

A Rare Combination of Persistent Left Superior Vena Cava and Partial Anomalous Pulmonary Venous Return



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Introduction

- A persistent left superior vena cava (PLSVC) is a form of anomalous venous drainage of the superior vena cava into the right atrium
- Its prevalence in up to 0.5 % of the general population, with the majority of cases undetected until the patient becomes symptomatic
- The presence of PLSVC with a partial anomalous pulmonary venous return (PAPVR) is extremely rare

Case Presentation

- A 57-year-old female with a history of tobacco abuse presented with dyspnea on exertion for 4 days
- Reported PND, orthopnea, difficulty completing a full sentence, and walking a few feet without getting short of breath

Physical Exam

- Vitals: 213/92 mmHg, 96 BPM, 98.1F, RR: 22, 94% on 5L
 Cardiac exam: 3/6 crescendo/decrescendo murmur with radiation to the right
- carotid
- Respiratory: Bibasilar crackles
- Other systems: Normal

Work up

- CBC, Mag, and BMP were normal
- BNP was normal at 41
- CXR showed no cardiopulmonary disease
- EKG showed NSR without ischemic changes
- TTE: LVEF 55% and aortic valve that is moderately stenotic with a peak velocity 3.66m/s, and mean gradient 34 mmHg. A bubble study showed LA filling before RA (Fig 1.), raising suspicion for PLSVC or PAPVR with no intracardiac shunt

Management

- The patient was diuresed and did well
- Further work up with heart catheterization (Fig 3.) as well as CT chest (Fig. 2A,B) confirmed PLSVC with no coronary sinus dilation
- She was discharged in stable condition with a follow up at an adult congenital cardiology clinic at a tertiary center
- Cardiac MRI (CMRI) (Fig 4B.) at a tertiary center showed PLSVC communicating with the coronary sinus with return into the right atrium and a small right-sided SVC (RSVC) draining into the superior right pulmonary vein with return into the left atrium.
- She remained asymptomatic with medical management for 2 years during regular cardiology follow up

