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Anesthetic management of a patient with progressive supranuclear palsy

Kazuyuki Sakai[1] and Koji Sumikawa[2]

[1] Department of Anesthesia, Nagasaki Medical Center of Neurology, Nagasaki, Japan
[2] Department of Anesthesiology, Nagasaki University School of Medicine, Nagasaki, Japan

Abstract
A 74-year-old woman with progressive supranuclear palsy (PSP) was scheduled for laryngotracheal separation surgery. Her neck showed severe backward tilt as a symptom of PSP. Magnetic resonance imaging (MRI) showed a significant airway stenosis due to the neck deformity. In the operating room, awake orotracheal intubation failed because of the neck deformity and airway stenosis. Therefore, tracheotomy was performed for airway management. General anesthesia was induced and maintained with sevoflurane (1.0%–2.5%) and fentanyl (total, 200 μg). Vecuronium (total, 5 mg) was used as a muscle relaxant. Monitoring of the train-of—four ratio in the ulnar nerve was impossible because of contracture of the fingers. Patients with PSP may have some serious associated deformities, and specific management, especially for the airway, may be necessary for general anesthesia.

Key words Progressive supranuclear palsy · Cervical backward tilt · Airway stenosis · General anesthesia

Introduction

Progressive supranuclear palsy (PSP) is an uncommon disease. The incidence is reported to be 4–5 persons per 100 000 [1]. It is an adult-onset neurodegenerative disorder. The characteristic is early postural instability, which leads to easy falls, and a vertical supranuclear-gaze palsy [2]. The degeneration occurs at supranuclear, brain stem, and cerebellar levels [1]. The disease is associated with gait disturbance, Parkinsonism, oculomotor disturbance, and dementia. The specific abnormality is the dystonia with cervical backward tilt [1]. We describe the anesthetic management of a patient with PSP undergoing laryngotracheal separation surgery.

Address correspondence to: K. Sakai, Department of Anesthesia, Isahaya General Hospital, 24-1 Eishouhigashi-machi, Isahaya, Nagasaki 854-0071, Japan
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Case report

A 74-year-old woman weighing 40 kg, and of unknown height, had been diagnosed with PSP. Her initial symptoms were slow movement and low levels of activity in daily life, which appeared at the age of 62. She was diagnosed with Parkinsons disease at the age of 68, but the drugs against Parkinsons disease were not effective. She became unable to walk at the age of 69, and was diagnosed with PSP at the age of 70. Dementia was also observed at that time. Subsequently, she suffered aspiration pneumonia several times. The laryngotracheal separation surgery was scheduled to prevent aspiration pneumonia.

Her past history showed that she was diagnosed with hypertension and diabetes mellitus at the age of 63.

On physical examination, she could not communicate with other persons. She rarely moved, but said meaningless words. Her neck showed severe backward tilt. A small percutaneous tracheal tube, for suction of sputum, was placed in the trachea. Because of the sputum, and airway stenosis, mucous rale was often heard at the neck, and suction was needed frequently. All four limbs showed severe contracture. Electrocardiogram revealed the frequent occurrence of supraventricular premature beats. Chest radiography showed regional pneumonia in the left upper lung. Biochemical tests showed a low concentration of serum protein ($5.9\,g \cdot d^{-1}$). Airway stenosis was identified on (MRI; Fig. 1. magnetic resonance imaging). The narrowest region was just below the vocal cords, and the diameter of this region was about 5.0 mm.

Induction and maintenance of anesthesia

Famotidine, 20 mg IV, was administered 2 h before the induction of general anesthesia, as premedication. We tried awake intubation with a reinforced tracheal tube (6.0 mm internal diameter [ID]), but failed because

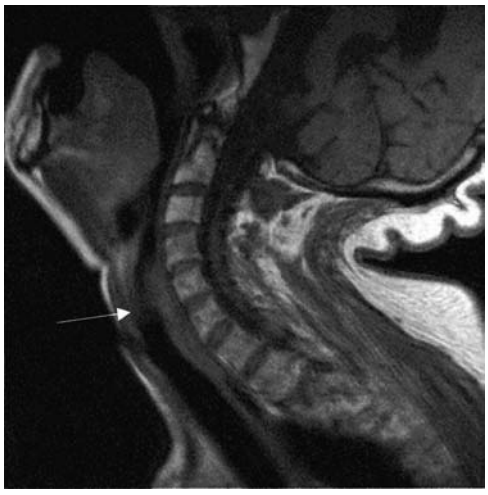

Fig. 1. Sagittal magnetic resonance imaging (MRI) of the neck. Cervical backward tilt and airway stenosis (*arrow*) are obvious

direct inspection of the pharynx was impossible. Awake fiberoptic intubation was not attempted because of the severe neck deformity and tracheal stenosis. Tracheotomy distal to the stenotic part was performed for airway management. General anesthesia was induced and maintained with sevoflurane (1.0%–2.5%) and fentanyl (total, 200 μg). Vecuronium (total, 5 mg) was used as a muscle relaxant. The operation time was 2 h 18 min, and the anesthesia time was 3 h. The blood pressure and heart rate were stable, and no sign of autonomic nervous system instability was observed during anesthesia. Spontaneous respiration appeared at the end of the operation. The tidal volume was 230 ml, and the respiratory rate was 18 times per min. The patient was admitted to the intensive care unit (ICU) without respiratory assistance, and she left the ICU next morning, uneventfully.

Discussion

There are few reports on general anesthesia for PSP patients. Epidural anesthesia for a patient with PSP was reported, but airway management was not needed in that patient [3]. Warwick and Popat [4] have reported that PSP is an unusual cause of trismus. The disease is characterized by early postural instability, with a notable tendency to tilt backwards, and a vertical supranuclear-gaze palsy [2]. The patient's head is retracted and the patient is unable to look down [2]. The cervical backward tilt, but not airway stenosis, is well known as a symptom of PSP. The importance of tau-protein accumulation has been noted [2], but the etiology of the cervical backward tilt is not known. In the present patient, we found airway stenosis before the operation, by MRI. We tried awake orotracheal intubation because orotracheal intubation would have provided a better surgical field than tracheotomy. However, orotracheal intubation was impossible because of the airway stenosis caused by the cervical backward tilt and neck contracture.

Warwick and Popat [4] reported that awake fiberoptic nasal intubation ensured safe anesthesia for PSP. In the present patient, oral or nasal intubation seemed to be impossible, because the diameter of the trachea at the narrowest part was about 5.0 mm.

The symptoms of PSP include Parkinsonism, and PSP is sometimes misdiagnosed as Parkinsons disease [1]. We should be careful with anesthesia for patients diagnosed with Parkinsons disease, and should check for symptoms and signs such as neck contracture and cervical backward tilt. Cervical MRI is useful to assess for the presence of airway stenosis.

The influence of anesthetics on PSP is unclear. There is no evidence that anesthetics exacerbate the neurological dysfunction in patients with degenererative neural disease. The present patient had an advanced stage of disease, and many functions had been already lost. Thus, no adverse effect of anesthetics on her neurological function was observed.

It is unknown that the action of muscle relaxants on patients with PSP is same or not. We monitored our patient's facial muscular relaxation (using train-of-four pulses), because the limbs showed contracture. The effect of vecuronium was not prolonged, and spontaneous respiration appeared after the operation, without a reversal procedure.

Patients with PSP usually show a cervical backward tilt, and this may cause airway stenosis. Specific airway management, such as awake fiberoptic nasal intubation or tracheotomy, may be necessary for general anesthesia in these patients.

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