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Anesthetic management of a patient with a double inferior vena cava and pulmonary alveolar proteinosis who underwent bilateral living-donor lobar lung transplantation

Hiroaki Murata, Tetsuya Hara, Shuhei Matsumoto, Haruka Inoue, Hikoma Yamashita, and Koji Sumikawa

Department of Anesthesiology, Nagasaki University School of Medicine, 1-7-1 Sakamoto, Nagasaki 852-8501, Japan

Abstract
A 43-year-old woman with pulmonary fibrosis secondary to pulmonary alveolar proteinosis was scheduled to undergo lung transplantation. Before the lung transplantation, she had undergone multiple whole-lung lavage procedures on extracorporeal circulation (ECC), which had caused scarring of the right femoral subcutaneous tissues. Preoperative examination revealed a double inferior vena cava (IVC) with interiliac communication, and the left IVC ended at the left renal vein. Surgical exposure of the right femoral vessels was performed immediately after anesthetic induction for emergent vascular access to establish an ECC. Cardiopulmonary collapse did not occur and the ECC was not required until lung resection. The lung transplantation was completed uneventfully. Congenital IVC anomaly is rare, but may make cannulation through the femoral vein difficult. Scarring of the subcutaneous tissue could result in a difficult "percutaneous" approach to the vessels. Evaluation of the vascular anatomy related to the establishment of an ECC is important before lung transplantation.

Key words Double inferior vena cava · Pulmonary alveolar proteinosis · Lung transplantation

Introduction

Some patients undergoing lung transplantation experience cardiac or respiratory instability during the procedure and require cardiopulmonary bypass (CPB) [1]. If the cardiopulmonary collapse occurs before the thoracotomy incision, emergent establishment of an extracorporeal circulation (ECC) through the femoral vessels is necessary [2,3]. Congenital inferior vena cava (IVC) anomalies are uncommon [4,5], but they may make cannulation through the femoral vein into the right atrium difficult or even impossible. Pulmonary alveolar proteinosis (PAP) is a disorder characterized by the excessive accumulation of lipoproteinaceous material within the alveoli [6]. Although extremely rare [7], progression from PAP to pulmonary fibrosis may require lung transplantation [8]. The existence of subcutaneous scarring due to multiple femoral skin punctures should be considered in patients with PAP who have undergone multiple whole-lung lavage (WLL) procedures as palliative therapy under extracorporeal membrane oxygenation (ECMO) support before lung transplantation. We describe the successful anesthetic management of a patient with a double IVC and PAP during bilateral living-donor lobar lung transplantation.

Address correspondence to: H. Murata
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Case report

A 43-year-old woman (163 cm, 45 kg, American Society of Anesthesiologists physical status III) with pulmonary fibrosis secondary to PAP was scheduled to undergo bilateral living-donor lobar lung transplantation after an 8-year history of progressive dyspnea. She had undergone WLL on full veno-arterial ECMO seven times. Home oxygen therapy had been initiated 3 years prior to the lung transplantation surgery. She had received granulocyte-macrophage colony-stimulating-factor therapy 2 years previously, but this treatment had been ineffective. Because the abdominal computed tomography (CT) image obtained before the last WLL suggested IVC stenosis, we performed venography of the IVC and its branches. As extensive scarring of subcutaneous tissue made a percutaneous approach to the right femoral vein difficult, venography was completed from the left femoral vein; this revealed a double IVC with interiliac communication and without IVC stenosis. The interiliac communication was large and ran from a low level of the left IVC to a high level of the right IVC (Fig. 1). The blood flow of the left IVC was cephalad to caudad (Fig. 1A) and the left IVC ended at

