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Anesthetic management of a pediatric patient with severe Williams-Campbell syndrome undergoing surgery for giant ovarian tumor

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Abstract
We performed anesthetic management for a patient with severe Williams-Campbell syndrome, which is a congenital deficiency of cartilage in the subsegmental bronchial tree. An 11-year-old girl with this syndrome had labored breathing because of abdominal distension caused by a giant ovarian tumor, and removal of the tumor was scheduled. Because she had been receiving home oxygen therapy for 10 years due to hypoxia, it was possible that positive-pressure ventilation may have increased the risk of perioperative pulmonary complications. So we selected combined spinal and epidural anesthesia with general anesthesia, using a ProSeal (Laryngeal Mask Company, Henley on Thames, UK) laryngeal mask airway. We placed an epidural catheter and induced spinal anesthesia blockade under general anesthesia as the main analgesia technique, in order to maintain spontaneous breathing. The surgery was completed uneventfully and the patient emerged from anesthesia without dyspnea. She had an uneventful recovery and was discharged home.

Key words Williams-Campbell syndrome · Home oxygen therapy · Regional anesthesia

Introduction

Williams-Campbell syndrome (WCS), a rare disorder, is characterized by a congenital deficiency of cartilage in the subsegmental bronchi, leading to distal airway collapse and bronchiectasis [1]. There are no reports in the literature of pediatric patients with WCS who have undergone anesthesia. We performed anesthetic management in a child with WCS.

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Case report

The patient was an 11-year-old girl who was 135 cm tall and weighed 35.9 kg. She had developed an abdominal tumor, which had increased in size rapidly in only 1 month and resulted in labored breathing, due to abdominal distension.

The child had been hospitalized frequently, after birth because of recurrent pulmonary infections. At age 1 year, she was diagnosed with WCS by bronchography and she had received home oxygen therapy (HOT) because of hypoxia (peripheral oxygen saturation [S_{PO_2}], 89% in room air). She was receiving tulobuterol and L-carbocysteine, and had medical examinations periodically. Chest radiography (Fig. 1) showed multiple cystic shadows in both lungs and mainly right pulmonary emphysema, and computed tomography (CT) scan (Fig. 2) showed calcification and hypertrophy of bronchi and cystic bronchiectasis. Ventilation-perfusion scintigraphy showed dysfunction of the left lung. Electrocardiogram showed right ventricular hypertrophy. Echocardiography showed normal left ventricular function and slightly elevated right ventricular pressure, which was estimated to be 50%–60% of the left ventricular pressure.

After admission to hospital for removal of the tumor, she became infected with respiratory syncytial virus (RSV) and received medical treatment with a nebulizing salbutamol-cromoglycate mixture, as well as receiving aminophylline and methylprednisolone. The surgery was postponed until her fever and productive cough subsided (1 week after admission). Preoperatively, S_{PO_2} was 95%–100% with 1 l·min^{-1} supplemental oxygen given via nasal cannulae in a head-up position. She had slight inspiratory and expiratory wheezes and no murmurs. The tumor was diagnosed as an ovarian cystic tumor ($30 \times 25 \times 20$ cm) by abdominal ultrasonography and CT scan (Fig. 3). On blood examination, the white blood cell count was 14 000 mm^{-3}, hematocrit was 39.9%,

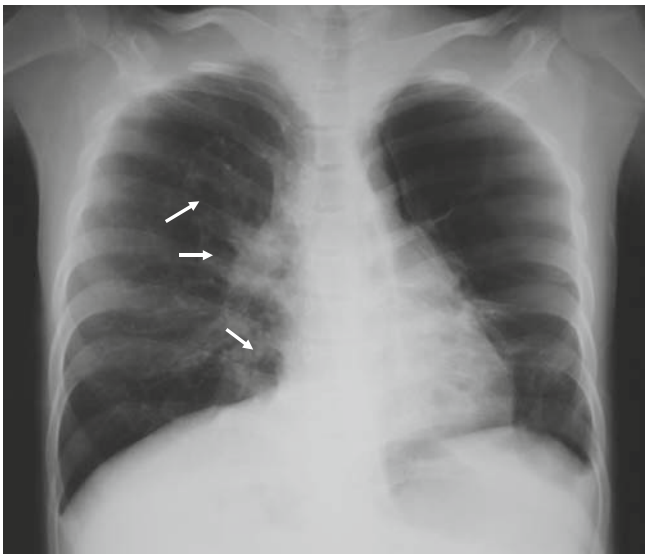

Fig. 1. Chest radiography shows multiple cystic shadows (*arrows*) in both lungs and mainly right pulmonary emphysema

