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Case report

Partial anomalous pulmonary venous return with dual drainage to the superior vena cava and left atrium with pulmonary hypertension

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A B S T R A C T

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital cardiovascular anomaly. A 68-year-old woman was referred to our hospital for detailed examination for pulmonary hypertension (PH). She had been diagnosed as having pulmonary artery dilation and suspected to have PH during a health check seven years prior. A contrast computed tomography showed that the right upper pulmonary vein (RUPV) returned to the superior vena cava (SVC) with a preserved normal connection to the left atrium (LA). Surgical repair was performed. We reported an extremely rare case of isolated PAPVR with PH showing dual drainage into the SVC and LA.

1. Introduction

Partial anomalous pulmonary venous return (PAPVR) is a congenital cardiovascular anomaly, which is characterized by abnormal connection of one or more, but not all, pulmonary veins (PVs) to systemic veins such as the superior vena cava (SVC), inferior vena cava (IVC), and/or the right atrium (RA) [1]. PAPVR arises from the failure of regression of primitive lung drainage when the pulmonary vascular bed and the common pulmonary vein from the left atrium (LA) establish a connection in the embryonic stage [2]. In common types of PAPVR, anomalous PVs connect to those systemic veins and lack a normal connection to the LA. In this case report, we describe a case of isolated PAPVR with a duplicated connection of the anomalous right upper pulmonary vein (RUPV) into both the SVC and LA, which preserved the normal connection and was unexpectedly accompanied by pulmonary hypertension (PH).

2. Case report

A 68-year-old Japanese woman with a history of hypertension and uterine fibroids was referred to our hospital for the evaluation of pulmonary hypertension (PH). Seven years prior to admission, an enlargement of the pulmonary arteries was found on a chest radiograph during a regular health check (the patient reported no symptoms). She was suspected to have PH based on the elevated tricuspid regurgitation pressure gradient (TRPG) (50–55 mmHg), measured using transthoracic echocardiography (TTE). She refused a detailed examination at that time. After treatment with beraprost 60 μg, warfarin, and losartan for 2 years, she discontinued them on her own judgement; exertional dyspnea and palpitation appeared shortly afterward. One year before admission, she had an episode of syncope, probably triggered by paroxysmal atrial fibrillation, atrial tachycardia, and PH, and she was carried to the previous hospital. After initiation of amlodipine 5mg, bisoprolol fumarate 2.5mg, rivaroxaban 15mg, and pilsicainide hydrochloride hydrate 50mg, she was then referred to our hospital for further examination and treatment.

On admission, her height and weight were 158 cm and 78 kg, respectively. The vital signs on admission were as follows: blood pressure: 133/71 mmHg; pulse rate: 64 beats per minute; and percutaneous oxygen saturation: 97% on room air. On auscultation, the rate and rhythm were regular, no heart murmur was detected, and the respiratory sounds were clear. Blood examinations were within normal range except for elevated brain natriuretic peptide (78.2 pg/ml). A chest radiograph showed pulmonary artery enlargement and cardiomegaly (cardiothoracic ratio: 57%). An electrocardiogram was normal with sinus rhythm. The TTE showed an elevated TRPG (52.8 mmHg), right heart enlargement, and interventricular septum displacement toward the left; however, no interatrial shunt flow, such as atrial septal

