Case of Refractory Idiopathic Pulmonary Arterial Hypertension

고려대학교 구로병원 내과[1]

황규일[1], *나진오[1]

Introduction: Idiopathic pulmonary arterial hypertension (IPAH) is a very rare disease. The diagnosis of IPAH is often delayed. Moreover, its registry data has been reported 5-yr survival rate is only 57% if remained untreated. Recently several IPAH-specific therapies has been developed. Nevertheless, the mortality rate of IPAH is still considerable. Here, we report case of a young male patient with IPH

Case: A 23-year-old man who had no medical histoy had been transferred from military hospital for newly developed dyspnea (NYHA II~III) while walking for three months. Initial chest X-ray showed severe cardiomegaly and following transthoracic echocardiography revealed severe pulmonary hypertension. NT-proBNP level was elevated to 2085 pg/ml. Chest CT and lung perfusion scan showed no evidence of pulmonary embolism nor perfusion defect. Because he complained arthralgia at time of first visit, studies for connective tissue disease was done but all negative. Right heart catheterization was performed for evaluation of etiology; mean pulmonary arterial pressure (MPA) 70 mmHg, pulmonary capillary wedge pressure (PCWP) 8 mmHg, pulmonary vascular resistance (PVR) 24.5 WU, cardiac output (CO) 2.35 L/min, and cardiac index (CI) 1.28 L/min/m^2. The patient was finally diagnosed with group 1 IPAH. Treatment was started with endothelin receptor blocker (macitentan) monotherapy. After ten months later, atrial flutter was developed and electrical ablation was done and add PDE5 inhibitor (sildernafil). Full dose oral prostacyclin (selexipag 1600 mg bid) was also added six month later. After triple combination therapy, his symptoms disappeared. After 5-yr treatment, however, following echocardiography and right heart catherization showed still D-shaped LV and only minimal improvement (MPA 81mmHg, PCWP 14 mmHg, PVR 21.7 WU, CO 3.18L/min, CI 1.77 L/min/m^2).

Discussion: We reported medically retractable IPAH case which has been known as a disease with high mortality. Although several PAH specific-drugs have been proven to improve symptom and survival rate, the early diagnosis, early combination therapy and careful follow-up is mandatory for medically refractory cases.