Surgical Correction of Total Anomalous Pulmonary Venous Return in an Adult Patient

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Case Report

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Abstract

Total anomalous pulmonary venous return (TAPVR) is a rare congenital heart disease. Most TAPVRs require surgical corrections in the neonatal period and survival to adulthood without surgical correction is extremely rare. Most untreated patients with large atrial septal defects and no pulmonary venous obstruction have pulmonary vascular damage from pulmonary over circulation. It is rare for TAPVR to be reported in adulthood, and we report TAPVR in a 44-year-old patient treated successfully with surgical correction. A snowman-shaped heart, including cardiomegaly and an increase in pulmonary blood flow, was seen in the chest X-ray, and A large-sized (around 3 cm) atrial septal defect (ASD) with dilated right atrium, right ventricle, and pulmonary artery was detected on echocardiography. Heart computed tomography was performed for further evaluation, and supra-cardiac type TAPVR without any obstructive lesion was identified. The patient is doing well, and sinus rhythm and mild mitral valve regurgitation have remained during 2.5 years of outpatient follow-up.

Case Report

A 44-year-old man was admitted for acute onset palpitations and dyspnea. On physical examination, clubbed fingers were observed. The patient's oxygen desaturation was 89% in room air, and atrial tachycardia was seen on electrocardiography. Blood chemistry evaluation showed elevated liver enzymes (aminotransferase/alanine aminotransferase: 2060/1649 U/L, total bilirubin/ direct bilirubin 4.41/0.84 mg/dL) and amino-terminal pro-brain natriuretic peptide (NT-proBNP) levels up to 6789 pg/mL. A snowman-shaped heart, including cardiomegaly and an increase in pulmonary blood flow, was seen in the chest X-ray (Fig. 1). Cardiomegaly on chest X-ray was detected 23 years ago and noted in his past medical history. However, there was no follow-up.

A large-sized (around 3 cm) atrial septal defect (ASD) with dilated right atrium, right ventricle, and pulmonary artery was detected on echocardiography. In addition, a D-shaped left ventricle (LV) and severe mitral valve regurgitation were also detected. Sinus rhythm recovered after digitalization and diuretic administration. The mitral valve regurgitation was improved to a mild degree on echocardiography, and the liver enzymes were also normalized. Heart computed tomography was performed for further evaluation, and supra-cardiac type total anomalous pulmonary vein return (TAPVR) without any obstructive lesion was identified. The pulmonary venous drainage course, including the superior vena cava was severely dilated. However, the left atrium and left ventricle sizes were relatively good (Fig. 1). The pulmonary arterial pressure was 43/22 mmHg (Mean: 29 mmHg) on right heart catheterization (Table 1).

Table 1
Preoperative cardiac catheterization data

Site	O2 saturation (%)	Pressure systolic/diastolic pressure (mean), mmHg
Superior vena cava	90.7	
Right atrium	85.4	12/4 (7)
Right ventricle	88.7	43/8 (20)
Pulmonary artery	88.7	43/22 (29)
Left atrium	89.6	12/5 (7)
Aorta	90	100/60 (73)

We performed a confluent vein left atrium direct anastomosis and ASD patch closure by the transverse sinus approach without total circulatory arrest. The vertical vein was ligated. The cardiopulmonary bypass (CPB) time and aorta cross-clamp time were 147 and 61 minutes, respectively. We also checked the mitral valve morphology. The mitral valve showed diffuse leaflet thickening at the anterior and posterior valves. However, the coaptation margin of the valve leaflet was good. Thus, we did not perform mitral valve surgery. CPB weaning was smooth. After CPB weaning, there was no problem with the pulmonary venous drainage course, and the mitral valve regurgitation was mild. The patient was extubated on postoperative day (POD) 0 and transferred to the general ward on POD 1. He was discharged on POD 8. The patient is doing well, and sinus rhythm and mild mitral valve regurgitation have remained during 2.5 years of outpatient follow-up (Fig. 2).

Discussion

TAPVR is a rare anomaly accounting for only 1.5–2.2% of congenital heart disease [1]. Almost all patients with TAPVR present with symptoms that include cyanosis and heart failure in the neonatal period, which require early surgical correction. A large ASD and no obstruction of the pulmonary venous drainage pathway are the two major factors affecting survival. However, most untreated patients with large ASDs and no pulmonary venous obstruction have pulmonary over-circulation, leading to pulmonary vascular damage [2]. Thus, it is unusual to encounter adult patients without symptoms.