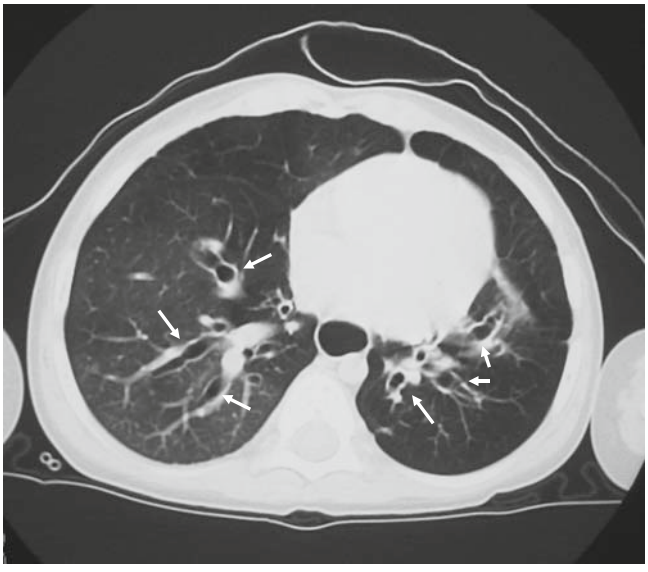

Fig. 2. Chest computed tomography (CT) shows calcification and hypertrophy of bronchi and cystic bronchiectasis (*arrows*)

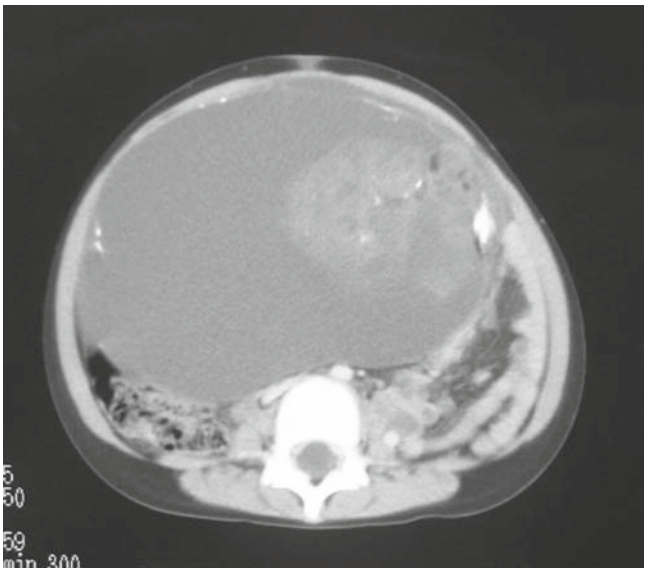

Fig. 3. Abdominal CT shows a giant cystic tumor

platelet count was 382 000 mm^{-3}, and blood chemistry was normal. A clotting screen was normal.

She fasted from the day before surgery. No premedication was given. Standard monitors were applied in the operating room. After preoxygenation, general anesthesia was induced with propofol 100 mg, and then a size 3 ProSeal (Laryngeal Mask Company, Henley on Thames, UK) laryngeal mask airway (PLMA) was placed, without complications, and a gastric tube was inserted through a side hole. There was very little gastric fluid. Anesthesia was maintained under spontaneous breathing with inspired sevoflurane (2%) with an oxygen-air mixture. An epidural catheter was inserted at the Th11 to Th12 interspace, using a loss-of-resistance technique with saline, and spinal anesthesia was induced with 0.5% hyperbaric bupivacaine 2 ml at the L2 to L3 interspace. During this procedure, the patient's respiratory and hemodynamic status was stable. After negative aspiration and a negative test dose of 1% lidocaine with 1:100 000 epinephrine, 1.5% lidocaine 5 ml was injected into the epidural space to obtain a faster onset of spinal anesthesia. Anesthesia was maintained while she remained breathing spontaneously. Sp_{O_2} was kept in the range of 97%–99% on fractional inspired oxygen ($F_{I_{O_2}}$) of 0.35, and end-tidal CO_2 remained in the range of 46–58 mmHg. Her hemodynamic status was stable. The ovarian tumor, which was 6850 g in weight, was removed completely without difficulty. The surgery took 108 min. The PLMA was removed uneventfully. Postoperative chest radiography did not show pneumothorax or atelectasis. The epidural catheter was left for 3 days, with continuous infusion of 0.2% ropivacaine with fentanyl 1.4 $\mu g \cdot ml^{-1}$ at 2 $ml \cdot h^{-1}$, with which her pain was well controlled. Her postoperative condition was satisfactory. Pathologically, the tumor was a teratoma.

