CASE REPORTS

Absent Right Superior Vena Cava with Persistent Left Superior Vena Cava Which Drains to Unroofed Coronary Sinus in a Child with Atrioventricular Septal Defect and Cor Triatriatum Sinister: Preop Correct Diagnosis and Successful Surgery in a Single Session

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ABSTRACT

We report a unique case of a 4-year-old boy with intermediate-type atrioventricular septal defect, cor triatriatum sinister, persistent left superior vena cava, unroofed coronary sinus, and absent right superior vena cava. Persistent left vena cava draining into the unroofed coronary sinus was demonstrated easily using the agitated saline-contrast echocardiography. After conformation with angiographic evaluation, surgery was performed at a single session. Roofing of the coronary sinus with polytetrafluoroethylene patch, mitral cleft repair, tricuspid annuloplasty, atrioventricular defect repair with pericardial patch, and resection of the membrane in the left atrium was succeeded without complication.

Key Words. Atrioventricular Septal Defect; Cor Triatriatum Sinister; Persistent Left Superior Vena Cava; Unroofed Coronary Sinus; Absent Right Superior Vena Cava

Introduction

Persistent left superior vena cava (PLSVC) is the most common variation in the systemic venous system. Its incidence increases in patients with congenital heart disease, which is estimated about 2–5%.[1,2] Absent right superior vena cava with PLSVC is extremely rare and commonly associated with cardiac situs anomalies.[3] Raghib et al.[4] first described the combination of a persistent left superior vena cava and unroofed coronary sinus (CS) in 1965. Right atrial isomerism and atrioventricular septal defect (AVSD) usually coexist with this complex, particularly if there is a large interatrial defect.[1,3] Cor triatriatum sinister is a rare congenital cardiac anomaly in which the left atrium is subdivided into two portions by a membrane. Association of cor triatriatum with AVSD is rare; hemodynamics of AVSD with cor triatriatum may lead to a diagnostic challenge in which misdiagnosis, such as inoperable pulmonary hypertension and inadequate surgical intervention, is possible.[5]

To our knowledge, coexistence of absent right superior vena cava, persistent left superior vena cava draining into the unroofed CS, AVSD, and cor triatriatum sinister has never been described.

Case

A four-year-old male patient referred for the evaluation of a murmur. Physical examination revealed an oxygen saturation of 94% and a 3/6 holosystolic murmur in left sternal border. There was left superior axis on electrocardiography (Car-

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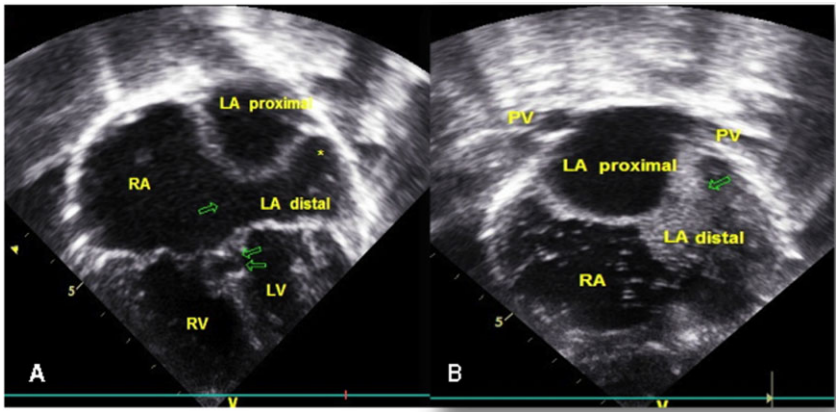

Figure 1. (A) Transthoracic echocardiograpy showing fibromuscular membrane that divides the left atrium into proximal and distal chambers, primum atrial septal defect (arrow), inlet ventriculoseptal defect (double arrow), and unroofed coronary sinus (*). (B) Contrast-agitated saline injection into the left antecubital vein opacifies the LA before the RA on echocardiographic image in subcostal view. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; PV, pulmonary vein.