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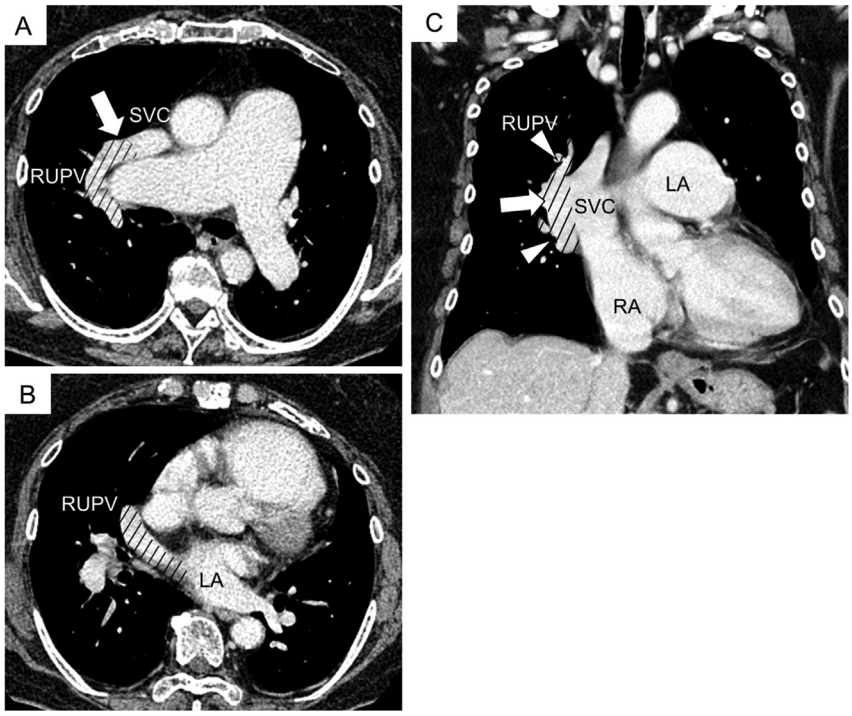

Fig. 1. (A) Contrast enhanced chest computed tomography showed abnormal connection of the right upper pulmonary vein (RUPV: shaded) and superior vena cava (SVC) (arrow). (B) The RUPV (shaded) preserved normal connection to the left atrium (LA). (C) Coronal. The RUPV (shaded) had an abnormal connection (arrow) to the SVC and returned to the LA. RUPV: right upper pulmonary vein, SVC: superior vena cava, LA: left atrium, RA: right atrium.

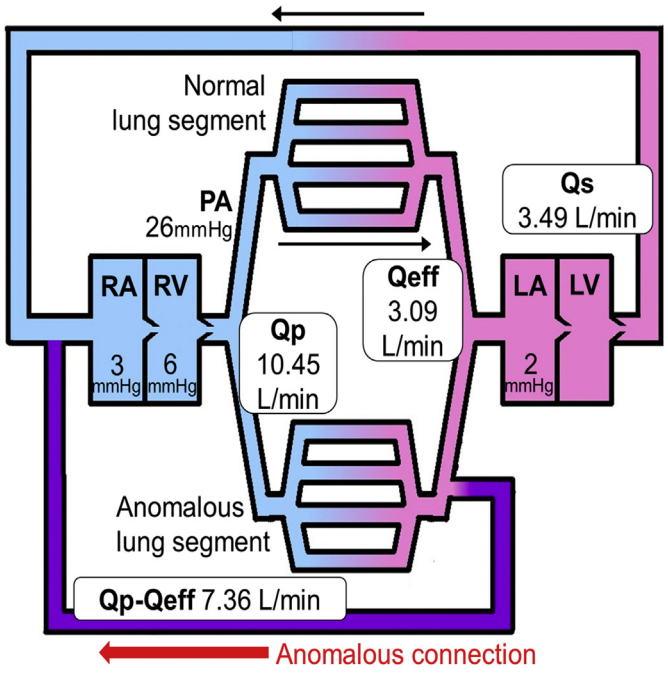

Fig. 2. Schema of pulmonary circulation. RA: right atrium, RV: right ventricle, PA: pulmonary artery, LA: left atrium, LV: left ventricle, Qp: pulmonary blood flow, Qeff: effective pulmonary blood flow, Qs: systemic blood flow.

