Journal of Community Hospital Internal Medicine Perspectives

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CASE REPORT

Isolated supra-cardiac partial anomalous pulmonary venous connection causing right heart failure

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Right heart failure (RHF) has been overlooked as left heart failure has predominated. One of the many causes of RHF is partial anomalous pulmonary venous connection (PAPVC), an extremely rare entity in nature. Physicians should consider the unusual causes of RHF after ruling out the common causes.

Keywords: *right heart failure*; *partial anomalous pulmonary venous connection*; *heart failure*; *ejection fraction*; *congenital disease*

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Received: 23 December 2015; Revised: 4 March 2016; Accepted: 4 March 2016; Published: 25 April 2016

One of the many causes of right heart failure (RHF) is partial anomalous pulmonary venous connection (PAPVC). Four variants of PAPVC exist: supra-cardiac, cardiac, infra-cardiac, and mixed. Herein, we report a rare entity of isolated supra-cardiac PAPVC, as the right pulmonary vein drains into the superior vena cava (SVC) with an intact atrial septum, precipitating RHF.

Case report

A 55-year-old man with hypertension, diabetes mellitus, coronary artery disease, pulmonary hypertension, and congestive heart failure presented with syncope. In the emergency department, the patient was alert and oriented with a blood pressure of 90/44 mmHg and a heart rate of 44 beats/minute. On examination, lungs were clear on auscultation, jugular venous distension was present, prominent S2 heart sound with a systolic ejection murmur 2/6 gradient auscultated loudest at the second left intercostal space, and bilateral pitting edema of the lower extremities reaching the knees was seen. Laboratory studies were significant for creatinine of 1.87 mg/dL, brain natriuretic peptide of 504 pg/mL, and troponin I of 0.06 ng/mL. Chest X-ray revealed cardiomegaly. Electrocardiography was significant for left atrial dilation and right ventricular hypertrophy. Transthoracic echocardiography showed severe right ventricular dilation decreasing the size of the left ventricle with left ventricular hypertrophy, dilated right and left atria, and severe tricuspid regurgitation with pulmonary artery systolic pressure of 85 mmHg. Ejection fraction was noted to be 55% with no evidence of atrial septal defect (ASD). Cardiac catheterization and computed tomography angiogram revealed severe pulmonary hypertension and drainage of the right pulmonary vein into the SVC (Figs. 1 and 2).

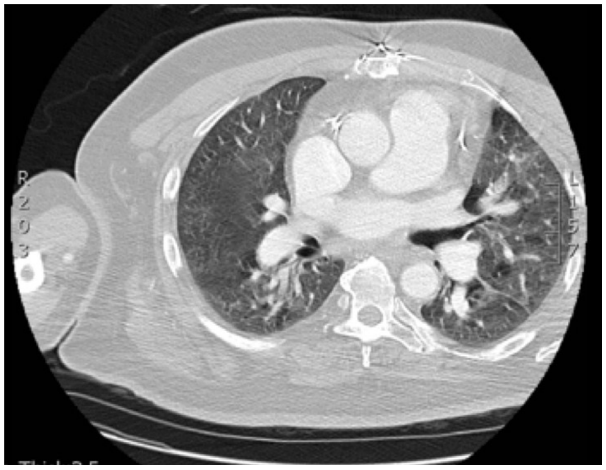

Fig. 1. Computed tomography with contrast demonstrating engorged pulmonary vasculature. Right pulmonary venous architecture adjacently positioned with a connected noted.

Citation: Journal of Community Hospital Internal Medicine Perspectives 2016, **6**: 30817 - http://dx.doi.org/10.3402/jchimp.v6.30817

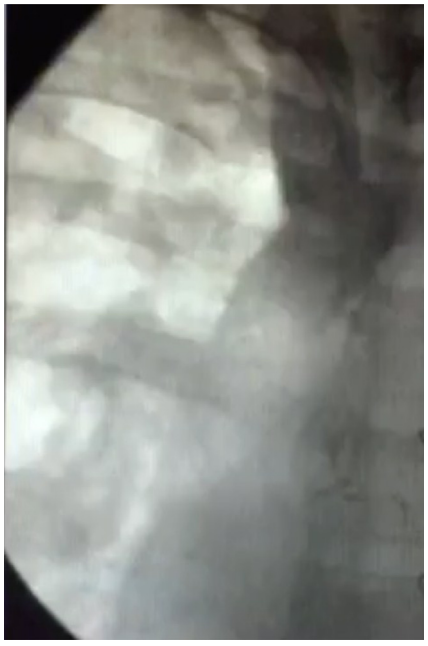

Fig. 2. Cardiac catheterization illustrating drainage from the right pulmonary vein anomalously into the superior vena cava.

Medications were titrated during hospital stay as patient was on a high dose of beta-blockers prior to presentation. Patient was discharged with bumetanide 2 mg BID and metoprolol 25 mg BID with options for elective pulmonary venous surgery.

Discussion

Anomalous of pulmonary venous connection is a rare congenital defect of the pulmonary veins which are drained into the right atrium instead of the normal return to the left atrium. This defect is classified into two subgroups: partial and total anomalous. PAPVC is characterized by abnormal return of one or more, but not all, of the pulmonary veins directly to the right atrium, or indirectly through venous connections from the anomalous pulmonary vein. Total anomalous pulmonary venous connection is a more severe form that differs from PAPVC in that all pulmonary venous vessels connect to the right atrium, and it is one of the cyanotic heart defects.

The incidence of PAPVC is found to be around 0.7% of all population, as it has been reported in autopsy data studies (1). Four variants of PAPVC have been reported; supra-cardiac, cardiac, infra-cardiac, and mixed. The most common form of PAPVC is supra-cardiac where there is a right upper pulmonary vein connecting to the right atrium or the SVC. In most patients with PAPVC, there is an associated cardiac defect, which is usually an ASD. Studies have indicated that 82% of patients with PAPVC have an ASD. However, PAPVC occurs in approximately 10% of patients with an ASD. In isolated PAPVC with intact atrial septum, patients are asymptomatic and generally are not diagnosed unless more than 50% of the pulmonary blood flow is recirculated back to the pulmonary flow (2) Our patient has isolated PAPVC that remains silent and diagnosed in his sixth decade.

Echocardiography is the most frequently used method for the diagnosis of PAPVC. Magnetic resonance imaging is now considered the procedure of choice for further investigation of PAPVC (3). Cardiac computed tomography can also be used to confirm the diagnosis of PAPVC. Cardiac catheterization can be a definitive diagnostic procedure as therapeutic intervention as well.

Conflict of interest and funding

The authors have not received any funding or benefits from industry or elsewhere to conduct this study.

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