Case Study

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Additional superior vena cava combined with abnormal inflow of the hepatic vein

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Abstract

We represent a case of successful surgical treatment of a rare congenital heart disease: abnormal inflow of an additional superior vena cava into the left atrium, combined with atrial septal defect, mitral and tricuspid valve regurgitation, and abnormal inflow of the left hepatic vein into a roofless coronary sinus.

Keywords

Coronary sinus, Heart atria, Hepatic veins, Vascular malformations, Vena cava superior

Introduction

Additional left superior vena cava (LSVC) occurs in 0.3%–0.4% of the population and accounts for 2%–5% of all congenital heart defects.[1,2] Isolated LSVC inflow into the left atrium (LA) is a rare abnormality that is often combined with a posteroinferior atrial septal defect (ASD) in the area of the coronary sinus,[3] an incomplete form of open atrioventricular canal, and absence of the inferior vena cava. We describe surgical correction of an additional LSVC draining into the LA, combined with a posteroinferior ASD and mitral and tricuspid valve regurgitation. We unexpectedly found abnormal inflow of the left hepatic vein into a roofless coronary sinus.

Case report

A 19-year-old female was hospitalized with complaints of shortness of breath on moderate exertion, cardiac pain, and fatigue. A secundum ASD had been diagnosed by echocardiography 2 years earlier. A systolic murmur was detected along the left sternal border. An electrocardiogram showed sinus rhythm, incomplete right bundle branch block, and hypertrophy of the right heart and left ventricle. Echocardiography demonstrated a 25-mm posteroinferior secundum ASD with an excessive left-right shunt, absence of the roof and expansion of the coronary sinus, an additional LSVC flowing into the LA, grade 2 mitral regurgitation with annulus dilatation up to 38 mm, grade 3 tricuspid regurgitation with annulus dilatation up to 38 mm, and significant right heart enlargement with right ventricular systolic pressure of 52 mm Hg. The left ventricular ejection fraction was 66% and right ventricular ejection fraction was 55%. Multislice computed tomography with contrast revealed the additional LSVC flowing into the LA near the base of the LA appendage, absence of the brachiocephalic vein, a posteroinferior ASD, and abnormal left hepatic vein inflow into the coronary sinus (Figure 1). After balloon obturation of the LSVC to identify collaterals between the left and right superior venae cavae, central venous pressure increased from 10 to 28 mm Hg. Retrograde catheterization of the coronary sinus through the LSVC and ASD showed abnormal drainage of the left hepatic vein into the coronary sinus, inflow of the collector of the left hepatic vein into the inferior vena cava, and shunting between the 2 hepatic veins (Figure 2). Under cardiopulmonary bypass, hypothermia, and cardioplegia, the heart was accessed via a median

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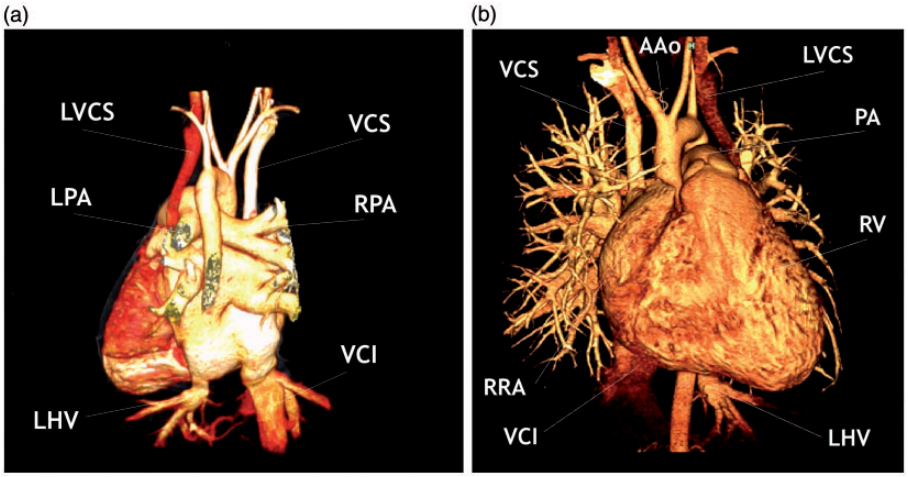

