

Complex congenital anomalies of superior vena cava and pulmonary veins with left-sided inferior vena cava

Sol yerleşimli inferior vena cava ile süperiyor vena cava ve pulmoner venlerin kompleks konjenital anomalileri

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Introduction

Development of inferior vena cava (IVC) begins at fifth week of gestation. Three paired venous channels that develop sequentially (posterior cardinal, subcardinal, supracardinal) anastomose and regress (1). Left sided IVC develops as a result of the persistence of the left supracardinal vein, and prevalence of this anomaly is estimated to be 0.2-0.5% (2). Persistent left superior vena cava (SVC) is a well-known anomaly, and found in 0.5-4% of general population. The most frequent anomaly accompanies with persistent left SVC is total anomalous pulmonary venous return (10%) (3). The incidence of pulmonary vein anomalies is about 0.5% (4).

We report a first case of complex venous system anomaly, which is a combination of three anomalies in systemic and pulmoner venous system.

Case report

A 20-year-old asymptomatic male was referred to our clinic for delineation of the cause of the enlarged cardiac silhouette on his chest X-ray obtained for routine check-up. His medical and family history and physical examination were unremarkable except 1/6-grade systolic murmur best heard at lower sternal region. Standard surface electrocardiogram and blood chemistry were in normal ranges. Echocardiographic examination revealed enlarged coronary sinus, right atrium and right ventricle. For identifying the cause of enlargement of right sided chambers, contrast echocardiographic examination with agitated saline was performed. During contrast injection via lower extremity veins primarily coronary sinus, secondarily right atrium filled with contrast (Fig. 1A). However, IVC was not visualized in substernal view (Fig. 1B). Instead of IVC, hepatic veins emptied into right atrium individually at the substernal echocardiographic view (Fig. 1B). This finding led us to suspect the absence of hepatic segment of the IVC and venous drainage of lower extremity is maintained via persistent left SVC. Further injection from the right antebrachial vein showed the right atrial filling from SVC (Fig. 1C), which also leads us to suspect persistent left SVC with intact right SVC.

Thoraco-abdominal computed tomography (CT) showed that right jugular and subclavian veins come together, form the right sided SVC, which drains to the right atrium, and left internal jugular vein with left

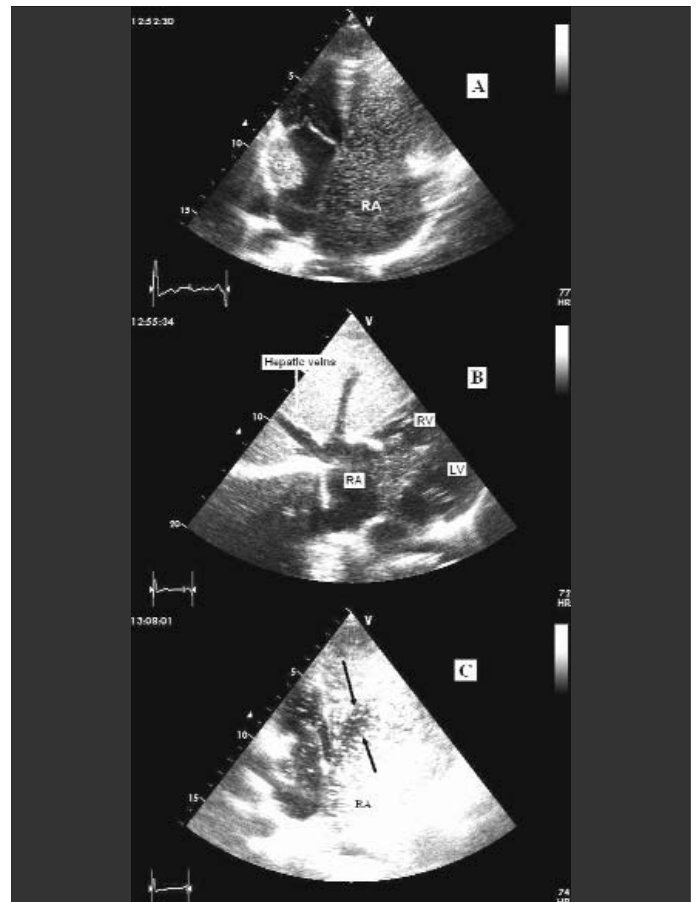


Figure 1. (A) Apical four-chamber view of agitated saline contrast echocardiography injected from lower extremity (B) Subcostal view during contrast echocardiography (C) Apical four-chamber view, contrast injected from right upper extremity filling right atrium from superior vena cava, coronary sinus flow negative contrast (arrow)

