Anesthetic management in corticobasal degeneration with central sleep apnea: A case report

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Corticobasal degeneration (CBD) is a rare neurodegenerative disease characterized by dystonia, cognitive deficits, and an asymmetric akinesic-rigid syndrome. Little information is available regarding anesthetic management for CBD patients. Our patient was a 55-year-old man with CBD complicated by central sleep apnea (CSA). Due to the risk of perioperative breathing instability associated with anesthetic use, a laryngeal mask airway was used during anesthesia with propofol. Spontaneous respiration was stable under general anesthesia. However, respiratory depression occurred following surgery, necessitating insertion of a nasopharyngeal airway. Since no respiratory depression had occurred during maintenance of the airway using the laryngeal mask, we suspected an upper airway obstruction caused by displacement of the tongue due to residual propofol. Residual anesthetics may cause postoperative respiratory depression in patients with CBD. Therefore, continuous postoperative monitoring of SpO₂ and preparations to support postoperative ventilation are necessary.

Keywords: Corticobasal Degeneration; Laryngeal Mask Airway; Propofol; Sleep Apnea.

INTRODUCTION

Corticobasal degeneration (CBD) is a rare neurodegenerative disease with a distinctive clinical picture, including features related to both cortical and basal ganglia dysfunction [1]. Some characteristic signs of CBD include asymmetric upper limb dystonia, cognitive deficits, myoclonus, gait abnormalities, and asymmetric akinesic-rigid syndrome [2]. CBD patients may develop rigidity and dystonia, thereby necessitating anesthetic management for dental treatment. Few reports have discussed anesthetic management in CBD patients. Central sleep apnea (CSA) encompasses a wide range of conditions resulting in the cessation of airflow without respiratory effort [3]. CSA can be provoked or exacerbated by the effects of sedative and anesthetic agents. This report describes our experience with general anesthesia for dental treatment in a patient with CBD complicated by CSA. We obtained informed consent from the patient’s legal guardian.

CASE PRESENTATION

The patient was a 55-year-old man with a body weight of 61 kg, height of 170 cm, and a body mass index of 21 kg/m². At age 50, he developed gait abnormalities, myoclonus, asymmetric dystonia in the left upper limb, and asymmetric akinesic-rigid syndrome on the left side.
Levodopa and dopamine agonists proved ineffective. His symptoms gradually worsened. By age 52, he could not walk. Cerebral blood flow scintigraphy revealed right frontoparietal atrophy, leading to CBD diagnosis. The clinical phenotype of CBD in this patient was diagnosed as cortical basal syndrome (CBS). At age 53, he developed sleep-disordered breathing at night. The patient's sleep condition was confirmed as breathing pauses without snoring. Polysomnography revealed patterns characteristic of CSA. At age 54, his dysphagia worsened, and he underwent gastrostomy insertion.

He visited our hospital due to pain in the oral cavity. He exhibited marked dystonia in the upper limb and orofacial areas. He was confined to a wheelchair and required assistance to eat, use the toilet, and get dressed and undressed. Brushing his teeth was also difficult; his toothbrush broke in his mouth due to oral rigidity. Since normal dental care was impossible, he underwent tooth scaling and several extractions under general anesthesia. His medications included baclofen (15 mg/day), dantrolene sodium hydrate (75 mg/day), and eszopiclone (1 mg/day).

His preoperative blood pressure (BP) and heart rate (HR) were 132/78 mmHg and 65 bpm, respectively. His SpO2 was 98% on room air, and arterial blood gas values were within the normal range. Blood tests revealed no abnormalities. However, respiratory function tests could not be performed. Supraventricular and ventricular extrasystoles were observed on electrocardiography (EKG), which also revealed a left ventricular ejection fraction of 51%.

The patient fasted for 8 h and refrained from drinking water for 2 h prior to general anesthesia. He was attached to a pulse oximeter, EKG leads, a sphygmomanometer, and a bispectral index (BIS) monitor. A 22-gauge intravenous catheter was placed in the left forearm, and 100% oxygen was initiated via a face mask (3 L/min). His initial vital signs were as follows: BP, 142/90 mmHg; HR, 68 bpm; and SpO2, 100%. After 3 min of preoxygenation, a propofol bolus was administered (2 mg/kg; 120 mg). After spontaneous breathing ceased, ventilation with a face mask was performed. A laryngeal mask airway (LMA) was inserted 90 s after propofol injection. Vital signs immediately after insertion were as follows: BP, 130/93 mmHg; HR, 62 bpm; SpO2, 100%; and BIS, 38. Ventilation was performed after LMA insertion. After 200 s, spontaneous respiration resumed. Anesthesia was maintained under the following conditions: oxygen, 2 L/min; air, 4 L/min; propofol, and 4-6 mg/kg/h. The LMA was fixed at the right corner of the mouth to secure an adequate surgical field for the surgeon. Thereafter, infiltration anesthesia was achieved using 3.6 mL of 2% lidocaine containing 1:80,000 epinephrine. Following scaling, four teeth were extracted. He was administered 1,000 mg of acetaminophen intravenously, 15 min before the end of the dental treatment. Spontaneous respiration was preserved with an SpO2 of 98-100%, respiration rate of 18 to 28 breaths/min, intraoperative tidal volume of 300-360 ml, and an end-tidal CO2 level of 44-50 mmHg.

