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Management of anesthesia in a case of Kennedy's disease undergoing laparoscopic gastric volume reduction surgery

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Case Report

The patient was male, 49 years old, height 168 cm, weight 86 kg, BMI 30.5 kg/m^2, and was admitted to hospital because of obesity and apnea syndrome. He has a family history of Kennedy's disease and has been diagnosed as Kennedy's disease through genetic testing. He walks with a "duck walk" and has normal muscle strength on both upper limbs. The proximal muscle volume of his both lower limbs was decreased. And the muscle strength of both lower limbs was grade IV, while the tendon reflex was disappeared and the pathological signs were not drawn out. He had a history of hypertension for 1 year, and olmesartan tablets were taken orally. His blood pressure was controlled at 110~130/70~90 mmHg (1 mmHg=0.133 kPa). He was diagnosed with Diabetes for 3 years, accompanied by blurred vision for 2 years. He had orally metformin hydrochloride tablets for treatment, and blood glucose control is good. The tongue muscles of the patient were asymmetrical, atrophied, dysphonia, and occasionally mispharyngeal, accompanied by severe cough, suggesting that the pharyngeal reflex was weakened. The head and neck movement of him was normal, and the mouth opening was 3 cm, Mallampati grade I, and the nail-chin distance was >6 cm. The head and neck CT examination showed no obvious abnormalities, indicating that there was no difficult airway. Blood routine examination, blood gas analysis and lung function showed no obvious abnormality and the lung function was close to the critical level. Laparoscopic sleeve gastrectomy is planned under general anesthesia.

Before surgery, BP 135/85 mmHg, HR 90 times /min, SpO_2 99%, TOF 100%, CVP 6 cm H_2O were monitored. Right radial artery catheterization and right internal jugular vein catheterization were performed under ultrasound guidance.

Anesthesia introduction

Etomidate 0.2 mg/kg, rocuronium 0.9 mg/kg, midazolam 0.05 mg/kg, sufentanil 0.6 μg/kg were injected intravenously. Endotracheal intubation was performed under visual laryngoscope, and the Mechanical ventilation parameters were set as tidal volume 6 mL/kg, respiratory rate 15 times/min, inhalation-exhalation ratio 1:2, oxygen concentration 60%, PEEP 5 cm H_2O.

Anesthesia maintenance

Anesthesia was maintained by inhalation of 2% sevoflurane and intravenous infusion of 2% propofol 40 $\mu g \cdot kg^{-1}Min^{-1}$ remifentanil 0.2 $\mu g \cdot kg^{-1}Min^{-1}$ and rocuronium 10 $\mu g \cdot kg^{-1}Min^{-1}$. During the operation, we kept BP 100~140/70~90 mmHg, HR 70~ 100 times/min and SpO_2 99%~100%, BIS 40~60, CVP 6~10 cm H_2O, $P_{ET}CO_2$ 35~45 mmHg and the Post-tetanic count (PTC) for 0. The operation was successful, and the patient was sent to ICU for further treatment without extubation.

After transfer to ICU, synchronized intermittent mandatory ventilation (SIMV) mode was used with tidal volume 6 mL/kg and frequency 13 times /min, and oxygen concentration 40%. 167 min later after surgery, the endotracheal intubation was removed and oxygen inhalation was replaced by nasal catheter. The SpO_2 was above 98%. He was discharged on the 5th day after surgery. One month after the operation, the body weight dropped 12 kg. Blood pressure and blood sugar returned to normal. Sleep snoring was significantly reduced, and the ventilator was no longer needed.

Kennedy's disease, also known as spinal cord and medulla muscular atrophy, is a motor neuron disease with x-linked recessive inheritance caused by abnormal amplification of CAG. The CAG repetition range of androgen receptor gene in normal people is lower than 36, while that in Kennedy's patients is 38-62 [1-4]. Kennedy's disease is characterized by slow progressive muscle weakness in the extremities. It usually happens between the ages of 30 and 50, with an incidence rate of 1 in 40 000. The common causes of death are pneumonia and respiratory failure, but so far there are no treatments that can slow or reverse the progression of Kennedy's disease [5].

