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

Short title: Lung transplantation and double inferior vena cava

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Abstract

A 43-year-old woman with pulmonary fibrosis secondary to pulmonary alveolar proteinosis was scheduled to undergo lung transplantation. She had undergone multiple whole lung lavage on extracorporeal circulation (ECC) before lung transplantation which caused scarring of right femoral subcutaneous tissues. Preoperative examination revealed a double inferior vena cava (IVC) with interiliac communication and left IVC ended at left renal vein. Then, surgical exposure of the right femoral vessels was performed immediately after anesthetic induction for the emergent vascular access to establish ECC. Cardiopulmonary collapse did not occur and ECC was not required until lung resection. Lung transplantation was uneventfully completed. 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 ECC is important before lung transplantation.

Introduction

Some patients undergoing lung transplantation experience cardiac or respiratory instability during the procedure and require cardiopulmonary bypass (CPB) [1]. When the cardiopulmonary collapse occurs before thoracotomy incision, emergent establishment of extracorporeal circulation (ECC) through the femoral vessels is necessary [2,3]. Congenital inferior vena cava (IVC) anomalies are uncommon [4,5], but might make cannulation through the femoral vein into the right atrium difficult or even impossible. Protein alveolar proteinosis (PAP) is a disorder characterized by excessive accumulation of lipoproteinaceous material within the alveoli [6]. Although extremely rare [7], progression from PAP to pulmonary fibrosis might 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) as a palliative therapy under extracorporeal membrane oxygenation (ECMO) support before lung transplantation. We describe successful anesthetic management of a patient with a double IVC and PAP during bilateral living donor lobar lung transplantation.

Case report

A 43-year-old woman, 163 cm, 45 kg, ASA 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 7 times. She

initiated home oxygen therapy 3 years prior to surgery. She received granulocyte-macrophage colony-stimulating-factor therapy 2 years previously, which was 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 toward the right femoral vein difficult, venography was completed from the left femoral vein, which revealed a double IVC with interiliac communication and without IVC stenosis. The interiliac communication is large and runs from the low level of the left IVC to the high level of the right IVC (Figure 1). Blood flow of the left IVC was cephalad to caudad (Figure 1A) and the Left IVC ended at the left renal vein (Figure 1B). We completed the last WLL under veno-arterial ECMO through the right femoral vein and right femoral artery by surgically exposing these vessels 8 months before lung transplantation.

Pulmonary function tests performed just before the last WLL showed severe restrictive lung disease, i.e., forced vital capacity 0.87 L (28.2% predicted), forced expiratory volume at one second ($FEV_{1.0}$) 0.83 L, and $FEV_{1.0\%}$ 92.2%. An arterial blood gas (ABG) analysis done on the day before lung transplantation under 8 $L \cdot min^{-1}$ of oxygen via a nasal cannula revealed PaO_2 92 mmHg, $PaCO_2$ 61 mmHg, pH 7.384, and SaO_2 96.7%. Electrocardiogram was normal and transthoracic echocardiography revealed mild tricuspid valve regurgitation and mild pulmonary hypertension with normal bilateral ventricular functions. Chest roentogenogram showed a bilateral reticular shadow, and chest CT revealed bilateral fibrous changes with multiple cysts.

On the day of lung transplantation, no anesthetic premedication was given. In the operating theater, pulse oximeter on bilateral index fingers, 5-lead electrocardiograph monitor, bispectral index sensor (Aspect A-2000; Aspect Medical Systems, Newton, MA), and 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 minutes showed PaO_2 502 mmHg and $PaCO_2$ 60 mmHg. Then, we decided to induce general anesthesia without surgical exposure of femoral vessels under local anesthetics.

Induction of anesthesia was performed with high-dose fentanyl, 50 μg/kg, and midazolam, 0.5 mg/kg. Neuromuscular block was provided by vecuronium bromide, 0.15 mg/kg. Intubation was performed using a 35 Fr 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 per minute, peak inspiratory pressure 33 cmH_2O and tidal volume 450 mL under 60% oxygen was PaO_2 190 mmHg, $PaCO_2$ 59 mmHg. Surgical exposure of the right femoral artery and vein was performed after anesthetic induction for immediate and assured vascular access to establish ECC.

