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A complex congenital cardiovascular anomaly: persistence of left inferior and superior caval veins in conjunction with atrial and ventricular septal defects

Kompleks bir konjenital kardiyovasküler anomali: Persistan sol inferiyor ve süperiyor kaval venlerin atriyal ve ventriküler septal defektler ile birlikteliği

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Introduction

Persistence of the left superior and inferior vena cava (SVC and IVC, respectively) is relatively rare cardiac anomalies, and their frequency varies from 0.2% to 0.5% in the general population (1-4). Some congenital cardiac abnormalities such as atrial or ventricular septal defect and unroofed coronary sinus may accompany the persistent left SVC, resulting in paradoxical embolism (1, 2). However, persistent left IVC or transposition of IVC is much rare congenital anomaly compared with persistent left SVC (3, 4). In the literature, several case reports about persistence of the left IVC with its azygos or hemiazygos continuation have been reported (3-8). However, no associated congenital cardiac defects such as atrial and ventricular septal defects (ASD and VSD, respectively) have been presented in those reports. Therefore, we describe a complex and rare congenital anomaly, which includes ASD, VSD, and persistent left IVC and SVC.

Case report

A 31-year-old woman presented with effort dyspnea. She had no history of previous cardiac disease. Her vital signs such as blood pressure and radial pulses were stable. Physical examination revealed holosystolic murmur at grade of 3/6 at the left sternal border radiating to other auscultation points. There were no other pathologic findings. Electrocardiogram was also normal. In transthoracic echocardiography, subaortic VSD and secundum type ASD were detected. There were also severe tricuspid regurgitation and mild aortic regurgitation. Pulmonary systolic pressure of 45-50 mmHg was calculated from tricuspid regurgitation. Pulmonary to systemic flow ratio of 1.7 was estimated by Doppler echocardiography. In addition, abnormal mosaic flow with undefined origin was observed in the right atrium. She underwent cardiac catheterization to confirm a significant left to right shunt and to exclude possible other cardiac abnormalities. During the catheterization, while forwarding the catheter via the right femoral vein, we realized that the direction of catheter was towards to the left arm of the patient and there was no normal IVC course. This vein was considered as persistent left IVC (Fig. 1A). When the catheter was further advanced, the right atrium was entered through persistent left SVC and its angiography was performed (Fig. 1B). We considered that persistent left IVC, which continues as vena hemiazygos drained into right atrium via persistent left SVC connecting to coronary sinus (Fig. 1A). Left innominate vein, was not viewed. There was a right superior caval vein which drained normally to the right atrium and normal pulmonary venous return in right atrial angiography. Superior vena cava and IVC related to the topography of the thorax and abdomen in this case is drawn schematically in Figure 2. Oxygen saturations of left IVC, persistent left SVC, right atrium and right ventricle were 66%, 70%, 75% and 85%, respectively. They showed a significant left-to-right shunting at both atrial and ventricular levels. Pulmonary artery could not be engaged via femoral vein because of failure in back-up of the catheter. Right internal jugular vein was used for pulmonary artery catheterization. The mean O2 saturation and pressure of main pulmonary artery were 84% and 55/30 mm Hg respectively. Persistent left SVC was also confirmed by administrating contrast media via left antecubital vein. Left ventriculography revealed the subaortic VSD (Fig. 3). By oxymetric method, pulmonary to systemic flow ratio of 1.85 was calculated. Thus, surgical closure for ASD and VSD was performed and those defects were confirmed. Abdominal ultrasonography demonstrated no additional abnormalities such as polysplenia and other visceral levoisomerism (liver, spleen and stomach) except hepatic veins drained directly into the right atrium. Liver was seen at right side, spleen and stomach were seen at left side of the abdomen. Cardiac chambers and great systemic arteries were at normal position (Fig. 4). So we considered abdominal situs solitus.

Discussion

Persistent left IVC and SVC are unusual congenital anomalies in the population. They result from the failure of regression of the right supra-cardinal vein and left anterior cardinal vein, respectively (1-4). Persistent left SVC is relatively more prevalent compared with the left IVC and may result in paradoxical embolism due to accompanying lesions such as ASD, unroofed coronary sinus and its directly drainage into the left atrium (1). Up to now, it has not been reported that persistent left SVC accompanies left IVC in a case with both ASD and VSD. To our knowledge, this is the first reported case of such an anomaly.

