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Anesthetic management of a patient with Alport-leiomyomatosis syndrome

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Abstract
We report the anesthetic management of esophagectomy for a patient with Alport-leiomyomatosis syndrome. A 23-year-old woman complained of dysphagia and severe chest pain. Her chest X-ray, computed tomography (CT), and magnetic resonance imaging (MRI) showed an enlarged esophagus, in contact with the trachea, heart, aorta, and large vessels. She frequently experienced severe asthma attacks. Because various risks in both respiration and circulation, especially in anesthesia induction, were of concern, her right femoral vessels were exposed, for the emergency use of percutaneous cardiopulmonary support (PCPS), prior to anesthesia induction. Anesthesia was induced and maintained with propofol, fentanyl, and vecuronium. Esophagectomy was performed uneventfully and no severe events were seen in anesthesia management. Alport-leiomyomatosis syndrome is a very rare disease. When we are involved in the anesthetic management of a patient with this disease, evaluation of the influence of the enlarged esophagus on both respiration and circulation, and careful preparation for emergence, are very important.

Key words Anesthetic management · Alport syndrome · Alport-leiomyomatosis syndrome · PCPS

Introduction

Diffuse esophageal leiomyomatosis is a very rare disease, involving multiple leiomyomas in the esophagus that are fused to each other, causing extensive thickening of the esophageal wall. Diffuse esophageal leiomyomatosis is a frequent complication of Alport-leiomyomatosis syndrome [1]; this syndrome involves progressive nephropathy accompanied by hematuria, eventually leading to renal failure. Herein, we report the anesthetic management of an esophagectomy for a patient with Alport-leiomyomatosis syndrome.

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Case report

The patient was a 23-year-old woman (height, 157 cm; weight, 45 kg). Her chief complaints were dysphagia, chest pain, and dyspnea. Because she was not able to lie in a supine position for a prolonged period due to chest pain, chest discomfort, and dyspnea, she always slept in a lateral position (in both directions). She had been treated for bronchial asthma for twenty years. Despite treatment with inhaled steroids, she frequently experienced severe asthma attacks and required β_2-adrenergic agonists, which were effective.

Chest X-ray images revealed a giant esophageal tumor, and immunostaining of a skin specimen provided the diagnosis of Alport-leiomyomatosis syndrome. On chest computed tomography (CT; Fig. 1) and magnetic resonance imaging (MRI; Fig. 2), the thoracic segment of the esophagus showed full-circumferential dilation and wall thickening and was in contact with the trachea, main bronchus, heart, aorta, and large vessels.

Respiratory function testing showed an obstructive pattern (forced expiratory volume [FEV] 1.0% 59.0 %, FEV1.0 1.34 l) and she was categorized as grade 3 in the Hugh-Jones classification.

In laboratory data, there were no abnormalities of hepatic or renal function (creatinine clearance 151.6 ml·min^{-1}) and no hematuria. Preoperative bronchofiberscopy showed no abnormal findings in the airway.

In regard to her past medical history, she had undergone open abdominal surgery for lower esophagectomy with a diagnosis of leiomyoma at another hospital at the age of 9 years, without diagnosis of the Alport-leiomyomatosis syndrome. She showed no symptoms suggesting lower esophageal sphincter dysfunction, such as heartburn. Her 2-year-old son was also diagnosed with Alport-leiomyomatosis syndrome (with the symptoms of proteinuria and hematuria) and bronchial asthma.

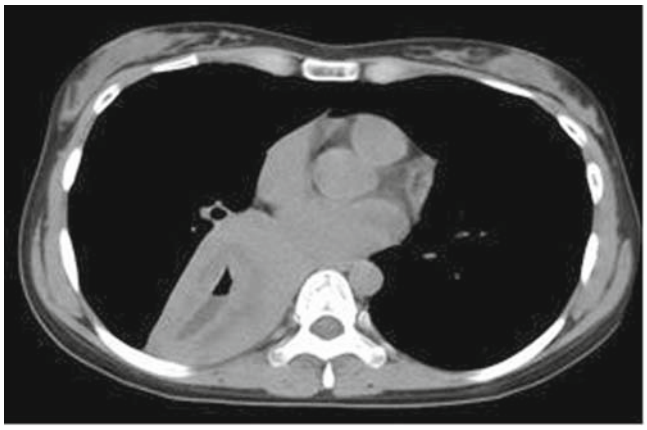

Fig. 1. Computed tomography (CT) image of the patient's chest, showing the dilated and enlarged esophagus, which was in contact with the heart, aorta, and major vessels

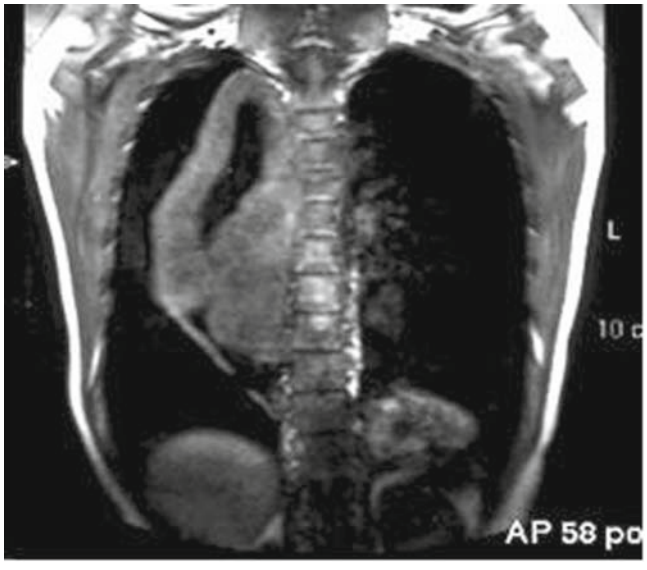

Fig. 2. Patient's chest magnetic resonance imaging (MRI) showing dilated and enlarged esophagus. *AP*, Anteroposterior

With the diagnosis of Alport-leiomyomatosis syndrome, the patient was scheduled to undergo subtotal esophagectomy and neck anastomosis with right colon.

