A Case of diffuse alveolar hemorrhage in a severe pulmonary arterial hypertension (PAH) patient

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Introduction: Pulmonary arterial hypertension(PAH) is a rare disease of the pulmonary vasculature. Accumulating evidence indicates suggests that dysfunction in the pulmonary vascular system of PAH patients can lead to an increased tendency for pulmonary hemorrhage, which can be life-threatening. We reported a case of severe PAH in a patient who underwent lung transplantation and experienced diffuse alveolar hemorrhage.

Case report: A 54-year-old man was referred to our hospital due to dyspnea and edema. He had previously been diagnosed with pulmonary hypertension at a local medical center but had not received advanced medications. Transthoracic echocardiography revealed severe resting pulmonary hypertension, with an estimated systolic pulmonary arterial pressure of 115mmHg, and severe right ventricle dysfunction (Figure 1). Further evaluation through right cardiac catheterization(RHC) showed normal pulmonary capillary wedge pressure (14 mmHg) but elevated mean pulmonary artery pressure (66 mmHg) and pulmonary vascular resistance (14.6 wood units). Based on the results of RHC, we diagnosed the patient with PAH. We started advanced medications including treprostinil, macitentan, and sildenafil, which resulted in symptom improvement. However, after 1 month, the patient suddenly developed coughing and dyspnea again. Chest computed tomography revealed areas of consolidation and patchy ground glass opacity in both lungs (Figure 2). In response to this critical situation, we applied extracorporeal membrane oxygenation (ECMO). Bronchoscopy was then performed, revealing evidence of bleeding in all bronchi, with blood present up to the trachea (Figure 3). As the patient's condition was worsened, our multidisciplinary team made the decision to proceed with a lung transplantation. Following the lung transplantation, the patient has been undergoing rehabilitation, and his symptoms have improved.

Conclusion: In PAH, diffuse alveolar hemorrhage is reported to occur rarely. Various treatments are available to improve bleeding, but lung transplantation is considered as one of the favorable options for simultaneously addressing pulmonary arterial hypertension and hemorrhage.

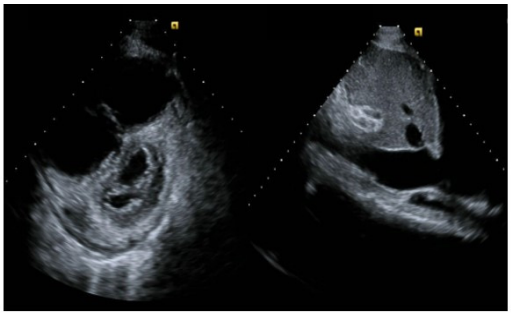

Figure 1. Transthoracic echocardiography(TTE). TTE showed D-shaped og left ventricle and dilated right ventricle. Tricuspid regurgitation peak velocity was 5m/s and complete inferior vena cava plethora was detected. Estimated systolic pulmonary arterial pressure was 115mmHg.

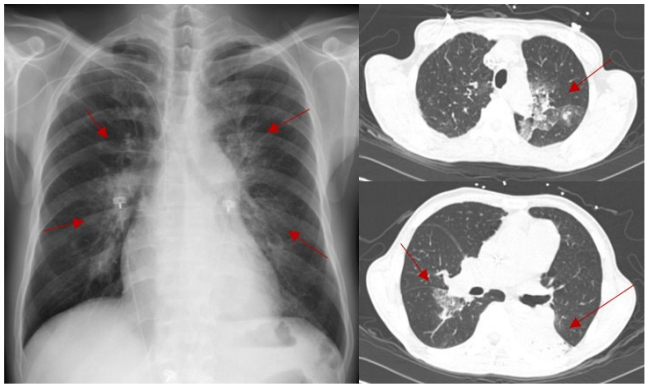

Figure 2. Chest X-ray and computed tomography(CT). Chest X-ray showed Increased hilar pulmonary vascular markings. Chest CT showed multifocal patchy areas of ground glass opacity on both lung and total atelectasis of left lower lobe(red arrows).

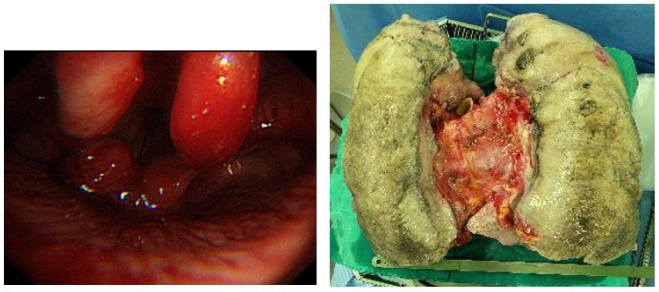

Figure 3. Brochoscopy and Explanted lung. The bronchoscopic findings showed active bleeding in all bronchi. Explanted lung showed pulmonary hemorrhage especially at left lower lobe.