An absent right superior vena cava with a persistent left superior vena cava combined with the presence of a ventricular septal defect in a pediatric patient: Implications for surgical correction

A persistent left superior vena cava in conjunction with an absent right superior vena cava (ARSVC) is a very rare anomaly of venous circulation which occurs in only 0.09-0.13% of patients with congenital heart defects.[1] This anomaly is usually seen together with situs inversus. However, it seldom occurs in situs solitus. In the literature, there have been many reported adult cases with this anomaly, but almost none involving pediatric patients. In this report we present the case of an ARSVC with a PLSVC which opened into a dilated coronary sinus in a four-year-old girl with situs solitus and a perimembranous outlet ventricular septal defect (VSD). The implications of this rare anomaly are also discussed.

CASE REPORT

A four-year-old female child weighing 13 kg with a height of 90 cm was admitted to our clinic after an angiographic examination performed by the pediatric cardiology department revealed heart failure. She had been followed since she was three months old due to a diagnosis of VSD and pulmonary hypertension. Her physical examination after transfer to our clinic disclosed a pansystolic murmur in the left third intercostal space. She was acyanotic, and her biochemical results were within the normal range. Transthoracic echocardiography revealed visceroatrial situs solitus along with a large outlet VSD and PLSVC. Her angiography showed an outlet VSD with a Qp/Qs of 3, and the pulmonary vascular resistance was measured.

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as 1 unit. In addition, the systemic vascular resistance was 8 units, and the pulmonary artery pressure was 64 mmHg maximum, 4 mmHg minimum (average 38 mmHg). The left ventricle was wide, and the superior vena cava (SVC) was absent with a PLSVC draining into the coronary sinus. After a joint decision with the pediatric cardiology department, an operation for VSD closure was planned.

The patient was premedicated with 0.15 mg/kg midazolam (Dormicum®, Roche, İstanbul, Turkey) intravenously after obtaining her parents’ written consent for anesthesiological intervention and surgery. She was then taken to the operating room. After routine monitorization (EKG, noninvasive blood pressure, partial oxygen pressure), the induction of anesthesia was performed. With the administration of lidocaine (Jetmonal®, Adeka Inc., Samsun, Turkey) 1 mg/kg and thiopenthal sodium (Pental®, IE Ulagay Inc., İstanbul, Turkey) 5 mg/kg. Subsequently, it was decided that mask ventilation was proceeding comfortably, and 0.1 mg/kg vecuronium (Norcuron®, Organon Teknika Inc., İstanbul, Turkey) was used to bring about muscle relaxation. A laryngoscopic examination revealed a grade 1 larynx according to the Cormack-Lehane classification system. The patient was intubated with a No. 4.5 cuffless endotracheal tube and experienced no complications. Invasive blood pressure was monitored via a radial artery catheter and central venous pressure was observed via the right internal jugular vein. Anesthesia was continued using sevoflurane (Sevorane®, Abdi İbrahim Inc., İstanbul, Turkey) and a 50/50 oxygen and air inhalation mixture, 5 mcg/kg/hour IV fentanyl (Fentanil®, Abbott Lab., İstanbul, Turkey) infusion, and 0.5 mg of vecuronium administered every 30 minutes.

Following a midline sternotomy, the patient was heparinized (3 mg/kg). Since the activated clotting time (ACT) was measured at 500 seconds, cardiopulmonary bypass (CPB) was commenced. Antegrade blood cardioplegia (15 ml/kg) was infused after cooling down to 30 °C. Upon opening the pericardium, the ARSVC was obvious (Figure 1). Before cannulation, the PLSVC was found (Figure 2). After standard aortic cannulation, the PLSVC was selectively cannulated extrapericardially, as was emphasized in the article by Kale et al.[2] The right atrial route brought us to the VSD, a perimembranous outlet-type defect of 4x5 cm in size which formed a tricuspid septal leaflet pouch. This defect was repaired with Teflon-buttressed polypropylene sutures using a Dacron patch. Weaning from CPB was uneventful. The cross-clamp time was 32 minutes, and the CPB time was 54 minutes. The heparin was neutralized using 3 mg/kg of protamine. The ACT was then measured at 120 seconds. After establishment of hemostasis and closure of the surgical wound, the patient was transferred to the intensive care unit (ICU) in an intubated condition. She was extubated after 12 hours in a very stable hemodynamic condition. The total 24-hour chest tube drainage was 130 ml. On the morning of her first postoperative day, the patient was transferred from the ICU to a normal ward. She was then discharged in stable condition on postoperative day four.

DISCUSSION

A PLSVC draining into the coronary sinus in situs solitus is the most common systemic venous anomaly.[3] Its estimated prevalence is almost 0.35% in normal subjects and 4.5% in patients with congenital heart diseases.[4] In a report by Biffi et al.,[5] a PLSVC was observed in 0.47% of a population of adults undergoing pacemaker implantation.
implantation. In an autopsy series, it was observed in 0.35% of the general population. An ARSVC is a rare anomaly in situs solitus with an incidence of about 0.07-0.13% among congenital cardiac anomalies. However, it is usually seen in situs inversus, and 10% of all people with congenital heart defects have an ARSVC. An even rarer anomaly is an ARSVC with a PLSVC with an average prevalence of about 0.1% in patients with congenital heart defects. It usually occurs with situs inversus. The total number of reported cases is less than 100 worldwide, and most of these are adults.

The significance of a PLSVC lies in the fact that its presence is associated with congenital heart defects and arrhythmia. Although it is usually asymptomatic, it may cause periprocedural difficulties and should be suspected when central venous access through the internal jugular vein is problematic. Our four-year-old female patient presented with the unique case a PLSVC together with an ARSVC.

This anomaly results from the persistent, defective regression of the fetal left anterior cardinal vein which causes the PLSVC to drain into the right atrium by way of the coronary sinus. The presence of a PLSVC with an ARSVC always indicates the need for further clinical investigation because of defects in the atrial, septal, and endocardial cushion. The tetralogy of Fallot presents during transvenous catheter placement, central venous line insertion, CPB, extracorporeal membrane oxygenation, total or partial cavopulmonary connection repair, and orthotopic heart transplantation procedures. Therefore, cardiologists, cardiac surgeons, and anesthesiologists should be well informed about the possible difficulties which may be imposed by the combination of PLSVC with ARSVC.

During pacemaker implantation, the presence of a PLSVC should be kept in mind when a guidewire takes a left turn and proceeds downward. An ARSVC should indicate an epicardial implantation.

In surgery, it would be judicious to cannulate the PLSVC extrapericardially, as in our case.

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**REFERENCES**