Interrupted Inferior and Double Superior Vena Cava Opening into the Left Atrium via Coronary Sinus in Polysplenia Syndrome: A Coincidence With Rheumatic Heart Valvular Disease

POLİSPLENİ SENDROMUNDA KORONER SİNÜS YOLU İLE SOL ATRİYUMA AÇILAN ÇİFT SÜPERİOR VENA KAVA VE KESİNTİLİ İNFERİOR VENA KAVA: ROMATİZMAL KALP KAPAK HASTALIĞI İLE KOİNSİDANS

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Summary

Purpose: Persistent left superior vena cava is a relatively common variant of the systemic venous return, occuring in 0.3% of the general population. In this paper, a patient with rheumatic valvular heart disease, interrupted inferior and double superior vena cava entering into the left atrium via coronary sinus in polysplenia syndrome was presented with reviewed literature.

Material Method: In a 40-year-old patient, following detection of a dilated coronary sinus in the transthoracic echocardiographic examination, diagnosis was verified by transesophageal echocardiography, cardiac catheterization, spiral computerized tomography and nuclear magnetic resonance imaging. In addition to these findings, situs ambiguous and polysplenia were detected.

Result: Persistent left superior vena and interrupted inferior vena cava which is a common systemic venous return anomaly have important clinical implications on some certain situations.

Key Words: Double superior vena cava, Inferior vena Cava, Rheumatic valvuler heart Disease

T Klin J Cardiol 2003, 16:391-395

Özet

Amaç: Persistan sol süperior vena kava %0.3 sıklıkta ve en sık saptanan torasik venöz dönüş anomalisidir. Bu yazıda koroner sinus yoluyla sol atriyuma açılan çift superior, kesintili inferior vena kavası olan polispleni sendromlu, romatizmal kalp kapak hastalıklı bir olgu sunulmuş ve literatür gözden geçirilmiştir.

Materyal ve Metot: 40 yaşında bayan bir hastada transtorasik ekokardiyografide genişlemiş koroner sinüs saptanması üzerine yapılan transözofagial ekokardiyografi, kardiyak kateterizasyon, spiral bilgisayarlı tomografi ve nükleer manyetik rezonans görüntüleme ile tanı doğrulandı. Bu bulgulara ek olarak situs ambigius ve polispleni saptandı.

Sonuç: Persistan sol süperior vena kava ve kesintili inferior vena kava, belirli klinik uygulamalar sırasında düşünülmesi gereken venöz sistem anomalisidir.

Anahtar Kelimeler: Çift süperiyor vena kava, İnferiyor vena kava, Romatizmal kalp kapak hastalığı

T Klin Kardiyoloji 2003, 16:391-395

Persistent left superior vena cava (PLSVC) is a relatively common variant of the systemic venous return, occurring in 0.3% of the general population and 3-34% of patients with congenital heart disease and the most common thoracic venous anomaly (1-5).

Case Report

A forty-year-old female patient with dispnea and fatigue lasting for three months was referred to our hospital. In her physical examination, blood pressure was 100/60 mmHg. Irregular, 110/min heart rate was detected. Jugular veins were promi-

nent. Cardiovascular examination revealed a loud S1, apical 2/6 pansystolic murmur and bilateral basilar crepitan rales in her lungs. Ascites, hepatomegaly and peripheral edema were found. Electrocardiogram showed rapid atrial fibrillation. The chest X-ray revealed mild cardiomegaly and pulmonary vascular enlargement.

Echocardiographic Examination

In the transthoracic echocardiographic examination (TTE) dilated coronary sinus (CS), thickened mitral leaflets and mitral stenosis (planimetric measurement of mitral valve area: 1.35 cm^2, peak/mean gradient 17/10 mmHg), mild aortic regurgitation, tricuspid stenosis (peak/mean gradient 9/5 mmHg) and severe tricuspid regurgitation were detected. Pulmonary arterial pressure was 35 mmHg. Left atrium (LA) and right atrium (RA) were dilated. PLSVC was suspected because of dilated CS in the TTE examination.

Patient underwent transesophageal echocardiographic (TEE) examination and dilated CS was demonstrated. A two-dimensional contrast echocardiogram using injection of agitated saline solution into the right antecubital vein revealed a prompt appearance of the contrast effect in the CS (Figure 1).

Figure 1. Injection of agitating saline to the antecubital vein revealing contrast effect in the coronary sinus before right atrium. CS: Coronary sinus, LA: Left atrium, RA: Right atrium.

Cardiac Catheterization

Cardiac catheterization for rheumatic valvular disease was performed by introducing a catheter into the right femoral vein, which could not be passed through the inferior vena cava (IVC) into the right atrium. Contrast injection into the IVC revealed the absence of hepatic portion of IVC and opacified the hemiazygous vein, PLSVC, CS and LA, respectively. A second catheter was introduced into the right cubital vein and passed into the SVC. Contrastmedia injected into the SVC then opacified the CS and LA. A PLSVC was present entering the CS.

Computerized Tomography

In the spiral computerized tomographic examination, cardiomegaly and SVC were detected (Figure 2). The PLSVC and hemiazygos vein joined at the level of left pulmonary artery and were draining into CS. The liver was enlarged and centrally located. Polysplenia was found on the right side (Figure 3).

Figure 2. Spiral computerized tomography image demonstrating left and right superior vena cava. RSVC: Right superior vena cava, LSVC: Left superior vena cava.