CS - coronary sinus, RA - right atrium, RV -right ventricle, LV - left ventricle

subclavian vein forms the left SVC, which drains to the right atrium via the coronary sinus (Fig. 2). Besides these anomalies left pulmonary veins drain to left atrium, right pulmonary veins drain to the right atrium (Fig. 3). Bilateral iliac veins converge and form the left sided IVC (Fig. 4). Left-sided IVC proceeds as hemiazygos vein (Fig. 5). Venous drainage of abdomen and lower extremities maintained by left subclavian vein via hemiazygos conduit. Computed tomography images also revealed malrotation of the spleen but not polysplenia.

Discussion

While each of left-sided IVC, double SVC and partial pulmonary venous return anomalies reported individually (5, 6), there was no knowledge about the presence of all these three anomalies at the same time except left atrial isomerism. However, our case cannot be diagnosed as left atrial isomerism because of absence of left sidedness such as two left lung and bronchi. A necropsy serial with double SVC declared that all the cases with left SVC had continuity with the coronary sinus (7). Ventricular septal defect and atrioventricular connection anomalies are the most frequent additional congenital anomalies (7). No accompanying cardiac anomaly was determined in

our patient. Although, absence of IVC is associated with polysplenia in 84% of cases (8), our case had not polysplenia, but malrotation of the spleen with a hilus located inferiorly was present. Gouley et al. (9) described a case of absent hepatic portion of the IVC with hemiazygos drainage direct into the left atrium, with questionable, persistent left

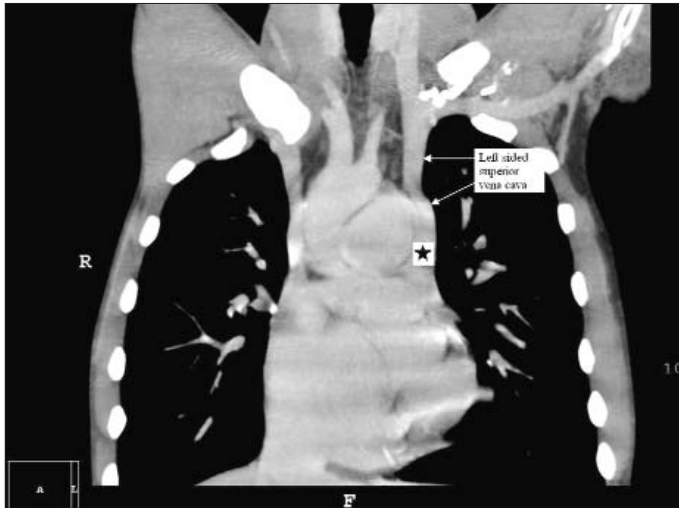


Figure 2. Left internal jugular vein and left subclavian vein come together and drain to the coronary sinus via persistent left superior vena cava

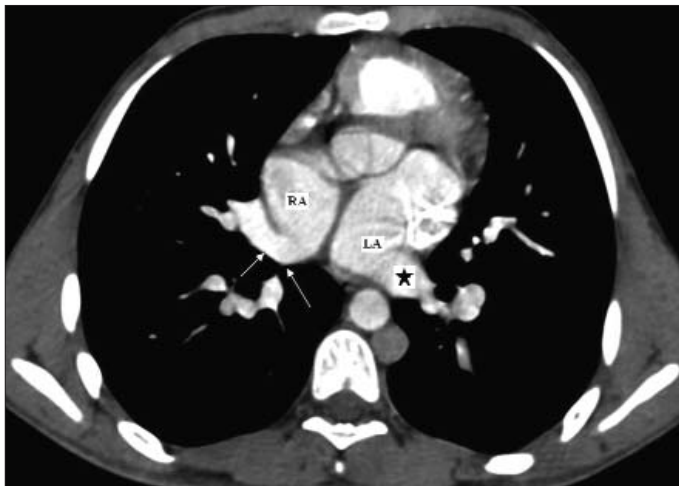


Figure 3. Right pulmonary veins drain to the right atrium (arrow), left pulmonary veins drain to the left atrium (star)

RA - right atrium, LA- left atrium



Figure 4. Coronal section of the left sided inferior vena cava localized left side of the abdominal aorta (arrow). Notice to the absence of the hepatic segment of the inferior vena cava (*)