Maintenance of anesthesia consisted of continuous infusion of fentanyl, 0.5 μg/kg/min, and midazolam, 2-4 mg/h. Dissection and mobilization of the lungs were performed through a transverse

thoracotomy (clamshell) incision, after which the recipient was cannulated for CPB at the right atrium and ascending aorta. Time elapsed between anesthesia induction and thoracotomy incision was about 140 minutes. Because cardiopulmonary function of the patient remained stable until lung resection, the femoral vein was not utilized to establish ECC. ABG analysis done before CPB under positive pressure ventilation with respiratory rate 12 per minute, peak inspiratory pressure 29 cmH_2O and tidal volume 425 mL under 75% oxygen was PaO_2 117 mmHg, $PaCO_2$ 67 mmHg. Pulmonary artery pressure and cardiac index between anesthesia induction and CPB establishment for lung resection were 29-46/13-27 mmHg and 2.7-4.9 $L/min/m^2$, respectively, with dopamine, 3-5 μg/kg/min, and prostaglandin E1, 0.02 μg/kg/min. 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 μg/kg/min; nitroglycerin, 0.5 μg/kg/min; prostaglandin E1, 0.02 μg/kg/min. 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 hematcrit (approximately > 30%). Post-CPB pulmonary artery pressure was 43/16 mmHg.

At the end of surgery, transesophageal echocardiography showed sufficient bilateral pulmonary vein flow. The patient's tracheal tube was changed to 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.

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 correction of coagulation parameters, a thoracic epidural catheter was placed on POD 4. Tracheal tube was removed on POD 5 with normal blood gases on 2 L/min of oxygen via a nasal cannula (PaO_2 139 mmHg, $PaCO_2$ 43 mmHg, pH 7.478) and falling PA pressures (35/13 mmHg). She was transferred to the surgical ward on POD 23 and discharged to 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 ECC in case of cardiopulmonary collapse before thoracotomy incision [9]. Cardiopulmonary function was preserved until CPB establishment for lung resection in the present patient. Regardless of preoperative cardiopulmonary functions, preparation for emergent ECC through femoral vessels is important for patients undergoing living donor lobar lung transplantation. In the present patient, IVC anomaly and scarring of femoral subcutaneous tissues were the problems related to the preparation of blood access through femoral vessels.

Duplication of the IVC results from persistence of bilateral supracardinal veins, the prevalence of which is 0.2-3% [10,11]. According to previously reported criteria, the present case is classified as

type BC [12] or type II-b-1 [13,14], which is the most frequent type of double IVC. The type II-b-1 is defined that the interiliac communication is large and runs from the low level of the left IVC to the high level of the right IVC. Because the left IVC ends at the left renal vein in this type of double IVC, the cannula inserted from the left femoral vein might migrate into the left IVC, which could result in delayed placement of venous cannula at the right atrium. If cardiopulmonary instability occurs during the surgical operation, delay of establishment of ECC results in tragedy. Therefore, sufficient anatomical evaluation of the IVC and its branches might be necessary when the anomalies of those vessels are suspected before lung transplantation. Although preferable, it may not be practically accepted to perform IVC venography in all of the patients who are scheduled to undergo lung transplantation. Thoracic and abdominal contrast-enhanced CT before lung transplantation is necessary to assess vessels such as pulmonary vessels, aorta and IVC which are related to surgical procedure and blood access during lung transplantation. IVC venography may be an option when its anomaly is suspected by preoperative CT.

Another concern about immediate establishment of ECC in the present case was the extensive scarring of the right femoral subcutaneous tissue, which could result in a difficult “percutaneous” approach to the femoral vessels. Subcutaneous scarring probably resulted from multiple WLL under ECMO support before lung transplantation. WLL has been established as 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 might all serve as alternative venous accesses for patients with an IVC anomaly or with a difficult percutaneous approach to the femoral vein [16]. Because of the DIVC anomaly with left IVC which ended at left renal vein and right femoral skin scarring, surgical exposure of right femoral vessels was necessary in the present patient to prepare for emergent establishment of ECC before thoracotomy incision.