Nuclear Magnetic Resonance Imaging

Nuclear magnetic resonance images of the heart confirmed the diagnosis of double SVC entering into the LA. It also demonstrated that the left

Figure 3. Spiral computerized tomography image demonstrating polysplenia and centrally located liver. S: spleen.

Figure 4. Nuclear magnetic resonance image showing hepatic veins draining into the RA directly.

sided IVC was interrupted at hepatic level and hepatic veins were draining into the RA directly (Figure 4).

The patient was referred to cardiac surgery for mitral and tricuspid valve reparation.

Discussion

We present a 40-year-old female patient with double SVC, interrupted IVC and rheumatic valvular heart disease. Extra-cardiac abnormalities associated with persistent left SVC include coarctation of the aorta, pulmonary arterial venous fistulae, patent ductus arteriosus and abnormalities of the IVC (6). Once a diagnosis of the SVC entering the LA is made, these commonly associated malformations should be excluded. In our case, none of the above mentioned abnormalities were detected except interrupted IVC continuing with hemiazygos continuation.

In the interrupted IVC, the hepatic portion of IVC is completely absent. Under these circumstances, the IVC remains a posterior structure and joins the azygos or hemiazygos vein and subsequently the right or left SVC. The hepatic veins converged to form a relatively narrow trunk, which joins the RA (7). In our patient above mentioned abnormalities of IVC were found. The hepatic portion of IVC was interrupted and joined the hemiazygos vein and subsequently to the left SVC.

The clinical presentation of this anomaly varies. Most patients had normal growth and development, and the majority had no cardiac murmur (6). Our patient complained of shortness of breath, palpitation, peripheral edema and abdominal discomfort, a decline in her performance, presumably a result of rheumatic valvular heart disease. PLSVC might have an association with disturbances of cardiac impulse formation and conduction (8). However, in a large study, during 10 year follow-up in 1139 patients undergoing pacemaker and defibrillator implantation, persistence of left SVC was similar to that in general population (9). In our case, atrial fibrillation due to mitral valve was observed.

Diagnosis of this anomaly has been assessed by different approaches such as contrast echocardiography (10,11), radionuclide studies (12,13), cardiac catheterization (14,15) and NMR. In the present case, the first diagnostic hint was a dilated CS in the TTE. Subsequently, the SVC could not be demonstrated by TEE and

a prompt appearance of the contrast effect in the CS. Cardiac catheterization, spiral CT and NMR verified the diagnosis.

Because of the nature of the atria and the position of abdominal organs, neither situs solitus nor situs inversus can be identified, situs ambiguous is said to be present. This usually applies in cases of asplenia or polysplenia. Polysplenia defined as two or more splenic masses and usually consisting of two somewhat larger spleens, tends to be characterized by levoisomerism or 'bilateral left sidedness'. The liver is abnormally symmetrical in about 25% of patients (16). Situs ambiguous is usually associated with systemic venous return anomalies (17). From 50 to 85% of patients have absence of infrahepatic to suprarenal portion of the IVC, with azygos or hemiazygos continuation of the suprarenal portion of the IVC to the ipsilateral SVC. Absence of the hepatic segment of the IVC with azygos continuation into the right or left SVC has been reported as an incidental finding at post-mortem examinations since 1793 (18). In a study of 46 post-mortem cases of visceral heterotaxy with polysplenia, an interrupted IVC was present in 39 of 46 (85%) patients (19). Ruscazio et al. discussed the autopsy findings of 6 asplenia cases with interrupted IVC found in literature review. Four of these 6 cases (67%) had cardiac malformations usually seen in the asplenia syndrome, whereas 2 of the 6 (33%) had heart defects usually encountered in polysplenia patients (20). Our case had characteristic findings of polysplenia syndrome with IVC abnormalities, symmetrical liver and multiple right sided spleens.

PLSVC and the interrupted IVC have important clinical implications in certain citations such as 1) transvenous pacemaker implantation impossible due to the abnormal angulations of the catheter because it is advanced from the left SVC by way of the CS to the RA and right ventricle. There is a substantial risk of catheter looping or knotting. Dislodgment of the catheter also occurs after its original implantation into the right ventricle (21). 2) Pulmonary artery catheter placement, monitoring is usually done without fluoroscopy. If the right SVC is absent, advancing the catheter into the right ventricle may require an acute angle of the catheter, and this may result in catheter looping or knotting in the RA. For such complications the main safeguard is the use of a fluoroscope screen, 3) Systemic venous cannulation for extracorporeal membrane oxygenation to achieve with the usual venous cannula (22), 4) In cases requiring cardiopulmonary bypass, the perfusion may be initiated through the IVC before the left SVC is cannulated retrogradely by way of the RA and CS 5) In partial or total cavopulmonary anastomoses, the preoperative knowledge of the systemic venous connections is essential (23).

The necessity for surgical correction of this abnormality varies. It is dependent on the patient's clinical presentation and should be considered in each individual case. Surgical correction has been performed in some of the reported cases (10-12,14,24). In a group of patients, the SVC was disconnected from the LA and anastomized to the RA (10,11,14). In other patients, a pericardial patch was fashioned so as to divert the SVC blood flow into the RA and the pulmonary venous return into the LA (12). In the present case, surgical intervention for severe valvular heart disease, but not for systemic venous abnormalities was performed.

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Geliş Tarihi: 10.04.2003

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