Images

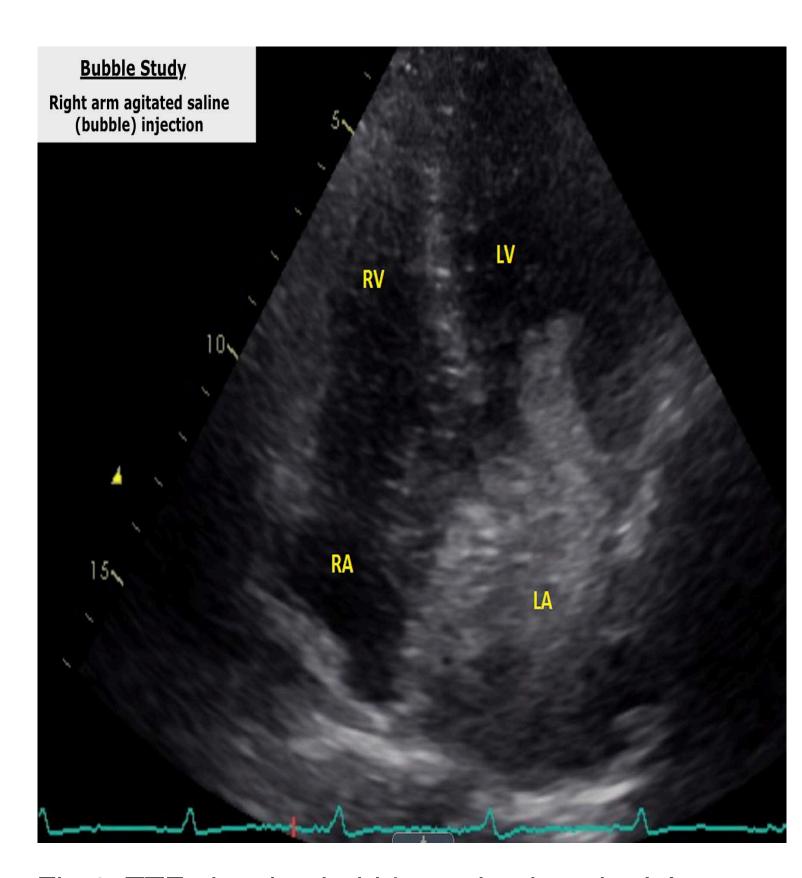


Fig 1. TTE showing bubbles going into the LA

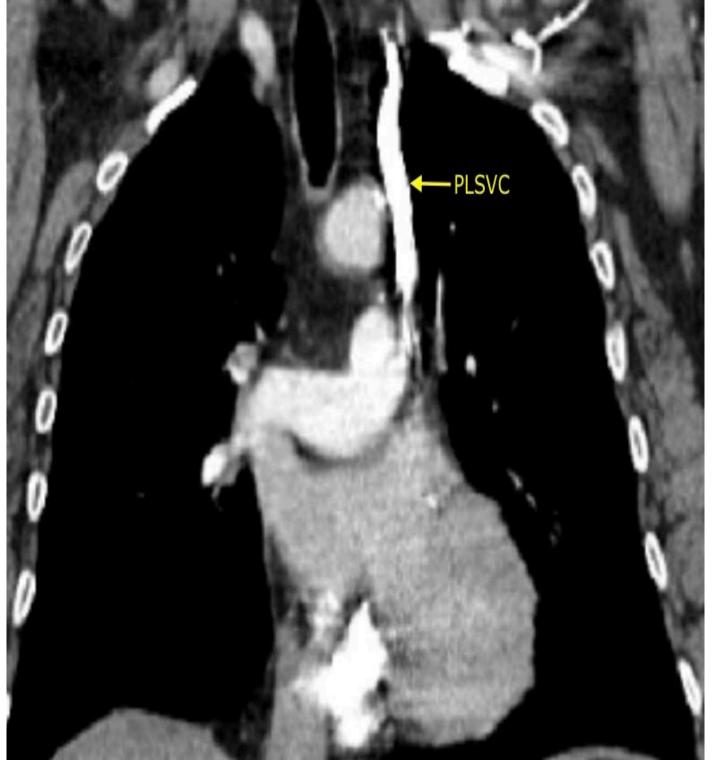


Fig 2A. CT Chest showing persistent LSVC

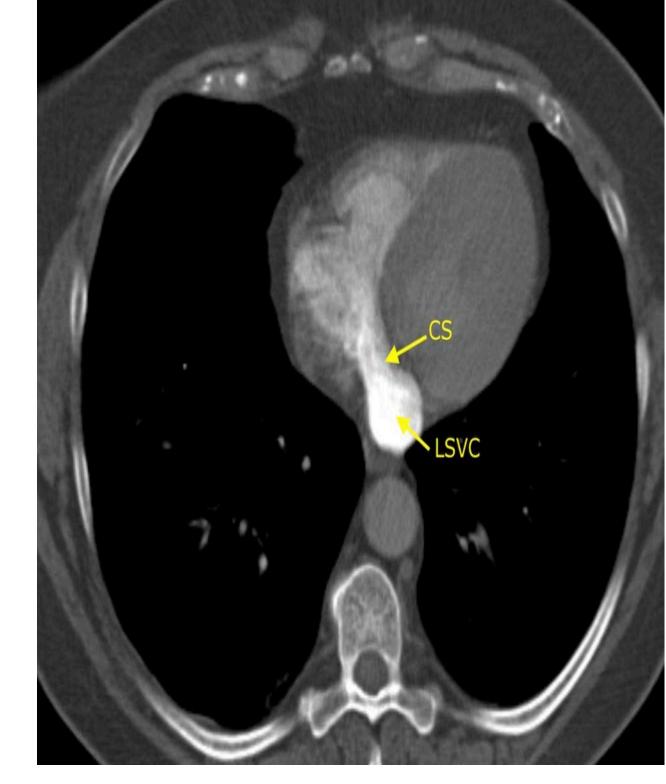


Fig 2B. CT Chest showing LSVC draining into CS

Discussion

- PLSVC results from a failure of obliteration of the left common cardinal vein and typically drains the left subclavian and jugular veins into the right atrium via the coronary sinus (CS)
- It can also associate with atrial and ventricular septal defects, endocardial cushion defects, tetralogy of Fallot, CS ostial atresia, and cor triatriatum
- Clinically, PLSVC has been associated with an increased risk of heart failure, and arrhythmias, most commonly atrial fibrillation
- Diagnosis of PLSVC is usually incidental in adults. In our case, the patient received a routine TTE with bubble study as a part of heart failure work up, Interestingly, TTE (Fig 1.) showed bubbles shown in the left chamber before the right chamber without atrial septal defect (ASD), which raised suspicion of PLSVC, coronary sinus ASD or PAPVR
- More interestingly, CMRI showed also showed PAPVR with RSVC draining into the right pulmonary vein with the return into the left atrium. Our patient did not have coronary sinus ASD which is extremely rare, but can be seen with PLSVC without dilated coronary sinus
- PLSVC is usually asymptomatic and the most common echocardiographic finding is a dilated coronary sinus, which was not present in our patient
- On the other hand, PLSVC can cause a higher incidence of complications such as arrhythmia, and cardiac tamponade when undergoing a pacemaker placement
- Our case demonstrates a rare combination of PLSVC and PAPVR



