CrossMark

CLINICAL REPORT

Peripheral nerve blocks as the sole anesthetic technique in a patient with severe Duchenne muscular dystrophy

Seung Uk Bang[1] · Yee Suk Kim[2,3] · Woo Jin Kwon[1] · Sang Mook Lee[1] ·
Soo Hyang Kim[1]

Abstract   General anesthesia and central neuraxial blockades in patients with severe Duchenne muscular dystrophy are associated with high risks of complications, including rhabdomyolysis, malignant hyperthermia, hemodynamic instability, and postoperative mechanical ventilation. Here, we describe peripheral nerve blocks as a safe approach to anesthesia in a patient with severe Duchenne muscular dystrophy who was scheduled to undergo surgery. A 22-year-old male patient was scheduled to undergo reduction and internal fixation of a left distal femur fracture. He had been diagnosed with Duchenne muscular dystrophy at 5 years of age, and had no locomotive capability except for that of the finger flexors and toe extensors. He had developed symptoms associated with dyspnea 5 years before and required intermittent ventilation. We blocked the femoral nerve, lateral femoral cutaneous nerve, and parasacral plexus under ultrasound on the left leg. The patient underwent a successful operation using peripheral nerve blocks with no complications. In conclusion general anesthesia and central neuraxial blockades in patients with severe Duchenne muscular dystrophy are unsafe approaches to anesthesia because of hemodynamic instability and respiratory depression. Peripheral nerve blocks are the best way to reduce the risks of critical complications, and are a safe and feasible approach to anesthesia in patients with severe Duchenne muscular dystrophy.

Keywords   Anesthesia · Duchenne · Malignant hyperthermia · Muscular dystrophy · Nerve block

Introduction

Duchenne muscular dystrophy (DMD) is an X-linked autosomal recessive disease that causes progressive muscle degeneration and results in proximal muscle weakness [1]. DMD can also affect the heart and respiratory muscles. General anesthesia is extremely hazardous in these patients, because it can lead to serious complications, including hyperkalemia, rhabdomyolysis, and malignant hyperthermia. Moreover, spinal or epidural anesthesia can cause serious hemodynamic instability and reductions in pulmonary function. Insufficient pulmonary function that is associated with muscle involvement exacerbates cardiopulmonary complications. Since general anesthesia and central neuraxial blockades in patients with severe DMD are associated with high risks of complications, they require a safer approach to anesthesia. We report the case of a patient with severe DMD who had a left distal femur fracture under peripheral nerve blocks (PNBs).

Case report

A 22-year-old male patient (167 cm tall, weight 47 kg) presented with a left distal femur fracture and needed to undergo reduction and internal fixation. The patient was diagnosed with DMD at 5 years of age. By the age of 13 years, he could not walk and had to use a wheelchair.

✉ Yee Suk Kim
    yskcmc4897@nate.com

1   Department of Anesthesiology and Pain Medicine, College
    of Medicine, The Catholic University of Korea, Seoul, South
    Korea

2   Department of Anesthesiology and Pain Medicine, College
    of Medicine, The Catholic University of Korea, Seoul, South
    Korea

3   327, Sosa-ro, Wonmi-gu, Bucheon City 14647, Kyunggido,
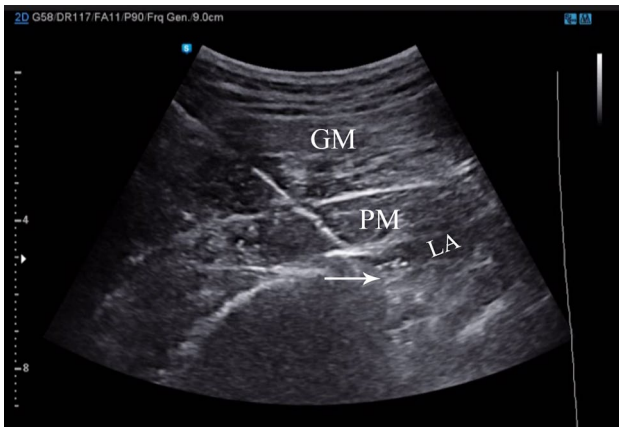   Republic of Korea

Fig. 1 Ultrasound-guided parasacral plexus block. The *white arrow* indicates the sacral plexus. *GM* Gluteus muscle, *LA* local anesthetic, *PM* pyriformis muscle

The disease progressed continuously, and 5 years before the operation he developed symptoms associated with dyspnea; he required non-invasive positive pressure ventilation with an inspiratory positive airway pressure of 14 cm H_2O and an expiratory positive airway pressure of 5 cm H_2O, which was delivered via a nasal interface because of breathlessness and the quality of his sleep. Electromyography showed there was no locomotive capacity, except for that in the finger flexors and toe extensors. Pulmonary function tests revealed severe restrictive lung disease with a forced expiratory volume at 1 s (FEV1) and a forced vital capacity (FVC) of 0.74 L (20 % of the predicted value) and 0.90 L (21 % of the predicted value), respectively, and an FEV1/FVC ratio of 82 %.