Fortunately, this patient had a relatively good LV size with a large ASD, no pulmonary venous pathway obstruction, and low pulmonary arterial pressure despite having no pulmonary stenosis. Thus, this patient had a good recovery. Severe mitral valve regurgitation was seen in this patient upon admission. However, the degree of mitral valve regurgitation improved after rhythm conversion. The mitral valve morphology in the operative field showed diffuse thickening with a good coaptation margin. There was no evidence of rheumatic history. Further follow-up is required.

There are few adult TAPVR patients worldwide. The cases reported previously showed a favorable prognosis in the postoperative phase. However, approximately 10–15% of the patients have evidence of late pulmonary vein obstruction, which tends to be recurrent and progressive [3]. In the three years of follow-up after surgery, this patient did not show any complications, including pulmonary venous obstruction or mitral valve regurgitation. However, long-term surveillance and monitoring are required.

Declarations

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Conflict of interest

The authors declare that they have no competing interests.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent

Informed consent was obtained from all individual participants included in the study.

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Figures

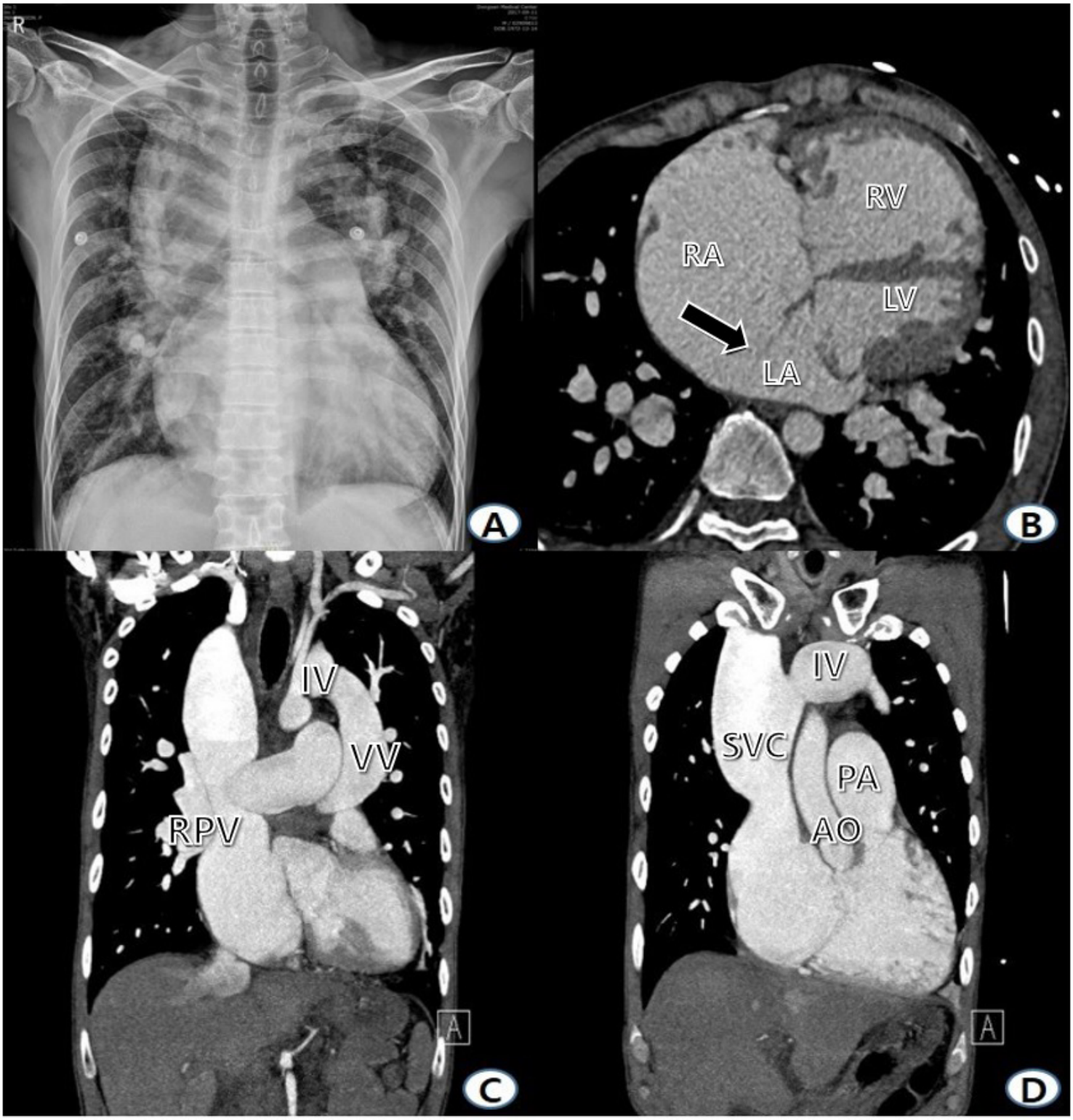

Figure 1

Preoperative chest X-ray and computed tomography (CT). (A) Preoperative chest X-ray. (B) Axial view on CT shows an enlargement of the RA and RV, secundum-type ASD (arrow), and left dislocation of the LA and LV. (C), (D) Coronal view on CT shows that the IV is connected to the VV and drains to the SVC.

RA right atrium, *RV* right ventricle, *ASD* atrial septal defect, *LA* left atrium, *LV* left ventricle, *IV* innominate vein, *VV* vertical vein, *RPV* right pulmonary vein, *SVC* superior vena cava, *PA* pulmonary artery, *AO* aorta.

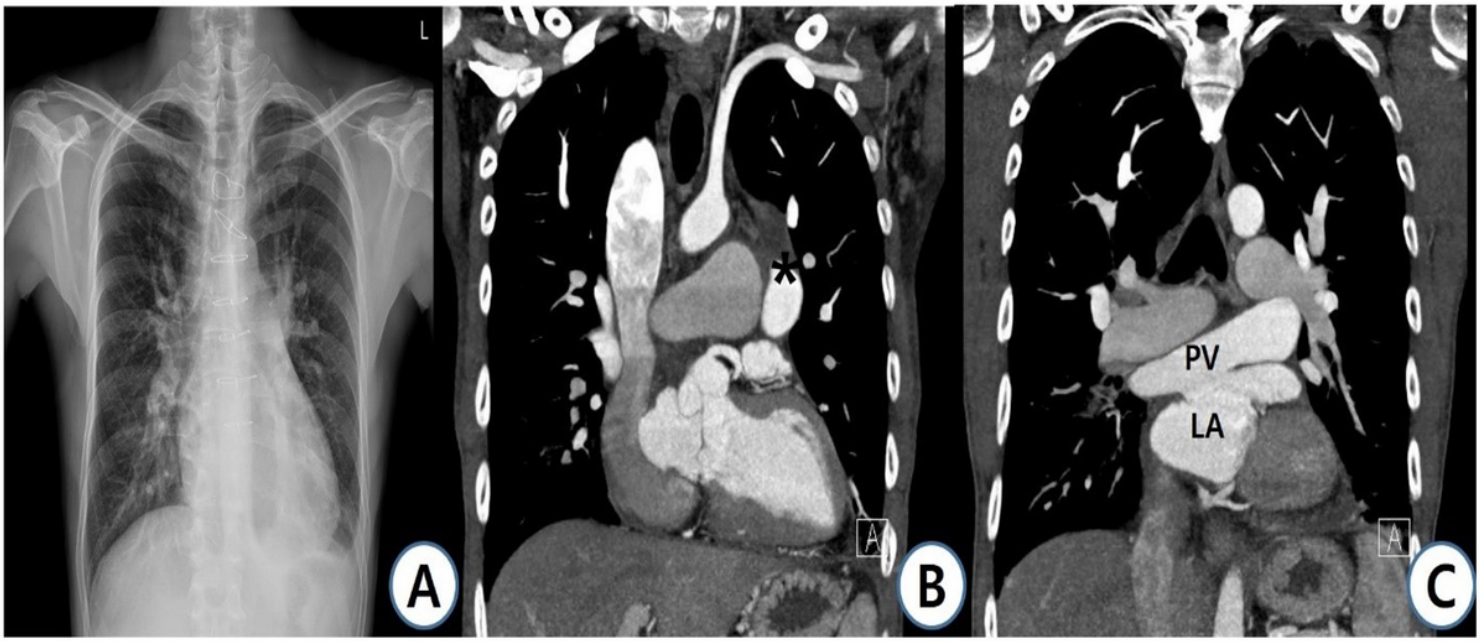

Figure 2

Postoperative computed tomography (CT). (A) Chest X-ray at discharge. (B), (C) coronal view on chest CT at 2.5 years after surgery. The PV was connected to the LA.

PV pulmonary vein, *LA* left atrium.

* ligated vertical vein.