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Safety of Spinal Anesthesia in A Patient with Kennedy's Disease: A Case Report

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Abstract **Case Report**

Kennedy's disease, otherwise known as spinal and bulbar muscular atrophy, poses significant anesthetic challenges in patients with bulbar dysfunction, respiratory muscle weakness, and potential sensitivity to neuromuscular blocking agents. This article documents a case involving a 53-year-old man diagnosed with Kennedy's disease who successfully underwent open reduction and internal fixation of a right ankle fracture under spinal anesthesia without perioperative respiratory or neurological complications. Careful titration of block height, avoidance of sedatives, and close postoperative monitoring contributed to a stable course, suggesting that spinal anesthesia can be safely administered in this particular patient population.

Keywords: Kennedy's disease; spinal and bulbar muscular atrophy; spinal anesthesia; neuromuscular disease.

INTRODUCTION

Kennedy's disease (KD), also known as spinobulbar muscular atrophy, is a rare X-linked recessive neurodegenerative disorder of the lower motor neurons. It is characterized by progressive proximal limb weakness, bulbar symptoms such as dysarthria and dysphagia, and endocrine manifestations, and typically presents in middle age [1]. Bulbar involvement and spontaneous laryngospasm can occur and may lead to respiratory failure, whereas underlying myopathy raises concerns regarding the use of neuromuscular blocking agents. These are important issues that anesthesiologists should be aware of.

Current expert recommendations suggest avoiding general anesthesia when significant amyotrophic weakness or bulbar dysfunction is present and favor regional or local anesthesia when feasible [2, 3]. However, there are few case reports addressing this rare disease [4-6], and a limited number of reports documenting perioperative considerations for spinal anesthesia. As such, the present case report describes the successful use of spinal anesthesia in a patient with KD undergoing lower extremity surgery and discusses the anesthetic considerations for this population.

CASE REPORT

The requirement for informed written consent was waived from the institutional review board of the authors' institution (SGPAIK 2025-12-012, confirmed on 2025.12.26). A 53-year-old man (height, 168 cm; weight, 64 kg) diagnosed with KD was scheduled to undergo elective open reduction and internal fixation for a right lateral malleolar fracture. KD was diagnosed by DNA testing 13 years previously, and his older brother was also diagnosed with KD. The patient's medical history was otherwise unremarkable, with no cardiopulmonary disease.

Neurologically, the patient exhibited mild bulbar dysfunction as dysphagia manifesting as cough or choking on liquids 2 to 3 times per week, mild sialorrhea during meals, and subtle tongue fasciculation. In addition, he had lower limb weakness and difficulty climbing stairs, although independent ambulation on level ground was preserved. Otherwise, the patient did not exhibit resting dyspnea, orthopnea, or a history of aspiration pneumonia. Pulmonary function testing had not been performed previously; however, the patient denied exercise intolerance. Preoperative blood tests, vital signs, and chest radiographs were within normal limits. Airway evaluation revealed a Mallampati classification II, with normal mouth opening and neck mobility. The preoperative coagulation profile was

within normal limits and there were no contraindications to neuraxial anesthesia. The patient provided informed consent to spinal anesthesia.

On arrival to the operating room, standard monitoring, including electrocardiography, noninvasive blood pressure measurement, pulse oximetry, and continuous monitoring of respiratory rate and end-tidal carbon dioxide, was performed. Supplemental oxygen was routinely administered at a rate of 3 L/min via nasal cannula. No premedications were administered. Non-invasive blood pressure was 123/86 mmHg, heart rate was 57 beats/min, and oxygen saturation was 100%. With the patient in the lateral decubitus position, a midline spinal puncture was performed in the L4–5 interspace using a 25-gauge Quincke-type needle. Bupivacaine hydrochloride (2.3 ml Marcaine Heavy, Mitsubishi Danabe Pharma, Osaka, Japan) was injected into the subarachnoid space after free flow of clear cerebrospinal fluid was confirmed. The patient was then positioned supine. Immediately after spinal injection, the sensory block extended to the temperature sensation at the T12 dermatome and pinprick at L1. After 15 min, the pinprick level stabilized at T10, which was adequate for the planned surgery. The motor block in the operated limb was complete, and hemodynamic parameters remained stable.