Figure 1. Multislice computed tomography with contrast: (a) rear view and (b) front view of the heart, showing the additional left superior vena cava (LSVC) flowing into the left atrium. AAo: ascending aorta; IVC: inferior vena cava; LHV: left hepatic vein; LPA: left pulmonary artery; PA: pulmonary artery; RAA: right atrial appendage; RPA: right pulmonary artery; RV: right ventricle; SVC: superior vena cava.

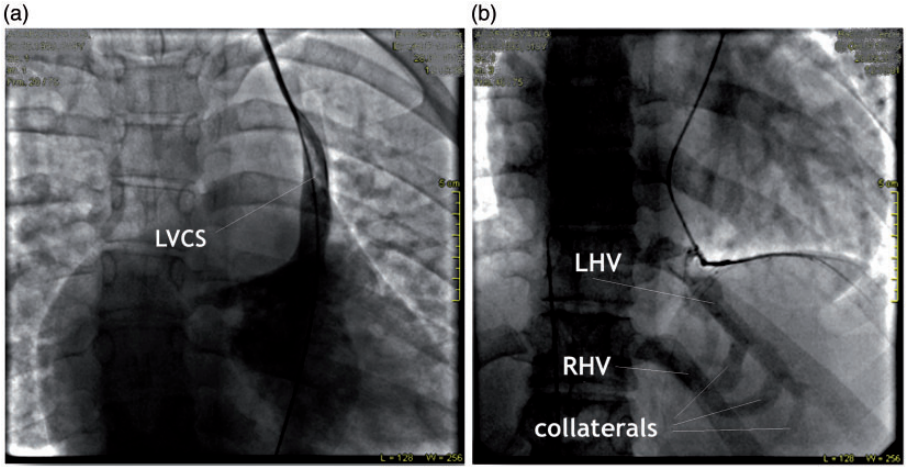

Figure 2. (a) Angiogram of the left atrium with contrast from a catheter inserted into the additional left superior vena cava (LSVC). (b) The left hepatic vein flowing into the coronary sinus is seen via a catheter inserted in the coronary sinus. The collaterals to the right hepatic vein are visualized.

sternotomy. The heart was significantly enlarged, particularly on the right side. Access to the LSVC revealed a diameter of 14 mm; the right-sided superior vena cava diameter was 10 mm. Separate cannulation of the aorta and the 3 venae cavae was performed with angled cannulas. The inferior vena cava and left hepatic vein were separately taped (Figure 3a). The right atrium (RA) was opened, revealing absence of the roof and significant expansion of the coronary sinus, and a typical central ASD above and behind the sinus (Figure 3b). The incision was extended to the dome of the LA. The atrial septum edges were fixed, and the LSVC was seen entering the LA close to the LA appendage above the left pulmonary veins of the superior lobe. A left-sided jugular venous catheter was inserted up to the LA edge as a benchmark. The mitral valve leaflets were thin and mobile and the annulus was dilated; after suture anuloplasty, a water test confirmed no regurgitation (Figure 3c). We tunneled a xenopericardial patch via the LSVC into the RA, with simultaneous plasty of the central ASD (Figure 3d). The tricuspid annulus was significantly dilated with incomplete leaflet coaptation; De Vega annuloplasty was performed. The cardiopulmonary bypass time was 220 min and aortic crossclamp time was 117 min at 22°C. Right ventricular pressure was 33 mm Hg with systemic pressure of 115 mm Hg. On terminating cardiopulmonary bypass, central venous pressure in the right and left superior venae cavae was 11 and 13 mm Hg, respectively. The patient was transferred to the intensive care unit with