Intraoperative BP, HR, and BIS values were maintained at 92-126/56-75 mmHg, 54-63 bpm, and 48-60, respectively. The intraoral procedures caused no movements or cough reflex. Propofol administration was discontinued after the final procedure. The total dose of propofol used was 580 mg. Eleven minutes later, the patient began to awaken. The LMA was removed, and oxygen was administered via a face mask at 3 L/min. The patient appeared somnolent after LMA removal and exhibited respiratory depression, which was alleviated via the placement of a nasopharyngeal airway. The patient fully awoke 20 min after LMA removal, and the nasopharyngeal airway was removed. His SpO2 was 100%, and his breathing was stable. The total time required for dental procedures was 70 min, and the duration of anesthesia was 95 min. He continued to receive oxygen and underwent pulse oximetry and EKG monitoring until the next morning. No nighttime respiratory depression was observed. He was discharged the following day. He experienced no complications following discharge.
CBD is a relentlessly progressive neurodegenerative condition with a mean disease survival rate of approximately seven years [1], although the shortest reported duration of survival was 24 months [4]. Previous criteria for CBD reflected a single phenotype only: CBS, which was thought to be pathognomonic for CBD. However, CBS is a CBD phenotype, and new criteria have been developed for other phenotypes [1]. CBD is associated with asymmetric upper limb dystonia, alien limb phenomenon, myoclonus, dysarthria, eye movement disturbance, gait abnormalities, and cognitive-behavioral and/or language disorders (i.e., CBS) [2]. The clinical diagnosis of CBS may be supported by the results of different neuroimaging and neurophysiological investigations, since asymmetric frontoparietal atrophy can be observed during magnetic resonance imaging [5]. Single-photon emission computed tomography can also be used to observe hypoperfusion and hypometabolism in the frontoparietal regions in CBS patients [6].

Dental procedures may be difficult in CBD patients due to dystonia in the orofacial area. Thus anesthetic management is necessary. However, there are no guidelines for anesthetic management in CBD patients. Given the progressive nature of CBD, it is difficult to predict responses to anesthetic agents. An additional complication of the present case was CSA, which is occasionally observed in patients with neurological disorders. CSA results from a reduction in output from the central respiratory generator in the brainstem, manifesting as apnea and hypopnea without discernible effort [3]. CSA patients experience hypoventilation during sleep; the ventilatory drive is reduced and compensatory mechanisms fail [7]. Therefore, administration of sedative and anesthetic agents may worsen CSA or induce sleep apnea [8]. Since the patient was prone to problems with respiratory status after general anesthesia and was at risk for hypercapnia and apnea, we chose anesthetic management by spontaneous breathing to avoid postoperative respiratory depression. Furthermore, his respiratory state after surgery could be predicted from the state of spontaneous breathing during anesthesia. An LMA was chosen because it can be inserted without the use of a muscle relaxant, allowing for rapid recovery of spontaneous breathing. Additionally, anesthetic management with LMA requires less total anesthetic dose. Although further studies are required to determine the comparative advantages/disadvantages of intravenous or inhaled anesthetic agents, we selected propofol due to its short, context-sensitive half-life [9] and ability to suppress pharyngeal reflex.

No apneas/hypopneas were observed during the procedures, and spontaneous respiration was stable. However, despite the removal of the LMA after adequate emergence, respiratory depression occurred after surgery. Since no respiratory depression was observed during airway maintenance using the LMA, CSA was unlikely to be the cause of the respiratory depression. Respiratory depression improved following insertion of a nasopharyngeal airway, suggesting that the patient experienced an upper airway obstruction caused by displacement of the tongue due to residual propofol. Our patient may have had mixed sleep apnea complicated by CSA and obstructive sleep apnea [10]. Although sleep apnea syndromes are frequently associated with other extrapyramidal syndromes, no reports have described such syndromes in CBD patients, likely due to the rarity of CBD. Nocturnal akinesia of the upper-airway muscles may be responsible for sleep apnea [11]. Our study highlights the importance of continuous postoperative oxygenation and SpO2 monitoring. Additionally, preparations for continuous positive airway pressure and non-invasive positive pressure ventilation may be necessary to support postoperative ventilation in CBD patients.

In a meta-analysis of the incidence of LMA and aspiration pneumonia, only three out of 112,901 patients experienced aspiration, suggesting that LMA use does not increase the risk of aspiration, unlike tracheal intubation [12]. Dysphagia is a frequent complication in advanced
cases of CBD. As a result, aspiration pneumonia is among the main causes of death in CBD patients [13]. We chose LMA because the benefits of preserving spontaneous breathing outweighed the risk of aspiration in this patient with CBD and CSA. Clinicians should decide carefully whether to use tracheal intubation or supraglottic airway devices for CBD patients.

**DECLARATION OF INTEREST:** There are no financial or other conflicts of interest to declare.

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