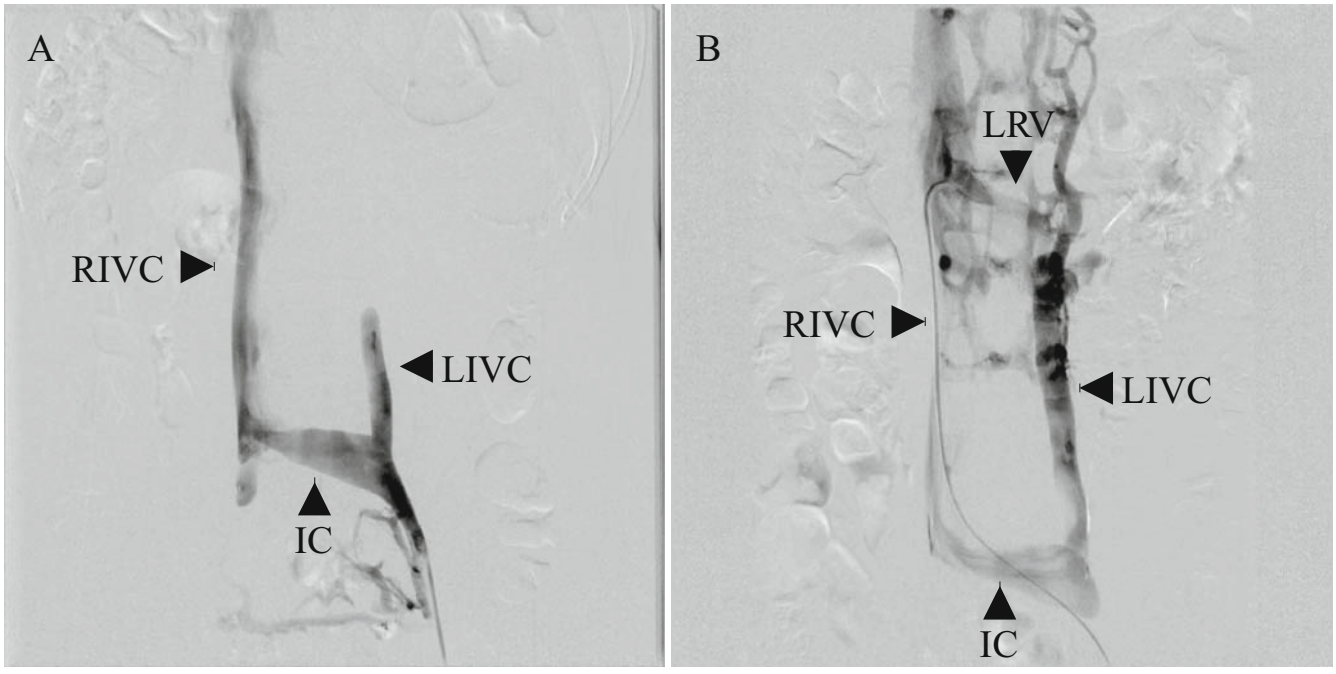

Fig. 1A,B. Venography shows a double inferior vena cava (IVC) with an infrarenal interiliac communication, which runs from a low level of the left IVC to a high level of the right IVC (*RIVC*, right IVC; *LIVC*, left IVC; *IC*, interiliac communication; *LRV*, left renal vein). **A** Injection of contrast agent into the LIVC revealed that the blood flow of the LIVC was from cephalad to caudad. Because of the direction of the blood flow, the cephalad portion of the LIVC was not contrast-enhanced. **B** Injection of contrast agent into the LRV demonstrated that the LIVC ended at the LRV. Retrograde enhancement of the LRV and blood flow of the LIVC from cephalad to caudad was confirmed

the left renal vein (Fig. 1B). Eight months before the lung transplantation we had completed the last WLL under veno-arterial ECMO, through the right femoral vein and right femoral artery, by surgically exposing these vessels.

Pulmonary function tests performed just before the last WLL showed severe restrictive lung disease, i.e., forced vital capacity (FVC), 0.87 l; %FVC, 28.2%; forced expiratory volume at 1 s ($FEV_{1.0}$), 0.83 l; and $FEV_{1.0\%}$, 92.2%. An arterial blood gas (ABG) analysis done on the day before the lung transplantation, under 8 l·min^{-1} of oxygen via a nasal cannula revealed Pa_{O_2} 92 mmHg, Pa_{CO_2} 61 mmHg, pH 7.384, and arterial oxygen saturation (Sa_{O_2}) 96.7%. Electrocardiogram was normal, and transthoracic echocardiography revealed mild tricuspid valve regurgitation and mild pulmonary hypertension, with normal bilateral ventricular functions. Chest roentgenogram showed a bilateral reticular shadow, and chest CT revealed bilateral fibrous changes with multiple cysts.

On the day of the lung transplantation, no anesthetic premedication was given. In the operating theater, a pulse oximeter was placed on bilateral index fingers, and a 5-lead electrocardiograph monitor, bispectral index sensor (Aspect A-2000; Aspect Medical Systems, Newton, MA, USA), and a near-infrared spectrometer (INVOS5100; Somanetics, Troy, MI, USA) were placed, followed by peripheral venous and right radial artery cannulations. ABG analysis after preoxygenation with 100% oxygen through a tightly fitted mask for 5 min showed Pa_{O_2} 502 mmHg and Pa_{CO_2} 60 mmHg. Then, we decided to induce general anesthesia without surgical exposure of the femoral vessels under local anesthetics.

