

Anesthesia Management of Patients with Kennedy's Disease Undergoing Cut-and-replace Internal Fixation of Fibular Fractures: A Case Report

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Abstract

Introduction: Kennedy disease is a late-onset X-linked recessive inherited neurodegenerative disorder characterized by progressive proximal limb and medullary muscle atrophy [1] with spontaneous laryngospasm [2-3]. Anesthesia for patients with Kennedy disease is extremely challenging because the nature of Kennedy disease exposes patients with the disease to anesthetic risks including acute laryngospasm, increased sensitivity to non-depolarizing muscle relaxants and postoperative respiratory failure or aspiration.

Case presentation: We report a 55-year-old male patient who suffers from Kennedy disease. He underwent open reduction and internal fixation of the left fibular fracture due to trauma. The patient first noticed muscle weakness in the proximal lower limbs at the age of 40 and was diagnosed with Kennedy disease after undergoing genetic testing two years later. After detailed discussion and with the patient's consent, the surgical procedure was completed under the method of "venous sedation combined with ultrasound-guided nerve block and acupuncture anesthesia".

Conclusion: This case report indicates that the internal fixation surgery for fibular fracture in patients with Kennedy disease can be safely and smoothly completed through intravenous sedation combined with ultrasound-guided nerve block and acupuncture anesthesia, without deterioration of neurological signs or symptoms.

Keywords: Acupuncture anesthesia • Ultrasound-guided nerve block • Kennedy disease • Muscle weakness • Balanced anesthesia.

Introduction

Kennedy disease is a late-onset X-linked recessive hereditary neurodegenerative disease, characterized by progressive proximal limb and bulbar muscle atrophy [1], accompanied by spontaneous laryngospasm [2-3]. Due to the fact that such patients often experience painful muscle spasms for many years before the onset of symptoms and their electromyogram often shows extensive damage to spinal cord anterior horn cells, anesthesia for these patients poses a high risk. Here, we describe a case of a patient with Kennedy disease who underwent open reduction and internal fixation of a fibular fracture under the method of "intravenous sedation combined with ultrasound-guided nerve block and acupuncture anesthesia". This indicates that for Kennedy disease patients who require surgery, this may be an effective anesthesia management method.

Case Presentation

A 55-year-old male (height 166 cm; weight 73 kg) was admitted to the hospital due to trauma and diagnosed with "left fibula fracture". He was scheduled to undergo open reduction and internal fixation for the left fibula

fracture. He first noticed muscle weakness in the proximal lower limbs at the age of 40 and the treatment with traditional Chinese medicine massage was ineffective. Two years later, he was diagnosed with Kennedy disease. Physical examination revealed no muscle atrophy in the lower and upper limbs, with lower limb muscle strength at grade 4 and normal upper limb muscle strength. There were no symptoms of slurred speech or facial tremor. He had no history of chronic systemic diseases such as hypertension, diabetes, coronary heart disease, allergies, or surgery. Laboratory tests revealed that the plasma creatine kinase concentration was 551 U/L (normal range: 55-170 U/L), creatine kinase isoenzyme was 6.5 ng/ml (normal value: <6.22 ng/ml) and myoglobin was 129.7 ng/ml (normal value: <70 ng/ml). After detailed discussion and with the patient's consent, intravenous sedation combined with ultrasound-guided nerve block and acupuncture anesthesia was chosen. Acupuncture treatment was administered to the patient the day before surgery, with points selected including Lianquan, Feishu, Pishu and Shuitu (bilateral). During the procedure, points Hegu, Neiguan and Taiyuan were selected and after needling to obtain qi sensation, an electroacupuncture device (model: G6805-2) was connected for electrical stimulation until the end of the procedure. The wave pattern was a sparse-dense wave, with a sparse wave of 4 Hz and a dense wave of 20 Hz and a peak current of 5 mA. Preoperative ultrasound-guided block of the femoral nerve (0.33% ropivacaine, 30ml) and the sciatic nerve above the popliteal fossa (0.33% ropivacaine, 10ml) were performed. To ensure patient calmness and stability, dexmedetomidine 50ug and medium-chain and long-chain propofol 2mg/kg were administered to induce deep sleep while preserving spontaneous breathing. Oxygen was administered through a nasopharyngeal airway at a rate of 5 liters per minute. Throughout the surgery, the patient remained calm, with a normal electrocardiogram, stable heart rate and blood pressure and SpO₂ at 100%. The patient did not complain of discomfort such as dyspnea or breathe holding. Postoperative recovery was good, with no deterioration in neurological signs and symptoms. Follow-up by telephone 3 months later revealed that the patient had returned to daily work without long-term postoperative adverse events.