His initial blood pressure (BP) was 136/76 mmHg, with a heart rate (HR) of 120 beats/min and oxygen saturation by pulse oximetry (SpO2) of 90 %. Supplemental oxygen was administered with a nasal cannula 2 L/min. The patient was placed in the lateral decubitus position, and the superior gluteal area was scanned caudal to the greater sciatic foramen using ultrasound (WS 80A; Samsung Medicine, Seoul, Korea) with a 2–5 MHz curved probe. The sacral plexus on was identified, and we injected 20 mL of 0.375 % ropivacaine into the area using a 21-gauge Sonoplex needle (Pajunk, Geisingen, Germany) (Fig. 1).

We then placed the patient in the supine position, and we identified the femoral nerve and the lateral femoral cutaneous nerve using a linear transducer on and around the inguinal ligament, and we blocked these nerves by administering 15 and 5 mL of 0.375 % ropivacaine, respectively (Fig. 2).

We checked that the innervated areas associated with the individual nerves were blocked using the pinprick tests after 30 min. We judged that the patient was in a suitable condition for surgery and began the operation. The total operation time and anesthesia time was 40 min and 85 min, respectively. The patient's vital signs remained stable during surgery. The patient lost 100 mL of blood during the operation, he received 450 mL of normal saline, and his urine output was 50 mL. The patient did not experience any discomfort and did not require any additional analgesics or vasoactive drugs. The operation was successful and the patient's sensory capability recovered approximately 12 h after surgery. His BP was 100/60–110/80 mmHg, his HR was 108–112 beats/min, and his SpO2 was 90–92 % without oxygen supply during the postoperative period. Intermittent mechanical ventilation was applied during sleep starting on postoperative day 1. He was discharged without any complications once he had met the criteria that follow surgical or anesthesiological procedures.

Discussion

DMD is caused by a mutation of the gene that produces dystrophin and muscle-stabilizing protein. The disease characteristically involves the proximal muscles, which
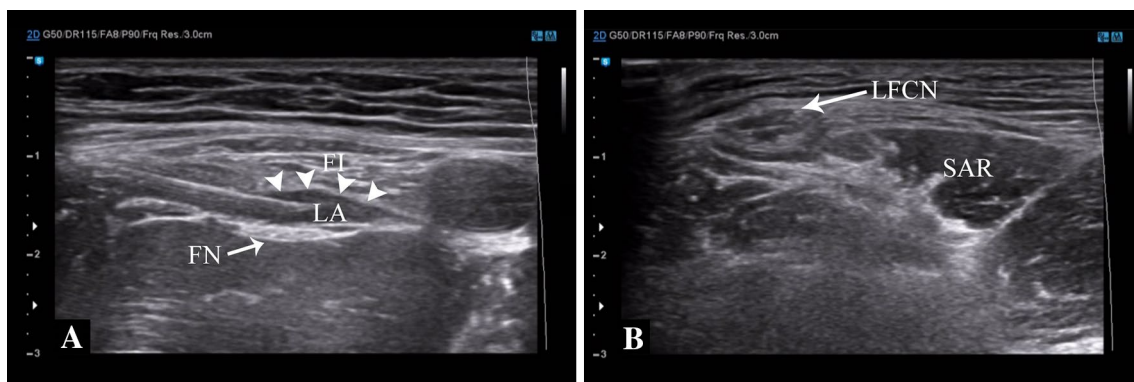

