A 37-year-old man presented at the emergency department with recent-onset dyspnea (New York Heart Association functional class IV) and abdominal distention. He had no prior history of exercise intolerance, shortness of breath, cyanosis, or palpitation. In the emergency department, tachycardia and engorged jugular veins were observed, and breath sounds were decreased on the left lower lung field, with no crackles. There was no cardiac murmur. The liver was palpable in the distended abdomen.

Chest radiography demonstrated marked cardiomegaly with a snowman appearance of increased pulmonary vascular markings (Figure 1, left). The ECG showed sinus tachycardia with evidence of right ventricular hypertrophy (Figure 1, right). Laboratory tests revealed hypoxia (PaO₂ 55.9 mm Hg, SaO₂ 89.1%) and elevated levels of N-terminal prohormone of B-type natriuretic peptide (15 125 pg/mL). A computed tomographic angiogram confirmed type I total anomalous pulmonary venous return (supracardiac type; Figure 2, left), with massive thromboembolism of the left pulmonary arteries (Figure 2, right). A transthoracic echocardiogram demonstrated a large atrial septal defect, a dilated and hypertrophied right ventricle compressing the left ventricle with biventricular dysfunction (ejection fraction 34%), loss of drainage of pulmonary venous flow into the left atrium, and large amounts of pericardial effusion (Figure 3 and online-only Data Supplement Movies I through IV). Pulmonary hypertension was suggested from the tricuspid regurgitation flow and inferior vena cava plethora (pulmonary arterial systolic pressure was estimated at 60 mm Hg with the modified Bernoulli equation). The pulmonary veins were traced and were found to have a connection into the superior vena cava through the vertical and innominate veins (online-only Data Supplement Movies II and III).

After pericardiocentesis for symptom relief, a corrective operation was performed, with subsequent procedures of thromboendarterectomy, direct anastomosis of left atrium–pulmonary venous trunk, atrial septal defect closure, and ligation of the vertical vein. The patient was discharged 2 weeks after the operation and has been followed up for 1 year without any discomfort (online-only Data Supplement Movies V and VI).

Disclosures

None.
**Figure 2.** CT angiogram. **Left,** The vertical vein (VV) drained into the superior vena cava (SVC) via the innominate vein (IV). **Right,** A large amount of pericardial effusion (PE) and massive thromboembolism (arrow) of left pulmonary arteries (PA) was seen.

**Figure 3.** Transthoracic echocardiography revealed a dilated right ventricle and right atrium (RA) and a large amount of pericardial effusion (PE), along with an atrial septal defect (ASD). The pulmonary veins (PV, white arrow) drained into the vertical vein (VV, red arrow), not into the left atrium. LV indicates left ventricle.
Atrial Septal Defect With Total Anomalous Pulmonary Venous Return in an Adult
Dong-Ho Shin, Hyo-Eun Park, Sang Il Choi, Kay-Hyun Park, Jung Yun Choi and Goo-Yeong Cho

_Circulation_. 2011;123:e612-e613
doi: 10.1161/CIRCULATIONAHA.110.986752
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2011 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the
World Wide Web at:
http://circ.ahajournals.org/content/123/21/e612

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2011/05/26/123.21.e612.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org//subscriptions/