Persistent left vertical vein: An unusual cause of pulmonary hypertension and cirrhosis in a patient with hypertrophic obstructive cardiomyopathy

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Introduction

The persistent vertical (levoatriocardinal) vein is an abnormal, persistent connection between the pulmonary venous system and the systemic venous system (1).

This variation accompanies total pulmonary venous return anomaly and rarely with pathologies such as atrial septal defect, ventricular septal defect, and mitral stenosis (1-3). Its association with hypertrophic obstructive cardiomyopathy (HOCM) has been shown in only one case (4). In this case report, we have presented the interventional treatment of both the pathologies in the same session.

Case Report

A 42-year-old female patient was referred to our hospital for HOCM. She had severe exertional dyspnea and palpitations during mild effort in the last months (New York Heart Association class 3).

Physical examinations showed blood pressure 130/75 mm Hg, heart rate 86/min, rhythmic, cyanosis on the lips, jugular venous distension, ascites, generalized edema, and severe bilaterally peripheral edema. Electrocardiography (ECG) indicated sinus rhythm, voltage criteria for left ventricular hypertrophy, and nonspecific ST/T wave changes. Laboratory findings included brain natriuretic peptide (BNP), 13,900 pg/mL; total bilirubin, 3.45 mg/dL; direct bilirubin, 2.3 mg/dL; alkaline phosphatase level (ALP), 24 IU/L; gamma-glutamyl transferase (GGT), 4 IU/L; and international normalized ration (INR), 1.6. Transthoracic ECG demonstrated hypertrophic obstructive cardiomyopathy, severe systolic anterior movement, moderate mitral regurgitation (MR), advanced tricuspid regurgitation, pulmonary hypertension, biatrial enlargement, stage-2 diastolic dysfunction, normal left ventricular function, global severe right ventricular dysfunction, intact interatrial and interventricular septum, and minimal pericardial effusion. The ECG measurements were left atrium (LA), 49 mm; left ventricular end-diastolic diameter (LVEDD), 36 mm; left ventricular end-systolic diameter (LVESD), 19 mm; right atrium (RA), 57 mm; right ventricle, 47 mm; septum, 25 mm; posterior wall (PW), 19 mm; ejection fraction (EF), 55%; left ventricle outflow gradient (LVOT) gradient, 196 mm Hg; and estimated pulmonary artery pressure (PAB), 85 mm Hg. In contrast-enhanced cardiac magnetic resonance imaging, hypertrophic cardiomyopathy (HCM), vertical vein between the LA and very dilated left brachiocephalic vein, biatrial enlargement, severe dilatation in the RV, vena cava inferior (VCI), pulmonary artery, patent foramen ovale (PFO), 3+ mitral insufficiency, significant pleural, peritoneal, and minimal pericardial effusions were detected (Video 1). Abdominal ultrasound revealed large and echogenic liver, moderate-severe free acid in the abdomen, VCI, and large portal vein.

Angiography showed a large vertical vein that connects the LA through the left upper pulmonary vein into the left brachiocephalic truncus (Fig. 1, Video 2). The coronary artery system was

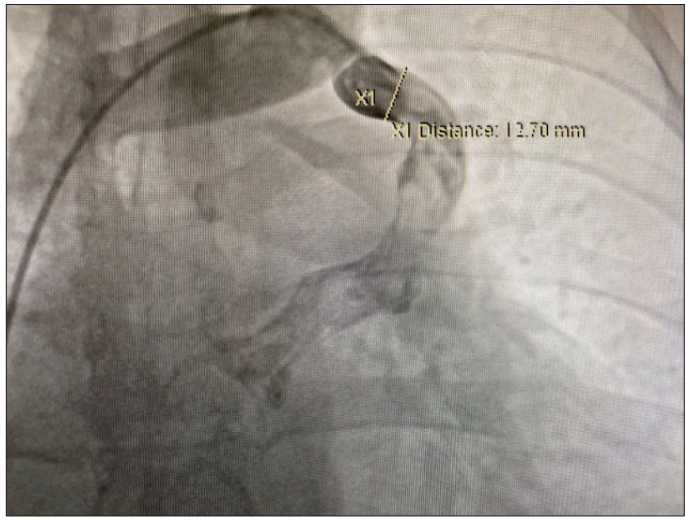

Figure 1. Angiography showed a large vertical vein that connects LA through the left upper pulmonary vein into the left brachiocephalic truncus
LA - left atrium

