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Incidentally Detected Pediatric Case with Absent Right Superior Vena Cava during Transcatheter VSD Closure

Transkateter VSD Kapama Sırasında Tesadüfen Saptanan Sağ Süperiyor Vena Kavası Olmayan Pediatrik Olgu

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ABSTRACT

Persistent left superior vena cava (PLSVC) with an absent right superior vena cava (RSVC) is an extremely rare congenital anomalie in normal atrial situs. Isolated cases are diagnosed incidentally during catheterization. In patients with PLSVC, due to frequent drainage of upper extremity, head and neck veins into coronary sinus, patients with enlarged coronary sinus should be suspected to have absence of the RVCS. In this report, we presented a 7-year-old girl who was diagnosed perimembranous VSD (ventricular septal defect), apparently enlarged coronary sinus, PLSVC with transthoracic echocardiography, and incidentally was recognized absence of RSVC during percutaneouse VSD closure. Consequently, we wanted to draw attention to echocardiographic and angiocardiographic findings of this rare venous anomaly.

Key Words: Absent right superior vena cava; Amplatzer device; Enlarged coronary sinus; persistent left vena cava; Transcatheter closure

ÖZET

Persistan sol vena kava süperiyor ile birlikte sağ vena kava süperiyorun olmaması, normai atriyal situs varlığında son derece nadir bir doğumsal anomalidir. İzole vakalar, kateterizasyon sırasında tesadüfen saptanır. Baş, boyun ve üst extremitenin venöz drenajı sol persistan vena kava aracılığıyla sıklıkla koroner sinüse olduğundan dolayı, genişlemiş koroner sinüsü olan hastalarda vena kava superiyor yokluğundan şüphelenilmelidir. Bu yazıda, transtorasik ekokardiyografiyle VSD, belirgin geniş koroner sinus ve sol persistan vena kava süperiyor tanısı konan ve transkateter kapama sırasında tesadüfen sağ vena kava superiyorunun olmadığı saptanan 7 yaşında kız hasta sunuldu. Böylece, bu nadir venöz anomalinin ekokardiyografik ve anjiyokardiyografik bulgularına dikkat çekilmek istendi.

Anahtar Kelimeler: vena kava süperiyor yokluğu, Amplatzer cihaz, genişlemiş koroner sinüs, sol persistan vena kava superiyor, transkateter kapama

INTRODUCTION

Persistent left superior vena cava (PLSVC) with an absent right superior vena cava (RSVC) is a rare anatomic variation among systemic venous return anomalies (0.07–0.13%) and more rare in atrial situs solitus. Isolated cases are typically asymptomatic and they are discovered incidentally during the open-heart surgery, central venous catheter insertion and catheterization such as pacemaker implantation. A PLSVC usually drains all upper extremity venous drainage into the right atrium through the dilated coronary sinus (CS) as in our case[1,2].

In this report, a 7-year-old girl, diagnosed with VSD (ventricular septal defect) is presented in whom PLSVC and absent RSVC was encountered during percutaneouse VSD closure. Due to the dislodgement of device into the right ventricle following 24 hours of device positioning, patient underwent open-heart surgery to remove the device and to close the VSD.

CASE

A seven-year-old girl was scheduled for transcatheter VSD closure to a pmVSD. Auscultation revealed pansystolic murmur best heard at the left lower sternal edge. A 12-lead electrocardiogram showed sinus rhythm with normal axis. Chest x-ray and routine laboratory tests were normal. Echocardiography showed pmVSD of 6.5 mm in diameter (in apical five chamber view), PLSVC and a enlarged CS. Aortic rim was 4 mm. The interventricular gradient was 48 mmHg continuous-wave Doppler by the modified Bernoulli equation.

She was taken to catheter laboratory for percutaneous VSD closure. Access to VCS from right atrium by various guide and catheters during hemodynamic study was unsuccessful. By using femoral venous route respectively vena cava inferior, right atrium, CS, PLSVC and finally innominate vein was entered. A radioopaque substance was given to innominate vein. No RSVC was observed (Fig. 1). The contrast medium descended into the large CS via the LPVCS. Thus absent RVCS was diagnosed and the CS drained all venous blood from the head, neck and upper extremities by the way of PLVCS from innominate vein. The inferior vena cava was normally located. The Qp/Qs was 1.8, PVR was 2.2. Perimembranous VSD was closed by 7 mm pmVSD Amplatzer device from transvenous route using arteriovenous loop without any complication. Intravenous injection of agitated saline from right brachiocephalic vein showed contrast enhancement of a enlarged CS before the right atrium by transthoracic echocardiographic subcostal view (Fig.2 and movie). This finding demonstrated a PLSVC and the absence of RSVC.