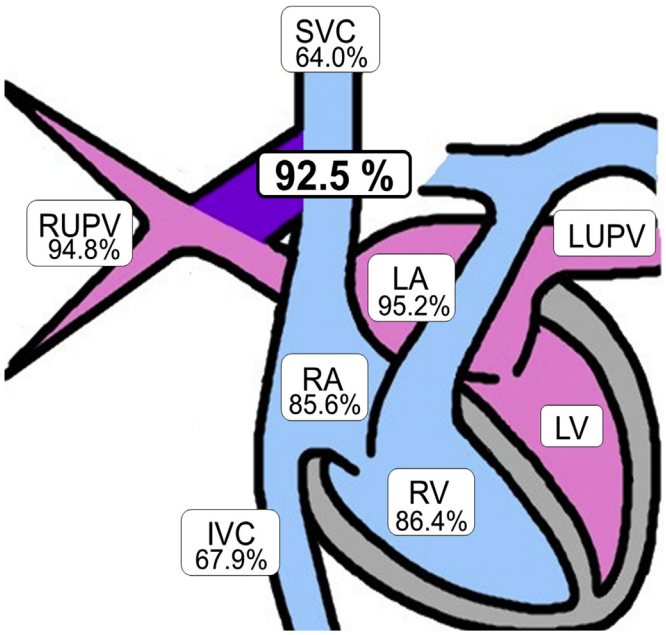

Fig. 3. Oxygen saturation of cardiac catheterization. The oxygen saturation elevated between the superior vena cava (SVC) and right atrium (RA). RUPV: right upper pulmonary vein, SVC: superior vena cava, LA: left atrium, RA: right atrium, RV: right ventricle, IVC: inferior vena cava, LV: left ventricle, LUPV: left upper pulmonary vein.

defect (ASD), was detected. A contrast chest CT revealed that the RUPV connected not only to the LA but also to the SVC (Fig. 1). Right cardiac catheterization (RHC) data were as follows: pulmonary arterial pressure (PAP) systolic/diastolic (mean): 47/12 mmHg (26 mmHg); left atrium pressure (LAP): 2 mmHg; cardiac index (CI) measured by Fick methods: 5.68 L/min/m^2; left-to-right shunt ratio: 70.5%; right-to-left shunt ratio: 11.5%; pulmonary blood flow (Qp): 10.45 L/min; systemic blood flow (Qs): 3.49 L/min; effective pulmonary blood flow (Qeff): 3.09 L/min; Qp/Qs 3.00; pulmonary vascular resistance (PVR): 122.3 dyn.-sec.cm^{-5} (Fig. 2). RHC also revealed increased oxygen saturation in the SVC (Fig. 3). The pulmonary angiography confirmed duplicated connection of the RUPV to both the SVC and LA (Fig. 4). The 6-min walk test (6MWT) revealed a walking distance of 337 m with lowest oxygen saturation of 93% and Borg scale value of 6. Based on all these findings, the patient was finally diagnosed with isolated PAPVR with a duplicated connection of the RUPV to the SVC and LA. Surgical repair was performed: disconnection and reinforcement with a pericardial patch between the RUPV and SVC. The hemodynamic data the day after surgery were as follows: PAP: 34/22 mmHg (26 mmHg); cardiac output:

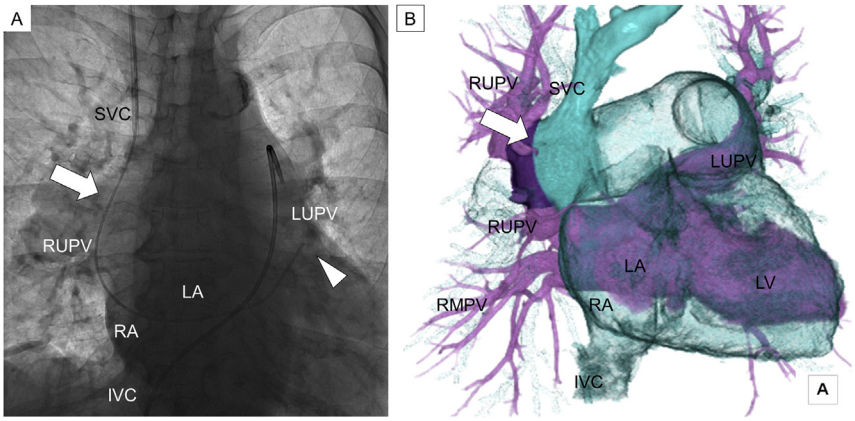

Fig. 4. (A) The catheter tip (arrowhead) is in the left atrium (LA) near the left upper pulmonary vein (LUPV) from the SVC via anomalous pulmonary vein and normal RUPV to the LA (arrow). (B) Three-dimensional computed tomography specifies the abnormal connection (arrow, purple) between the RUPV and SVC. The right cardiac system is shown in blue, the left cardiac system is shown in transparent pink. SVC: superior vena cava, RUPV: right upper pulmonary vein, RA: right atrium, IVC: inferior vena cava, LA: left atrium, LV: left ventricle, LUPV: left upper pulmonary vein. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Table 1
Literature review of cases of dual drainage PAPVR (RUPV-SVC/LA).