When we were considering her anesthetic management preoperatively, various risks were of concern, especially in anesthesia induction, i.e., (1) circulatory collapse due to compression of the heart and/or large vessels, (2) difficulties in ventilation after anesthesia induction with muscle relaxants due to compression of the airway, and (3) the possibility of a severe asthma attack with the stimulation of tracheal intubation.

Accordingly, prior to anesthesia induction, the right femoral vessels were exposed, under local anesthesia, for the emergency use of percutaneous cardiopulmonary support (PCPS).

Epidural anesthesia was not selected in combination with general anesthesia, because it was possible that an anticoagulant would have been used in the event of emergency PCPS.

Preanesthetic medication was not administered. Anesthesia was induced with propofol (target-controlled infusion; effect site concentration) 4 $\mu g \cdot ml^{-1}$ and fentanyl (200 μg) in the supine position. After noting that the lungs were easily ventilated, 6 mg vecuronium was administered. Intubation was easily achieved with a 35-Fr double-lumen tracheal tube (Bronchocath; Mallinckrodt Medical; Hazelwood, MO, USA); however, it was difficult to place the tube in the correct position. Another attempt at intubation and replacement with a 32-Fr Bronchocath was successful. Bronchofiberscopy after tracheal intubation did not show thickening or deformity in the surface of the airway smooth muscle. Maintenance of anesthesia was achieved mainly with propofol (2–4 $\mu g \cdot ml \cdot ml^{-1}$), and sevoflurane (0.5%–2%; end-tidal concentration) was added temporarily when the airway pressure increased, to take advantage of the strong bronchodilatory effect of volatile anesthetics. Fentanyl and vecuronium were administered continuously. Aminophylline was administered before and during anesthesia (250 mg each time). During the operation, with one-lung ventilation in the left lateral position, pH, Pa_{O_2}, and Pa_{CO_2} were monitored continuously with a Paratrend7 device (Phillips Medical Systems, Best, Netherlands) with the sensor placed at the left radial artery. The operation was performed uneventfully. There were no serious changes in hemodynamics or respiration throughout the operation. The operation time was 930 min, the duration of anesthesia was 1050 min, and the blood loss was 900 ml.

Because the patient's asthma was uncontrolled preoperatively, the neuromuscular blockade was not reversed and the patient underwent subsequent mechanical ventilation in the intensive care unit (ICU). Patient-controlled analgesia with both continuous and bolus administration of fentanyl was successful for the management of postoperative pain.

In the postoperative course, mechanical ventilation with tracheostomy was required for several weeks in the ICU due to recurring mediastinitis, pneumonia, and bronchial asthma. Eighty-four days after the operation, the patient was discharged from the hospital without any complications.

Discussion

Alport-leiomyomatosis syndrome is very rare and only about 50 cases have been reported. [2–6]. This syn-

drome involves progressive nephropathy accompanied by hematuria, which eventually leads to renal failure. The association of this syndrome with the partial deletion of genes encoding type IV collagen α5 and α6 chains on the X chromosome has been reported [7,8].

Fortunately, critical changes in hemodynamics and/or respiration, which were of concern preoperatively, were not observed in our patient. However, it was really difficult to predict how her giant tumor would affect respiration and circulation especially during the period of anesthesia induction and postural change. The incidence of airway complications with general anesthesia in patients with mediastinal masses has been reported to be 7%–18% [9], and in this case, the patient exhibited uncontrolled asthma. Therefore, we prepared PCPS as an emergency standby. However, we do not believe that every patient with Alport-leiomyomatosis syndrome requires PCPS as standby.

In general, it is considered safe to induce anesthesia in the lateral position for a patient with a giant mediastinal tumor, especially if the patient is more comfortable with the lateral than with the supine position. However, in our patient, we selected the supine position because we believed that this position would have advantages in assuring ventilation and intubation with a double-lumen tube, with the PCPS as standby.

Although the patient was diagnosed with Alport-leiomyomatosis syndrome, her renal function was normal. Therefore, we did not require special consideration for the selection or the dose of drugs administered throughout the perioperative period.

This syndrome is often accompanied by leiomyoma of the trachea, pudendum, or uterus (esophago-vulvar syndrome). However, in the present patient, no abnormalities such as stenosis or an elevated lesion in the surface of the airway (from the upper airway to the bronchus), were observed during preoperative bronchofiberscopy. Therefore, the difficulty in the placement of the 35-Fr tube appears to have been due to the compression of the airway by the enlarged esophagus after anesthesia induction with the muscle relaxant.

Although the patient had uncontrolled asthma, we were able to prevent a severe attack by the administration of aminophylline before and during the operation, the use of gentle procedures (especially in tracheal intubation), and the use of volatile anesthetic in addition to propofol in anesthesia maintenance.

In conclusion, we successfully managed the anesthesia of a patient with Alport-leiomyomatosis syndrome and bronchial asthma. For the safe management of patients with this disease, appropriate evaluation of the influence of the enlarged esophagus on the vital organs in the mediastinum and careful preparation should be done.

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