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Case Report

Peripheral nerve blocks in patients with Ehlers-Danlos syndrome, hypermobility type: a report of 2 cases☆

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Abstract Ehlers-Danlos syndrome is an inherited disorder of collagen production that results in multiorgan dysfunction. Patients with hypermobility type display skin hyperextensibility and joint laxity, which can result in chronic joint instability, dislocation, peripheral neuropathy, and severe musculoskeletal pain. A bleeding diathesis can be found in all subtypes of varying severity despite a normal coagulation profile. There have also been reports of resistance to local anesthetics in these patients. Several sources advise against the use of regional anesthesia in these patients citing the 2 previous features. There have been reports of successful neuraxial anesthesia, but few concerning peripheral nerve blocks, none of which describe nerves of the lower extremity. This report describes 2 cases of successful peripheral regional anesthesia in the lower extremity. In case 1, a 16-year-old adolescent girl with hypermobility type presented for osteochondral grafting of tibiotalar joint lesions. She underwent a popliteal sciatic (with continuous catheter) and femoral nerve block under ultrasound guidance. She proceeded to surgery and tolerated the procedure under regional block and intravenous sedation. She did not require any analgesics for the following 15 hours. In case 2, an 18-year-old woman with hypermobility type presented for medial patellofemoral ligament reconstruction for chronic patella instability. She underwent a saphenous nerve block above the knee with analgesia in the distribution of the saphenous nerve lasting for approximately 18 hours. There were no complications in either case. Prohibitions against peripheral nerve blocks in patients with Ehlers-Danlos syndrome, hypermobility type, appear unwarranted.
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1. Introduction

Ehlers-Danlos syndrome (EDS) is an inherited connective tissue disease of collagen production or organization that results in several organ derangements, most notably joint and skin hypermobility as well as tissue and vessel fragility. EDS is classified into 6 major and other types according to the Villefranche classification from 1997, with EDS, hypermobility type (EDS-HT) being the most common. Although the genetic cause is known for most of the subtypes of EDS, the genetic defect that causes EDS-HT has yet to be definitively elucidated. As a consequence, patients may sustain recurrent joint dislocations, instability, peripheral neuropathy, and pain along with increased risk of bleeding and hematoma formation [1-4]. Several reports advise against the use of regional

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anesthesia in these patients because of reported insufficient anesthesia with local anesthetics and possible hematoma formation [5-7]. There have been several reports of successful neuraxial anesthesia in patients with EDS, but only 2 regarding peripheral nerve anesthesia, both in the upper extremity [8,9]. This is a report of 2 cases of successful lower extremity peripheral regional anesthesia in 2 separate patients with EDS-HT presenting for orthopedic surgery.

2. Case report

2.1. Case 1

A 16-year-old adolescent girl presented for operative management of osteochondral lesions of the talus with osteochondral allograft transplantation. Consent to report her case was obtained from her mother after the procedure. She had been diagnosed several years earlier with EDS-HT based on generalized joint hypermobility and positive family history. Her mother also has EDS-HT and interestingly reported receiving successful regional anesthesia for shoulder surgery without complication. The patient reported a history of chronic joint instability with several joint dislocations, although did not report chronic pain in her joints. She denied any history of easy skin bruising or excessive bleeding with any prior procedures or with intramuscular vaccinations as a child. The benefits, risks, and alternatives of regional anesthesia, specifically that there is a potential higher risk of developing hematoma, were discussed with her and her mother. However, several mitigating steps would be taken during the procedure to include the use of ultrasound guidance, using needle approaches that avoid muscle and vessel trespass, prophylactic pressure application to the sites immediately after the procedure, monitoring for signs of hematoma, as well as placement of a continuous catheter for extended postoperative analgesia. The patient decided to proceed with regional anesthesia as her main anesthetic with intravenous sedation in the operating room. After obtaining informed consent, she was transported to our regional block area where standard American Society of Anesthesiologists monitors with supplemental oxygen were applied. She was placed in the prone position, and after a timeout was performed, 2 mg midazolam and 100 μg fentanyl were administered for sedation. Using an M-Turbo ultrasound with an HFL38x 13-6 MHz linear probe (FUJIFILM SonoSite, Inc, Bothell, WA), the sciatic nerve at its division into the common peroneal and tibial nerves was imaged in the popliteal fossa. A medial-lateral tract was identified from skin to nerve that was free of vessels (imaged with color Doppler) and muscle. After skin disinfection, the skin and subcutaneous tissues were carefully anesthetized with 2% lidocaine under ultrasound guidance using a 25G needle. Next, a 21G × 90 mm block needle (StimuQuik, Arrow Int'l, Inc, Reading, PA) was inserted in-plane and advanced toward the nerve. The common perineural sheath around both component nerves was visualized and penetrated with 0.5-1 mL aliquots of normal saline (3 mL total) demonstrating injectate spreading around both nerves within the sheath. Thirty milliliters of 0.5% ropivacaine with 1:400,000 epinephrine was injected with slight needle redirections to ensure complete circumferential spread around both nerves. The block needle was removed, and an 18G Tuohy needle was inserted into the perineural sheath along the same track as above. A catheter was placed and injection through the catheter could be seen spreading around both nerves. She was placed in the supine position for the femoral nerve block after an additional 2 mg midazolam and 100 μg fentanyl were given. The femoral nerve was imaged between the inguinal ligament and crease as it lay superficial to the iliopsoas muscle. Color Doppler showed no vascular structures in a lateral-medial in-plane needle path. The block was performed in the same manner as above with deposition of 15 mL of the above anesthetic solution above and below the nerve. She tolerated the procedure well, and pressure was held over the block sights. After 10 minutes, the block sights were reexamined with ultrasound with no evidence of deep hematoma; however, she did have small (<5 mm), superficial skin bruising at the needle puncture sites, not commonly seen in other patients. She endorsed progressive diminishing/absent sensation to light touch below the knee and was taken to the operating room where she underwent her procedure with intravenous sedation as she preferred not to be awake. She required no analgesics in the operating room or the postanesthesia care unit with a visual analog scale of 0/10 for pain. Her first analgesic request was 15 hours from the completion of the blocks. She was admitted overnight for postoperative observation and discharged the next morning with a continuous infusion of 0.2% ropivacaine at 10 mL/h. She removed her catheter on postoperative day 3 when the infusion was complete with return of normal motor and sensory function.

2.2. Case 2

An 18-year-old woman with EDS-HT presented for left medial patellofemoral ligament reconstruction with allograft for chronic patella instability. She consented to have her case reported after her procedure. She was diagnosed at age 14 years based on generalized joint hypermobility, chronic joint pain, spondylosis/spondylolisthesis, dislocation of several joints, and ptosis/myopia. She had several patella dislocations and endorsed being prone to easy musculoskeletal injury, bruising, and bleeding. On the day of surgery, we discussed her option of receiving a saphenous nerve block for postoperative analgesia along with general anesthesia. Of note, her surgeon routinely refuses regional anesthesia of the femoral and sciatic nerves, due to potential for motor nerve injury, but is comfortable with patients receiving a saphenous nerve block. After obtaining informed consent, she was transported to our regional block area where standard American Society of Anesthesiologists monitors with supplemental oxygen were applied. After a timeout was performed, 2 mg midazolam and 100 μg fentanyl were administered for sedation. Using the same ultrasound equipment as the previous case, a saphenous nerve block as