During surgery, the patient was comfortable and was not administered sedatives. The surgical procedure proceeded uneventfully over a total operative time of approximately 100 min. Estimated blood loss was 50 ml, and 500 ml of balanced crystalloids (Plasma solution A; HK Inno. N, Cheongju, Korea) was administered intravenously. At the conclusion of the operation, the patient reported that if he stayed in the same position for a long time, his arms felt numb and he wanted to move both arms. After unbuckling the arm belt, the patient immediately felt fine.

After surgery, the patient was transferred to the post-anesthesia care unit. Regression of the sensory block to the L1 level occurred within the expected time frame, and motor function returned without delay or asymmetry. Postoperatively, the patient did not experience any new weaknesses, dysphagia, or respiratory symptoms. Postoperative analgesia was initially managed with intravenous non-opioid analgesics followed by oral administration. The patient's postoperative course was uneventful, without any neurological deterioration. The patient was discharged on postoperative day 7, with no complications observed at the outpatient follow-up on postoperative day 14.

DISCUSSION

KD is a lower motor neuron disease caused by a trinucleotide repeat expansion (cytosine-adenine-guanine) in the androgen receptor gene, producing a toxic polyglutamine tract that leads to the degeneration of spinal and bulbar motor neurons with associated myopathy [7, 8]. It usually presents in the third to fifth decades of life with slowly progressive proximal and bulbar weakness accompanied by tremors, gynecomastia, testicular atrophy, diabetes, and impotence due to androgen receptor dysfunction, and many patients ultimately require supportive care despite a near-normal life expectancy [7, 8].

Several anesthetic concerns have been highlighted in patients diagnosed with KD. Bulbar dysfunction, with dysphagia and impaired laryngeal reflexes, increases the risk for aspiration and upper airway obstruction during general anesthesia, and laryngospasm may occur spontaneously or be triggered by airway manipulation [3, 9]. Altered neuromuscular transmission raises concerns about hyperkalemia with succinylcholine and prolonged blockade with non-depolarizing neuromuscular blocking agents. In addition, sedatives and systemic opioids can impair protective reflexes in the airway. Postoperative respiratory failure remains a concern, leading to failure of timely extubation and prolonged mechanical ventilation in patients with KD undergoing general anesthesia [2, 3, 10, 11].

Although neuraxial anesthesia (spinal, epidural, and combined spinal–epidural) appears to be feasible, it is not entirely without risk in patients diagnosed with neuromuscular disease. Excessive cephalad spread of local anesthetic may impair intercostal muscle function and reduce pulmonary reserve. Therefore, careful dose selection and attention to patient positioning are important to limit the sensory block level. Many authors advocate avoiding blocks higher than the midthoracic region, when possible, especially in patients with documented pulmonary dysfunction [12].

There is also an ongoing theoretical concern about performing neuraxial anesthesia in patients with pre-existing neurological diseases due to the possibility of exacerbating deficits or attributing new symptoms after the block. However, available reports of spinal, epidural, and combined spinal epidural anesthesia in KD and other lower motor neuron disorders have not demonstrated a clear increase in neuraxial related neurological complications [2, 13-15]. Hemodynamic instabilities, such as hypotension and bradycardia, may be more pronounced in frail patients with neuromuscular conditions, and prompt treatment with fluids and vasopressors is necessary to maintain adequate organ perfusion.

Several reports have documented the safety of sugammadex in patients with neuromuscular diseases such as myasthenia gravis. There is also a reported case in which general anesthesia was safely administered to a patient with KD after the introduction of sugammadex [6]. Nevertheless, the use of neuromuscular blocking agents continues to carry the risk for exacerbating pre-existing weakness of the lower limbs and bulbar

musculature, as well as increasing the potential for postoperative airway compromise [2, 10].

The case described herein demonstrates that spinal anesthesia in patients with KD can be administered without introducing new neurological or respiratory deficits. By avoiding or minimizing sedation, limiting block height, and ensuring close monitoring, spinal anesthesia can be safely administered to patients with KD. We support neuraxial anesthesia as a reasonable option for lower-limb procedures in this population because it yields meaningful advantages by avoiding airway manipulation and neuromuscular blocking drugs. Additional case reports and series are needed to refine the recommendations and clarify the long-term neurological outcomes.

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