cally, the tumor cells were positive diffusely for vimentin, and less intensely for desmin, smooth muscle antigen, muscle specific antigen, epithelial membrane antigen, caldesmon, and negative for pankeratin, S-100, CD-31, CD-34, and D2-40. In addition, p53 protein was positive in 20% and Ki-67 was positive in 80% of tumor cells. The study revealed the morphologic aspect characteristic of leiomyosarcoma (Figs 2A, 2B).

The postoperative course was uncomplicated and the patient was discharged on the eighth postoperative day. Although adjuvant therapy was recommended to the patient after the operation, the patient did not agree to it. Eight months after the operation without the adjuvant therapy, she was asymptomatic, and echocardiography and a chest x-ray revealed no evidence of recurrence.

Comment

The important features of this case report are early diagnosis before surgical treatment and complete removal of the leiomyosarcoma. According to the literature, there seems to be a female predominance and leiomyosarcomas are more often observed in the inferior vena cava [4]. In our patient, the tumor was located in the main pulmonary artery and branches. It is hoped that these factors will increase the patient's chances of survival.

Diagnosis of this tumor is often difficult and late. The symptoms are insidious and nonspecific. Patients with pulmonary artery leiomyosarcoma commonly present with atypical symptoms or symptoms suggestive of chronic pulmonary thromboembolism. Differentiation between pulmonary embolism and primary PA leiomyosarcoma preoperatively can be difficult. Computed tomography scanning, magnetic resonance imaging, and angiography cannot differentiate between thrombus and tumor. Two-dimensional echocardiography can be a useful diagnostic method for the diagnosis of intracardiac tumors [5].

Progressive pulmonary dysfunction and right ventricular failure occur due to pulmonary hypertension. The mean survival time of patients with leiomyosarcoma is 6 months after diagnosis [6]. Early diagnosis and surgical intervention in operable patients may significantly improve clinical symptoms. It is well established that early and primary surgical resection is the best treatment [1]. In our patient, surgical intervention was performed within 2 days of the beginning of the complaints.

The role of adjuvant therapy has not yet been clearly defined in the literature. The limited experience of any center in the treatment of these neoplasms makes it difficult to evaluate the relative importance of surgical excision and adjuvant therapy. Some investigators are in favor of adjuvant therapy and describe encouraging results [2, 3]. However, our patient, despite her refusal to undergo adjuvant therapy, was asymptomatic and there was no evidence of recurrence according to chest x-rays and echocardiography at the follow-up at 8 months. Early diagnosis and complete surgical resection could offer the chance for long-term survival without adjuvant therapy.

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FEATURE ARTICLES

Delayed Chest Closure Assessed by Transesophageal Echocardiogram in Single-Lobe Lung Transplantation

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A 6-year-old girl with idiopathic interstitial pneumonia successfully underwent living-donor right single-lobe lung transplantation from her mother. Her mother's right lower lobe was 207% bigger than her right chest cavity, and attempting chest closure caused significant compression and narrowing of the right pulmonary vein anastomosis, as assessed by transesophageal echocardiogram. Her chest was temporarily closed without rib approximation. The following day, her chest could be completely closed. The pulmonary vein anastomosis, confirmed by transesophageal echocardiogram, was now patent. Currently, 7 months after the transplantation, she is well without limitations.

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Single-lobe living-donor lobar lung transplantation (LDLLT) has been performed occasionally when the recipient is a small child or only one donor is available [1]. Because an adult lower lobe may be too large for a small child, size matching in single-lobe LDLLT poses a critical challenge. Intraoperative transesophageal echocardiogram (TEE) is an important tool for optimizing patient care in lung transplantation [2]. Herein, we report successful right single-lobe LDLLT with a delayed chest closure assessed by

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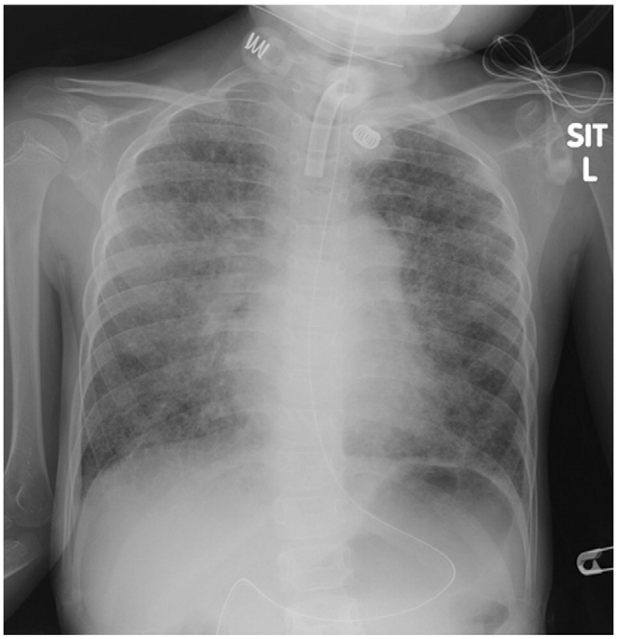

Fig 1. Chest roentgenogram of a 6-year-old girl with idiopathic interstitial pneumonia on admission.

