Case Report

Sciatic Nerve Hemangioma, a Challenge in Klippel–Trenaunay Syndrome for Perioperative Analgesia

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Abstract

Children with Klippel–Trenaunay syndrome have vascular malformations, varicosities at unusual distribution, and unilateral soft and skeletal tissue hypertrophy of the lower extremity. Corrective surgery of the deformity is necessary to improve gait and wear acceptable footwear. Perioperative pain relief for debulking of the right great toe in a known case of KTS with sciatic hemangioma was achieved with ultrasound-guided sciatic nerve block in an avascular area.

Keywords: Klippel–Trenaunay syndrome, magnetic resonance imaging, ultrasound-guided sciatic nerve block, vascular malformations

Introduction

The condition was first described by French physicians Maurice Klippel and Paul Trenaunay in 1900; they referred to it as nevus vasculosus osteohypertrophicus.[1] Klippel–Trenaunay syndrome (KTS) is a classical triad that consists of vascular malformations, varicosities at unusual distribution, and unilateral soft and skeletal tissue hypertrophy, usually of the lower extremity.[2,3] Children need corrective orthopedic surgeries of the lower limb. Apart from subcutaneous hemangiomas in the abdominal area, neuraxial hemangiomas are a known occurrence in KTS.[4,5] We report a case of KTS for debulking of the great toe under general anesthesia with perioperative pain relief achieved with an ultrasound-guided peripheral nerve block. Ultrasound may be preferable to landmark technique when aberrant anatomy is suspected.[6,7]

Case Report

An 8-year-old boy weighing 26.7 kg with KTS presented with right great toe hypertrophy along with soft-tissue swelling in the lower abdomen and medial aspect of the right thigh [Figure 1a]. Hypertrophy of the great toe hindered his gait and wearing proper footwear. He, therefore, was posted for osteotomy and debulking of the great toe. Laboratory reports and chest radiograph were normal. Examination revealed multiple compressible swellings on the right hypochondrium, lumbar and iliac quadrants of the abdomen, and right upper medial thigh [Figure 1b]. Magnetic resonance imaging (MRI) revealed multiple hemangiomas in the liver, spleen, and rectum.

Written informed consent was obtained from parents for surgical procedures to be performed under general anesthesia and peripheral nerve block for perioperative pain relief. Anesthesia was induced with intravenous (IV) fentanyl 1 μg/kg and propofol 2 mg/kg, and a laryngeal mask airway (2.5) was inserted. Anesthesia was maintained with nitrous in oxygen and sevoflurane through a closed circuit. Since the surgical incision was planned on the medial side of the right great toe and forefoot, an adductor canal block and sciatic nerve block were planned for postoperative pain relief. In the left lateral decubitus position, the sciatic nerve was scanned in real-time posteriorly from the greater trochanter level until the popliteal fossa, with a linear probe (Sonosite M-Turbo, USA – 6–13 mHz). The scan revealed an elliptical sciatic nerve surrounded by several pulsatile vascular structures [Figure 1c] encircling the sciatic

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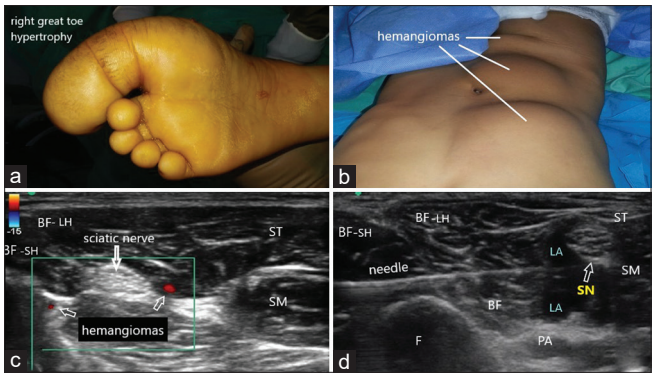

Figure 1: (a) Hypertrophy of the right great toe. (b) Subcutaneous hemangiomas on the right abdominal quadrants. (c) Ultrasound detection of vascular malformations around the sciatic nerve. (d) Ultrasound-guided mid-thigh sciatic nerve block. ST: Semitendinosus muscle; SM: Semimembranosus muscle; BF-LH: Biceps femoris long head muscle, BF-SH: Biceps femoris short head, F: Femur, SN: Sciatic nerve, PA: Popliteal artery, LA: Local anesthetic

nerve throughout its course from the gluteal area to the popliteal fossa, except in one area at mid-thigh level. This was abnormal and considered a part of the KTY syndrome. Scan around the ankle, too, revealed anomalous vascular structures. The nonvascular zone at the mid-thigh level of the sciatic nerve was considered a safe zone to administer an in-plane sciatic nerve block. No vascular malformations were observed in the entire adductor canal. US-guided and neurostimulation-aided sciatic nerve block was performed in the identified safe zone [Figure 1d and Video 1], and 0.2% of 11-ml ropivacaine and 15 µg of clonidine were injected after eliciting plantar flexion at 0.4 mA. A saphenous nerve block was administered in the adductor canal in the mid-thigh area with 8 ml of 0.2% ropivacaine. At surgical incision, there was no rise in heart rate and blood pressure more than 20% of baseline. No additional analgesics were required in the intraoperative period. IV paracetamol 500 mg was administered 30 min before extubation.

Postoperatively in the recovery room, the patient was pain free with a Visual Analog Scale (VAS) score of 1–2. No motor block was observed. The child complained of pain in the foot for the first time at 14.5 h after the surgery at a VAS = 4. IV paracetamol 500 mg was injected every 8 h thereafter until the patient was discharged on the 3rd postoperative day. At discharge, sensations were preserved in the distribution of the femoral and sciatic nerve in the foot. Plantar and dorsiflexion of the foot and movements of toes were normal.

Discussion

Regional anesthesia is a challenge in KTS because of associated vascular malformations reported in the spinal canal.[4,5] Although MRI scanning can detect central neuraxial vascular malformation, it is prudent to avoid central neuraxial blocks when peripheral nerve blocks can provide equivalent analgesia.[8] We could detect vascular malformations around the entire length of the sciatic nerve [Figure 1c] from the gluteal area to the popliteal fossa, except in one area at the mid-thigh level [Figure 1d]. This was considered a safe zone to administer sciatic nerve blocks. With vascular malformations around the ankle joint, the ankle block was excluded. Vascular hemangiomas have been reported around the ankle joint.[9] This emphasizes the role of ultrasound in diagnosing vascular malformations before implementing a block in diagnosed KTY patients. A saphenous nerve block was added as a supplemental nerve block considering the incision on the medial aspect of the great toe and forefoot.

Conclusion

This report emphasizes the role of the US in diagnosing vascular malformations in peripheral nerves, assisting regional anesthesia for improved analgesia, and an overall better outcome. Further, we would like to caution regional anesthesia enthusiasts to refrain from administrating neuraxial anesthesia and landmark-guided blocks in KTY syndromes or similar disorders associated with hemangiomas elsewhere.

Declaration of patient consent

The authors certify that they have obtained appropriate patient's guardian consent form. In the form, the guardian has given the consent for the child's images and other clinical information to be reported in the journal. The guardian understands that the child's name and initial will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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