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

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Abstract

A persistent left superior vena cava (LSCV) is a form of anomalous venous drainage of the superior vena cava into the left atrium. We present a case of LSCV and partial anomalous pulmonary venous return (PAPVR) in a patient with progressively worsening dyspnea on exertion. A 57-year-old female with a history of aortic valve stenosis, and tobacco abuse presented with dyspnea on exertion for 4 days. On presentations her vitals were unremarkable except elevated blood pressure and oxygen saturation of 94% on 5L oxygen. Physical exam was pertinent for 3/6 crescendo-decrescendo murmur with a radiation to the right carotid, mild bibasilar crackles, and trace pitting edema in the lower extremity bilaterally. EKG showed normal sinus rhythm without ischemic changes. Echocardiogram showed preserved ejection fraction of 55% and moderately stenosed trileaflet aortic valve with a peak velocity 3.66 m/s and mean gradient 34 mmHg. A bubble study showed left atrium filling before right atrium raising suspicion for PLSVC or PAPVR with no intracardiac shunt. Further work up with heart catheterization as well as CT chest confirmed PLSVC with no coronary sinus dilation. Cardiac MRI (CMRI) 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.

Keywords

PLSVC, Congenital anomalies, PAPVR

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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 left atrium. Its prevalence is up to 0.5 % of the general population, with the majority of cases undetected until the patients become symptomatic.¹ We present a case of PLSVC and partial anomalous pulmonary venous return (PAPVR) in a patient with progressively worsening dyspnea on exertion.

A Case Presentation:

A 57-year-old female with a history of moderate aortic valve stenosis and tobacco abuse and without family history of congenital cardiac anomalies presented with dyspnea on exertion for 4 days. She reported paroxysmal nocturnal dyspnea, orthopnea, difficulty completing a full sentence, and inability to walk more than a few feet without getting short of breath.

On presentation, her blood pressure was elevated at 213/92 mmHg, heart rate was regular at 96 beats per minute, temperature was normal, respiratory rate was 22 and oxygen saturation was 94% on 5L of supplemental oxygen via nasal cannula. Physical exam revealed a 3/6 crescendo-decrescendo murmur with radiation to the right carotid, mild bibasilar crackles, and trace pitting edema in the lower extremities bilaterally. The rest of the examination was unremarkable. Laboratory workup was within normal range with BNP of 41 pg/mL. Chest radiograph showed no cardiopulmonary disease. EKG showed normal sinus rhythm without ischemic changes. Echocardiogram showed preserved ejection fraction of 55% and moderately stenosed trileaflet aortic valve with a peak velocity 3.66 m/s and mean gradient 34 mmHg. A bubble study was ordered on admission as she had no baseline echocardiogram in the setting of possible new onset heart failure. Surprisingly, it showed left atrium filling before right atrium (Fig 1.), raising suspicion for PLSVC or PAPVR with no intracardiac shunt. Further work up with CT chest (Fig. 2A, B, 4A) and cardiac catheterization (Fig 3.) 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 the 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.

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).¹ It can also be associated with atrial and ventricular septal defects, endocardial cushion defects, tetralogy of Fallot, CS ostial atresia, and cor triatriatum.² Clinically, PLSVC has been associated with an increased risk of heart failure, and arrhythmias, most commonly atrial fibrillation (AF).¹ 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

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. Also, Kim, Y.G. et al.'s study demonstrates how PLSVC plays a role in the induction and maintenance of AF.³ Thus, identifying PLSVC prior to invasive procedures, such as pacemaker implantation, cardiac resynchronization therapy, or central venous catheter placement is very important to avoid possible complications.⁴ Lastly, imaging modalities prior to catheter ablations for AF are not only useful for evaluating pulmonary venous anatomy, but also guide electrophysiologists to ablate PLSVC if detected, as it can trigger AF. Our case demonstrates a rare combination of PLSVC and PAPVR.

Conclusion:

A combination of PLSVC and PAPVR is a rare congenital anomaly. PLSVC and PAPVR are usually asymptomatic and can be found incidentally. PLSVC and PAPVR can both be associated with other congenital anomalies. PLSVC also has been associated with an increased risk of heart failure, and arrhythmias, most commonly atrial fibrillation. Identifying PLSVC on radiological study is important because the existence of PLSVC and PAPVR can change the surgical or minimally invasive approach.