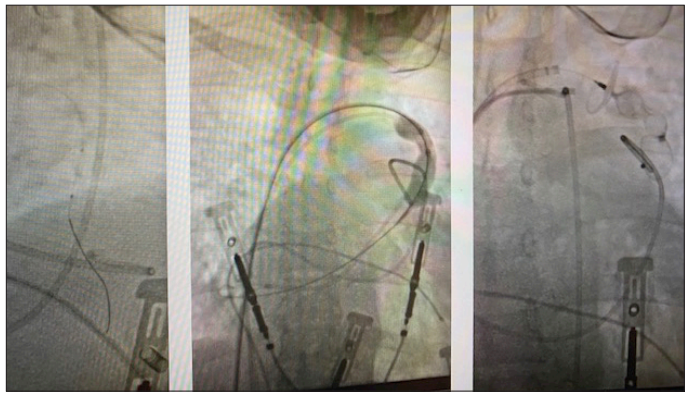

Figure 2. Simultaneous percutaneous intervention: ablation of the first septal artery with 2.5 mL of alcohol. The occlusion of the vertical vein with a 22 mm Amplatzer vascular plug (AGA Medical Corporation, Plymouth, United Kingdom). The mean LA and PA pressure values were 36 and 55 mm Hg at the end of the procedure
LA - left atrium; PA - pulmonary artery

Table 1. Right heart catheterization findings before and after the procedure

Blood gas O_2 and pressure (mm Hg)	Baseline (%, min/mean/max)		After procedure (%, min/mean/max)	
Left brachiocephalic vein	85.5		59	
Vena cava superior	74.2		58.3	
Right atrium	51.9	24/37/38	58	7/13/22
Vena cava inferior	51.9		59.7	
Right ventricle	53	21/57/104	59	10/17/49
Pulmonary artery	48.7	44/62/90	59.9	23/33/45
Left atrium	93.8	29/38/50	96	22/30/46
Pulmonary capillary wedge	94	27/34/48	95.8	23/27/40
Left ventricle	93.7	10/24/252	93.7	2/21/114

max - maximum; min - minimum

intact. Subsequent catheterization of the right side of the heart showed pulmonary hypertension [mean systolic pulmonary artery (PA) pressure, 62 mm Hg and arterial oxygen tension (aPO_2), 48.7%], and blood gas oximetry revealed an O_2 step-up at the left brachiocephalic venous truncus [pulmonary flow/systemic flow (Qp/Qs) shunt ratio, 1.98] and a pulmonary vascular resistance (PVR) of 8 WU. The PO_2 in the left vertical vein was 94.6%. The LA pressure was 38 mm Hg, and RA pressure was 38 mm Hg with a PO_2 of 51.9%.

A myocardial biopsy was performed. Histopathology of the biopsy specimen demonstrated left ventricular hypertrophy and mild disarray of cardiomyocytes and interstitial fibrosis, which is compatible with HCM. It was decided to perform percutaneous closure of the vertical vein and septal alcohol ablation for HOCM in the same session. First, the vertical vein was temporarily occluded with a balloon to evaluate how much the vein's closure would affect the patient's hemodynamics. The LA and PA pressures were re-evaluated. As the mean LA pressure increased to 41 mm Hg, septal ablation was performed to relieve LVOT. The first septal artery was ablated with 2.5 mL of alcohol. The LVOT gradient turned to normal; mean LA and PA pressures decreased to 32 mm Hg and 57 mm Hg, respectively. The vertical vein was then occluded with a 22 mm Amplatzer vascular plug (AGA Medical Corporation, Plymouth, United Kingdom). The mean LA and PA pressure was found as 36 mm Hg and 55 mm Hg, respectively, at the end of the procedure (Fig. 2). Before and after the vertical vein closure, the detailed blood gas and pressure measurements were listed in Table 1.

