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Aggravated sleep apnea after general anesthesia in a patient with Shy-Drager syndrome

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Introduction

Shy-Drager syndrome is a rare, chronic, progressive disease characterized by multiple system atrophy and autonomic failure [1]. Anesthetic literature has mainly focused on cardiovascular management during surgery [2,3], but respiratory problems in the perioperative period have rarely been mentioned [4,5]. In addition to upper airway obstruction during sleep, manifested as loud snoring and stridor [6,7], impaired central generation of respiratory movements has been reported [6,8] in patients with Shy-Drager syndrome. We report a case in which intraoperative management was not eventful, but remarkably aggravated sleep apnea was observed in the postoperative period.

Case report

A 62-year-old, 45-kg woman was admitted to the hospital for repair of an accidentally fractured femoral neck 6 months after a diagnosis of Shy-Drager syndrome was made. At the time of admission the patient showed urinary incontinence, sweating disturbance, dysarthria, and orthostatic hypotension in which blood pressure decreased from 150/102 mmHg in the supine position to 102/80 mmHg on standing without a compensatory rise in the heart rate. There was no alteration in heart rate during immersion of a limb in iced water. A Valsalva maneuver demonstrated a blocked response with no pressor overshoot and no bradycardia. Neurological examination revealed positive pyramidal signs and cerebellar dysfunction, and laryngoscopy revealed left abductor vocal cord paralysis. Magnetic resonance imaging (MRI) revealed atrophy in the brainstem and the vermis of the cerebellum. Her only medication was midodrine hydrochloride, a selective α_1 agonist, at the time of admission.

The patient was orally premedicated with diazepam 6 mg, and roxatidine acetate 75 mg 2 h before surgery. Preoperative arterial blood gas analysis values were: pH 7.414, $Paco_2$ 42.8 mmHg, and Pao_2 86.1 mmHg. Anesthesia was induced with ketamine 90 mg i.v.. Tracheal intubation was facilitated by succinylcholine 40 mg i.v. and controlled ventilation was instituted, during which there was no increase in blood pressure or heart rate. Anesthesia was maintained with diazepam 10 mg i.v., fentanyl 300 μg in total (6.7 $\mu g \cdot kg^{-1}$), continuous intravenous infusion of ketamine 1.5 $mg \cdot kg^{-1} \cdot h^{-1}$ in FIo_2 0.40 with air. Muscle relaxation was achieved with vecuronium bromide, 4.5 mg in total during the procedure of right femoral head replacement lasting 2 h. Intraoperative hemodynamics were stable. Systolic blood pressure and heart rate fluctuated within a range of 120 and 170 mmHg, 70 and 92 beats·min^{-1}, respectively, throughout anesthesia. No sympathomimetics were used. The tracheal tube was removed 45 min after the discontinuation of ketamine. Changes in blood pressure and heart rate did not occur.

The patient was transferred to the intensive care unit (ICU). She was slow in response to verbal commands, and did not phonate clearly, but she breathed smoothly and adequately while she was awake. During sleep, however, loud snoring and frequent irregular apneic episodes were seen. The $Paco_2$ fluctuated widely from 46.2 mmHg to 62.9 mmHg during the night. The lowest Pao_2 was 110 mmHg with 3–5 l·min^{-1} of supplemental oxygen through a face mask. She complained of hoarse-

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ness, aggravated dysarthria, and difficulty in sputum clearance. Frequent suctioning of sputum was needed throughout the night. Systolic blood pressure once decreased to 70 mmHg due to postoperative bleeding, which was treated with rapid blood transfusion. We observed no compensatory rise in the heart rate. Her body temperature rose to 39.6°C and remained at around 38.5°C during the following night despite the administration of diclofenac sodium suppositories (an antipyretic and nonsteroidal antiinflammatory drug (NSAID) and surface cooling with ice bags. We scarcely noticed her perspiration.

On the 2nd postoperative day, the patient was able to expectorate without help, and her phonation had returned to the preoperative level. However, supplemental oxygen was continuously provided to treat her hypoxemia, which was Pao_2 53.8 mmHg when breathing room air. Her $Paco_2$ was 45.9 mmHg. No obvious cause for hypoxemia was found at that time. Postoperatively, her oxygen saturation (Spo_2) was monitored with a pulse oximeter on consecutive nights. The 8-h trend graphs in Spo_2 during sleep, breathing room air, and breathing 0.5 l·min^{-1} of supplemental oxygen through a nasal cannula demonstrated that the low flow of additional oxygen had effectively decreased the incidence and the magnitude of arterial desaturation (Fig. 1). The patient was successfully weaned from oxygen therapy on the 8th postoperative day. Thereafter, we observed not more than three periodic decreases in Spo_2 less than 90% at night. She started ambulation training 15 days after surgery.

Plasma epinephrine levels (normally less than 0.12 ng·ml^{-1}) were 0.02, 0.03, and 0.02 ng·ml^{-1} before induction of anesthesia, during surgery, and 1 day after surgery, respectively. Plasma norepinephrine levels (normally within a range of 0.10 and 0.41 ng·ml^{-1}) were 0.12, 0.11, and 0.18 ng·ml^{-1}, respectively.

