Persistent left superior vena cava associating with anomalous right superior vena cava drainage, atrial septal defect and atrial fibrillation: a case report

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To the Editor: A 53-year-old man recently presented to our hospital with a history of palpitations, lightheadedness, dyspnea and chest tightness over the past half month. In addition, the patient had been prone to activity-induced fatigue since childhood, and had a history of being susceptible to respiratory tract infection. Following physical examination, there was a detected an irregular pulse and cardiac murmur. Auscultation of the pulmonary valve revealed accentuation of the second heart sound at the left side of the sternum, between the first and second ribs, and grade 3/6 systolic murmur. Basic laboratory investigations were unremarkable. Electrocardiogram (ECG) showed atrial fibrillation (AF). Chest radiograph showed mild cardiomegaly. Transthoracic echocardiogram (TTE) was suggestive of atrial septal defect (ASD) from left to right shunt; dilated left atrium, right atrium and right ventricle; moderate tricuspid regurgitation; mild pulmonary arterial hypertension and a markedly dilated coronary sinus due to persistent left superior vena cava (PLSVC). Cardiac computed tomography (CT) was considered for better delineation of the venous anomaly and to define the location of the shunt. CT was performed on a Philips Brilliance 64 slice multidetector CT (MDCT) scanner, which confirmed the TTE findings of PLSVC [Figure 1A], and defined the cardiac shunt as sinus venosus-type atrial septal defect (SVASD) [Figure 1B]. The opening of right superior vena cava (SVC) drains into the roof of left atrium [Figure 1B]. Left-sided persistent SVC was seen draining into the coronary sinus which then was grossly dilated, measuring at 3.0 cm. A defect measuring 2.0 cm was seen in the inter-atrial septum. Moreover, CT helped to rule out some cardiac anomalies more commonly reported in association with PLSVC such as ventricular septal defect, aortic coarctation, transposition of the great vessels, Tetralogy of Fallot, and anomalous connections of the pulmonary veins. SVASD, if left unrepaired, may eventually lead to right-heart volume overload and subsequent hypertensive pulmonary vascular disease. The PLSVC drains into the right atrium via the coronary sinus, resulting in no hemodynamic consequence. However, when the right SVC drains directly into the roof of left atrium, it may result in right-to-left shunt or hemodynamic overload on the left atrium with the risk of atrial fibrillation or paradoxical embolization. Therefore, the patient was advised to undergo surgery. The cardiac surgery involved opening the right heart chamber and requiring drainage of the PLSVC by a separate venous cannula. The modified Cox-Maze AF surgical procedure is concomitant with the SVASD repair. The surgical patch is used to repair SVASD and isolate the opening of right SVC to the right atrium. We did not ligate the PLSVC.

PLSVC is the most common anomaly of the thoracic venous system and occurs approximately in 0.5% of the general population.[1] It is due to the failed regression of the left anterior cardinal vein that the Marshall ligament generally forms. Most commonly, PLSVC coexists with a right SVC in up to 80%–90% of cases.[2] In the instance of bilateral SVCs, a left innominate vein may be completely absent in up to approximately 65% of such cases.[3] In approximately 80%–92% of PLSVC cases, the PLSVC drains into the right atrium through the coronary sinus, resulting in no hemodynamic consequences.[2,3] Conversely, in approximately 10%–20% of cases of PLSVC, the PLSVC can drain via the left atrium, either through an unroofed coronary sinus or through the left superior pulmonary vein or in a straight line fashion into the roof of the left atrium.[4] In the instance of bilateral SVCs, the right SVC generally drains normally into the right atrium. However, the right SVC drainage into the roof of left atrium in our patient is very rare. PLSVC is usually asymptomatic and is an incidental finding on imaging.