Written informed consent was obtained from the patients and their families for the publication of this report.

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Discussion

Kennedy disease, named after neurologist Dr. William R. Kennedy, is a late-onset X-linked recessive hereditary neurodegenerative disease characterized by progressive proximal limb and medullary muscle atrophy [1], accompanied by spontaneous laryngospasm [2-3]. The incidence of Kennedy disease is approximately 1 in 40,000 males [4]. The etiology of this disease is caused by the unstable amplification of the CAG triplet repeat (40-62 repeats) in exon 1 of the Androgen Receptor (AR) gene on chromosome Xq11-12 [5]. On May 11, 2018, the National Health Commission of China and other four departments jointly included this disease in the "First Batch of Rare Diseases List". We searched Medline data from 1806 to 2025 and found only two articles reporting on this topic. Issues related to anesthesia management in patients with Kennedy disease have rarely been reported. However, given that Kennedy disease is characterized by progressive proximal limb and bulbar muscle atrophy, there are several potential anesthetic risk factors, including acute laryngospasm, hyperkalemia caused by the use of succinylcholine, increased sensitivity to non-depolarizing muscle relaxants and postoperative respiratory failure or aspiration [6]. Complications reported to date include glottic edema, respiratory distress and tension pneumothorax. Additionally, general anesthesia can inhibit the swallowing reflex and may further increase the risk of pulmonary aspiration.

Skeletal muscle relaxants are a class of drugs that selectively act on the N2 cholinergic receptors on the motor nerve end plate membrane, leading to skeletal muscle relaxation by blocking nerve impulse transmission. In conventional general anesthesia for open reduction and internal fixation of fibular fractures, the use of muscle relaxants is essential as they can relax skeletal muscles and optimize surgical conditions. This patient suffered from Kennedy disease and had preoperative symptoms of thigh muscle weakness, but no significant clinical manifestations such as dysphagia or laryngospasm were observed. However, we must consider the risks associated with the use of muscle relaxants [7], such as intraoperative laryngospasm, postoperative worsening of muscle weakness and the need for ventilator support due to postoperative respiratory muscle weakness, prolonged hospital stay and potential ICU admission. Therefore, we adopted ultrasound-guided nerve block combined with acupuncture anesthesia techniques and used sedatives to induce sleep in the patient, maintaining spontaneous breathing and avoiding the use of muscle relaxants during surgery. This facilitated the smooth progress of the surgery.

Ultrasound-guided nerve block is a revolutionary technology in the fields of anesthesiology, pain management and emergency medicine. Its core advantage lies in achieving real-time dynamic visualization, significantly enhancing the safety, effectiveness and scope of application of nerve block. This technology ensures that the needle tip accurately reaches the vicinity of the target nerve due to its advantages of precise positioning and real-time guidance, significantly reducing the uncertainty of traditional anatomical landmark positioning or nerve stimulator positioning methods and avoiding positioning failure caused by individual variation. It significantly improves safety, makes the blocking effect more complete and greatly promotes the development of precise anesthesia and comfortable medical care.

Acupuncture-assisted anesthesia is a vibrant interdisciplinary field that combines acupuncture, surgery, anesthesiology, neurophysiology and other disciplines, abbreviated as acupuncture anesthesia [8]. Its technology and theory are based on the valuable experience of traditional acupuncture in treating painful diseases and inhibiting and preventing injurious pain. It is an innovative anesthesia method with Chinese medicine characteristics, combining acupuncture therapy with surgery. It is an effective supplement to modern anesthesia [9]. Acupuncture-assisted anesthesia has been proven to enhance the sedative and analgesic effects of drugs, reduce the occurrence of perioperative complications, help regulate the internal environment of the

human body, improve organ function and maintain the body in a balanced state during the perioperative period [10].

Our study has one key limitation. Due to the combined application of three different anesthesia methods, it is difficult to clearly distinguish the specific contribution of each anesthesia method to the observed overall anesthesia effect. Therefore, when generalizing the conclusions, it is necessary to explicitly state that these results were obtained under the specific background of combined anesthesia.

Conclusion

There are very few reports on perioperative anesthesia management for patients with Kennedy disease. Our case report provides an effective perioperative anesthesia method for patients with Kennedy disease. However, further research on anesthesia management for such patients is still needed.

Acknowledgement

None.

Conflict of Interest

None.

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