Induction of anesthesia was performed with high-dose fentanyl, 50 µg·kg^{-1}, and midazolam, 0.5 mg·kg^{-1}. Neuromuscular block was provided by vecuronium bromide, 0.15 mg·kg^{-1}. Intubation was performed using a 35-French left-sided Bronchocath double-lumen tube (Mallinckrodt, Hazelwood, MO, USA). Transesophageal echocardiography was then introduced. Central venous and pulmonary arterial cannulations were performed with a strict aseptic technique. Pulmonary artery pressure after anesthetic induction was 40/20 mmHg. ABG analysis done 30 min after anesthetic induction under positive-pressure ventilation with respiratory rate 14 breaths per min, peak inspiratory pressure 33 cmH_2O, and tidal volume 450 ml under 60% oxygen, showed Pa_{O_2} 190 mmHg and Pa_{CO_2} 59 mmHg. Surgical exposure of the right femoral artery and vein was performed after anesthetic induction, for immediate and assured vascular access to establish an ECC.

Maintenance of anesthesia consisted of continuous infusion of fentanyl, 0.5 $\mu g \cdot kg^{-1} \cdot min^{-1}$, and midazolam, 2–4 $mg \cdot h^{-1}$. Dissection and mobilization of the lungs were performed through a transverse thoracotomy (clamshell) incision, after which the recipient was cannulated for CPB in the right atrium and ascending aorta. The elapsed time between anesthesia induction and the thoracotomy incision was about 140 min. Because the cardiopulmonary function of the patient remained stable until lung resection, the femoral vein was not utilized to establish an ECC. ABG analysis done before CPB under positive-pressure ventilation with respiratory rate 12 breaths per min, peak inspiratory pressure 29 cmH_2O, and tidal volume 425 ml under 75% oxygen, showed Pa_{O_2} 117 mmHg and Pa_{CO_2} 67 mmHg. The values for pulmonary artery pressure and the cardiac index between anesthesia induction and CPB establishment for lung resection were 29–46/13–27 mmHg and 2.7–4.9 $l \cdot min^{-1} \cdot m^{-2}$, respectively, with dopamine, 3–5 $\mu g \cdot kg^{-1} \cdot min^{-1}$, and prostaglandin E1, 0.02 $\mu g \cdot kg^{-1} \cdot min^{-1}$. Bilateral sequential lung transplantation was uneventful and adequate gas exchange was easily achieved. CPB time was 252 min without an aortic cross-clamp. Separation from CPB was achieved with dopamine, 5 $\mu g \cdot kg^{-1} \cdot min^{-1}$; nitroglycerin, 0.5 $\mu g \cdot kg^{-1} \cdot min^{-1}$; and prostaglandin E1, 0.02 $\mu g \cdot kg^{-1} \cdot min^{-1}$. Nitric oxide, 20 parts per million, was inhaled to prevent pulmonary hypertension after lung implantation. Homologous blood was transfused as needed to maintain an adequate hematocrit (approximately >30%). Post-CPB pulmonary artery pressure was 43/16 mmHg.

At the end of the surgery, transesophageal echocardiography showed sufficient bilateral pulmonary vein flow. The patient's tracheal tube was exchanged for a single-lumen tube with an 8.0-mm internal diameter, and the patient was transferred in a stable condition to the intensive care unit.

The femoral incision for vascular exposure was closed on postoperative day (POD) 3. The patient required inhaled nitric oxide treatment for persistent pulmonary hypertension until POD 4. After the correction of coagulation parameters, a thoracic epidural catheter was placed on POD 4. The tracheal tube was removed on POD 5, with normal blood gases on 2 $l \cdot min^{-1}$ of oxygen via a nasal cannula (Pa_{O_2} 139 mmHg, Pa_{CO_2} 43 mmHg, pH 7.478) and falling pulmonary artery pressure (35/13 mmHg). She was transferred to the surgical ward on POD 23 and discharged home without requiring oxygen therapy on POD 76.

Discussion

We experienced the anesthetic management of a patient with a double IVC and PAP who underwent living-donor lobar lung transplantation. Surgical exposure of the right femoral artery and vein was performed after anesthetic induction for immediate and assured vascular access to establish an ECC in case of cardiopulmonary collapse before thoracotomy incision [9]. In the present patient, cardiopulmonary function was preserved until CPB establishment for lung resection. Regardless of the patient's preoperative cardiopulmonary function, preparation for an emergent ECC through femoral vessels is important for patients undergoing living-donor lobar lung transplantation. In the present patient, an IVC anomaly and the scarring of femoral subcutaneous tissues were problems related to the preparation of blood access through the femoral vessels.

