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Case report / Olgu Sunumu

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

Pediatrik bir hastada ventriküler septal defekt varlığında persistan sol superior vena kava eşliğinde sağ superior vena kava yokluğu: Düzeltme cerrahisi için öneriler

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A persistent left superior vena cava in conjunction with a absent right superior vena cava is a very rare anomaly of venous circulation. In this article, we present a four-year-old girl with a persistent left superior vena cava opening into a dilated coronary sinus and an absent right superior vena cava accompanied by a perimembraneous outlet ventricular septal defect which were corrected using cardiopulmonary bypass surgery.

Key words: Absent right superior vena cava; persistent left superior vena cava; ventricular septal defect.

Kalıcı sol superior vena kava, sağ superior vena kavanın yokluğu ile birlikte, venöz dolaşımın çok nadir rastlanan bir anomalisidir. Bu yazıda genişlemiş bir koroner sinüse açılan persistan sol superior vena kava ve kardiyopulmoner baypas cerrahisi ile düzelttiğimiz perimembranöz bir çıkım yolu ventriküler septal defekti eşliğinde görülen sağ superior vena kava yokluğunun olduğu dört yaşındaki bir kız olgu sunuldu.

Anahtar sözcükler: Sağ superior vena kava yokluğu; persistan sol superior vena kava; ventriküler septal defekt.

A persistent left superior vena cava (PLSVC) 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®, İE 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 perimembraneous 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 CBP 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

Figure 1. The absence of right superior vena cava is seen.

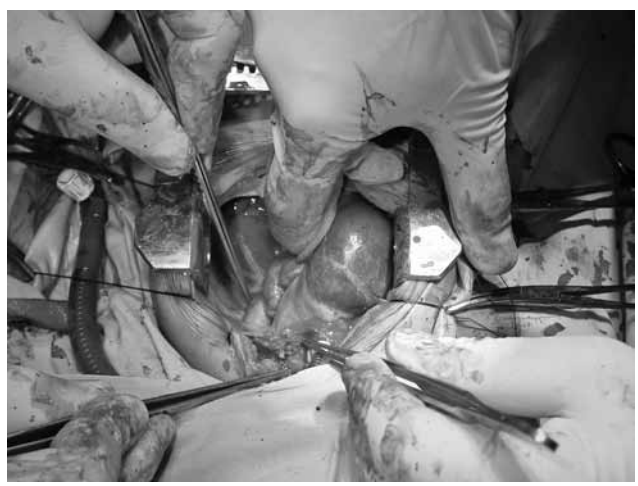

Figure 2. The persistent left superior vena cava is seen.

implantation. In an autopsy series, it was observed in 0.35% of the general population.[5] An ARSVC is a rare anomaly in situs solitus with an incidence of about 0.07-0.13% among congenital cardiac anomalies.[6] However, it is usually seen in situs inversus, and 10% of all people with congenital heart defects have an ARSVC.[5] An even rarer anomaly is an ARSVC with a PLSVC with an average prevalence of about 0.1% in patients with congenital heart defects.[1,7,8] It usually occurs with situs inversus.[9] The total number of reported cases is less than 100 worldwide, and most of these are adults.[9]

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.[7] 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 with an isolated PLSVC in 46% of cases and more anomalies, such as VSD, as seen in our patient, aortic coarctation, and transposition of the great arteries, may exist when it is paired with an ARSVC.[7]

In a review by Bartram et al.,[6] more than half of the patients with an ARSVC and situs solitus had no other congenital heart disease. However, since this anomaly is asymptomatic when it exists without accompanying congenital anomalies, its incidence may be even higher in patients without any evidence of cardiac disease. Insertion of central venous catheters and pacemaker implantation may increase thrombogenesis, cannula size limitation, electrode displacement and pacemaker syndrome.[5,7,9]

Even though a PLSVC draining into the right atrium via the coronary sinus without additional cardiac anomalies has no hemodynamic consequence, it may be a hindrance when a patient with these conditions undergoes cardiac surgery with CPB. Central vein insertion may be problematic causing unforeseen arrhythmia as the catheter goes into the right atrium via the coronary sinus. It may also cause a pacemaker syndrome-type ailment in patients with pacemakers when the guidewire enters the coronary sinus via the PLSVC.[4]

This anomalous complex also causes difficulties during transvenous catheter placement, central venous line insertion, CPB, extracorporeal membrane oxygenation, total or partial cavopulmonary connection repair, and orthotopic heart transplantation procedures.[2,5,8] 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.[8] 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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