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Case report

Veno-venous extracorporeal membrane oxygenation bridged living-donor lung transplantation for rapid progressive respiratory failure with pleuroparenchymal fibroelastosis after allogeneic hematopoietic stem cell transplantation

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ABSTRACT

Cases of extracorporeal membrane oxygenation (ECMO) bridged lung transplantation (LTx) are rare in Japan because an allocation system to prioritize patients based on urgency remains to be established. For critically ill patients who cannot wait for a brain-dead donor LTx, ECMO bridge to living-donor LTx may be the only practical option. A 21-year-old woman with pleuroparenchymal fibroelastosis after hematopoietic stem cell transplantation was admitted to our hospital with rapidly progressive respiratory failure. She was waitlisted for 6 months before admission, but veno-venous ECMO was initiated. She was

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transported under ECMO support via a jet plane and underwent successful living-donor LTx.

1. Introduction

Use of extracorporeal membrane oxygenation (ECMO), as a bridge to lung transplantation (LTx), is increasing significantly across Europe and the United States [1]. These increases are attributed to technological and experiential advances related to ECMO use and the modern lung allocation scoring systems, which prioritize patients according to urgency [2]. In contrast, the Japanese lung allocation system is based on accrued time on the transplantation list. Consequently, patients with rapidly progressive disease, who need ECMO support, often miss the opportunity to undergo LTx from a brain-dead donor. Here, we report a case of progressive respiratory failure and interstitial pneumonia after hematopoietic stem cell transplantation, successfully treated by ECMO bridged living-donor LTx.

2. Case report

The patient was a 21-year-old woman who was diagnosed with femur B-precursor cell lymphoma with humerus, retroperitoneum, and central nervous system involvement at 4 years of age. After chemotherapy combined with allogeneic umbilical cord blood stem cell transplantation, she achieved complete disease remission. Although late effects after the treatment, including developmental disorders related to thyroid, growth, and ovarian hormones, had been observed since the patient was 8 years of age, her respiratory state was stable. However, 10 years after treatment, she began complaining of shortness of breath. By 19 years of age, her respiratory symptoms had substantially worsened. Chest X-ray and CT revealed bilateral reticular shadow along the bronchi and traction bronchiectasis, predominantly in the upper lobe (Fig. 1A-D). She was diagnosed with pulmonary fibrosis after hematopoietic stem cell transplantation. Although corticosteroids were started, her symptoms worsened along with recurrent pneumothoraces. Pulmonary function testing showed severe restrictive respiratory impairment (forced vital capacity [FVC]: 0.38 L, %FVC: 14%). She was registered for LTx from a brain-dead donor at 20 years of age. However, she was admitted to our hospital with progressive dyspnea at 21 years of age. Chest X-ray and CT demonstrated diffuse ground-glass opacities mainly in the left lower lobe (Fig. 1-E-G). Laboratory results showed elevated C-reactive protein concentrations and elevated KL-6 (Table 1, Fig. 2). BNP was elevated from 73.8 pg/ml in the previous month to 237.0 pg/ml on admission; her body weight increased from 22 kg in the previous month to 24 kg on admission. Echocardiography on admission showed low ejection fraction (44.3%), mild tricuspid regulation (pressure gradient 36 mmHg), right ventricular dilatation, and normal inferior vena cava diameter (4 mm) with respiratory changes, all of which were not a significant change compared with results from the previous year. Cardiologists assessed that she had chronic heart failure that slightly worsened with tachycardia and hypoxia, but was not mainly responsible for her respiratory failure. Infectious pneumonia and heart failure were excluded, leaving acute exacerbation of interstitial pneumonia as the most likely diagnosis. Although steroid pulse therapy was introduced, her respiratory symptoms and chest images gradually worsened, and she was mechanically ventilated on the eighth day of admission (Fig. 2). Due to severe restrictive lung dysfunction, her carbon dioxide levels did not improve, and we introduced veno-venous ECMO that day.

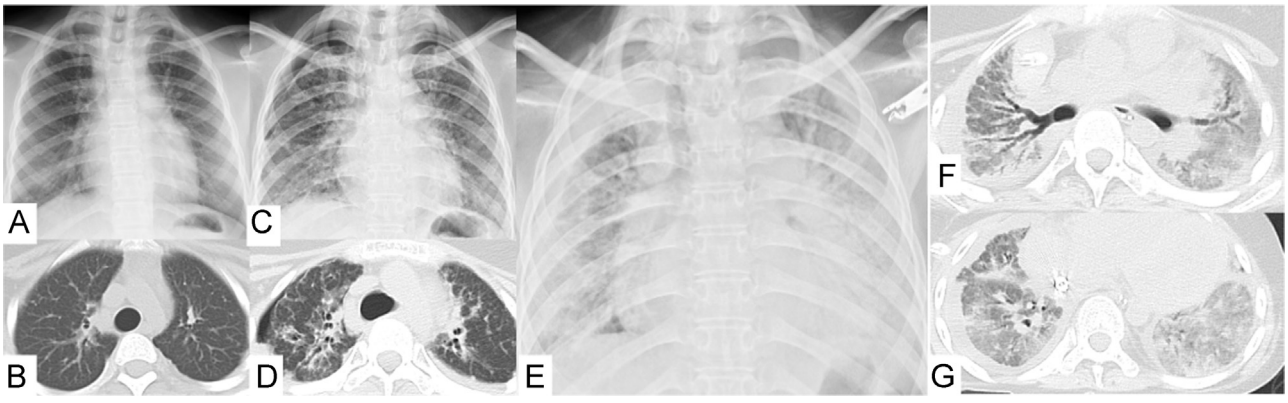

Fig. 1 – Chest images. (A) Chest X-ray and (B) CT at 14 years of age are almost normal, (C) Chest X-ray at 20 years of age, with pneumothorax apparent in both lungs. (D) CT at 20 years of age; fibrotic changes along the bronchi are apparent. (E) Chest X-ray on the day of admission. (F, G) CT showed ground-glass opacity in broad area of both the lungs. Abbreviations: CT: computed tomography.

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