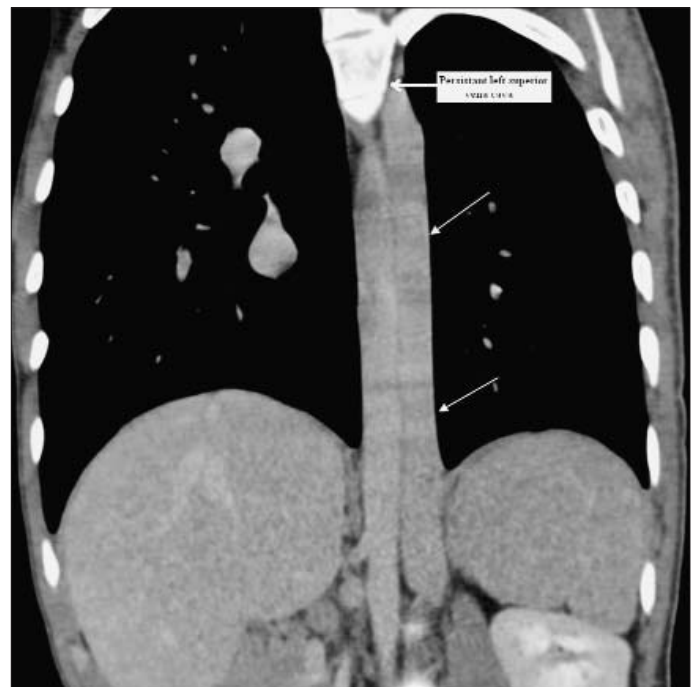


Figure 5. Coronal section of the hemiazygos vein localized left side of the thoracic aorta (arrows) and drained to the persistent left superior vena cava.

SVC. In other series, the hemiazygos drained into the left SVC, which mostly drains into the coronary sinus as in our case (10). Although the left sided IVC has been described previously, we have not found any report having three different anomalies all together. We suspected the drainage of subdiaphragmatic venous return to persistent SVC and absence the hepatic segment of IVC by agitated saline contrast echocardiography, and confirmed the diagnosis by multidetector CT.

Conclusion

Our case is unique for the presence of all three congenital systemic and pulmonary venous anomalies, come together. Determining of venous anomalies is of particular importance before implantation of cardiac pacemakers and implantable cardioverter defibrillators.

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Incidentally diagnosed congenital left ventricular aneurysm: report of two cases

Tesadüfen tanı konulmuş konjenital sol ventriküler anevrizma: İki olgu sunusu

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Introduction

Congenital ventricular diverticulum and aneurysm are two distinct rare entities with different morphologic and histological characteristics and outcomes. Congenital ventricular diverticulum and aneurysm have a reported prevalence ranging from 0.4% to 0.8% (1, 2). Although congenital diverticulum is usually associated with other anomalies such as Cantrell pentalogy, a congenital aneurysm is frequently detected as standalone finding (3). The differential diagnosis of these two conditions is cumbersome for the clinician. Nevertheless, a wide neck, the absence of normal ventricular layers and paradoxical contraction point to the presence of an aneurysm (4).

We report here incidentally diagnosed two congenital left ventricular aneurysm cases in which different imaging modalities were used.

Case 1

A 24-year-old male was admitted to our clinic with a complaint of atypical chest pain. His physical examination was unremarkable except apical mild late-systolic murmur. Electrocardiography and routine blood

chemistry were also within normal limits. Transthoracic echocardiographic examination revealed a mild mitral regurgitation and an outpouching located at posterior wall on parasternal long axis view (Video 1. See corresponding video/movie images at www.anakarder.com). This akinetic structure was also observed at the inferior wall (Video 2. See corresponding video/movie images at www.anakarder.com).

We thought that the patient had coronary artery disease and hence coronary angiography was performed. However, it revealed a normal coronary artery anatomy and a non-contractile structure suggesting an aneurysm approaching 1.5 cm diameter (Fig. 1). We performed a cardiac scintigraphy for better delineation of the problem, which confirmed an inferiorly-located outpouching without any evidence of ischemia (Fig. 2). Based on these findings we decided that the patient has a congenital left ventricular aneurysm located at basal ventricular region and should be followed with regular sixth month intervals.

Case 2

A 21-year-old male patient was studied for exertional dyspnea. His physical examination was unremarkable except leftward displacement of