TEE for a critically ill 6-year-old girl, using an oversized graft from her mother.

A 2-month-old girl with a normal birth presented with poor feeding and tachypnea. She was diagnosed with idiopathic interstitial pneumonia by open lung biopsy. Various intensive treatments, including steroid pulse therapy, immunosuppressant therapy, artificial surfactant administration, and hydrochloroquine treatment, slowed progression of the disease. A tracheostomy was performed at 1 year, and she was able to be weaned from the ventilator. After a period of years with home oxygen therapy, she again deteriorated and was referred to Kyoto University Hospital for possible lung transplantation at 6 years of age. On admission, she was receiving supplemental oxygenation with 10 L/min through a tracheostomy. Arterial blood gas analysis with supplemental oxygenation with that condition revealed a pH of 7.41, PaO_2 of 54.7 mm Hg, and $PaCO_2$ of 46.6 mm Hg. A chest radiograph (Fig 1) showed bilateral diffuse reticulonodular shadows. An LDLLT was the only realistic option for the girl because only brain death was accepted for potential donors older than 15 years in Japan at that time. The girl was 105 cm in height and weighed 13.4 kg. Her mother was 162 cm in height and weighed 60 kg. Three-dimensional computed tomographic volumetry indicated the recipient's right and left chest cavity volumes to be only 552 mL and 455 mL, respectively. The evaluated volume of the mother's right lower lobe was 1,142 mL, indicating the graft would be 207% bigger than the right chest cavity of the recipient (Fig 2). The case was carefully discussed and approved by the Ethical Committee of Kyoto University Hospital.

The operation was performed through a right anterior thoracotomy. The patient was placed on standard cardiopulmonary bypass. After a right pneumonectomy, the right lower lobe of the mother was implanted using cardiopulmonary bypass. When the graft was reperfused and reventilated, it became obvious that the graft was large. The patient was weaned from cardiopulmonary bypass without difficulty. The cardiopulmonary bypass time was 169 minutes, and the ischemic time of the graft was 88 minutes. Systolic pulmonary artery pressure was approximately 30 mm Hg and PaO_2 was 240 mm Hg with 100% oxygen inhalation. The TEE demonstrated that the pulmonary arterial anastomosis was patent, but that the pulmonary venous (PV) anastomosis was stenotic under a positive end-expiratory pressure (PEEP) of 5 cm H_2O (minimum diameter of 2.2 mm) (Fig 3A). After reduction of the PEEP to 3 cm H_2O, the diameter improved to 3.8 mm (Fig 3B), suggesting that the PV patency depended

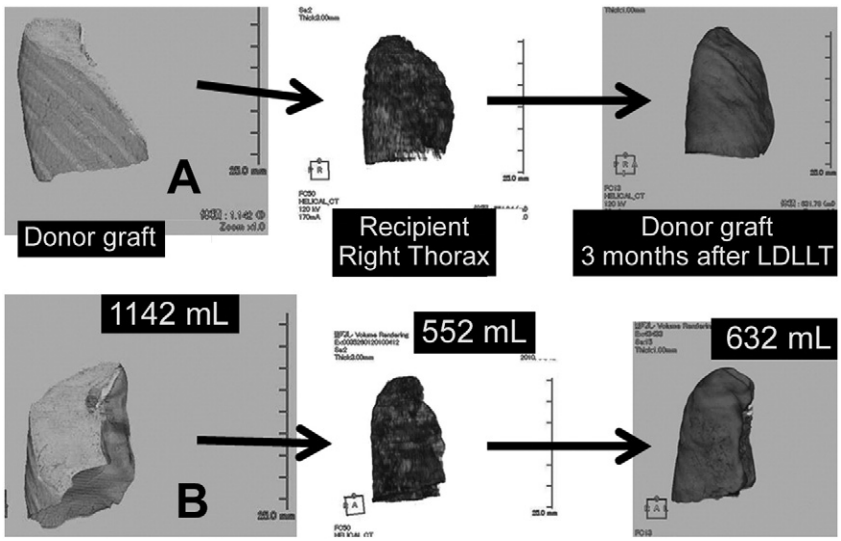

Fig 2. The trend of perioperative three-dimensional (3D) computed tomographic (CT) volumetry. (A) Lateral view (upper 3 panels) and (B) anterior view (lower three panels). 3D-CT volumetry can visualize size matching between the donor and the recipient. Preoperative 3D-CT volumetry indicated the recipient's right chest cavity volume to be only 552 ml. The evaluated volume of the mother's right lower lobe was 1,142 ml, indicating the graft would be 207% bigger than the right chest cavity of the recipient. Postoperative 3D-CT volumetry showed that the implanted graft volume was 632 ml, which was 55.2% of its original size.