	Peynircioglu R et al. [3]	Karacus, G et al. [4]	The current report
Sex	Female	Female	Female
Age	73	66	68
Symptom	Dyspnea on effort	Dyspnea	Dyspnea
Comorbidities	Hypertension, Arthritis, irritable bowel syndrome, carotid endarterectomy, and paroxysmal arterial fibrillation		Hypertension, uterine fibroid
Echocardiography			
eRVSP (mmHg)	61	39	50–55
Hemodynamics			
PAP (mmHg)	29/9 (mean 18)	No data	47/12 (mean 26)
PCWP (mmHg)	7	No data	2
PVR (Wood units)	1.2	No data	2.3
CO (L/min)	9.05	No data	10.5
Qp/Qs	2.2	No data	3.0
Diagnosis	RHC, PAG	TTE, MRI	RHC, PAG
Treatment	Surgery	Surgery	Surgery

eRVSP: estimated right ventricular systolic pressure; PAP: pulmonary arterial pressure; PCWP: pulmonary capillary wedge pressure; PVR: pulmonary vascular resistance; CO: cardiac output; Qp/Qs: a ratio of pulmonary to systemic flow; RHC: right heart catheterization, PAG: pulmonary angiography, TTE: transthoracic echocardiography, MRI: magnetic resonance imaging.

3.90 L/min/m^2; CI: 2.21 L/min/m^2; Qp/Qs: 1.0. One month after surgery, the cardiothoracic ratio was 46%, the interventricular septum displacement disappeared, and the TRPG was 39 mmHg as seen on TTE. The classification of WHO functional assessment for pulmonary hypertension improved from class III to II.

3. Discussion

We describe an extremely rare case of isolated PAPVR, which showed a duplicate connection of the RUPV to the LA and SVC. The abnormal connection of the RUPV could have resulted in increased pulmonary blood flow and PH, although the normal connection had been preserved.

This subtype of PAPVR, with a duplicated connection of the RUPV to both the SVC and LA, is extremely rare [3]. The precise incidence of dual-drainage PAPVR is unclear; to the best of our knowledge, only three patients, including our case, have been reported [3,4] (Table 1). All three patients were female; their age at diagnosis was 73, 66, and 68 years, respectively. In all patients, progressive symptoms were the clue to precise examination and diagnosis. In two of three cases, RHC revealed that the pulmonary blood flow was highly increased, although the mean PAPs were normal or slightly higher than normal.

In this case, the left-to-right shunt, which was estimated by measuring the Qp-Qeff [5], was extremely high. It was therefore suggested that the left-to-right shunt flow could be responsible for increased pulmonary blood flow. It is seen that congenital heart disease, including PAPVR, brings about a left-to-right shunt and often induces PH [6,7]. It is reported that around 5% of adults with congenital heart disease develop PH [8,9]; the prevalence of PH associated with congenital systemic-to-pulmonary shunts is estimated to be between 1.6 and 12.5 cases per million adults [10], although the incidence of PH development in patients with PAPVR was unclear. Coexisting congenital heart defects such as ASD, and the number of the anomalous pulmonary vein connections could be related to the development of PH in patient with PAPVR [11]. In dual-drainage PAPVR, it seemed that blood flow through the normal connection might reduce the left-to-right shunt flow, although the blood flow through the normal connection was not evaluated in the current case.

Asymptomatic PAPVR might be difficult to detect. The electrocardiogram and echocardiography of patient with PAPVR having less shunt flow could be normal [11,12]. It was reported that careful interpretation of chest CT scan might be useful for detecting PAPVR and differential diagnosis of PH [7]. In this case, the CT scan clearly showed the abnormal connection between the RUPV and SVC.

We here report a case of dual-drainage PAPVR with extremely increased pulmonary blood flow and PH. Precise diagnosis contributed to the definitive surgical repair. The disease could progress silently; therefore, careful interpretation of CT scans for suspicion of PAPVR is inevitable.

Conflicts of interest

The authors have no conflicts of interest to declare.

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