Twenty-four hours after closure in the transthoracic echocardiography device had tendency to embolise to the right ventricle. Therefore device was planned to be snared and retrieved percutaneously which failed subsequently. The patient was taken to operation room for surgically removing of amplatzer device. Median sternotomy and pericardiotomy was made. The right-sided appendage displayed the morphology of right atrium and left-sided one of left and inferior vena cava entered the right-sided atrium. The innominate vein ran toward left and connected to the LPSVC, which flowed to the coronary sinus in the right atrium. The RSVC was completely absent. Surgical procedure comprised of standard cardiopulmonary bypass established through aortic and bicaval cannulation, through PLSVC and IVC (L-shaped cannulas). Antegrade cardioplegia was used for myocardial protection. Device was tired to remove with atriotomy. Because of device jammed in the chords of tricuspid valves it could not removed. Therefore right ventriculotomy was performed and device taken out without damage tricuspid valves. In the same session, VSD was closed with Dacron patch. The patient tolerated the procedure well and was discharged without any complications.

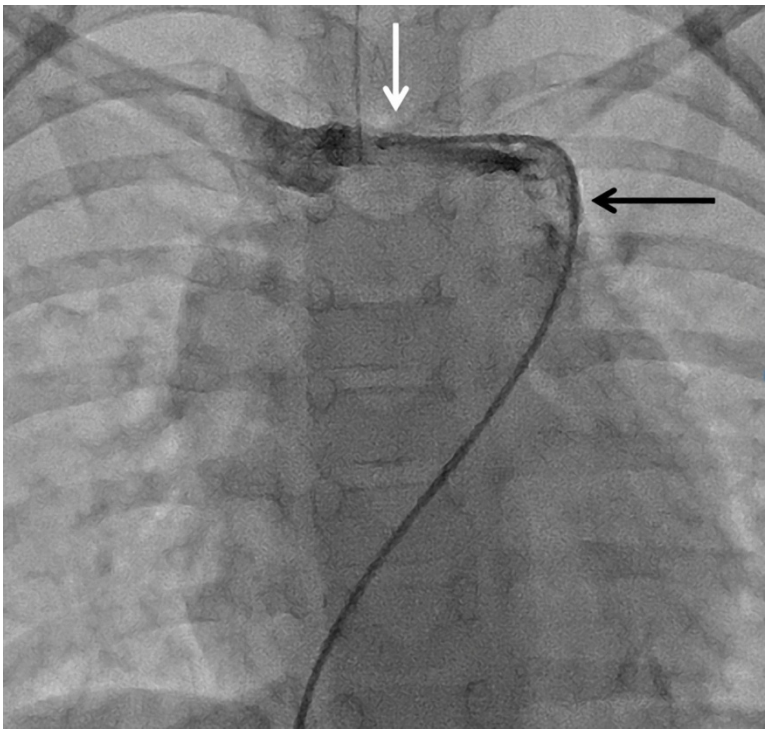

Figure. 1. Following femoral venous access; CS, PLVCS and innominate vein were reached respectively. A venogram was performed in the innominate vein (white arrow). The RSVC was absent and the CS drained all venous blood from the head, neck and upper extremities by the way of PLVCS (black arrow).