Fig. 2 Ultrasound-guided nerve block. **a** Femoral nerve. **b** Lateral femoral cutaneous nerve. *FI* fascia iliaca, *FN* femoral nerve, *LA* local anesthetic, *LFCN* lateral femoral cutaneous nerve, *SAR* sartorius muscle

results in progressive muscle degeneration. Furthermore, it can affect the respiratory and cardiac muscles, and it can cause respiratory failure, dilated cardiomyopathy, and arrhythmia [1, 2]. The type of anesthesia and the anesthetic agents used in these patients must be chosen very carefully, because cardiomyopathy or respiratory failure often causes mortality in these patients. Indeed, the most dangerous complications associated with general anesthesia in this patient group are malignant hyperthermia and anesthesia-induced rhabdomyolysis, which have mortality rates of approximately 30 %. Anesthesia-induced rhabdomyolysis is a condition that causes rhabdomyolysis and hyperkalemia but it is not malignant hyperthermia. Clinically, we cannot distinguish between them. Furthermore, the trigger factors for these pathological conditions are similar, and include inhaled anesthetic agents and succinylcholine [2–4].

An additional problem is that the standard doses of neuromuscular blocking agents (NMBAs) might delay the onset of the maximum blockade or cause prolonged recovery in DMD patients. Therefore, residual muscular blockades will occur after surgery which will increase the risk of respiratory complications. Consequently, some patients have undergone total intravenous anesthesia without the use of NMBAs for intubation or surgery, but the excessive doses of anesthetic may increase the incidence of myocardial depression or hypotension [5–7]. Furthermore, patients with preoperative FVC of <30 % who are under general anesthesia are more likely to require postoperative ventilator support [8].

As mentioned above, regional anesthesia is recommended for patients with severe DMD, because general anesthesia is associated with so many complications in these patients [9]. Moreover, we cannot categorically state that central neuraxial blockades are safe for patients with severe DMD. Indeed, in the case of high-level blockade, central neuraxial blockades are associated with critical complications.

First, inadvertent high-level blocks can cause phrenic nerve blocks, which can cause restrictions in the diaphragmatic muscle, low maximal inspiratory volumes, and low negative intra-pleural pressures during maximal inhalations. Consequently, they can cause a variety of complications. Furthermore, central neuraxial blockades might cause respiratory arrest, because decreasing cardiac outputs reduce the blood flow to the medulla of the brain, i.e., negatively influence expiratory respiration as well as inspiration. Although the level of the block is not high, it has been reported that central neuraxial blockades also reduce the vital capacity. This is caused by reductions in the expiratory reserve volume, which is a consequence of abdominal muscle paralysis rather than diaphragmatic or phrenic nerve dysfunction. Furthermore, the blockade of

the anterior abdominal muscles results in a reduced ability to actively cough, a lower maximum breathing capacity, and a low maximal expiratory volume [10]. These conditions contribute to a high risk of respiratory complications because of the reduced toileting function of the tracheobronchial secretions.

Second, most patients with DMD have accompanying scoliosis, which causes deficiencies in chest wall compliance and this adversely affects the uniform distribution of the tidal volume [8, 11]. The factors that aggravate the morbidity and mortality of DMD patients who have severe respiratory compromise must be acknowledged.

Finally, hypotension is the most common immediate side-effect of central neuraxial blockades, and it occurs in 33–40 % of cases [12, 13]. Hypotension is caused by blockade of the sympathetic fibers, which results in a reduction in cardiac output, and systemic vascular resistance. When the spinal denervation level encroaches on the cardiac accelerator fibers, the cardiac output may accentuate the degree of hypotension. This hemodynamic instability causes an imbalance between the delivery and uptake of oxygen in the tissues, and detrimental effects, including myocardial ischemia and hypoperfusion of other organs, ensue.

PNBs have several advantages compared with central neuraxial blockades. PNBs during surgery on the lower extremities have tenuous effects on the cardiovascular system, and they provide a higher level of hemodynamic stability, because the sympathetic fibers are blocked to lesser extents and efficient homeostatic vascular mechanisms in the unblocked areas compensate for the vasodilatation in the blocked areas. However, lumbar plexus blocks cannot reduce the incidence of hypotension because of epidural spread of local anesthetics [14]. Although rare, lumbar plexus blocks have been associated with other serious complications, including cardiac toxicity caused by accidental intravascular injections, renal hematoma, and lumbar plexopathy [15]. Therefore, the block should preferably be administered below rather than at the level of the plexus to prevent these complications.

Moreover, PNBs have less impact on respiratory mechanisms than central neuraxial blockades, and they do not block the abdominal muscles or the muscles that control respiration, so they have the advantage of maintaining normal pulmonary physiology, especially in high-risk patients. In addition, PNBs have considerable advantages in relation to postoperative analgesia, because they can minimize the side-effects of the opioids, which include respiratory depression, postoperative nausea, and vomiting, by decreasing the postoperative pain score and the amounts of opioids consumed.

