Isolated persistent left superior vena cava associated with anomalous left hepatic vein drainage into the right atrium accidentally discovered after sternotomy

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A 57-year-old woman with a medical history of arterial hypertension and atrial fibrillation was admitted for mitral valve surgery because of moderate mitral regurgitation registered on transthoracic echocardiography. Due to a borderline indication for surgical treatment, a transesophageal echocardiographic examination was performed preoperatively. Although its focus was on mitral regurgitation, which was confirmed as moderate (regurgitation grade 2/3, the mitral valve annulus 3.8 cm), the bicaval view could not be seen despite multiple attempts. No other abnormalities were registered.

After induction of anesthesia, which proceeded without difficulties, median sternotomy was performed. After pericardiotomy, normocardia, situs solitus and the presence of venous anomalies were noticed by the surgeon. At that time, the presence of the persistent left superior vena cava (PLSVC), agenesis of the right superior vena cava (RSVC) and double inferior vena cava were suspected. The operation was abandoned for additional diagnostics. Contrast-enhanced computed tomography (CECT) of the heart showed the presence of a PLSVC draining into the right atrium (RA) via the dilated coronary sinus (CS), along with the agenesis of the RSVC (Figure 1A, B). CECT of the abdomen showed anomalous left hepatic vein (LHV) drainage into the RA (Figure 1C, D). To the best of our knowledge, a combination of these anomalies has not been described so far. Another curiosity is that it was discovered in such a clinical scenario. Considering the discovered congenital anomalies on the one hand and a moderate grade of mitral regurgitation on the other, the Heart Team and the patient made a joint decision to continue with conservative medical treatment.

The PLSVC is an embryologic remnant of the left superior cardinal vein seen in 0.1% to 0.3% of healthy adults [1]. Most cases of PLSVCs are associated with RSVC, a condition known as duplication of the superior vena cava [1, 2]. The PLSVC with the agenesis of an RSVC is extremely rare, and it is known as an isolated PLSVC [3]. The diagnosis can be easily missed unless specific preoperative testing is carried out, such as a cardiac computed tomography scan. It is often discovered incidentally during an examination of heart disease, central venous line insertion, or pacemaker implantation. The PLSVC usually drains into a dilated CS, and its catheterization can cause hypotension, angina, and perforation of the heart, causing tamponade and cardiac arrest [1]. Therefore, when congenital systemic venous anomalies are suspected, accurate anatomical assessment of the venous system to detect possible coexistent cardiac anomalies is useful before insertion of central venous catheters, pulmonary artery catheters, pacemakers, or defibrillator leads.

Abnormal drainage of the LHV into the RA is very rare. Its occurrence may be explained

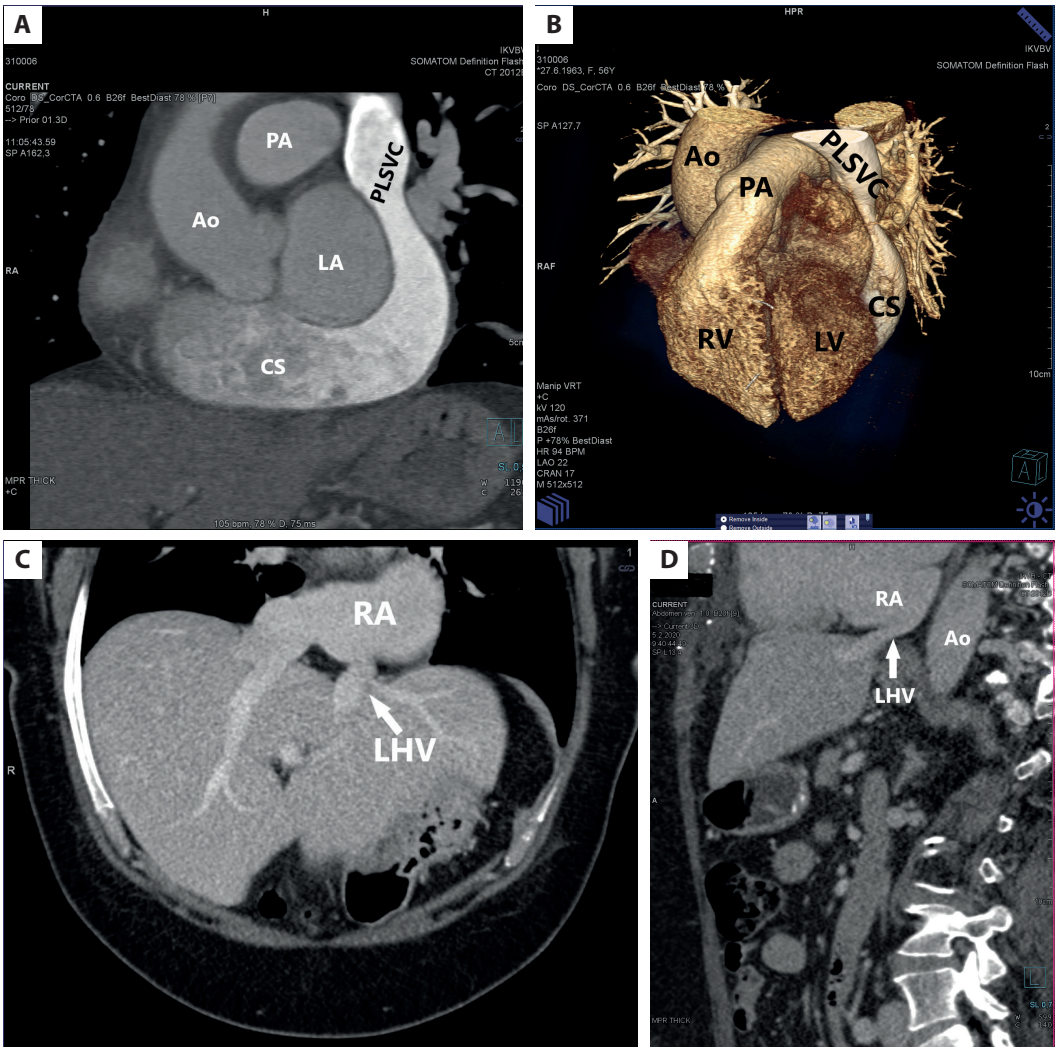

Figure 1. A, B. Contrast-enhanced CT of the heart: the presence of a PLSVC draining into the right atrium via the dilated CS, along with agenesis of the right superior vena cava **C, D.** Contrast-enhanced CT of the abdomen: anomalous LHV drainage into the RA

Abbreviations: Ao, ascending aorta; CS, coronary sinus; CT, computed tomography' LA, left atrium; LHV, left hepatic vein; LV, left ventricle; PA, pulmonary artery; PLSVC, persistent left superior vena cava; RA, right atrium; RV, right ventricle

by the persistence of the left vitelline connection with the left sinus horn and the ductus venosus during the fetal period [4]. Such abnormal hepatic vein drainage usually has no clinical significance, but this anomaly may present potentially fatal challenges to the donor operation if not determined preoperatively, especially when the left lobe is the choice for explantation [4].

By retrospectively analyzing our patient's echocardiographic examination results and knowing her anatomical abnormalities, we could conclude that the inability to see the bicaval view on transesophageal echocardiography should have raised suspicion of potential venous anomalies.

Article information

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