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Treatment of Acute Pain Crisis in a Patient with Sickle Cell Disease Using Continuous Regional Anesthesia: A Case Report

LCDR Henry R. Deyoung, MD[1], LT Blake A. Morgan, MD[1], and LCDR Jacob H. Cole, MD[2,3]

Background: Sickle cell disease (SCD) is a predictor of both chronic pain and acute pain crises (APC). Vaso-occlusive APC typically require an escalation of home analgesic therapy, often requiring inpatient large doses of opioid pain medications. The analgesia and distal vasodilation provided by regional anesthesia could be of theoretical benefit when treating patients with SCD who present with vaso-occlusive APC.

Case Report: This case describes the treatment of a patient with SCD who presents with upper extremity APC. Her pain crisis was otherwise refractory to medical analgesic therapy and was successfully treated using continuous supraclavicular regional anesthesia. The patient remained pain-free even after discontinuation of the supraclavicular catheter 72 hours after placement.

Conclusion: Regional anesthesia could be considered as a reasonable non-opioid treatment modality for refractory APC in patients with SCD.

Key words: Sickle Cell Disease, regional anesthesia, pain management, vasoocclusive, local anesthesia, inpatient, opioid sparing

BACKGROUND

Vaso-occlusive acute pain crisis (APC) is a common reason for inpatient admission in patients with sickle cell disease (SCD) and is often challenging to manage (1). The CDC estimates that sickle cell-related disease represents more than 113,000 hospitalizations per year and $488 billion in health care expenditures (2). Even with prompt recognition and aggressive multimodal pain management, patients often require escalating doses of opioids that place them at high risk of complications such as respiratory depression, sedation, constipation, and rebound hyperalgesia.

There are several studies that show promising adjuncts for the treatment of APC. These include the use of subanesthetic ketamine infusions, lidocaine infusions, magnesium, and possibly cannabinoids (3-5). In addition to these analgesic medications, peripheral nerve blockade (PNB) may have theoretical benefit in patients with a single point of pain from their APC or particularly painful presentation of this pathology.

In general, there is no one single generator of pain in a vaso-occlusive APC such as is seen in patients with SCD. Instead, it is a conglomeration of multiple different factors that lead to the ultimate presentation of an APC.

From: [1]Department of Anesthesiology and Pain Medicine, Navy Medicine Readiness and Training Command Portsmouth, Portsmouth, VA; [2]Department of Anesthesiology, Perioperative, and Pain Medicine, Brigham and Women's Hospital, Boston, MA; [3]Department of Anesthesiology, Uniformed Services University of the Health Sciences, Bethesda, MD

Corresponding Author: Jacob H. Cole, MD, E-mail: jcole0@bwh.harvard.edu
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When the sickled hemoglobin cells occlude capillary beds, nociceptive receptors are triggered along with inflammation due to local ischemic cellular injury. Factors that induce vasoconstriction, such as hypothermia and sympathetic activation due to pain, serve to worsen the vaso-occlusion resulting in a vicious cycle of positive feedback. Also, patients with SCD and recurrent APC tend to have some degree of central sensitization and hyperalgesic responses to pain that is only made more difficult to treat by prior chronic exposure to opioids and potential opioid tolerance.

As shown in Fig. 1, PNB has potential to treat many of these contributing factors to a vaso-occlusive APC. By blocking the nociceptive signal to the central nervous system, PNB can absolutely decrease nociception and minimize central sensitization. By minimizing nociception, PNB can also help to mitigate tolerance by decreasing the baseline pain signal generated by the crisis. Interestingly, PNB may even play a therapeutic role in treating APC due to its profound vasodilatory effect on the circulation in the limb distal to the blockade.

Indeed, there are case reports on the use of PNB in patients with SCD who presented with lower extremity APC (6,7). Here, we present a case of an adult patient with SCD who had an APC in the right forearm and who received continuous supraclavicular nerve blockade for 72 hours, which resulted in complete resolution of pain in that extremity. The patient has provided written consent for publication of this case, and written authorization has been obtained in accordance with the Health Insurance Portability and Accountability Act.

Fig. 1. Diagram of theoretical states that contribute to refractory pain in patients with acute pain crisis. Peripheral nerve blockade has the potential to stop vasoconstriction, block nociception, is not affected by tolerance, and can help to decrease central sensitization.

CASE

This case describes a 40-year-old woman with a history of SCD, venous thromboembolism, and mild asthma. She presented to the emergency department with severe pain in her bilateral forearms typical of previous APC episodes. This pain was refractory to her typical outpatient oral pain control regimen of sustained release oxycodone 80 mg 3 times daily and immediate release morphine 60 mg every 4 hours as needed for pain. At the time of presentation, she stated that her right forearm was much more painful than her left.

On admission, the patient was awake, alert, and appropriately oriented. Her upper extremities were tender to palpation and light touch, with the right being more tender than the left. Specifically, the pain and tenderness spread in a glove-like distribution from just distal to the elbow to the fingertips. Per report this was stereotypical of her past APCs. Despite this pain, she was found to have brisk capillary refill, and intact motor function and sensation throughout the bilateral upper extremities. Her radial pulses were strong bilaterally. She was clinically hypovolemic with dry mucus membranes and poor skin turgor on admission. Otherwise, the remainder of her cardiopulmonary and abdominal exams were unremarkable.