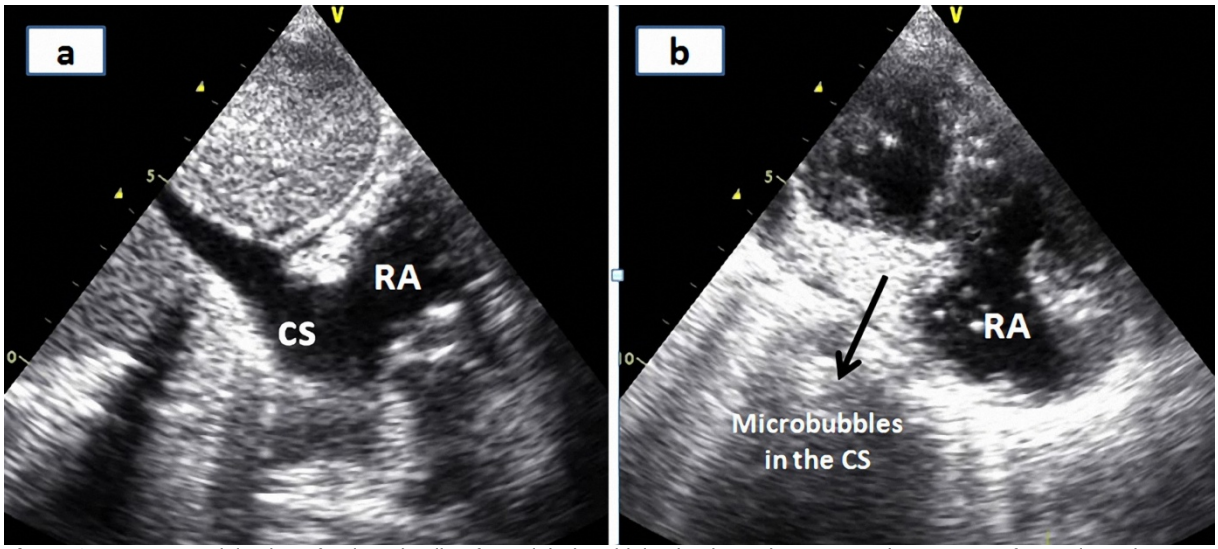

Figure 2. Intravenous injection of agitated saline from right brachial vein showed contrast enhancement of an enlarged CS, before the right atrium by transthoracic echocardiographic subcostal view. This finding demonstrated a PLSVC and the absence of a right superior vena cava.

DISCUSSION

During normal fetal development, the left-sided anterior venous cardinal system regresses, leaving the CS and ligament of Marshall. Failure of the regression leads to the existence of PLSVC[2]. Usually, PLSVC is associated with RSVC. A PLSVC has an incidence of 0.3-0.5% in the general population. In the presence of another congenital heart disease, the incidence of PLSVC increases to 3-10%. Persistant Left Superior Vena Cava with absent RSVC is a very rare venous malformation and drains into the right atrium via a dilated CS in most cases[1,2].

Absent RSVC with PLSVC is very rare in the setting of visceroatrial situs solitus, but is seen in visceral heterotaxy and associated atrial situs inversus. Our patient had visceroatrial situs solitus with VSD (1-6). This venous malformation occasionally can be accompanied by several heart defects but it has not specific patterns of associated heart anomalies. The most common malformations associated with this anomaly such as ASD (16%), endocardial cushion defect (11%), and tetralogy of Fallot (9%). Although VSD is rare, it can be accompanied with absent RSVC like in our case[3,4].

Isolated cases are frequently asymptomatic. Before any invasive intervention, it is important to establish an accurate diagnosis due to possible complications related to the procedures like implantation of a transvenous pacemaker or defibrillator, placement of pulmonary artery catheter for intraoperative or intensive care unit monitoring, systemic venous cannulation for cardiopulmonary bypass or extracorporeal circulation, cavopulmonary anastomosis, and orthotopic heart transplantation[4,5]. If the patient had not absence RVCS, removing of the device would have tried via VCS.

It should be suspected when PLSVC associates an enlarged CS in echocardiography. Diagnostic transthoracic echocardiographic criterias are as follows: 1) presence of a dilated CS on two-dimensional echocardiography, 2) intravenous injection of agitated saline into the left antecubital vein for contrast enhancement imaging of the giant CS and subsequently the right atrium, 3) for ruling out concomitant absence of a RSVC, in which case intravenous injection of agitated saline into the right antecubital vein should be performed. In that case the CS contrasts first and is followed by the right atrium like in our case (movie). We underline the importance of right antecubital vein agitated saline injection for confirming the diagnosis of absence RSVC[5,6].

In conclusion, clinicians should be alert and diagnosis of the disease should be done properly in order to avoid possible complications before various surgical and transcatheter procedures.

Abbreviations: CS: coronary sinus, RA: right atrium, VSD: ventricular septal defect, RSVC: Right superior vena cava, PLSVC: persistent left superior vena cava, VCS: vena cava superior

REFERENCES

1. Heye T, Wengenroth M, Schipp A, Johannes Dengler T, Grenacher L, Werner Kauffmann G. Persistent left superior vena cava with absent right superior vena cava: morphological CT features and clinical implications. Int J Cardiol 2007;116:103–5.
2. Jacob M, Sokoll A, Mannherz HG. A case of persistent left and absent right superior caval vein: An anatomical and embryological perspective. Clin Anat 2010;23:277-286.
3. Korkmaz L, Akyüz AR, Erkuş ME, Topal C. Isolated persistent left superior vena cava with absent right superior vena cava in two cases. Turk Kardiyol Dern Ars. 2011;39:501-4.
4. Bartram U, Van Praagh S, Levine JC, Hines M, Bensky AS, Van Praagh R. Absent right superior vena cava in visceroatrial situs solitus. Am J Cardiol 1997;80:175-83.

5. Yuce M, Kizilkan N, Kus E, Davutoglu V, Sari I. Giant coronary sinus and absent right superior vena cava. Vasa. 2011;40:65-7.

6. Gümüş A, Yildirim SV. Absent right superior vena cava with persistent left superior vena cava: two case reports. Turk J Pediatr. 2012;54:545-7.

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