Interruption of the IVC or persistent left IVC with azygos or hemiazygos continuation is an unusual but well-known anomaly of the IVC (3-8). In case of persistent left IVC, possible routes for the return of blood to the right atrium are via the azygos vein to the SVC, via the hemiazygos vein to persistent left SVC or via the left brachiocephalic vein to right SVC (4). Infrequently, this anomalous vein may accompany ASD and open directly

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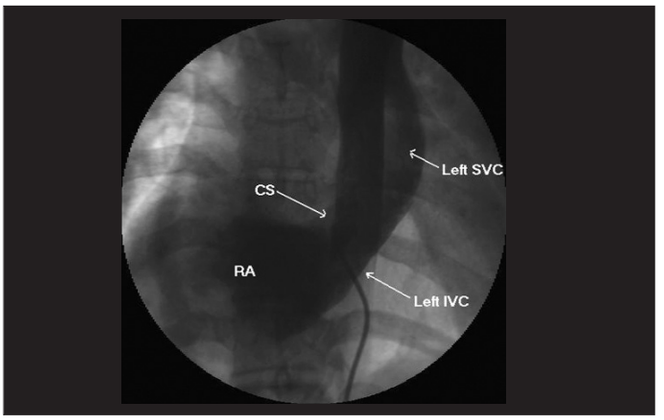

Figure 1A. Antero-posterior view showing contrast injected into the left inferior vena cava with hemiazygos continuation, which drains into the persistent left superior vena cava and then into the right atrium via coronary sinus

CS - coronary sinus, IVC - inferior vena cava, RA - right atrium, SVC - superior vena cava

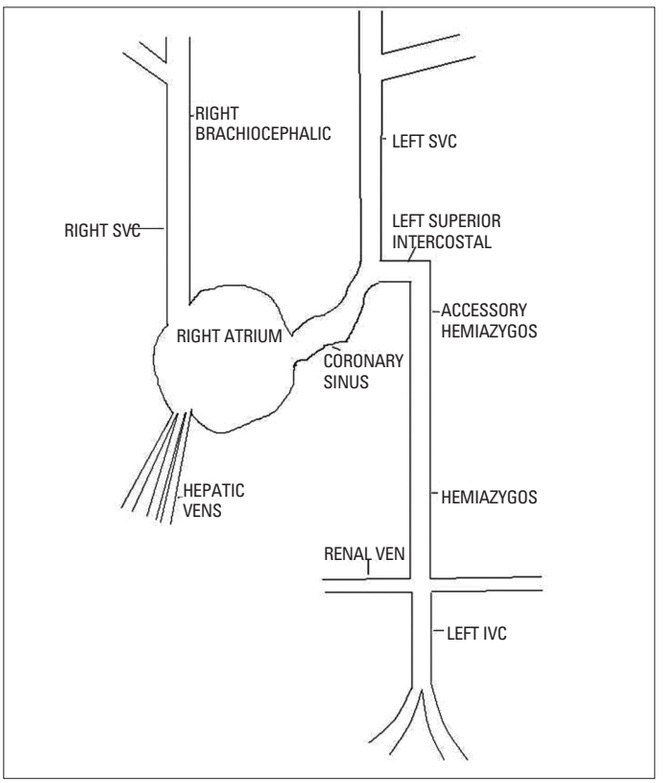

Figure 2. Schematic diagram of the routes of venous drainage in this case. IVC-inferior vena cava, SVC - superior vena cava

into the left atrium, resulting in right-to-left shunting without pulmonary hypertension (9). In general, the return of blood of left IVC to the right atrium is usually provided with its hemiazygos continuation connecting to left SVC. In the literature, a case with ASD, patent ductus arteriosus and a left IVC with hemiazygos continuation and drainage into the coronary sinus was reported (8).

Hemiazygos continuation of left IVC has been reported previously by computed tomography in the literature (3-6). We did not perform the tomography with contrast medium. Instead, we passed directly the mentioned course of venous return with catheter under the guidance of fluoroscopy.