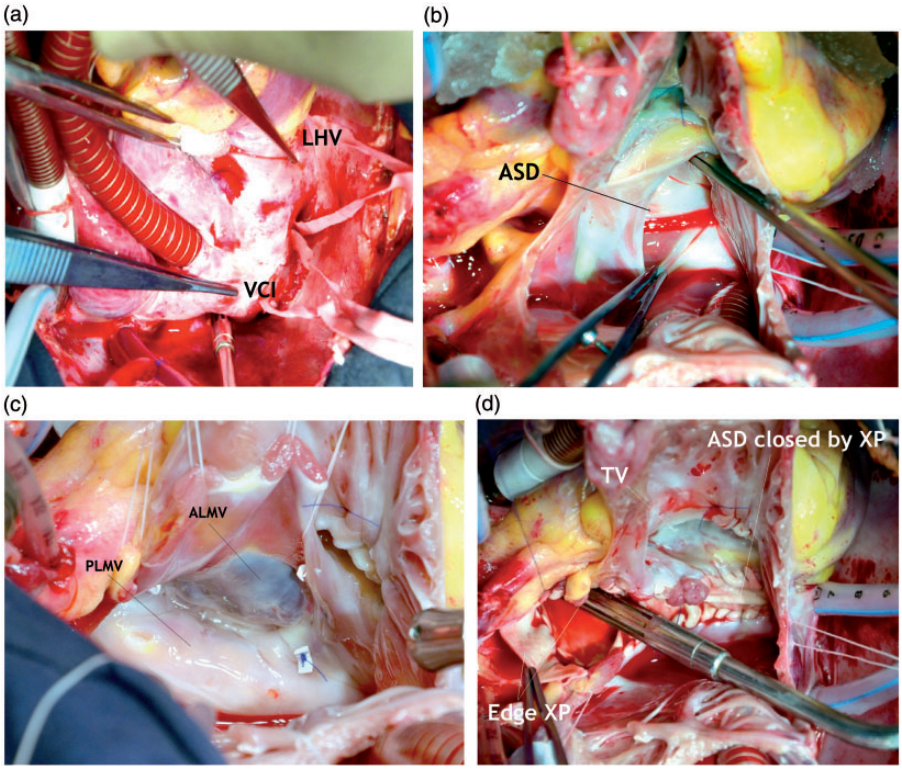

Figure 3. Operative photographs. (a) A view of the heart showing the separately cannulated venae cavae and aorta. The forceps indicate the inferior vena cava (IVC) and left hepatic vein (LHV), which are separately secured with tape. (b) The opened right atrium revealing the posteroinferior atrial septal defect (ASD); the edges of the defect are separated with forceps. The tapes on the left hepatic vein and inferior vena cava are tightened. (c) Suture annuloplasty of the mitral valve using 3 Gore-Tex pads. A water test confirmed tight closure of the anterior (ALMV) and posterior (PLMV) mitral valve leaflets. (d) A coronary suction line was inserted into the intraatrial tunnel created by the xenopericardial patch (XP). One edge of the patch is held by forceps before attachment to the opposite edge. TV: tricuspid valve.

inotropic support, and extubated on postoperative day 2. Heparin 2500 U was given 4-hourly (changed to warfarin for one year after discharge). Postoperatively, venous pressure in the right and left superior venae cavae was 7 and 9 mm Hg respectively. The tunnel from the LSVC inside the LA, which was transferred to the RA, had an orifice diameter of 11 mm (Figure 4a–4c). The left ventricular ejection fraction was 70%. The patient was discharged on postoperative day 8 in good condition with sinus rhythm and no shunt flow on echocardiography.

Discussion

LSVC results from disrupted obliteration of the left anterior cardiac vein that feeds via a large cardiac vein and the coronary sinus into the RA during embryological development. Usually, it is additional to normal development of the right superior vena cava. In 82%–92% of cases, the additional LSVC drains through the coronary sinus into the RA,[4] but in 18%–20%, it drains into the LA, usually via a completely or partially roofless coronary sinus. Because there are no specific clinical signs, diagnosis of additional LSVC is complex, and it is often discovered incidentally on insertion of a central venous catheter or endocardial electrodes for pacing,[5] or during thoracic surgical interventions. Transthoracic echocardiography does not provide anatomical diagnosis of LSVC. We used multislice computed tomography with intravenous contrast injection, which precisely defined the anatomical characteristics of the LSVC, its drainage into the LA, and collaterals between the venae cavae, which are important in deciding the method of surgical correction.