On the first day in the coronary care unit, nonsustained ventricular tachycardia attacks developed that did not disturb the hemodynamics. During follow-up, peripheral edema and ascites increased despite sildenafil and multiple diuretic therapies; the urine output decreased, and hypotension developed. In the echocardiographic re-evaluation, it was found that RV enlargement increased, and the systolic function significantly decreased compared with the preprocedure; PA systolic pressure could not be measured reliably owing to the advanced tricuspid regurgitation. Therefore, patient was connected to ultrafiltration, and the fluid was withdrawn for 4 days. The RV parameters partially recovered in the echocardiographic control. On the last day of the ultrafiltration, ventricular fibrillation developed, but the heart rate returned to sinus rhythm after defibrillation. Because there were also recurrent ventricular tachycardia attacks, an automatic implantable cardioverter-defibrillator implantation was decided and performed 10 days after the first procedure. N-terminal (NT)-pro hormone BNP (NT-proBNP) level after the closure was 2430 pg/mL before the hospital discharge in the early period. The patient was discharged with apixaban, tadalafil, metoprolol, verapamil, furosemide, and spironolactone treatment. Pretibial edema had completely regressed, but acid continued in the abdomen at the fourth-month control. Right heart catheterization was performed again for reliable pulmonary pressure measurements. Oxygen saturation was as follows: subclavian, 59%; vena cava superior (VCS), 58.3%; RA upper, 58.4%; RA middle, 57.7%; RA lower, 59%; VCI, 59.7%; PA, 59.9%; and aorta, 96%. The mean pulmonary capillary occluded pressure was 26 mm Hg, and mean pressures of the PA and RV were 33 mm Hg and 17 mm Hg, respectively. The PVR was calculated as 1.2 WU.

There was no rhythm problem in the long-term follow-up, except for short-term atrial fibrillation attacks. In the first year, control, dyspnea, ascites, and hepatomegaly significantly regressed, and peripheral edema disappeared. In the third year of control, physical examination was normal, and liver enzymes turned back to normal levels.

Discussion

It is known that diastolic filling is impaired, and restrictive filling patterns may develop with preserved systolic functions in HOCM (5). Initially, the vertical vein acts as a compensation mechanism. The LA pressure decreased, whereas the RA pressure and pulmonary blood flow increased. However, if the shunting continued, the pulmonary vasculature's capacity will be overwhelmed, LA pressures will return to the same levels, and right-sided heart failure will develop. This mechanism resulted in liver failure in this patient. Therefore, we aimed to correct both pathologies.

Surgical or percutaneous closure of the persistent vertical vein is controversial (6). There is no current guideline recommendation or expert consensus on the indications for vertical vein closure in adult patients, and case series are generally at the level of advice for vertical vein closure in infants with total anomalous pulmonary venous drainage (6, 7). So, there is no recommendation for routine closure of the vertical vein. Therefore, an individual approach is the best approach for these patients and similar patients according to the patient's clinic, comorbidities, and catheterization findings.

Although it is known that approximately 40%-50% of patients with HOCM have pulmonary hypertension, there are no accurate data about the reason (8, 9). Therefore, the vertical vein

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should be investigated, especially in patients with pulmonary hypertension with increased pulmonary capillary wedge pressure, and should be closed because it is a type of total pulmonary venous filling anomaly causing pulmonary hypertension.

In addition, the selection of treatment strategy is crucial in patients with HOCM, because the symptomatic benefit from septal ablation is variable. For this reason, detailed examinations, and noninvasive and invasive measurements were performed on this patient before both intervention decisions. There are 2 main reasons for preferring septal ablation first in this patient. First, when the left atrial and pulmonary pressures were measured after a temporary balloon occlusion of the vertical vein, the left atrial pressure increased to 55/35/41 mm Hg. Therefore, it was thought to relax the left ventricular outflow tract. The second reason is the decrease in PVR, PA pressure, and MR in addition to left ventricular filling pressures with septal ablation.

It is not surprising that the patient developed clinical worsening and low cardiac output early after the intervention. Vertical vein occlusion results in significantly elevated LA pressure and negative impacts on left ventricular function and cardiac output, suggesting, for a while, a small and poorly compliant left ventricle in the patient (6). It was observed that the clinical findings improved with the change in physiology over time. Septal ablation is more logical than medical treatment of pulmonary hypertension and vertical vein occlusion in another session. Our motivation for simultaneous intervention was only cost benefits for the patient. There was no option to pay the costs of intervention with different sessions for the patient. Furthermore, we could not reference another tertiary clinic because there was no experience of this procedure. The staged procedure seems more reliable for similar cases.

Conclusion

This case report is the first in the literature to show the simultaneous percutaneous treatment of both pathologies, to the best of our knowledge.

Informed consent: Written informed consent of the patient was obtained.

Video 1. HCM in the contrast-enhanced cardiac MR
HCM - hypertrophic cardiomyopathy; MR - mitral regurgitation

Video 2. Large vertical vein that connects LA through the left upper pulmonary vein into the left brachiocephalic truncus in the angiography
LA - left atrium

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