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LETTER TO THE EDITOR

Living-donor lobar lung transplantation for interstitial pneumonia associated with dermatomyositis

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Collagen diseases, such as dermatomyositis (DM) and systemic sclerosis (SSc), often cause interstitial pneumonia (IP), which leads to severe pulmonary dysfunction. However, very few cases have been reported involving lung transplantation for DM, and outcomes were often unsatisfactory. All previously reported patients received cadaveric lung transplantation for acute forms of IP. Herein, we report bilateral living-donor lobar lung transplantation in a patient with IP associated with DM and SSc.

A 54-year-old female patient was diagnosed with SSc and IP at the age of 43, when she developed Raynaud's phenomenon and dyspnea on effort. Steroid therapy (prednisolone) was initiated. She was then diagnosed with DM at the age of 47 years, with Gottron's papule, Shawl sign, and increased serum creatine kinase (400 IU/l). Under oral steroid medication, SSc symptoms gradually improved, but DM and IP gradually worsened. Continuous supplemental oxygen inhalation and cyclosporine were initiated at the age of 52 and 53 years, respectively. Although the symptom of DM was controlled with treatment, her respiratory status further deteriorated because of IP progression.

The patient was transferred to Kyoto University Hospital on August 7, 2008. On admission, her vital capacity was 0.9 l (39.2% predicted) and arterial blood gas revealed a pH of 7.44, PaO_2 of 68.6 mmHg, and $PaCO_2$ of 42.8 mmHg with 6 l/min oxygen administered via a nasal cannula. Muscle strength was normal for all extremities on physical examination. There was no sign of esophageal dysmotility. A computed tomographic scan of the chest demonstrated severe fibrosis of the bilateral lungs (Fig. 1). The level of serum creatine kinase was 61 IU/l. Antinuclear, anti-Scl-70, and anti-Jo-1 antibodies were negative. Preoperative evaluation including a 2-deoxy-18F-fluoro-glucose-positron emission tomography (FDG-PET) scan revealed no evidence of coexisting malignant diseases. Moreover, the results of blood examinations including serum levels of tumor markers were within normal ranges.

Her life expectancy was estimated to be quite limited without lung transplantation. On August 21, 2008, she underwent living-donor lobar lung transplantation with a left lower lobe from her first daughter (25 years old) and a right lower lobe from her second daughter (22 years), who desired to be lung donors and were eligible by medical and psychological preoperative evaluations. The surgical aspects of the donor lobectomy, donor backtable preservation technique, and recipient bilateral pneumonectomy and lobar implantation have been previously described by Starnes' group [1]. We performed simultaneous lung lobe donation from the both donors with three teams including recipient team. Ischemic times of right lower and left lower lobes were 144 and 203 min, respectively.

The postoperative course was relatively uneventful. Two episodes of acute rejection required high-dose methylprednisolone intravenously. Because of the patient's weak respiratory muscle, postoperatively, we performed the weaning from the ventilator very carefully. We started weaning on postoperative day (POD) 26, increasing the duration without ventilatory support gradually. The patient was completely weaned from a ventilator on POD 48. In spite of long-term preoperative steroid use (prednisolone: 15 mg/day), excellent bronchial healing was

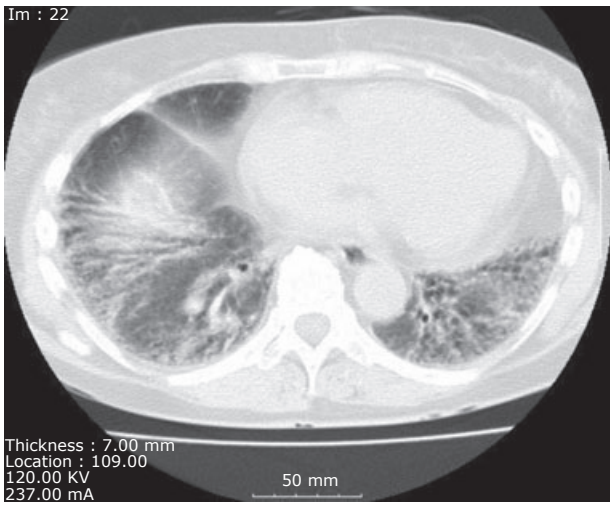

Figure 1 Computed tomographic scan of the chest demonstrated severe fibrosis in the bilateral lower lobes.

observed bilaterally. The patient was discharged from the hospital on POD 124. At that time, her vital capacity was 2.43 l (73.8% predicted) and arterial blood gas in room air revealed a pH of 7.47, PaO_2 of 85.3 mmHg, and $PaCO_2$ of 33.2 mmHg. Fifteen months postoperatively, she has resumed her normal life without oxygen inhalation, and is able to perform daily activities.

Lung transplantation for the treatment of respiratory failure caused by systemic diseases remains controversial. Interstitial pneumonia associated with collagen disease is a rare indication for lung transplantation. Because of the various manifestations of collagen disease, each patient should undergo individual consideration. According to the guidelines for the selection of lung transplant candidates proposed by the International Society of Heart and Lung Transplantation (ISHLT), evidence for quiescent systemic disease is recommended, and any evidence of active vasculitis should preclude referral [2]. Shitrit *et al.* recently reported a review of the literature on lung transplantation for scleroderma. In their review of 54 patients with scleroderma, there was no difference in infection, rejection, and 2- and 5-year survival rates between patients with scleroderma and other lung transplant recipients [3].

Dermatomyositis is well known that DM is often associated with malignancies. It is for these reasons that there are very few reports of lung transplantation for DM [4]. In performing lung transplantation for DM, two major issues should be carefully discussed. First, DM should be well controlled irrespective of IP. Secondly, the possibility of coexisting malignancy should be excluded.

In the present case, inflammatory myositis and scleroderma were well controlled by steroid and cyclosporine administration. However, IP was progressive and clearly life-threatening. Selva-O'Callaghan *et al.* reported two cases of lung transplantation for acute IP associated with DM, which were unsuccessful, but the cause of death could not be determined [5]. Recently, Kim *et al.* reported successful bilateral lung transplantation for a patient with acute form of IP associated with DM from a cadaveric donor [4]. The patient did not show recurrent DM or IP at 11 months. Patients with the acute form of IP are often in a very unstable condition. Two of three reported patients with the acute form of IP died after cadaveric lung transplantation. In contrast, our present case did not show an acute form of IP.

In the present case, 7 years passed without malignant diseases since the diagnosis of DM had been made. Careful systemic preoperative examination including whole-body high resolution CT, FDG-PET, and blood tumor markers revealed no coexisting malignancy before lung transplantation.

In 1997, Japanese law for system of donation for organ transplantation from the patient with brain death was established. However, the number of lung transplantation performed from brain-dead donors has been less than 10 per year in the whole country and the realistic option for this patient was only living-donor lobar lung transplantation, as obtaining brain-dead donors is extremely difficult.

To our knowledge, this case represents the first living-donor lobar lung transplantation for a DM patient. Close post-transplant follow-up is mandatory for the possible occurrence of malignancy in the future, with whole-body high-resolution CT, FDG-PET, and blood tumor markers.

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