

Successful bilevel positive airway pressure therapy in a patient with amyotrophic lateral sclerosis after emergency laparotomy: A case report

ABSTRACT

Patients with amyotrophic lateral sclerosis (ALS) present an increased risk of postoperative respiratory failure after general anesthesia. We report the case of a 71-year-old man with ALS who underwent emergency laparotomy for small bowel strangulation. After surgery, he remained intubated and was transferred to the high care unit under mechanical ventilation, due to unstable hemodynamics requiring inotropic support. On postoperative day (POD) 3, he was extubated under stable hemodynamics and respiratory status. Immediately after extubation, bilevel positive airway pressure (bilevel PAP) was prophylactically applied to prevent postoperative respiratory failure, which may have been caused by respiratory muscle fatigue, attributed to general anesthesia and surgical stress. On POD 7, bilevel PAP was smoothly weaned off because no signs and symptoms of respiratory failure were observed. On POD 10, he achieved 30 m-walk without rest. No postoperative complications were observed up to one month after surgery. Postoperative respiratory failure may lead to death in patients with neuromuscular disorder. Non-invasive ventilation (NIV) reduces respiratory muscle fatigue, resulting in easy sputum expectoration, promoting CO₂ washout, and better oxygenation. Consequently, the prophylactic use of NIV to avoid postoperative respiratory insufficiency should be considered in patients with ALS after emergency operation under general anesthesia.

Key words: Amyotrophic lateral sclerosis; bilevel positive airway pressure; emergency laparotomy; general anesthesia; postoperative respiratory failure

Introduction


Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that primarily affects the upper and lower motor neurons and eventually leads to death due to respiratory failure.^[1] Degeneration of motor neurons of bulbar muscles leads to upper airway and respiratory symptoms such as dysphagia, dysarthria, and dyspnea.^[2] General anesthesia

may cause fatal respiratory depression in patients with ALS because of their aberrant sensitivity to neuromuscular blocking agents (NMBA).^[3] In patients with ALS, noninvasive ventilation (NIV) helps in the management of respiratory muscle weakness.^[4] For example, NIV improves the lung volumes without interfering with the contribution of the chest wall compartment in ALS patients.^[5]

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We report successful postoperative management of a patient with ALS who underwent emergency laparotomy using prophylactic bilevel positive airway pressure (bilevel PAP) therapy as a step-down alternative.

Case Report

A 71-year-old man (body weight 53 kg; height 160 cm) with a one-year history of ALS and mild chronic obstructive pulmonary disease (COPD) presented at our emergency room with complaints of severe abdominal pain. Enhanced computed tomography (CT) revealed that he had small bowel strangulation. Surgeons planned immediate operation to release intestinal obstruction. He had advanced bulbar symptoms with mild dysphagia and severe dysarthria. However, he had almost no weakness in his lower and upper limbs (manual muscle test [MMT] 4/5) without any respiratory symptoms. He was taking 100 mg riluzole daily for the treatment of ALS. His COPD was well-controlled with inhaled ciclesonide. His preoperative laboratory tests showed white blood cells (WBC) 9130/ μ L, Hb 9.0 g/dL, C-reactive protein (CRP) 18.9 mg/dL, and activated partial thromboplastin time (APTT) 44.5 sec. His preoperative blood gas analysis (BGA) at room air showed PaO₂ 102 mmHg, PaCO₂ 40 mmHg, and pH 7.49. Because of severe abdominal pain and the acceptable results of BGA, preoperative pulmonary function test was not performed.

In the operating room, his blood pressure (BP), heart rate (HR), respiratory rate (RR), and SpO₂ at room air were 100/60 mmHg, 110 bpm, 20 per min, and 98%, respectively. Because we evaluated that he was in impending shock status and his stomach was assumed to be full, awake intubation was performed after preoxygenation and subsequent administration of 50 mcg fentanyl. After tracheal intubation, 30 mg propofol and 30 mg rocuronium were administered slowly and then anesthesia was maintained via 1 L/min oxygen and 2 L/min air with 1.5% sevoflurane and intravenous fentanyl. No additional rocuronium was administered after intubation. During surgery, his hemodynamic status was unstable despite continuous infusion of noradrenaline. At the end of surgery, considering his hemodynamic instability and neuromuscular comorbidity, he remained intubated and was transferred to high care unit (HCU) under mechanical ventilation.