Whether to perform surgical exposure of femoral vessels before anesthesia induction was a critical problem in the present patient. Judging from the ABG analysis just before anesthesia induction (PaO_2/FiO_2 ratio > 500) and preoperative preserved cardiac function, we considered that severe hypoxia, carbon dioxide retention or cardiac collapse that requires emergent ECMO or cardiopulmonary support is not likely by anesthesia induction and following positive pressure ventilation. Although the surgical exposure of femoral vessels under local anesthesia before anesthesia induction might be a safer procedure in the present patient, it might put the patient in a stress condition, which induces the cardiopulmonary instability. Then, we advanced to anesthesia induction without surgical exposure of femoral vessels under local anesthesia. Cardiopulmonary function was remained stable until CPB establishment for bilateral lung resection for more than two hours. Therefore, we think that our decision of surgical exposure of femoral vessels after anesthesia induction was clinically acceptable.

If the pulmonary insufficiency did happen before thoracotomy incision, we needed to determine the blood access for ECC, that is, via surgically exposed femoral vessels, through ascending aorta and right atrium after immediate thoracotomy incision or combination of these vessels. Although surgically exposed femoral vessels were in easily accessible condition, placement of catheters in proper position might require longer time than expected especially in patients with IVC anomaly, and the smaller diameter of femoral catheters can restrict extracorporeal blood flow, which resulted in insufficient cardiopulmonary support. CPB through ascending aorta and right atrium might provide quicker blood access with sufficient extracorporeal blood flow than using femoral vessels though it requires a previous thoracotomy incision. In the present patient, we planned femoral access as first choice because ECMO support during the last WLL prior to lung transplantation was properly achieved. However, in case of emergency cardiopulmonary collapse, cooperation of anesthesiologists, surgeons and perfusionists is necessary to determine the best way to establish 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, the status of subcutaneous scar formation due to multiple femoral skin punctures should be considered in patients with PAP before lung transplantation.

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Figure legend

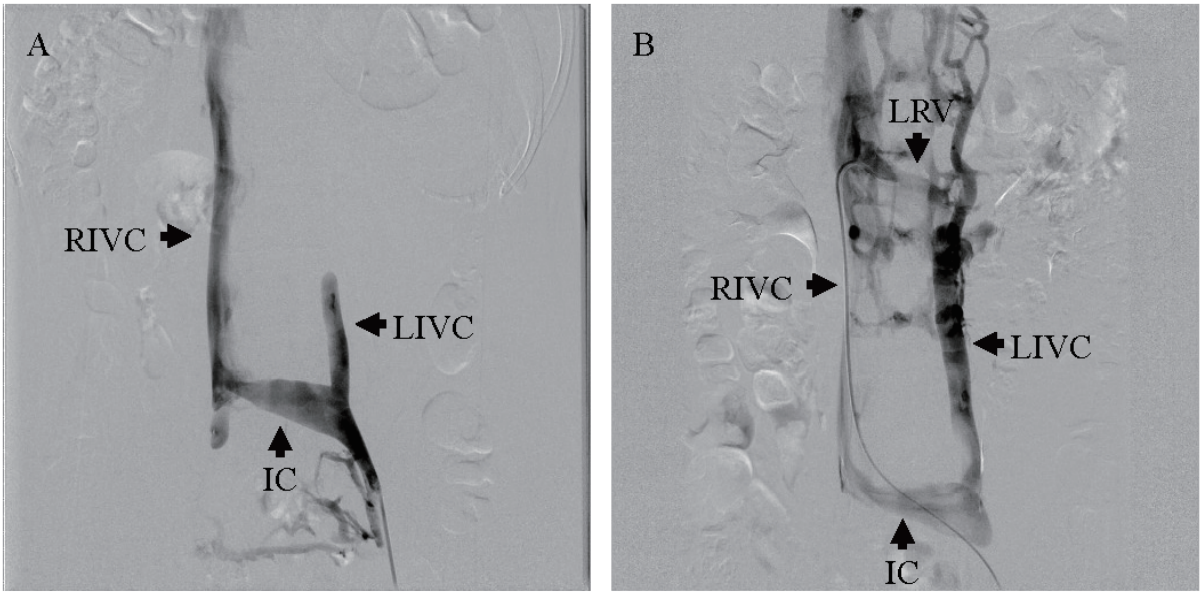

Figure 1: Venography shows a double inferior vena cava (IVC) with an infrarenal interiliac communication (RIVC = right IVC; LIVC = left IVC; IC = interiliac communication, LRV = left renal vein). (A): Injection of contrast agent into LIVC revealed that blood flow of LIVC is from cephalad to caudad. Because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