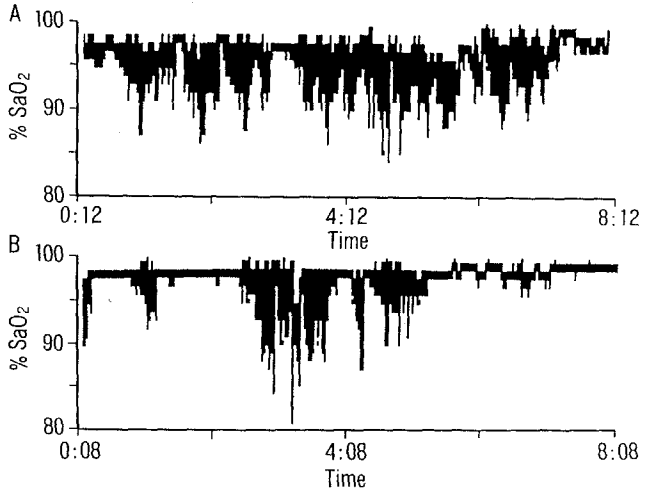

Fig. 1A,B. The 8-h trend graphs from midnight to 8 a.m. in oxygen saturation with a pulse oximeter shows that the low flow of 0.5 l·min^{-1} of supplemental oxygen through a nasal cannula (**B**) effectively decreased the incidence and the magnitude of arterial desaturation compared to those in room air (**A**). Trend graphs **A** and **B** were recorded on the 4th and 5th postoperative days, respectively

Discussion

Loud snoring and stridor during sleep are the most commonly observed clinical manifestations of upper airway obstruction in Shy-Drager syndrome [6,7]. In such cases, laryngoscopy has usually revealed either unilateral or bilateral vocal cord paralysis [7–9]. Degeneration in the nucleus ambiguus and in the retrofacial nucleus is likely to be responsible for the paralysis as well as for the loss of control over laryngeal muscles, presumably the cause of snoring [7]. Our patient could not abduct her left vocal cord, and she could have developed bilateral paralysis at any time which would be an acute emergency. Sudden death has been reported in patients with bilateral vocal cord paralysis [10,11]. Early tracheostomy in the presence of mild obstruction during sleep could be justified because it is at present the only effective treatment for bilateral paralysis.

The poor expulsive mechanism for clearing sputum in the present case was due to the residual hypnotic effect of diazepam and ketamine, and to prolonged inhalation of cold, dry gas which decreases ciliary activity [12]. This was aggravated by the transient deterioration in laryngeal function induced by anesthesia, which was clinically manifested as hoarseness and dysarthria. Laryngeal edema after tracheal extubation, weakness of inspiratory muscles [7], and the difficulty in sputum clearance would increase the risk of total glottic obstruction even in unilateral cord paralysis, as in this case. Minitracheostomy or minicricothyrotomy might have been performed if the difficulty in sputum management had persisted.

During sleep, behavioral and state-related ventilatory excitation is minimized or abolished, and breathing is highly dependent on the metabolic control involved in blood-gas homeostasis and acid-base balance [13]. Because the metabolic control system is a component of the autonomic nervous system, a defective respiratory pattern during sleep can occur in cases with Shy-Drager syndrome. This is supported by the data of markedly impaired ventilatory responses to hypoxia and a reduced response to hypercapnia in those with autonomic failure [8]. It is also reported that such patients are highly sensitive to tranquilizers and opioids [14,15]. Therefore, patients with Shy-Drager syndrome are at

significantly greater risk for catastrophic arterial desaturation during sleep after general anesthesia. In the present case, supplemental oxygen was provided for 7 days after surgery, during which we continuously monitored arterial oxygenation using a pulse oximeter. As seen in Fig. 1, a very low flow of oxygen successfully diminished the incidence and the magnitude of arterial desaturation during sleep. Since additional oxygen is innocuous under most circumstances [16], supplemental oxygen should be provided for sufficient length of time in the postoperative period.

During surgery and the following stay in the ICU, the preexisting autonomic dysfunction in our patient was presumably further impaired. Plasma catecholamine levels were extremely low before surgery, and showed no elevation either during surgery or 1 day after. Her heart rate was relatively fixed, and the changes in blood pressure were primarily dependent on her blood volume. Disturbed perspiration played a role in the persistent high fever in the postoperative period. Although the administration of NSAIDs, adding to the residual analgesic effect of ketamine and its metabolites, afforded excellent pain relief in our patient, the use of parenteral opioid agonists or opioid agonist-antagonists for pain control in the postoperative period might be needed in case of abdominal or thoracic surgery which would then increase the risk of additional impairment of autonomic function. Extended monitoring of the respiratory and cardiovascular systems would also be mandatory in the postoperative period as well as during surgery in patients with autonomic failure.

In conclusion, considerably impaired central generation of respiratory movements and aggravated upper airway obstruction due to the difficulty in sputum clearance were observed in a patient with Shy-Drager syndrome. A low flow of supplemental oxygen successfully diminished the incidence and the magnitude of arterial desaturation during sleep. In these patients, not only must their circulatory instability be anticipated, but the risk of sudden or slowly developing respiratory problems as well.

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