A full laboratory evaluation was obtained that found the patient to be in her baseline state of anemia (hemoglobin level of 8.6 g/dL). She was also found to have an elevated bicarbonate level, indicative of volume contraction. Chest radiography and bedside ultrasonography were unremarkable.

Following admission for pain control, her analgesic regimen was escalated to include patient-controlled analgesia (PCA) with intravenous hydromorphone 0.4 mg given up to every 15 minutes. Other adjunctive pain medications started at the time of her presentation included oral gabapentin, acetaminophen, and ibuprofen, as well as intravenous magnesium repletion. These interventions provided limited relief and she was also started on a ketamine infusion at 5 mcg/kg/min and a lidocaine infusion at 30 mcg/kg/min with minimal relief. Despite aggressive titration of these medications, none ultimately controlled her pain.

On the morning of the 10th day of her admission, she

reported that the pain in her right forearm was much worse than her left, and she was offered a placement of a right-sided supraclavicular peripheral nerve block catheter. Supraclavicular blockade of the brachial plexus was chosen to provide a high likelihood of adequate sensory blockade throughout the right upper extremity with a planned continuous peripheral nerve block catheter. After explaining the potential risks and benefits of this procedure, an ultrasound-guided PNB catheter was placed in the right supraclavicular fossa in the region of the divisions of the brachial plexus under sterile technique without complication. Prior to placement of a 19-gauge multi-orifice, wire-reinforced catheter (B. Braun Medical, Melsungen, Germany), the space was dilated via a 17-gauge Touhy needle (B. Braun Medical, Melsungen, Germany) with 10 mL of 0.5% ropivacaine. The catheter was secured using skin adhesive (Dermabond, Ethicon, Raritan, NJ) at the puncture site and a sterile adhesive dressing. Programmed intermittent boluses of 7 mL of 0.1% ropivacaine were then given via a programmed CADD-Solis Epidural Infusion pump (Smiths Medical, St. Paul, MN) every 45 minutes.

Given the novel nature of this therapy for vaso-occlusive APC, this approach to regional anesthesia was chosen to allow a stepwise assessment of analgesia. First, 0.5% ropivacaine was administered as a single-injection trial. However, with the success of the trial, the PNB catheter was placed concurrently with the injection of 0.5% ropivacaine to facilitate a follow-on continuous regional technique using 0.1% ropivacaine. Upon assessment of the patient 20 minutes following the single-injection trial of 0.5% ropivacaine, her pain had entirely resolved in her right arm. The decision was made at that time to utilize the in-situ PNB catheter for 72 hours. If the patient had not had significant relief with the single-injection trial, the catheter would have been removed at that time.

Following placement of the PNB catheter, the patient reported complete resolution of her pain in that extremity for the duration of its use. Following the planned removal of the catheter 72 hours after placement, the patient continued to report complete resolution of the pain in her right forearm for the remaining 8 days of her inpatient hospital stay. When prompted, she described satisfaction with the pain control that was provided from the regional nerve block technique. Of note, the lesser pain in her left forearm remained unchanged throughout this period.

DISCUSSION

PNB is often utilized for perioperative pain control. Benefits that include distal vasodilation, reduced pain, and reduced inflammation are well documented (8-12) and could have direct effects on the generators of pain in vaso-occlusive APCs. These effects may have theoretical benefit in APC in patients with SCD, potentially stopping the vicious cycle of vaso-occlusion, inflammation, pain, sympathetic activation, and further vasoconstriction.

The patient in this case report not only had complete pain relief while the PNB catheter was in place, but also for days after its removal. In the setting of unchanged pain in her contralateral forearm that did not receive the PNB catheter, this suggests a potential therapeutic effect of regional anesthesia. The theoretical mechanism of this could be related to the vasodilation imposed by the sympathetic blockade of the extremity by the regional anesthetic technique. This vasodilation could lead to less vaso-occlusion and in turn break the pain cycle described above. This mechanism is theoretical but represents a potential step forward in the care of patients with vaso-occlusive APC. It may also prompt future investigations into the use of stellate ganglion blockade to achieve a similar vasodilatory effect. Most standard therapies for this condition are supportive, and future research into the therapeutic efficacy of regional anesthesia could be warranted given the marked and lasting response seen by the patient in this case report.

CONCLUSION

In conclusion, APC in patients with SCD is a difficult syndrome to treat, often requiring rapid escalation of opioid pain medication. In patients with SCD who have pain that is localized to a single joint or region and who are otherwise amenable to regional anesthetic technique, PNB could be considered for the treatment of APC.

Contributions

HD: Resident who performed the procedural technique, manuscript preparation

BM: Background research for manuscript, manuscript preparation

JC: Attending anesthesiologist who oversaw procedural technique, manuscript preparation

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