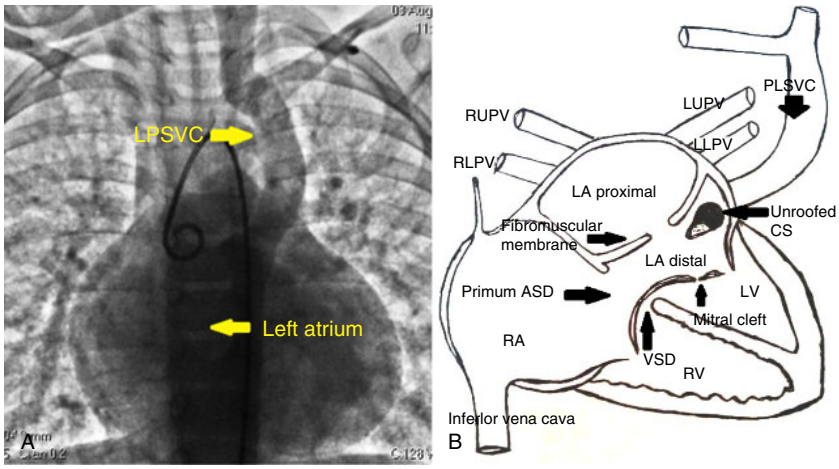

Figure 2. (A) Angiography showing upper extremity venous return to the LPSVC and absence of right SVC. (B) Schematic drawing of the preoperative findings that were confirmed in the operation. ASD, atrial septal defect; CS, coronary sinus; LA, left atrium; LLPV, left lower pulmonary vein; PLSVC, persistent left superior vena cava; LUPV, left upper pulmonary vein; LV, left ventricle; RA, right atrium; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; RV, right ventricle; VSD, ventricular septal defect.

diofax, Nihon Kohden, Tokyo, Japan). Pulmonary congestion with marked pulmonary conus was present on chest x-ray. On echocardiography (Vivid S6, General Electric Healthcare, Milwaukee, WI, USA), intermediate-type AVSD, cor triatriatum sinister, mitral cleft, and moderate tricuspid valve regurgitation were noted (Figure 1A). Saline-contrast echocardiography was arranged as superior vena cava was not seen on echocardiography. Agitated saline injected into the left antecubital vein first appeared in the left atrium, then in the right atrium (Figure 1B). Absent right superior vena cava, persistent left superior vena cava draining into the unroofed CS, AVSD, and cor triatriatum sinister were confirmed by catheter angiography (Innova 2100-IQ, General Electric Healthcare) (Figure 2). During heart catheterization, mean pulmonary artery pressure was 20 mm Hg, pulmonary capillary wedge pressure was 13 mm Hg, Qp/Qs > 2, and the ratio between pulmonary vascular resistance and systemic vascular resistance (PVR/SVR) was 0.018. Complete corrective surgery was performed. LSVC was used for cannulation due to atretic right superior vena cava (Figure 3). Exploration revealed a cor triatriatum sinister, and the aberrant membrane was resected. Unroofed CS was noted in the left atrium. Orifice of the PLSVC

was located above the left atrial appendix. Roofing was performed using a 0.6-mm polytetrafluoroethylene (PTFE) (GORE-TEX Soft Tissue Patch, Gore Medical, Flagstaff, AZ, USA) patch. Mitral cleft was repaired with single-prolene sutures. Subsequently, atrioventricular defect repair was achieved with autologous pericardium. Annuloplasty was performed to the tricuspid valve. No postoperative complication occurred. Postoperative echocardiography revealed mild mitral valve regurgitation and tricuspid valve regurgitation (Figure 4). Patient was discharged on the eight postoperative day. He is clinically well and has sinus rhythm, and we observe only trivial mitral-tricuspid regurgitation after 6 months postoperative follow-up on echocardiography.

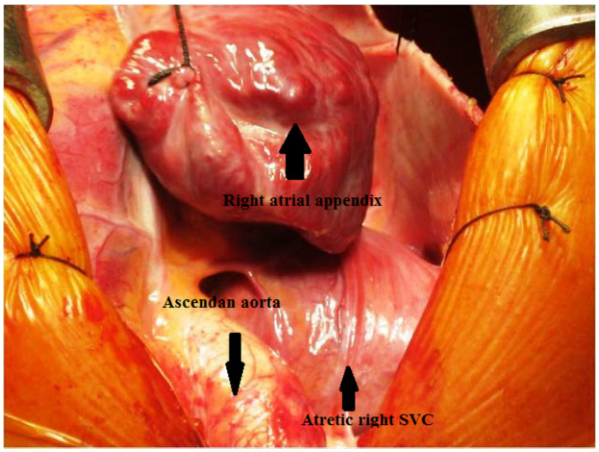

Figure 3. Operational view of the atretic right SVC.