On postoperative days (PODs) 1 and 2, he was sedated with continuous infusion of propofol and mechanically ventilated. On POD 3, due to stable hemodynamic status, he was weaned off inotropic support followed by sedation. After confirming that this respiratory status (PaO₂/FiO₂ ratio: 250, rapid-shallow breathing index (RSBI): 40 breaths/min/L, and peak cough flow: 60 L/min) met the criteria for extubation, the patient

was extubated. After extubation, to avoid postoperative respiratory failure, bilevel PAP therapy with spontaneous (S) mode was prophylactically applied immediately using V60[®] ventilator (Philips Respironics, Murrysville, Pennsylvania, United States) with oronasal mask. Initial settings included inspiratory positive airway pressure (IPAP) 7 cm H₂O, expiratory positive airway pressure (EPAP) 4 cm H₂O, and FiO₂ 0.4. On POD 3, bilevel PAP was applied only during day time owing to the patient's preference. During night-time sleep, bilevel PAP was temporarily weaned off and oxygen was administered via face mask. During the morning of POD 4, his BGA under 5 L/min of oxygen showed mild CO₂ retention (PaCO₂ 47 mmHg). Therefore, bilevel PAP was applied during both daytime and night-time sleep during PODs 4–6. During the morning of POD 7, bilevel PAP was completely weaned off, and his BGA under 3 L/min of oxygen showed PaO₂ 94 mmHg, PaCO₂ 43 mmHg, and pH 7.44 in the evening. Because no signs of respiratory failure were observed, oxygen supply was also weaned off on POD 8. On POD 10, he achieved 30 m-walk without rest in a ward. During the postoperative period in HCU, his chest radiograph showed no abnormalities. No postoperative complications were observed up to one month after surgery.

Discussion

In the present case, we described the efficacy of the prophylactic application of NIV in ALS patients after emergency operation under general anesthesia. Patients with ALS usually die from respiratory failure, which is due to respiratory muscle weakness caused by loss of bulbar, cervical, and thoracic motor neurons.^[6] Because of diminished respiratory function and aberrant sensitivity to NMBA, general anesthesia with NMBA increases the risk of postoperative respiratory failure.^[7] Some reports have described successful anesthetic management of ALS patients undergoing elective surgery by using regional anesthesia and general anesthesia without NMBA.^[8,9] However, general anesthesia with NMBA could not be avoided in our case. Postoperative respiratory failure was highly expected in our case because he had neuromuscular disorder with COPD. We hypothesized that after extubation, respiratory collapse may have been caused mainly by difficult sputum expectoration due to his coexisting COPD and respiratory muscle fatigue, triggered by general anesthesia and surgical stress. Furthermore, considering the possibility of ventilator-associated pneumonia, we considered that earlier extubation should be performed in our case. However, our ALS patient with COPD had required inotropic support due to hemodynamic instability until POD 3. Considering the possibility of postoperative respiratory failure often observed in ALS patients and hemodynamic

instability in the present case, weaning from mechanical ventilation was performed carefully to avoid reintubation. As a result, mechanical ventilation was continued until POD 3.

Noninvasive ventilation (NIV) reduces respiratory muscle fatigue, promoting CO₂ washout, and better oxygenation.^[10] Furthermore, it can help facilitate expectoration of respiratory secretions. NIV after extubation is effective in avoiding respiratory failure and improving survival in patients with chronic respiratory disorders.^[11] Koyama *et al.* reported the effectiveness of NIV on postoperative respiratory failure in a patient with myotonic dystrophy.^[12] Olivieri *et al.* described that the use of NIV after elective surgery under general anesthesia safely prevents postoperative respiratory failure in ALS patients.^[7] Consequently, we chose prophylactic bilevel PAP therapy immediately after extubation as a step-down strategy to protect from postoperative respiratory failure. Our patient appeared to be in a mild form of ALS at the time of surgery. However, because of his coexisting COPD, we strictly applied the prophylactic strategies and then stepped down these therapies carefully. In our case, we prophylactically used low-pressure NIV. This may be a potential concomitant effect on helping with atelectasis after laparotomy. Because postoperative respiratory failure in such cases may lead to death, prophylactic application of NIV immediately after extubation may be critical to avoid potentially fatal postoperative respiratory failure.

In conclusion, prophylactic application of NIV should be considered in patients with ALS after emergency operation under general anesthesia with NMBA.

Consent for publication

Written informed consent was obtained from the patient and his brother for publication of this case report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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