Discussion

WCS is an unusual form of bronchiectasis, with cough, wheezing, and recurrent pulmonary infections [1, 2]. Autopsy studies reveal a deficiency of cartilage in the subsegmental bronchial tree [2]. Diagnosis requires a

clinical history indicating WCS, the characteristic expiratory airway collapse on radiological investigation, and the exclusion of other causes of congenital and acquired bronchiectasis [1–3]. Some patients progress rapidly to respiratory failure and death, while others survive into adulthood with variable degrees of recurrent pulmonary infections and respiratory limitations [4, 5]. Although there is no specific therapy for WCS, antibiotics and chest percussion are employed to treat the recurrent pulmonary infections associated with bronchiectasis [2]. Surgical resection of severely bronchiectatic lobes has been described, but without significant improvement being noted [5]. Palmer et al. [3] reported that bilateral sequential lung transplantation might not be an effective therapeutic option in patients with WCS.

The anesthetic management of patients with pulmonary impairment is associated with several risks and challenges. In particular, the respiratory management of these patients is most critical perioperatively. Regional anesthesia should be considered whenever applicable. However, although some authors have reported that abdominal surgery, coronary artery bypass grafting, and aortic aneurysm repair could be safely performed using regional anesthesia alone in adult patients with pulmonary impairment [6–9], regional anesthesia should be carefully considered in children, because depression of abdominal and intercostal muscle function by a thoracic level of regional anesthesia may not be tolerated, and many children who are extremely frightened and anxious about the surgery need some sedation. Instead, general anesthesia, supplemented by regional anesthesia for abdominal or thoracic procedures, can be much better tolerated, is safer, and provides good operative conditions, rapid emergence, and a pain-free postoperative state.

Although endotracheal intubation with muscle relaxation may be mandatory in patients with pulmonary impairment, we should be careful not to excessively inflate lungs in which air is trapped in patients with WCS. Positive-pressure ventilation or positive airway pressure can also exacerbate the emphysema. Kirse et al. [10] reported a patient with a subclinical form of WCS who died after outpatient adenotonsillectomy. They hypothesized that a postoperative decrease in airway pressure associated with the adenotonsillectomy, along with the flaccidity of the bronchial walls, contributed to bronchial collapse, leading to subsequent alveolar hypoventilation and possible asphyxiation. Patients with WCS may need some degree of elevated airway pressure in order to keep their airways patent. We thought that it would be difficult to control the airway pressure for a patient with WCS, although low airway pressure ventilation and spontaneous breathing are suitable for patients with emphysema. Moreover, our patient became infected with RSV only 1 week prior to the planned date of surgery. As residual airway reactivity may persist for months or even years after RSV infection [11], it is unclear how long surgery should be postponed [12]. As our patient had extremely labored breathing due to abdominal distension, we scheduled the surgery immediately after the infectious symptoms subsided. One study has suggested the possible advantages of a laryngeal mask airway (LMA) over endotracheal intubation in patients with upper respiratory infections [13]. As our patient had very few secretions, the risk of airway trouble associated with secretions was thought to be low.

Considering the risk of aspiration, endotracheal intubation is generally the choice of airway for general anesthesia in patients with abdominal distension. However, in our patient, the stomach was not filled with food or secretions. Although Griffin and Hatcher reported a case of aspiration pneumonia after elective cholecystectomy performed with the patient under general anesthesia using a classical LMA [14], some studies have reported that an LMA is an effective alternative to endotracheal intubation in some patients having abdominal surgery [15–18]. The PLMA provides a better seal around the glottic aperture than other LMAs and permits high peak airway pressure without leakage, and the drain tube through the PLMA provides easy access for deflation of the stomach and reduction of gastric fluid volume. Of note, the induction of anesthesia with muscle relaxants in patients with abdominal distension can cause difficult ventilation with a face mask. Therefore, for our patient, we thought that the PLMA would provide a significant benefit, considering her fragile, but relatively dry airway, relatively empty stomach, and the risk of difficult ventilation with a face mask.

In patients with pulmonary impairment, it is important to shorten the anesthesia time. In our patient, a local anesthetic was injected into the epidural space, because it has been reported that injecting local anesthetics or saline into the epidural space after spinal anesthesia speeds the onset of anesthesia and raises the analgesic level [19, 20]. Although spinal anesthesia alone might have been effective in our patient, such anesthesia is insufficient for patients with pulmonary impairment because it does not provide postoperative analgesia.

Patients receiving HOT for respiratory disorders may have secondary cardiac dysfunction. Our patient had no cardiac symptoms and only right ventricular pressure was slightly elevated; therefore, it was concluded that she would tolerate general anesthesia and surgery.

In summary, the respiratory management of patients with WCS is associated with several risks and challenges, including hypoxia, tension pneumothorax, difficult weaning from positive-pressure ventilation, and difficult extubation. General anesthesia under sponta-

neous breathing, using the PLMA and the combined spinal and epidural anesthesia (CSEA), was performed uneventfully for abdominal surgery in a pediatric patient with WCS.

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