Fig 3. Coronary angiogram showing PLSVC and SVC

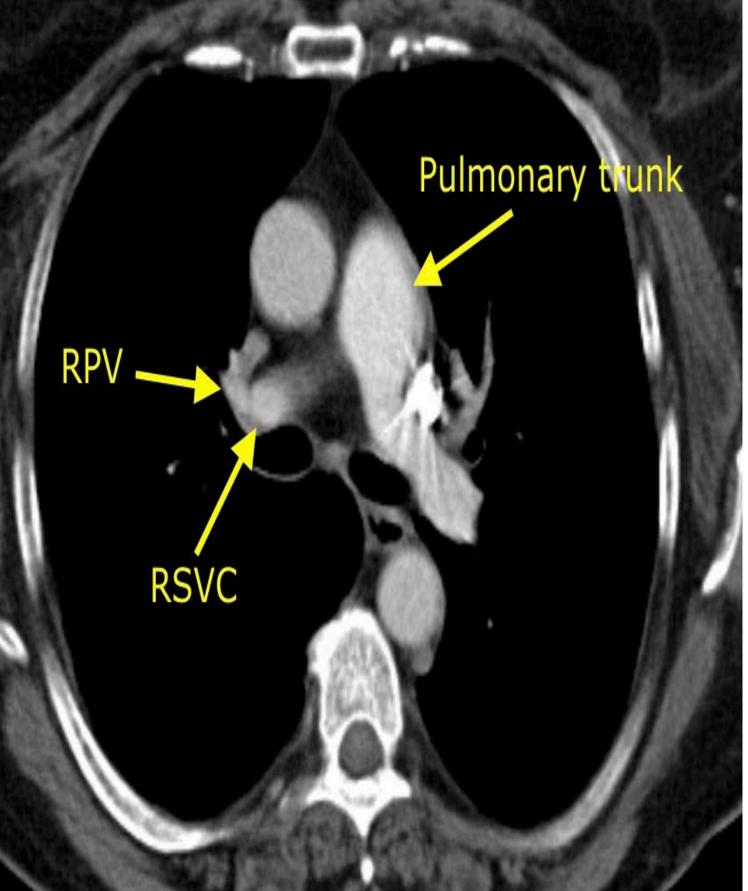


Fig 4A. CT Chest showing RPV draining into RSVC

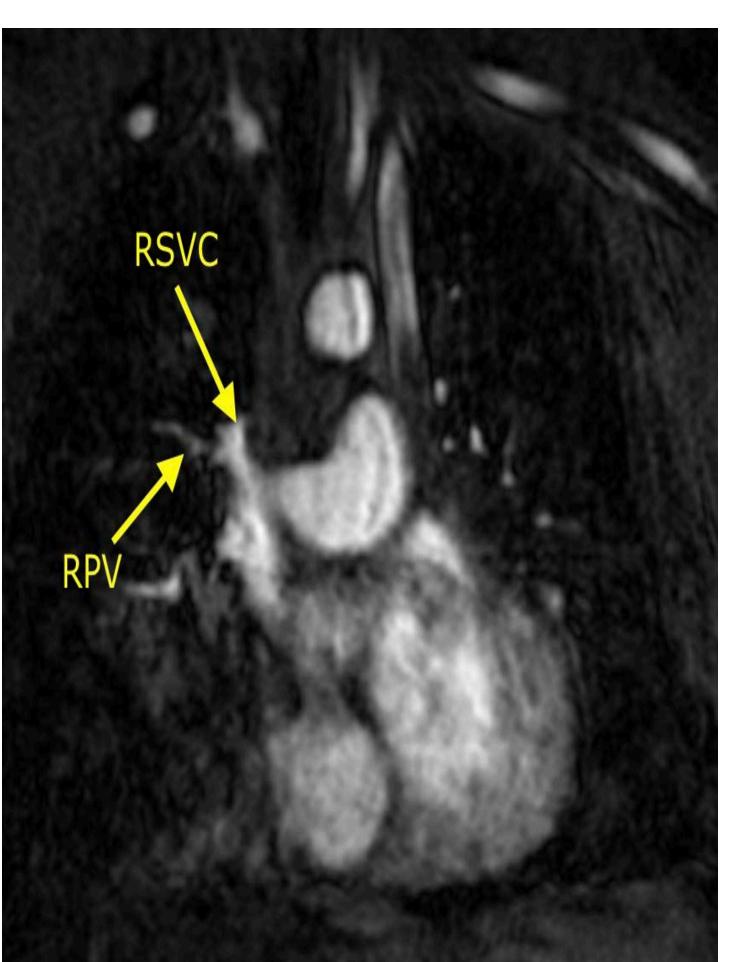


Fig 4B. CMRI showing RPV draining into RSVC

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