References:

- 1 Irwin, R.B., Greaves, M., Schmitt, M. Left superior vena cava: revisited. *European Heart Journal – Cardiovascular Imaging* 2012; 13: 284-291.
2. Bartram U., Van Praagh S., Levine J.C., Hines M., Bensky A.S., Van Praagh R. Absent right superior vena cava in viscerotrial situs solitus. *Am J Cardiol* 1997;80:175 –83.
3. Kim YG, Han S, Choi JI, Lee KN, Baek YS, Uhm JS, Shim J, Kim JS, Park SW, Hwang C, Kim YH. Impact of persistent left superior vena cava on radiofrequency catheter ablation in patients with atrial fibrillation. *Europace*. 2019 Dec 1;21(12):1824-1832.
4. Azizova A, Onder O, Arslan S, Ardali S, Hazirolan T. Persistent left superior vena cava: clinical importance and differential diagnoses. *Insights Imaging*. 2020 Oct 15;11(1):110 Erratum in: *Insights Imaging*. 2021 Apr 16;12(1):49.

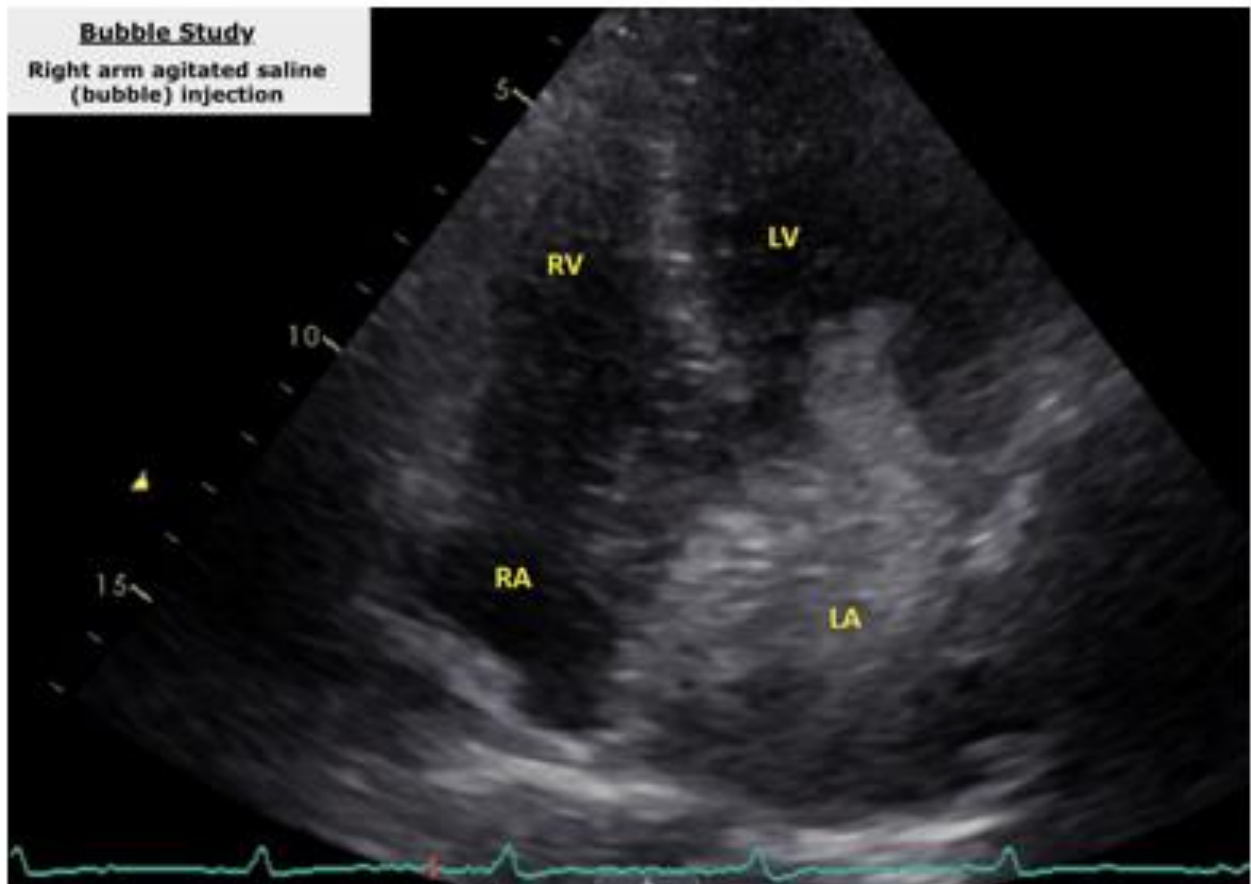


Fig 1. TTE showing bubbles going into the LA

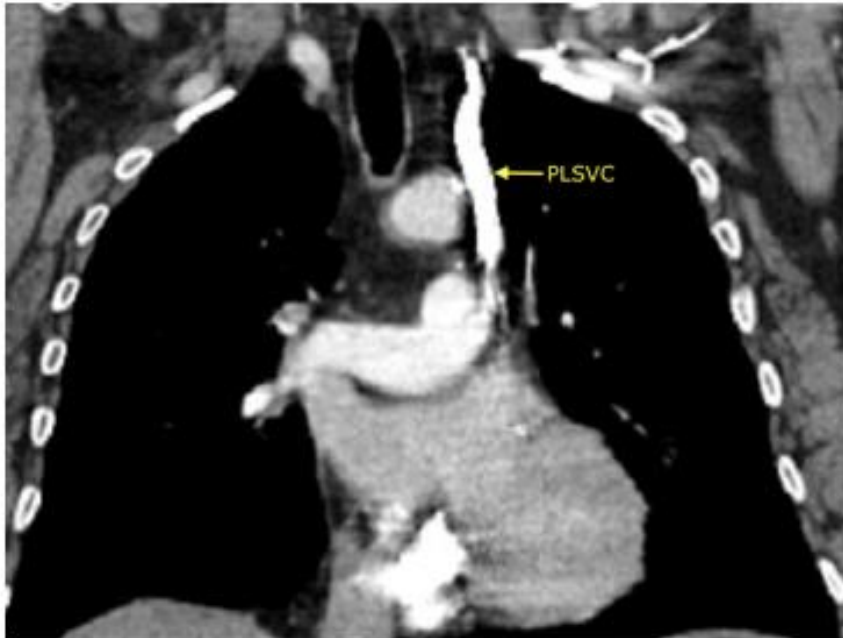


Fig 2A. CT Chest showing persistent LSVC

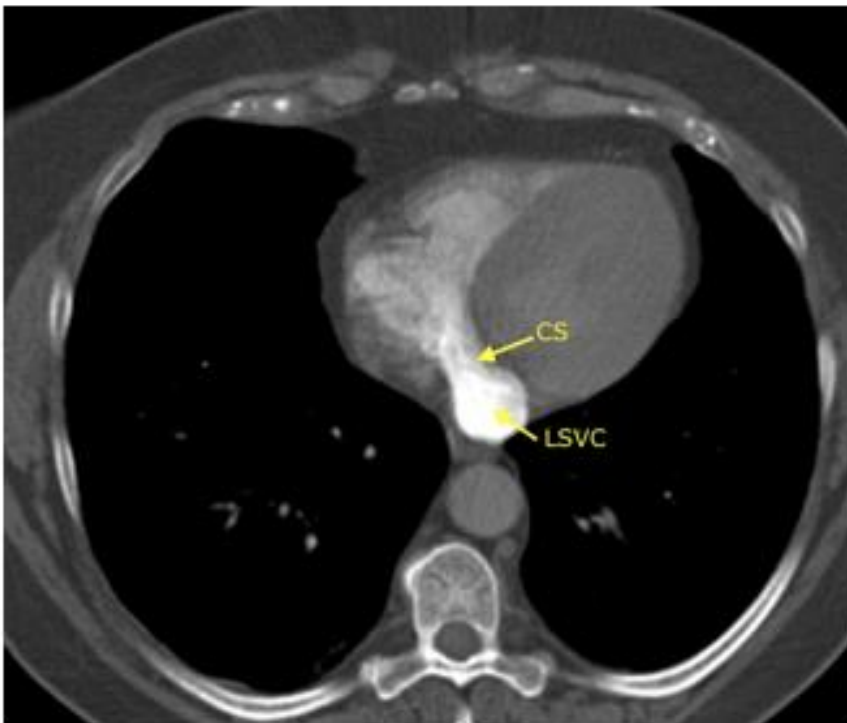


Fig 2B. CT Chest showing LSVC draining into CS



Fig 3. Coronary angiogram showing PL SVC and SVC

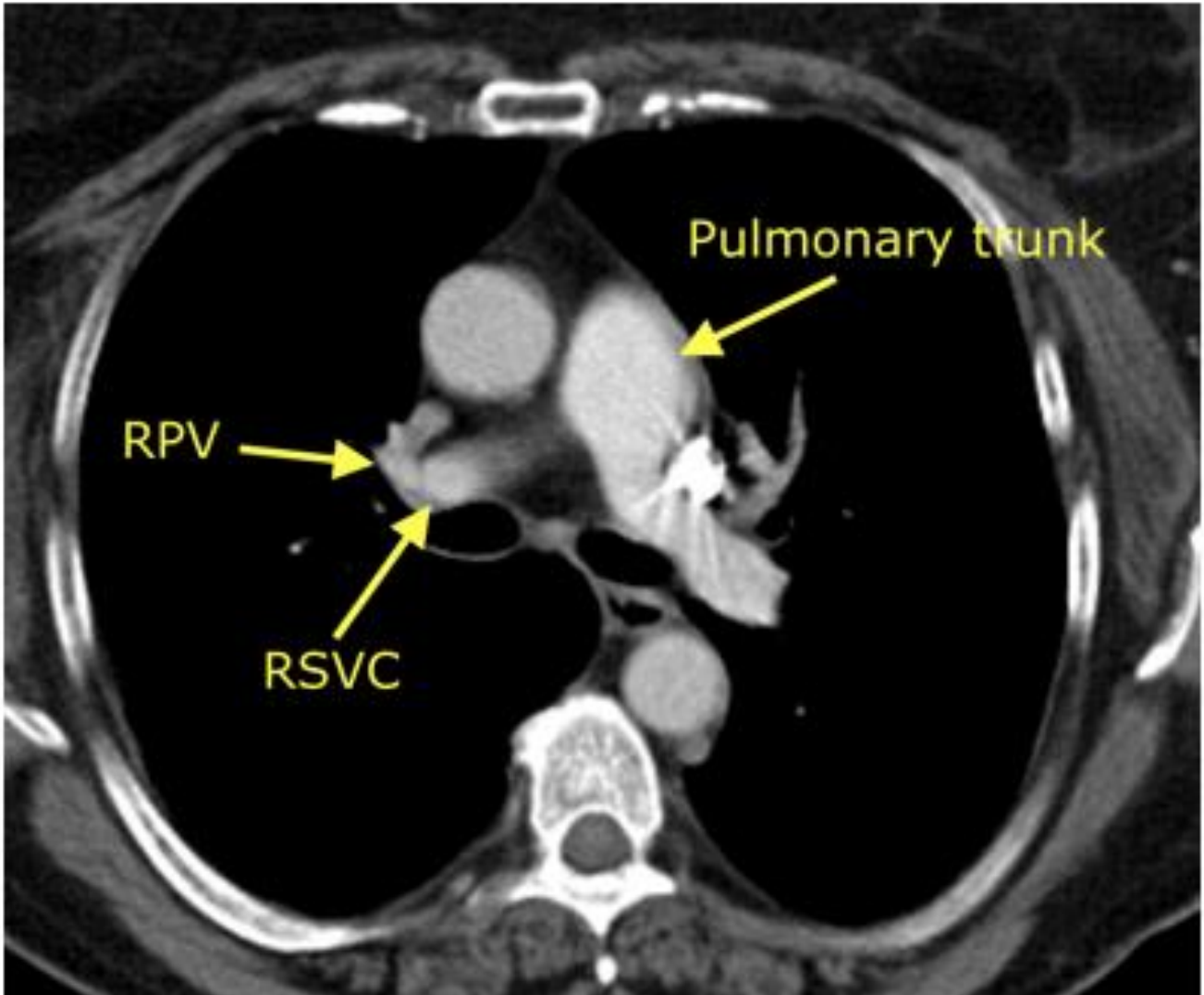


Fig 4A. CT Chest showing RPV draining into RSVC