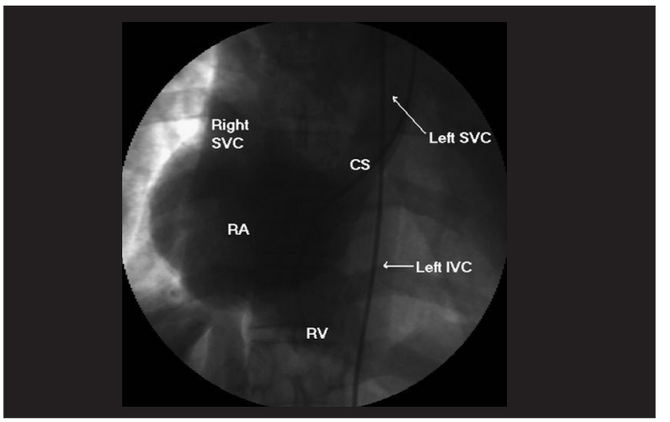

Figure 1B. Angiographic view of the right atrium in the antero-posterior projection. It was obtained through persistent left superior vena cava connecting to coronary sinus via the left inferior vena cava

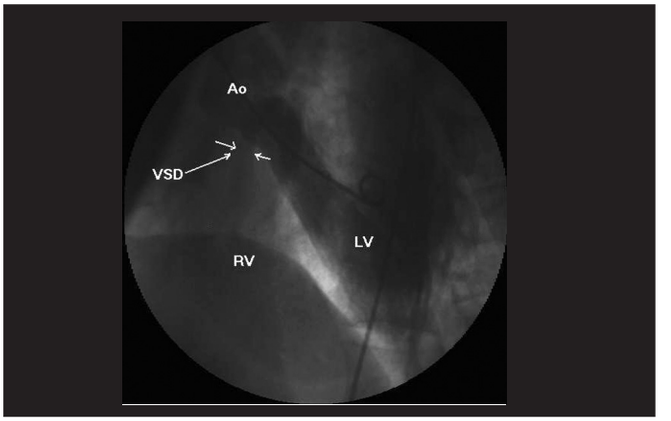

Figure 3. Left ventriculography in the left anterior oblique projection with cranial angulation Arrows demonstrate the subaortic ventricular septal defect. Ao- aorta, LV- left ventricle, RV- right ventricle, VSD-ventricular septal defect

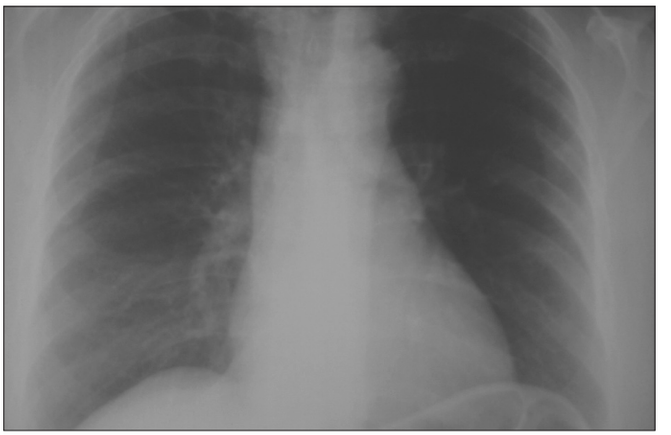

Figure 4. Chest X-ray-showing absence of dextrocardia

Left IVC and persistent left SVC are usually asymptomatic, being detected incidentally during routine thoraco-abdominal imaging by ultrasonography or computed tomography or cardiac catheterization. Their presence may complicate some surgical procedures such as Glenn shunt, Mustard repair and abdominal surgery (10). Their injuries by surgery may result in severe hemorrhage. In addition, percutaneous inter-

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ventions such as implantation of pacemaker, cardioverter-defibrilator or caval filter may be difficult in such patients. Similarly, these anomalies can also make percutaneous closure of ASD or VSD difficult.

Persistent left IVC can make the right heart catheterization difficult via femoral vein as we encountered. We were not able to advance the catheter into the pulmonary artery via femoral approach, and used the right internal jugular vein for this purpose.

Persistent SVC usually drains into the coronary sinus, resulting in its dilatation. By stretching the atrioventricular node and His bundle, rhythm disturbances may occur in such patients (1, 2). Our patient had no rhythm abnormality.

Conclusion

In conclusion, it should be kept in mind the possibility that any congenital cardiac anomaly may accompany other cardiovascular abnormalities. A detailed investigation should not be neglected.