Our patient's DMD involved the respiratory muscles, and the patient had dyspnea and required intermittent

mechanical ventilation. General anesthesia in this patient would have been associated with a critical respiratory complication risk, even if a risky drug was not used, and we were unable to predict the optimal dose and the duration of the muscle relaxant. Because we were unable to use neuromuscular monitoring due to severe muscular distrophy, it wasdifficult to decide the proper time of extubation. Central neuraxial blockades are associated with several complications, including hemodynamic instability and respiratory depression. The lateral femoral cutaneous nerve, posterior femoral cutaneous nerve (PFCN), femoral nerve, and sciatic nerve should be blocked with consideration of the dermatome and osteotome because internal fixation is performed through an incision of the lateral side of the thigh. If the medial aspect of the thigh, hip joint, or knee joint is involved, the obturator nerve should be blocked. In addition, a parasacral plexus block efficiently blocks the PFCN and sciatic nerve, while a distal sciatic nerve block, including a subgluteal and popliteal approach, is inadequate to block the PFCN.

Hence, we conclude that PNBs in severe DMD patients are a safe and feasible approach to reduce the risks of the critical complications caused by anesthesia. We also recommend that the anesthetic injection is administered distal to the level of lumbar plexus rather than at the level of the lumbar plexus.

Compliance with ethical standards

References

1. Biggar WD, Klamut HJ, Demacio PC, Stevens DJ, Ray PN. Duchenne muscular dystrophy: current knowledge, treatment, and future prospects. Clin Orthop Relat Res. 2002;401:88–106.

2. Gurnaney H, Brown A, Litman RS. Malignant hyperthermia and muscular dystrophies. Anesth Analg. 2009;109:1043–8.

3. Hayes J, Veyckemans F, Bissonnette B. Duchenne muscular dystrophy: an old anesthesia problem revisited. Paediatr Anaesth. 2008;18:100–6.

4. Goresky GV, Cox RG. Inhalation anesthetics and Duchenne's muscular dystrophy. Can J Anaesth. 1999;46:525–8.

5. Wick S, Muenster T, Schmidt J, Forst J, Schmitt HJ. Onset and duration of rocuronium-induced neuromuscular blockade in patients with Duchenne muscular dystrophy. Anesthesiology. 2005;102:915–9.

6. Yemen TA, McClain C. Muscular dystrophy, anesthesia and the safety of inhalational agents revisited; again. Paediatr Anaesth. 2006;16:105–8.

7. Muenster T, Schmidt J, Wick S, Forst J, Schmitt HJ. Rocuronium 0.3 mg kg^{-1} (ED95) induces a normal peak effect but an altered time course of neuromuscular block in patients with Duchenne's muscular dystrophy. Paediatr Anaesth. 2006;16:840–5.

8. Almenrader N, Patel D. Spinal fusion surgery in children with non-idiopathic scoliosis: is there a need for routine postoperative ventilation? Br J Anaesth. 2006;97:851–7.

9. Smith CL, Bush GH. Anaesthesia and progressive muscular dystrophy. Br J Anaesth. 1985;57:1113–8.

10. Braun DL. Spinal, epidural, and caudal anesthesia. In: Miller RD, editor. Miller's Anesthesia. 7th ed. Philadelphia: Churchill Livingstone; 2009. p. 1616–8.

11. Harper CM, Ambler G, Edge G. The prognostic value of preoperative predicted forced vital capacity in corrective spinal surgery for Duchenne's muscular dystrophy. Anaesthesia. 2004;59:1160–2.

12. Carpenter RL, Caplan RA, Brown DL, Stephenson C, Wu R. Incidence and risk factors for side effects of spinal anesthesia. Anesthesiology. 1992;76:906–16.

13. Moore DC, Bridenbaugh LD. Spinal (subarachnoid) block. A review of 11,574 cases. JAMA. 1966;195:907–12.

14. de Visme V, Picart F, Le Jouan R, Legrand A, Savry C, Morin V. Combined lumbar and sacral plexus block compared with plain bupivacaine spinal anesthesia for hip fractures in the elderly. Reg Anesth Pain Med. 2000;25:158–62.

15. Parkinson SK, Mueller JB, Little WL, Bailey SL. Extent of blockade with various approaches to the lumbar plexus. Anesth Analg. 1989;68:243–8.