Duplication of the IVC results from the persistence of bilateral supracardinal veins, the prevalence of which is 0.2%–3% [10,11]. According to previously reported criteria, the present case was classified as type BC [12] or type II-b-1 [13,14], which is the most frequent type of double IVC. In the report by McClure, Type BC means persistence of right and left supracardinal veins. Type II-b-1 is defined when the interiliac communication is large and runs from a low level of the left IVC to a high level of the right IVC. Because the left IVC ends at the left renal vein in this type of double IVC, a cannula inserted from the left femoral vein might migrate into the left IVC, which could result in the delayed placement of the venous cannula in the right atrium. If cardiopulmonary instability occurs during the surgical operation, delay of ECC establishment results in tragedy. Therefore, sufficient anatomical evaluation of the IVC and its branches seems to be necessary when anomalies of these vessels are suspected before lung transplantation. Although preferable, it may not be practicable to perform IVC venography in all patients who are scheduled to undergo lung transplantation. Before lung transplantation, it is necessary to perform thoracic and abdominal contrast-enhanced CT to assess vessels such as the pulmonary vessels, aorta, and IVC, which are related to the surgical procedure and blood access during lung transplantation. IVC venography may be an option when an IVC anomaly is suspected by preoperative CT.

Another concern about the immediate establishment of an ECC in the present patient was the extensive scarring of the right femoral subcutaneous tissue, which could have resulted in a difficult "percutaneous" approach to the femoral vessels. The subcutaneous scarring probably resulted from the multiple WLL procedures under ECMO support that the patient had undergone before the lung transplantation. WLL has been established as a palliative treatment for PAP [6]. The median duration of clinical benefit from lavage has been reported to be 15 months [15]. Although extremely

rare [7], progression from PAP to pulmonary fibrosis might require lung transplantation [8]. Therefore, the existence of subcutaneous scarring due to multiple femoral skin punctures should be considered in patients with PAP before lung transplantation. The axillary, internal jugular, and subclavian veins could all serve as alternative venous accesses for patients with an IVC anomaly or those with a difficult percutaneous approach to the femoral vein [16]. In the present patient, because of the double-IVC anomaly with the left IVC ending at the left renal vein, and because of the right femoral skin scarring, surgical exposure of the right femoral vessels was necessary to prepare for the emergent establishment of an ECC before the thoracotomy incision.

Whether to perform surgical exposure of the femoral vessels before anesthesia induction was a critical problem in the present patient. Judging from the ABG analysis just before anesthesia induction (Pa_{O_2}/fractional inspired oxygen ($F_{I_{O_2}}$) ratio > 500) and the preoperative preserved cardiac function, we considered that severe hypoxia, carbon dioxide retention, or cardiac collapse that would have required emergent ECMO or cardiopulmonary support was not likely to be caused by anesthesia induction and the following positive-pressure ventilation. Although the surgical exposure of femoral vessels under local anesthesia before anesthesia induction may have been a safer procedure in the present patient, it may have put the patient in a stress condition, which would have induced cardiopulmonary instability. Therefore, we advanced to anesthesia induction without surgical exposure of the femoral vessels under local anesthesia. The patient's cardiopulmonary function remained stable for more than 2 h until CPB establishment for bilateral lung resection. Accordingly, we believe that our decision to perform surgical exposure of the femoral vessels after anesthesia induction was clinically acceptable.

If pulmonary insufficiency had occurred before the thoracotomy incision, we would have needed to determine blood access for the ECC; that is, via the surgically exposed femoral vessels before the thoracotomy incision, and through the ascending aorta and right atrium or a combination of these vessels, immediately after the thoracotomy incision. Although the surgically exposed femoral vessels were in an easily accessible condition, the placement of catheters in proper positions may have required a longer time than expected, especially in this patient with an IVC anomaly, and the smaller diameter of femoral catheters could restrict extracorporeal blood flow, which would result in insufficient cardiopulmonary support. CPB through the ascending aorta and right atrium, although it requires a previous thoracotomy incision, may provide quicker blood access with sufficient extracorporeal blood flow than using the femoral vessels. In the present patient, we planned femoral access as the first choice, because ECMO support during the last WLL prior to the lung transplantation had been properly achieved. However, in the case of an emergency cardiopulmonary collapse, the cooperation of anesthesiologists, surgeons, and perfusionists is necessary to determine the best way to establish an ECC.

In conclusion, we experienced the anesthetic management of a patient with a double IVC and PAP who underwent living-donor lobar lung transplantation. Sufficient evaluation of the vascular anatomy related to the establishment of ECC should be conducted before lung transplantation. Additionally, before lung transplantation, the status of subcutaneous scar formation due to multiple femoral skin punctures should be considered in patients with PAP.

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