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Atrial Septal Defect With Total Anomalous Pulmonary Venous Return in an Adult

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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 (Pao_2 55.9 mm Hg, Sao_2 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).

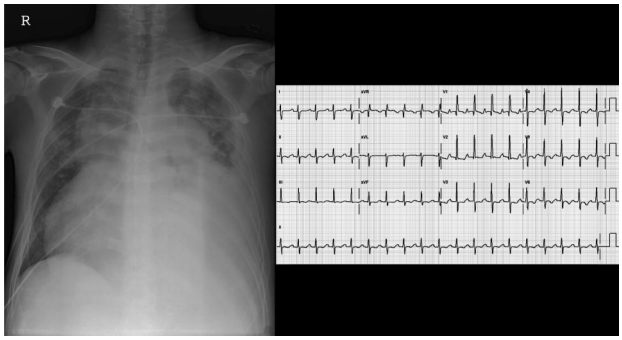

Figure 1. Left, Chest radiograph revealed marked cardiomegaly with snowman appearance of increased pulmonary vascular markings. **Right**, ECG indicating sinus tachycardia with evidence of right ventricular hypertrophy.

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.

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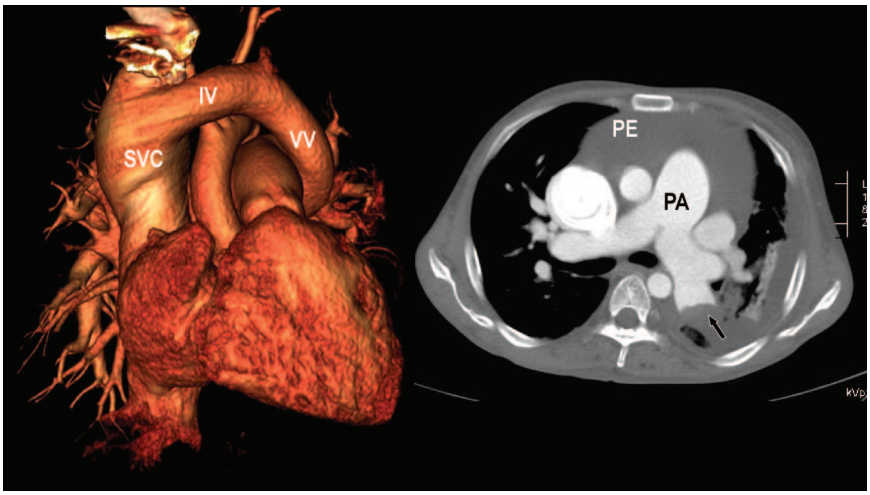

Figure 2. CT angiogram. **Left**, The vertical vein (VV) drained into the superior vena cava (SVC) via the innominate vein (IV). **Right**, A large amount of pericardial effusion (PE) and massive thromboembolism (arrow) of left pulmonary arteries (PA) was seen.

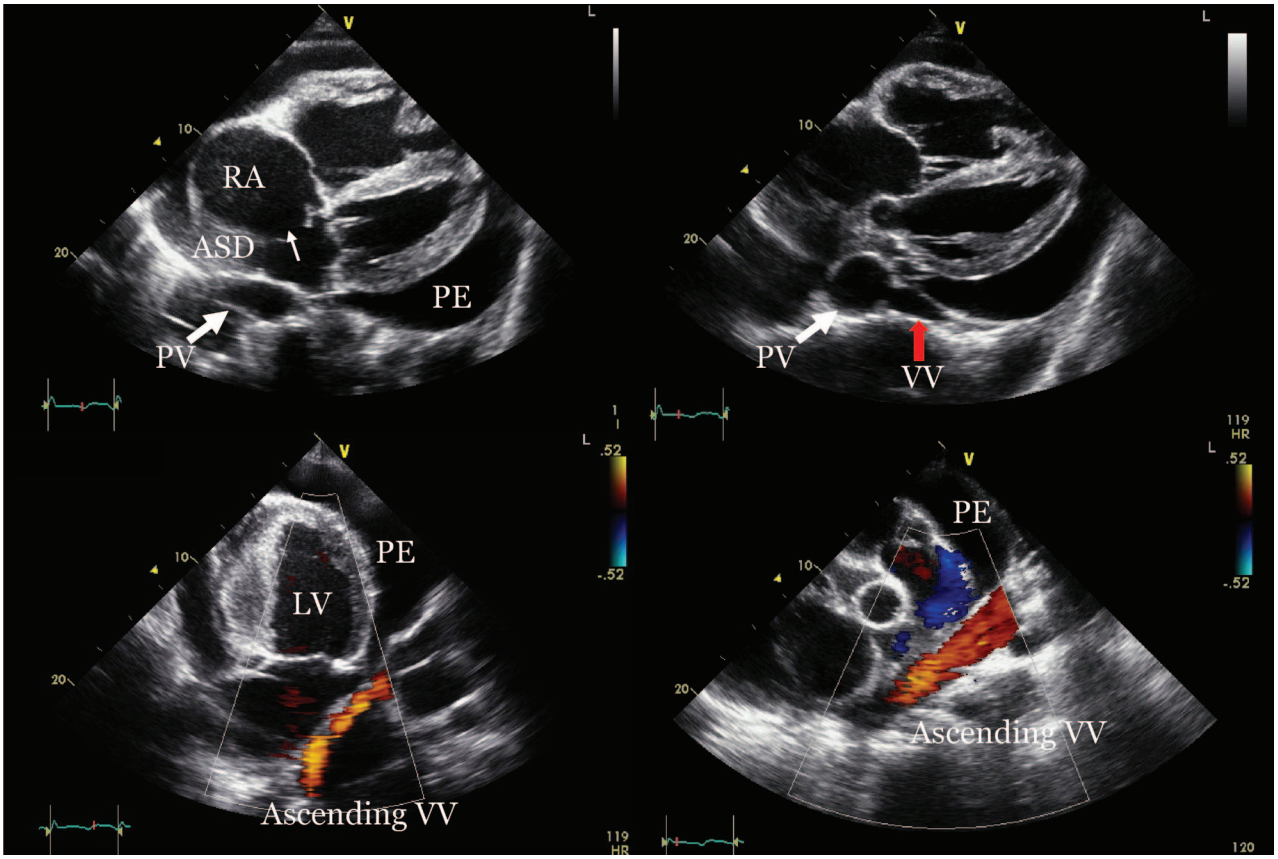

Figure 3. Transthoracic echocardiography revealed a dilated right ventricle and right atrium (RA) and a large amount of pericardial effusion (PE), along with an atrial septal defect (ASD). The pulmonary veins (PV, white arrow) drained into the vertical vein (VV, red arrow), not into the left atrium. LV indicates left ventricle.