Peripheral Nerve Block is Safely Administered in a Patient with Kearns–Sayre Syndrome

Woo-Jin Kwon, Seung-Uk Bang, Sae-Cheol Oh, Jung-Woo Shim

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

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Kearns–Sayre syndrome (KSS) is an extremely rare form of mitochondrial myopathy characterized by progressive external ophthalmoplegia, bilateral pigmentary retinopathy, and cardiac conduction abnormalities. In addition, KSS patients might present with proximal muscle weakness, diabetes mellitus, hearing loss, cerebellar ataxia, and elevated cerebrospinal fluid protein levels.[1]

Ensuring adequate general anesthesia can be very challenging for KSS patients; they can have severe hemodynamic instability and respiratory complications because of the complete atrioventricular (AV) block, congestive heart failure due to dilated cardiomyopathy, respiratory muscle weakness, and the prolonged effects of neuromuscular blocking. Neuraxial blockade might exacerbate hypotension by decreasing preload and systemic vascular resistance and inducing bradycardia.

Peripheral nerve block (PNB) may be useful in subjects with KSS because it does not block the abdominal muscles and diaphragm. It also minimizes sympathetic blockade, allowing the maintenance of hemodynamic stability. We report successful PNB anesthesia in a KSS patient who underwent multiple muscle biopsies.

An 18-year-old male patient underwent muscle biopsy of the anterior thigh muscle to confirm KSS. He had been diagnosed with diabetes mellitus a year earlier and was admitted to the emergency room due to sudden loss of consciousness 5 days prior. After a third-degree AV block was diagnosed, the cardiologist inserted a pacemaker and consulted other specialists regarding a possible KSS diagnosis. His ophthalmologic consult showed bilateral pigmentary retinopathy, and a muscle biopsy was suggested to confirm KSS. The orthopedic surgeon suggested that local anesthetic (LA) infiltration was insufficient given the size and number of biopsies required.

The patient was placed in the supine position, and the inguinal area was scanned using ultrasound (WS 80A, Samsung Medicine, Seoul, Korea) with a 7–12 MHz linear probe. The femoral and lateral femoral cutaneous nerves were blocked with 1% lidocaine (15 and 5 ml, respectively) before biopsying the vastus lateralis and quadriceps femoris [Figure 1].

We confirmed that the innervated were blocked using pinprick tests after 30 min. The patient was in a suitable condition, and the biopsies proceeded without sedation. His vital signs were stable, and he did not require additional analgesics or vasoactive drugs. The total operating and anesthetic times were 30 and 55 min, respectively. A total of five samples were obtained from the vastus lateralis and quadriceps femoris. The patient’s sensory capability recovered about 3 h after the operation, and the patient was able to walk alone 5 h after the PNB. He was discharged without any complications, and the biopsies confirmed KSS.

Anesthesia types and agents must be chosen very carefully for KSS patients because they are vulnerable to cardiomyopathy or respiratory failure. The most dangerous complication in this patient group is malignant hyperthermia (MH), although it is controversial whether

Address for correspondence: Dr. Seung-Uk Bang, Department of Anesthesiology and Pain Medicine, College of Medicine, The Catholic University of Korea, 64, Daheung-ro, Jung-gu, Daejeon, 301-723, Seoul, Republic of Korea
E-Mail: seungukb@naver.com

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this is caused by KSS. The trigger factors for MH in KSS are inhalation agents and succinylcholine. Inhalation agents suppress mitochondrial function by inhibiting complex I respiration (NADH dehydrogenase). In normal patients, mitochondria are only affected by higher clinical doses, but KSS patients are more susceptible.[2]

KSS patients might have heart conditions including dilated cardiomyopathy, congestive heart failure, and AV block. Some anesthetics induce hypotension and exacerbate organ perfusion because of their cardiac depressant effects. Organ hypoperfusion aggravates metabolic acidosis, and KSS patients have increased pyruvate-to-lactate conversion because their mitochondria do not properly metabolize glucose. This leads to metabolic acidosis and affects the actomyosin cross-bridge cycle, which translates into less force production and delayed contraction velocity.[3]

Due to complications related to general anesthesia in KSS patients, regional anesthesia is recommended. However, we couldn't confirm that central neuraxial blockades were safe because unintended high-level blocks could cause phrenic nerve palsy, which restricted the diaphragmatic muscle, decreased maximal inspiratory volumes, and led to low negative intrapleural pressures for maximal inhalations. Even with a low-level central neuraxial blockade reduces the expiratory reserve volume and vital capacity. This is a consequence of abdominal muscle paralysis rather than phrenic nerve dysfunction. Abdominal muscle paralysis decreases the ability to cough and lowers maximum breathing capacity and maximal expiratory volume. These conditions contribute to a high risk of respiratory complications because they reduce secretions.[4]

Hypotension is the most common immediate side effect of central neuraxial blockades due to reductions in heart rate, stroke volume, and systemic vascular resistance. If cardiac accelerator fibers are blocked, low cardiac output may further exacerbate low blood pressure. Although hypotension is generally overcome by an increased heart rate, patients with KSS cannot compensate their blood pressure due to cardiac conduction abnormalities. This hemodynamic instability causes an imbalance between oxygen delivery and uptake, and detrimental effects, including myocardial ischemia and organ hypoperfusion ensue. KSS patients already have elevated lactate levels, and anoxegen delivery deficiency can exacerbate lactic acidosis.

PNBs have several benefits compared with central neuraxial blockades. They have minimal effects on the cardiovascular system and provide hemodynamic stability due to less sympathetic fiber blockade. Moreover, efficient homeostatic vascular mechanisms in the unblocked areas compensate for vasodilatation in the blocked areas. PNBs do not block the abdominal or respiratory muscles; therefore, they have less impact on pulmonary function than central neuraxial blockades. In addition, PNB can decrease the postoperative pain score and opioid consumption. This is important in KSS patients as opioids can cause respiratory depression, although a small opioid dose can minimize PNB side effects including nausea, vomiting, and pruritus.

LAs for PNB should use be used carefully in patients with KSS because LAs affect mitochondrial function. Propacaine, cocaine, and tetracaine dissipate the mitochondrial membrane potential. Ropivacaine interferes with mitochondrial energy transduction, and bupivacaine directly affects mitochondrial function by depolarizing the mitochondrial membrane and oxidizing pyridine nucleotides. Mitochondrial toxicity was reported in one patient with KSS who received local anesthesia with articaine.[5]

We conclude that PNB is a safe anesthesiology procedure that can reduce complications associated with general anesthesia or central neuraxial blockade in KSS patients.

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There are no conflicts of interest.

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