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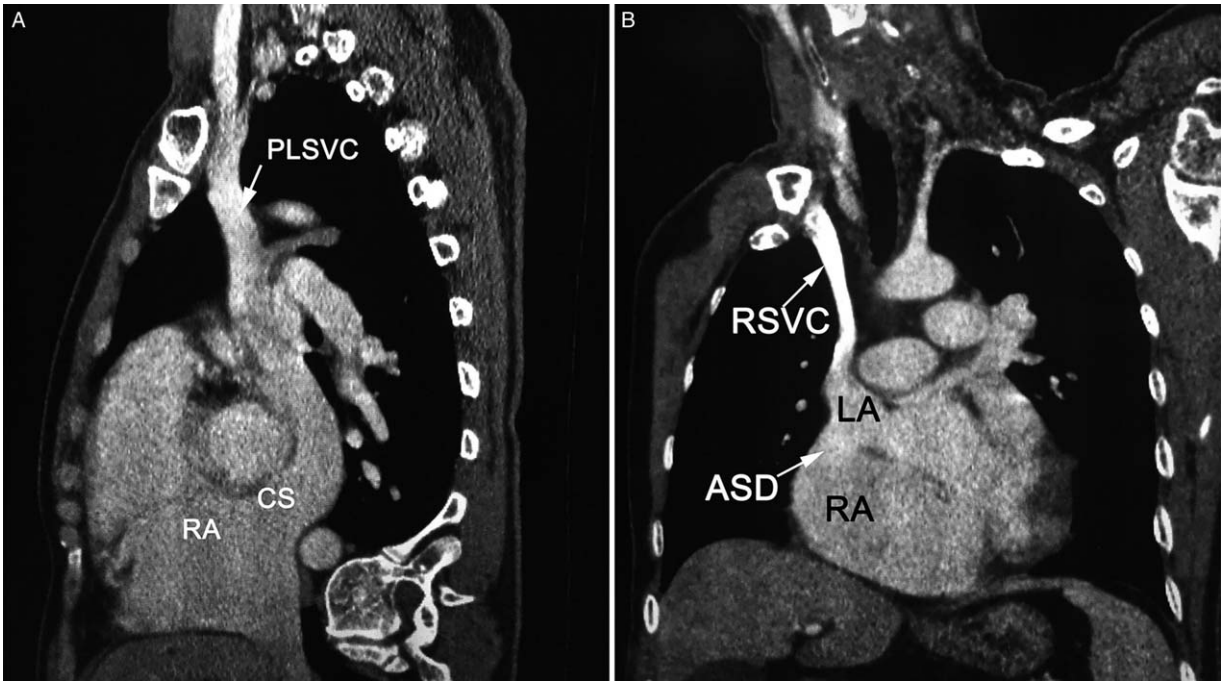

Figure 1: Cardiac computed tomography confirming the findings of persistent left superior vena cava (PLSVC; arrow; A). Cardiac computed tomography confirming the cardiac shunt (CS) as sinus venosus-type atrial septal defect (ASD; arrow) and the opening of right superior vena cava (RSVC) drainage into the roof of left atrium (LA; arrow; B). RA: right atrium.

Conversely, when the right SVC drains into the left atrium, it may result in right-to-left shunt or in hemodynamic overload on the left atrium with the risk of atrial fibrillation or paradoxical embolization. Our patient of PLSVC drained into the right atrium via the coronary sinus, resulting in no hemodynamic consequence.

In approximately 0.3%–0.5% of cases, PLSVC coexist with congenital heart disease (CHD).[5] During the diagnostic procedure for our patient, SVASD was found. ASD develops due to absence or maldevelopment of atrial infolding that normally separates the 2 atria.[6] SVASD accounts for 5%–10% of all atrial septal defects (ASDs). SVASD is located along the superior aspect of the atrial septum, near the entry of the right SVC in our patient. SVASD may be asymptomatic in childhood but may become symptomatic with age. Unrepaired SVASD leads to right heart volume overload and can eventually lead to hypertensive pulmonary vascular disease. Therefore, our patient was treated with surgical repair of SVASD once the diagnosis was made. The association of PLSVC and SVASD is very rare in the reported literature.

PLSVC have practical implications when performing procedures such as permanent pacemaker placement, implantable cardioverter defibrillator placement and right-heart catheterization. Serious complications such as arrhythmia, cardiogenic shock, cardiac tamponade, and coronary sinus thrombosis have been reported when pacemaker leads or catheters have been inserted via PLSVC.[2] It is critical to confirm the presence of PLSVC, to fully characterize the pattern of cardiac venous return, and to find other potential coexisting congenital heart abnormalities prior to initiation of use of central venous access device and the thoracic surgery.[7] TTE, transesophageal echocardiography (TEE), Cardiac CT can be used to assess PLSVC and potential coexisting congenital cardiac malformation.

Atrial arrhythmias (AAs), including AF and/or flutter (AFL), are significantly increased in patients with ASDs. The left-to-right shunt enabled by the presence of an ASD results in cardiac remodeling secondary to long-standing hemodynamic overload. It is this geometrical remodeling that plays a vital role in the pathogenesis of AAs.[8] The presence of AAs should be considered an indication for closure of an ASD. The surgeons will perform an AF surgical procedure, such as the modified Cox-Maze procedure, prior to the surgical repair of ASDs.[9]Although the AAs may not revert to sinus rhythm after the combination of the 2 surgical procedures, there are likely to improve mortality and symptoms.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has give his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initial will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

Author contributions

Li L collected important background information and drafted the manuscript. Ji KQ collected important

background information and carried out the manuscript editing. You CY provided the assistance for images acquisition, images analysis and preformed manuscript review.

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