Discussion

This case is unique, which possesses characteristics of four congenital cardiac anomalies that include absent right superior vena cava with PLSVC, unroofed CS, AVSD, and cor triatriatum sinister. Up to now, this complex of anomalies has never been documented. Fortunately, diagnosis of all these anomalies was made before the corrective surgery in this case. Unroofed CS is a rare congenital cardiac anomaly in which shunt occurs between the left atrium and the CS due to partial or complete defect in the CS roof.[1,2] It is often associated with PLSVC and other forms of complex congenital heart disease, usually heterotaxia syndromes.[6] The diagnosis of unroofed CS can be suspected if the patient has cardiac murmur, right-sided heart enlargement, transient cyanosis or hypoxia, or paradoxical embolism.[6] In this case, we used agitated saline echocardiogram as a part of diagnosis. Contrast injected into the arm veins opacifies the CS if the patient has isolated PLSVC. Contrast injection into either arm opacifies left atrium in the combination of unroofed CS and PLSVC.[1,6] Cardiac catheterization is usually performed for diagnostic confirmation and to identify associated cardiac anomalies.[5] The basic repair consists of roofing the CS (using pericardial patch) from within the left atrium.[6,7]

Embryologically, the unroofed CS with persistent LSVC is supposed to derive from the failure of complete formation of the left atriovenous fold, which normally forms the inferior portion of the

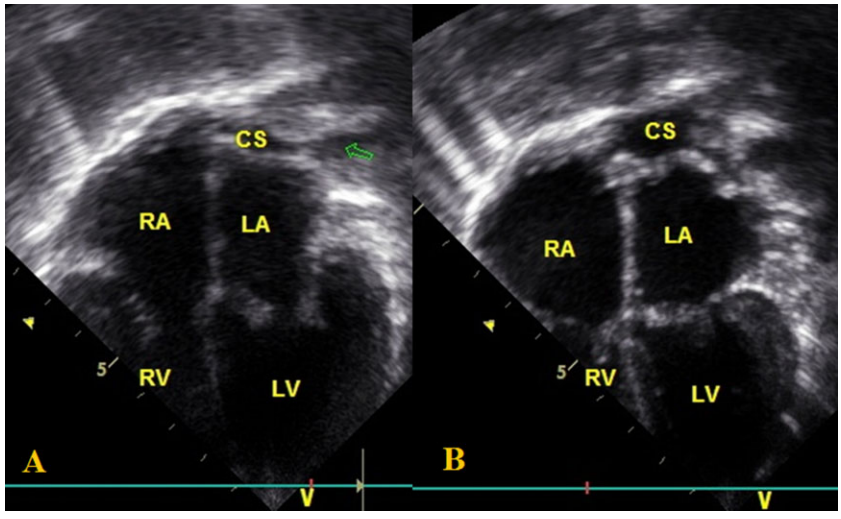

Figure 4. Apical four-chamber views after surgery. Coronary sinus (CS) is created above the roof of the left atrium (LA) with adequate flow to the right atrium (RA). RV, right ventricle; LV, left ventricle; arrow, persistent left superior vena cava.

CS.[4] Left atriovenous fold also supports interatrial primum septum. One of the embryological factors responsible for the development of cor triatriatum, which was proposed by Gharagozloo et al.,[8] is the induction of an aberrant membrane by impaction of a PLSVC.

In conclusion, as in this case, the unique combination of cardiac anomalies including AVSD, cor triatriatum sinister, absent right superior vena cava, and PLSVC draining into unroofed CS is never presented before. Several pathophysiological mechanisms including left-to-right shunting, right-to-left shunting, and pulmonary venous hypertension may influence the clinical presentation in this patient, owing to this extraordinary combination of cardiac anomalies. Along with transthoracic echocardiography, saline-agitated contrast echocardiography is very useful in the patient who has unexplained cyanosis and dilated CS.

Author Contributions

Önder Doksöz: Design, data analysis/interpretation, drafting article, approval of article.

Barış Güven: Data analysis/interpretation, critical revision of article, approval of article.

Yılmaz Yozgat: Design, acquisition, critical revision of article.

Rahmi Özdemir: Design, acquisition, critical revision of article.

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