Pulmonary Arterial Hypertension in a Patient with Connective Tissue Disease and ALCAPA Syndrome

Konnektif Doku Hastalığı ve ALCAPA Sendromu olan Bir Hastada Pulmoner Arteriyal Hipertansiyon

Wang Kin Wong¹, Wan-Jing Ho², Jaw-Ji Chu³, Shue-Fen Luo⁴

Abstract
A 43-year-old female with mixed connective tissue disease was incidentally discovered to be an adult survivor of an anomalous left coronary artery originating from the pulmonary artery during a work-up for her pulmonary hypertension. We describe here the management strategy of severe pulmonary hypertension prior to and after the surgical correction of the congenital anomaly.

Key words: Pulmonary hypertension, pulmonary arterial hypertension, anomalous left coronary artery originating from the pulmonary artery, ALCAPA, connective tissue disease.

Özet
Mikst konnektif doku hastalığı olan 43 yaşındaki kadında, pulmoner arteriyel hipertansiyon araştırması sırasında, yaşayan bir yetişkinde, pulmoner arterden orijin alan sol koroner arter anomalişini saştırdı. Konjenital anomalinin cerrahi tedavisi öncesi ve sonrası, ciddi pulmoner hipertansiyon yönetim stratejisinin açıklыoruz.

Anahtar Sözcükler: Pulmoner arteriyal hipertansiyon, pulmoner hipertansiyon, pulmoner arterden kaynaklanan sol koroner arter anomalişisi, ALCAPA, konnektif doku hastalığı.

¹School of Medicine, Chang Gung University, Chang Gung Memorial Hospital, Tao-Yuan, Taiwan
²Department of Cardiology, Chang Gung Memorial Hospital, Chang Gung University, Tao-Yuan, Taiwan
³Department of Cardiothoracic and Vascular Surgery, Chang Gung Memorial Hospital, Chang Gung University, Tao-Yuan, Taiwan
⁴Department of Rheumatology, Allergy and Immunology, Chang Gung Memorial Hospital, Chang Gung University, Tao-Yuan, Taiwan

Submitted (Başvuru tarihi): 20.12.2020 Accepted (Kabul tarihi): 06.01.2021

Correspondence (İletişim): Wan-Jing Ho, Department of Cardiology, Chang Gung Memorial Hospital, Chang Gung University, Tao-Yuan, Taiwan

e-mail: auditory@cgmh.org.tw
Anomalous left coronary artery originating from the pulmonary artery (ALCAPA) is a rare congenital heart disease (CHD) that can be classified into infant and adult types (1). Most of those with the infant type die from heart failure during infancy secondary to myocardial ischemia. Patients with the adult type survive due to collateral development between the left and right coronary arteries, although they may be susceptible to chronic myocardial ischemia, pulmonary hypertension (PH) and sudden cardiac death. Pulmonary hypertension (PH) is classified into five subgroups: Pulmonary arterial hypertension (PAH), PH due to left heart disease, PH due to chronic lung disease (CLD) and/or hypoxia, PH due to pulmonary artery obstruction, and PH with an unclear and/or multifactorial mechanism. PAH fulfills the hemodynamic criteria of an mPAP > 20 mmHg, PAWP ≤ 15 mmHg, and PVR ≥ 3 WU (2). PH may be idiopathic or secondary to other diseases such as connective tissue disease (CTD), CHD, chronic liver disease, HIV infection, or schistosomiasis.

CASE
We report here on the case of a 43-year-old woman with a 3–4-month history of progressive dyspnea on exertion, with a WHO function class (FC) of III. A physical examination revealed a regular pulse of 92 beats/min, blood pressure of 136/93 mm Hg and respiratory rate of 16 breaths/min. Cardiac auscultation revealed an accentuation of P2 without murmur. Edema was absent in both legs.

The patient had long-term mixed connective tissue disease (CTD), including systemic sclerosis and systemic lupus erythematosus. Her surgical and family history was unremarkable.

Serologic studies showed the level of C3 and C4 to be within normal limits and the anti-dsDNA antibody was negative. The serum brain natriuretic peptide (BNP) level was 43.5 pg/ml. To assess whether the pulmonary hypertension (PH) was related to CLD, a pulmonary function test (PFT) was carried out, revealing a forced expiratory volume in 1 s and a forced vital capacity (FVC) ratio of 80%, a FVC of 71%, a diffusing capacity of the lung for carbon monoxide (DLCO) of 54%, and a DLCO/alveolar volume ratio of 85%. The 6-min walk distance (6MWD) was 559 m and chest radiography (Figure 1a) revealed moderate cardiomegaly. A high-resolution computed tomography (HRCT) of the chest revealed interstitial lung disease (ILD) limited to the lower lung fields (Figure 1b). Echocardiography revealed severe PH, a hypertrophic right ventricle, a dilated main pulmonary artery (MPA), normal left ventricular systolic function (ejection fraction = 79%) and a large amount of pericardial effusion. A lung perfusion scan revealed a low probability of pulmonary emboli. To confirm the diagnosis of pulmonary arterial hypertension related to CTD (CTD-PAH), cardiac catheterization was performed and the hemodynamic data obtained were as follows: right atrial pressure (RAP), 8 mmHg; mean pulmonary artery pressure (mPAP), 57 mmHg; pulmonary artery wedge pressure (PAWP), 15 mmHg; cardiac output (CO), 3.4 l/min; cardiac index (CI), 2.4 l/min.m²; and pulmonary vascular resistance (PVR), 12 WU. A 7% step-up in oxygen saturation was observed in the MPA with a pulmonary-systemic flow ratio (Qp/Qs) of 1.3. An anomalous left coronary artery originating from the pulmonary artery (ALCAPA) was detected during a right coronary angiography (Figure 2a and b, Video 1 and 2 ▶), and a 3D-CT angiography was performed to reveal the ALCAPA and adjacent structures (Figure 3a and b).

Figure 1a and b: Frontal chest x-ray showing moderate cardiomegaly (a); an axial image from high-resolution computed tomography (CT) demonstrating interstitial lung fibrosis (b)
CONCLUSION

The identification of the main cause of PAH was challenging in this patient due to the coexistence of ALCAPA, CTD and ILD. Managing this complex condition requires multidisciplinary care.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS


YAZAR KATKILARI


REFERENCES