

# Anesthetic Management of a Patient With Spinocerebellar Ataxia Type 1

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Spinocerebellar ataxia type 1 (SCA1) is one of the autosomal dominant spinocerebellar degeneration (SCD) diseases characterized by progressive cerebellar ataxia, muscle atrophy, and peripheral neuropathy. We report the management of a 43-year-old man with SCA1 who underwent general anesthesia for open reduction and internal fixation of a mandibular fracture. Although anesthesia-induced vocal cord paralysis has been reported in patients with SCD, nasotracheal intubation was performed uneventfully with video laryngoscope. After taking into consideration the increased risk of postoperative respiratory depression in patients with SCD, rocuronium dosing was titrated carefully, and fentanyl was not used during surgery. Preparation for an anticipated difficult airway and avoiding significant respiratory depression are crucial when providing general anesthesia for patients with SCA1.

**Key Words:** Spinocerebellar ataxia type 1; Mandibular fracture; General anesthesia.

Spinocerebellar degeneration (SCD) is a neurodegenerative disease characterized by progressive cerebellar ataxia. Spinocerebellar ataxia type 1 (SCA1) is one of the autosomal dominant inherited SCD diseases, and its frequency is relatively rare at 5.8%. Patients with SCA1 exhibit problems with balance and coordination but can also have dysphonia, dysphagia, cognitive impairment, and muscle spasticity and atrophy. These issues should be carefully taken into consideration when managing anesthesia for patients with SCD/SCA1.

## CASE PRESENTATION

A 43-year-old man (height 171 cm; weight 54 kg; body mass index 18.5 kg/m<sup>2</sup>) with SCA1 fell while attempting to walk independently and fractured his mandible, leading to the need for open reduction and internal fixation under general anesthesia. His clinical signs and symptoms included gait disturbance, dysarthria, dysphagia, oculomotor dysfunction, and anxiety disorder, although vocal cord paralysis was not observed on

preoperative flexible fiber-optic laryngoscopy performed by an otorhinolaryngologist. Because anesthesia-induced vocal cord paralysis has been reported in patients with SCD, a video laryngoscope and flexible fiber-optic bronchoscope were prepared before induction of anesthesia. General anesthesia was induced with an intravenous (IV) bolus of propofol (50 mg) and a continuous infusion of remifentanyl (0.12 µg/kg/min) after a radial arterial line was placed. Rocuronium (total: 30 mg) was carefully titrated to effect as guided by neuromuscular monitoring, and nasotracheal intubation was uneventfully performed with a McGRATH MAC video laryngoscope. General anesthesia was maintained with desflurane (5%) and a remifentanyl infusion (0.2-0.36 µg/kg/min), and continuous infusions of etilefrine (1-4 mg/h), an α1- and β1-adrenoceptor agonist, and phenylephrine (0.1-0.4 mg/h) were administered to avoid hypotension that could be induced by autonomic neuropathy. To avoid postoperative respiratory depression, fentanyl was not used during surgery, and IV acetaminophen (1000 mg) was used for postoperative pain instead. Local anesthetics were also not used during this case. After fully emerging from general anesthesia with full recovery of spontaneous breathing, the patient was extubated after cuff-leak test was performed. The absence of vocal cord paralysis and upper airway obstruction was confirmed by video laryngoscopy. The patient was transferred to the intensive care unit and discharged uneventfully.

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## DISCUSSION

SCD belongs to a group of hereditary progressive neurodegenerative diseases. SCA1 is classified as one of the many genetic SCD types. With SCA1, the 3-base CAG repeat in the exon of the causative ATXN1 gene is abnormally extended, affecting the production of the protein ataxin-1.<sup>1</sup> The CAG repeat is translated into continuous glutamine chains that form higher-order structures and produce abnormal polyglutamine chains in the brain.<sup>1</sup>

The clinical symptoms of SCA1 can include cerebellar ataxia, pyramidal and extrapyramidal symptoms, eye movement disorders, sensory disorders, dysphagia, hyperactive tendon reflexes, and muscle atrophy. In addition, autonomic nervous system disorders such as orthostatic hypotension, dysuria, sweating disorders, and depression are also often observed. SCA1 patients are at high risk for maxillofacial fractures because of their ataxia. In patients with SCA1, there can also be neurogenic atrophy of the intrinsic laryngeal muscles and an increase in the activity of laryngeal adductors, which could induce vocal cord paralysis.<sup>2</sup> These symptoms are considered to be potentiated by general anesthetics. Therefore, we prepared a video laryngoscope and flexible fiber-optic bronchoscope before induction of anesthesia.

Peripheral neuropathy occurs in 82% of patients with SCA1 and may prolong the effects of muscle relaxants.<sup>3</sup> In patients with SCD, train-of-four values may be inaccurate if there is peripheral neuropathy affecting the site of electrode placement for the peripheral nerve stimulator. Because the patient's extrapyramidal symptoms were more intense on the right side than on the left, we placed the nerve stimulator electrodes on the left forearm. In addition, etilefrine and phenylephrine were continuously administered to avoid potential hypotension, which could also be induced by autonomic neuropathy.

Considering the risk for postoperative respiratory depression in patients with SCD, fentanyl was not used during surgery, and general anesthesia was maintained with the ultra-short-acting opioid remifentanyl. Furthermore, acetaminophen was selected to provide postoperative analgesia instead of fentanyl. Local anesthetics are another option for controlling perioperative pain in patients with SCA1; however, we did not perform any mandibular nerve blocks or infiltrate any local anesthetics during this case.

## CONCLUSION

In the anesthetic management of SCA1 patients, it is important to consider the potential for difficulty managing the airway as well as minimizing the risk of respiratory depression postoperatively. Conducting a thorough perioperative airway assessment, carefully titrating any neuromuscular blocking agents, and avoiding/minimizing non-ultra-short-acting opioid agonists are recommended.

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