References

1. Tak T, Crouch E, Drake GB. Persistent left superior vena cava: Incidence, significance and clinical correlates. Int J Cardiol 2002; 82: 91-3.
2. Sorodia BD, Stoller JK. Persistent left superior vena cava: Case report and literature review. Respir Care 2000; 45: 411-6.
3. Bricker ME, Eichhorn EJ, Netto D, Cigarroa RG, Brogan WC, Simonsen RL, et al. Left-sided inferior vena cava draining into the coronary sinus via persistent left superior vena cava: A case report and review of the literature. Catheter Cardiovasc Diagn 1990; 20: 189-92.
4. Kim HJ, Ahn IO, Park ED. Hemiazygos continuation of a left inferior vena cava draining into the right atrium via persistent left superior vena cava: Demonstration by helical computed tomography. Cardiovasc Intervent Radiol 1995; 18: 65-7.
5. Allen HA, Haney PJ. Left-sided inferior vena cava with hemiazygos continuation. J Comput Assist Tomogr 1981; 5: 917-20.
6. Dudiak CM, Olson MC, Posniak HV. CT evaluation of congenital and acquired abnormalities of the azygos system. Radiographics 1991; 11: 233-46.
7. Munechika H, Cohan RH, Baker ME, Cooper CJ, Dunnick NR. Hemiazygos continuation of a left inferior vena cava: CT appearance. J Comput Assist Tomogr 1988; 12: 28-30.
8. Benrey J, Williams RL, Reul GJ. Hemiazygos continuation to coronary sinus with normal left innominate vein. Cardiovasc Dis 1975; 2: 325-30.
9. Hallali P, Tcheng P, Davido A, Leriche H, Corone P. Atrial septal defect and cyanosis. Apropos of 6 cases related to abnormal drainage of the inferior vena cava into the left atrium. Arch Mal Coeur Vaiss 1988; 81: 783-6.
10. Brener BJ, Darling RJ, Frederic PL, Linton RR. Major venous anomalies complicating abdominal aortic surgery. Arch Surg 1974; 108: 159-6.

Triküspit kapakta kitle: Manyetik rezonans görüntüleme bulguları

Tricuspid valve mass: magnetic resonance imaging findings

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Giriş

Manyetik rezonans (MRI) ile görüntüleme tekniği, radyoaktif yayılım olmadan, noninvazif bir şekilde manyetik radyo dalgaları ile yapılan bir görüntüleme tekniğidir. Diğer standart noninvazif metodlar arasında ekokardiyografi ve bilgisayarlı tomografi (BT) sayılabilir. Bazı vakalarda ekokardiyografik incelemelerde yanlış pozitif veya negatif sonuçlara da rastlanmaktadır (1, 2). Bilgisayarlı tomografi tetkikinde iyonize radyasyon ve intravenöz kontrast kullanımı söz konusudur (3). Ayrıca tümör ve trombüs ayrımı yapmak her zaman mümkün olmayabilir.

Kalp kapakçıklarında kitle lezyonu nadir olarak görülmektedir.

Olgu Sunumu

Altmış dokuz yaşında erkek hasta, baş dönmesi, fenalık hissi şikâyetleri ile doktora müracaat ediyor. Fizik muayenesinde, tansiyonu 150/70mmHg nabız dakika sayısı 66/ ritmik, sistem muayenelerinde bir özellik yok. Elektrokardiyografide sinüzal ritm, efor testi normal bulunuyor. Transtorasik ekokardiyografi tetkikinde; sağ atriyum ve sağ ventrikül normalden hafif geniş. Triküspit kapak septal leafletin atriyal yüzünde 1.3x0.9 cm çapında hiperekojen, nodüler kitle imajı gözleniyor. Kapak açılımı normal. Anlamlı gradiyent saptanmıyor. Triküspit kapaktan sistolde sağ atriyum içine hafif derecede (1+) regurjitan akım saptanıyor. Kardiyak MRI tetkikinde; triküspit kapağın septal yaprağı üzerinde yaklaşık 1 cm boyutta nodüler kitle lezyonu tespit edilmiş olup, iv kontrast enjeksiyonu sonrası kontrast tutulumu gösteriyor (Resim 1, 2. Video 1. Video/hareketli görüntüleri www.anakarder.com`da izlenebilir).

Noninvazif kardiyak görüntüleme tetkikleri arasında ekokardiyografi, kardiyak BT ve kardiyak MRI'ı sıralayabiliriz. Kalp içi ve dışı kitle lezyonlarının belirlenmesinde kardiyak MRI'ın yerini belirlerken yaygın olarak kullanılan ekokardiyografiye kıyasla kitlenin doku özelliklerini tanımlamada daha kolaylık yarattığını belirtmek gerekir. (4-6). Buna karşılık uzun sürmesi, hasta açısından sıkıntılı bir tetkik olması (klostrofobi vs.), elektrokardiyografi takibi ile beraber stabil bir kardiyak ritm gerektirmesi dezavantajıdır (7). Vakamızda yapılan ekokardiyografi ve MRI tetkiklerine ait sonuçlar birbiri ile uygunluk göstermiştir. Hasta asemptomatik olduğun-

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