesthesia allows the practitioner to see key anatomic structures beneath the skin, visualize needle movements in real time, and observe local anesthetic spread. These factors have resulted in faster onset times, longer block durations, better predictability of block success, and reduced needs for supplemental analgesia.[1]

In this particular case, a pre-procedure scan was used to rule out the presence of aberrant vascular structures and to confirm the enlarged fascicle size of the tibial and peroneal nerves. Because the patient was the first patient with CMT seen by the provider, the abnormal appearance of the tibial and peroneal nerves greatly influenced the decision to abandon the block. Given the patient's already limited mobility, it was feared any additional nerve injury to the peroneal nerve would result in permanent foot drop that could be devastating. Performing the infiltration proximal to the proposed surgical site afforded the patient the benefits of analgesia while minimizing the motor component of the regional procedure.

There are multiple reports in the literature describing successful regional anesthesia with minimal or no postoperative complications in patients with CMT.[7-9,16-18] Brock and colleagues[16] described the successful use of an epidural block with catheter placement in the management of an obstetric patient with CMT. The epidural was initially placed for labor management but was then used for cesarean delivery secondary to fetal intolerance. The patient experienced prolonged numbness and tingling of the right lower extremity and foot after the procedure. Although this sequela is consistent in patients with CMT, the authors noted "it is important to consider that neurologic complications following labor occur in the absence of anesthesia and are actually 5 times more common after childbirth itself independent of regional blockade."[16]

Ritter et al[17] described a subarachnoid block in combination with an ultrasound-guided sciatic nerve block as part of the anesthetic plan in a patient undergoing gastrocnemius recession with anterior tibialis tendon transfer. When describing the block, the authors noted, "Per ultrasound, the anatomic relationships, nerve structure, and size appeared as they do in normal patients without CMT."[17] The patient required minimal narcotic on postoperative day 1 and received twice daily treatment with oral diclofenac for analgesia before discharge on postoperative day 4. The authors found "the use of neuraxial blockade in combination with ultrasound-guided peripheral nerve block to be a safe and effective method for providing anesthesia and post-operative analgesia in this setting."[17]

Barbary and colleagues[9] presented a case study of a rescue sciatic nerve block in a patient with CMT experiencing severe postoperative pain following realignment surgery with multiple foot and ankle osteotomies under general anesthesia. Following an ultrasound-guided sciatic block in the midthigh, the patient noted relief within 15 minutes, and the effects lasted for 10 hours. No

neurologic complications were observed at the 6-month follow-up. During the scan, the authors noted the sciatic appeared as a "hyperechoic [bright] structure, with some small rounded uncompressible hypoechoic [dark] areas, similar to those described in neurofibromatosis."[9]

Bui and Marco[7] described a supraclavicular block in a morbidly obese patient with multiple comorbidities, including CMT, undergoing arthroscopic rotator cuff repair and subacromial decompression. The block was accomplished with 30 mL of bupivacaine, and the case was performed using general anesthesia with propofol induction and desflurane. The authors noted that "save for a prolonged duration of analgesia lasting approximately 30 hr [hours], telephone follow-up revealed that the patient's complete recovery was unremarkable with eventual complete resolution."[7]

Dhir and colleagues[8] described 3 ultrasound-guided axillary brachial plexus blocks performed in patients with CMT because of limited motor response noted with nerve stimulation. In all 3 cases the surgeries proceeded without complications, and no exacerbation of the neurologic condition was noted in any patient.[7]

Schmitt et al[18] performed a small series of distal sciatic blocks with catheters in patients with CMT undergoing foot surgery. Using nerve stimulation under ultrasound guidance, the authors found that patients who received a nerve block had lower analgesic requirements than those who did not.[18] In addition, the block recipients were contacted to report their status by self-assessment approximately 1 year after surgery. There were no anesthetic-related complications reported.[18] In their article, Schmitt et al[18] noted, "Many anesthesiologists fear permanent neurological injury or aggravation of the underlying disease when performing regional anesthesia in patients with pre-existing neuropathy. In contrast, as far as the authors know, there is no case published documenting any adverse outcome following peripheral nerve block in CMT patients."

Conclusion

Currently the ASRA Practice Advisory on Neurogenic Complications offers no definitive recommendations for the performance of regional anesthesia in patients with preexisting neuropathy other than "if a regional technique is chosen; extra precautions should be taken to minimize other surgical or anesthetic risk factors."[10] Ultrasound guidance provides the anesthetist a tool to avoid limitations associated with landmark and nerve stimulation techniques. The decision in this case not to perform the peripheral nerve block was based on a lack of available information regarding the degree and extent of the patient's condition, concerns over the abnormal appearance of the nerves compared with those described in previous reports, and the substantial amount of neuropathy already exhibited by the patient. However, multiple authors have demonstrated successful outcomes following regional anesthetic administration in patients with CMT.

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