Abnormal drainage of the left hepatic vein into a dilated coronary sinus is very rare and occurs as a result of disruption of bile duct obliteration with the left anterior horn of the coronary sinus. There are only 2 reported cases of left hepatic vein draining into the RA with an additional LSVC draining into the coronary sinus.[6,7] Such abnormal hepatic vein drainage usually has no clinical significance, but lack of a preoperative diagnosis may lead to technical difficulties during the operation. The presence of collaterals between the right and left hepatic veins provided the

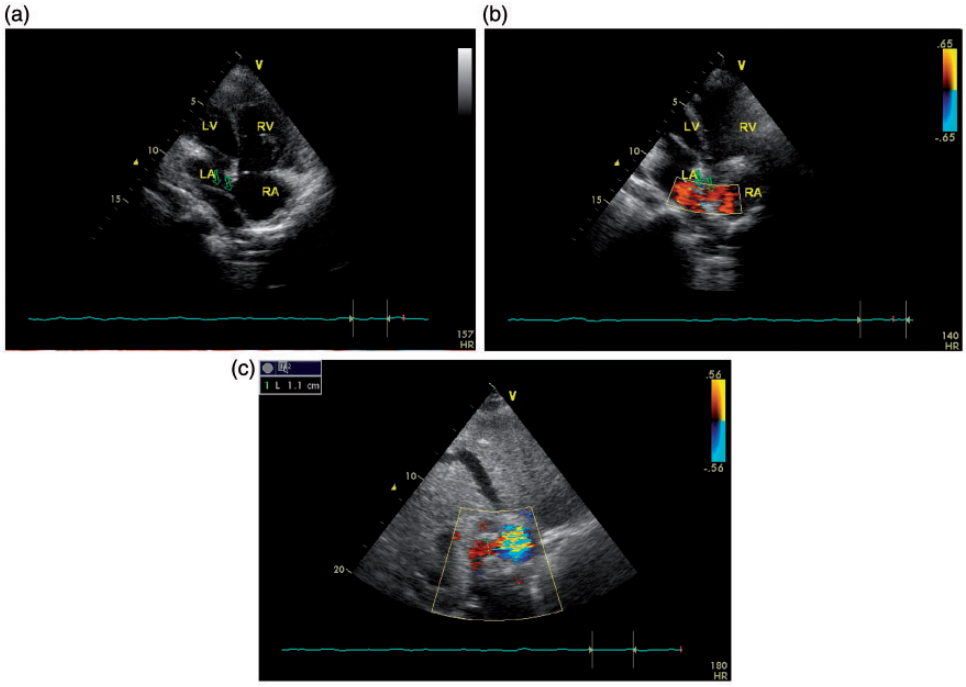

Figure 4. (a, b) Parasternal long-axis view of the left ventricle, showing the tunnel inside the left atrium (LA) from the additional left-sided superior vena cava to the right atrium (RA). (c) Subcostal view; the diameter of the tunnel mouth at the inflow to the right atrium was 1.1 cm.

opportunity to clamp the left hepatic vein without using an additional venous cannula.

The preferred method of surgical treatment is extra-cardiac translocation of the LSVC root into the RA appendage or the right-sided superior vena cava (pre- or postaortic). In our case, none of the various methods of reimplantation could be used due to the short length of the LSVC and the anatomical distance to the RA. Thus we employed intraatrial tunneling of the LSVC mouth through the LA roof after dissection of the atrial septum, using a xenopericardial patch, with a good clinical result. In this case, the patient's age precludes the phenomenon of overgrowth of the tunnel in the future.

Declaration of conflicting interests

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