Preoperative assessment (1) Disease assessment: Referring to the evaluation of the disease degree of Kennedy patients by the Department of Neurology, the Kennedy Disease 1234 scale was evaluated before surgery, which mainly assessed the medullar function, upper limb function, lower limb function and respiratory function [6]. (2) Anesthesia risk assessment: The most important assessment is medulla oblongata function and respiratory function. In accordance with the "Kennedy disease 1234 scale" in medulla oblongata function evaluation [LANGUAGE: normal for 3 points, dysarthria, but can be understood for 2 points, the language is hard to be understood for 1 point, language cannot be understood for 0 point; SWALLOWING: normal for 3 points, early dysphagia (occasionally, choking cough a lot) for 2 points, eating speed significantly slower (often, choking cough) for 1 point, need stomach tube, parenteral nutrition for 0] and respiratory function assessment [after breathing normal three points; mild activities (such as walking, talking, etc) or felt shortness of breath 2 points, 1 points resting felt shortness of breath; need help breathing machine 0]. The patient's medulla oblongata function was 4 points (dysphonia, inability to pronounce certain words correctly, and lisp was rated as 2 points; the respiratory function was rated as 3 points (at present, there was no difficulty in short distance walking on the ground was rated as 3 points), and the total score was 7 points (the full score of the two items was 9 points), indicating that the less damage to the patient's medulae oblongata and respiratory function, the lower the risk of anesthesia.

Anesthesia methods

The patient in this case had obesity and apnea syndrome, which would increase the difficulty of mask ventilation and endotracheal intubation. And the obese patient had reduced functional residual capacity and oxygen reserve [7]; Kennedy disease is accompanied by more medulla oblongata dysfunction caused by pharyngeal muscle activity obstacle. So, before intubation and extubation, the patient is placed by the beach chair position. This position can effectively keep the throat unobstructed, reduce the risk of glossoptosis. And at the same time can make the diaphragm go down, which could increase oxygen reserves and reduce the influence on pulmonary ventilation [8]. Airway assessment and head and neck CT scanning suggested that the patient did not have a difficult airway, so rapid sequential induction was used. Anesthesia management was referred to patients with obesity, sleep apnea syndrome, and peripheral nervous system disease.

Airway management

In patients with Kennedy's disease, lung function is reduced, and respiratory muscle fatigue exists [9]. Decreased pulmonary ventilation function can increase postoperative complications. The lung function of the patient in this case was still in the normal range, but lower than the predicted value. Therefore, lung protection strategy should be adopted during the operation, using small tidal volume +PEEP ventilation mode to reduce the influence on postoperative lung function. Ventilators can be set at 6-8 mL/kg for ideal body weight. And according to $PaCO_2$ pressure, we set the ventilation frequency so that the peak airway pressure would not exceed 30 cm H_2O; there is no regulation on the value of PEEP, as long as reasonable physiological status can be maintained [7].

Tracheal extubation and related complications

In this patient, muscle relaxation monitoring instrument was routinely used to monitor muscle relaxation, but the signs and symptoms of neurodegenerative diseases in some patients with Kennedy disease may be aggravated after surgery, and there is a possibility of Re-intubation. It has been reported that the tracheal tube was re-intubated 3 days after the removal of the tracheal tube by cardiopulmonary bypass in patients with concomitant Kennedy disease [5]. It has also been reported that 2 of the 6 patients with concomitant Kennedy disease underwent re-intubation and continued to be treated with ventilator after receiving general anesthesia [10]. Therefore, in this case, the tracheal tube was not immediately removed after surgery, but was transferred to ICU for ventilator and the tracheal tube was removed very circumspectly.

After tracheal extubation in patients with Kennedy's disease, pharyngeal muscle abnormalities can occur, manifested as dysarthria, progressive dysphagia, and progressive loss of consciousness, which can occur on the second postoperative day. Dysphagia can be characterized by choking during feeding, increasing the risk of regurgitation and aspiration, requiring a nasogastric tube for nutrition. The exacerbation of these neurodegenerative signs took about 6 weeks to return to preoperative status [5]. For the prevention of possible complications, the "Kennedy disease 1234 Scale" was continued to be used for postoperative evaluation. If the patient developed progressive pharyngeal muscle dysfunction, re-intubation could be considered as an alternative option. It is recommended that the patient be followed up regularly in neurology outpatient department after surgery until the function is restored to the preoperative status. Luckily, there were No dysphagia or other complications occurred in this patient.

In conclusion, the Kennedy disease scale 1234 can effectively evaluate the medulla oblongata and respiratory function in patients with Kennedy disease during perioperative period. Reasonable preoperative airway assessment, the best way of endotracheal intubation, effective lung protection strategy, prevention of postoperative endotracheal re-intubation, and avoidance of inhalation pneumonia are the key points of anesthesia management in patients with Kennedy disease.

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