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Left persistent superior vena cava with large coronary sinus: A case report

Sol persistant vena kavaya bağlı geniş koroner sinus: Olgu sunumu

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Abstract

Persistent left superior vena cava (LPSVC) is a rare and important congenital venous anomaly. It is caused by a defect in the closure of the left anterior cardinal vein during cardiac development. The LPSVC drains into the right atrium via the coronary sinus (CS) in 90% of cases, connects to the left atrium in 10 % of them. When cardiac anomalies are present, LPSVC is usually linked directly to left atrium. Thus, LPSVC which drains in the CS is generally isolated and asymptomatic. In our case, patient presented a heavy respiratory symptomatology without any diagnosis since all of the respiratory tests were normal. After realization of a computed tomography (CT), LPSVC had been discovered inducing a huge dilatation of CS, which its diameter was three times more than reported in literature and without any associated congenital heart disease. LPSVC seems to be a complex anatomic variation with different clinic and anatomic shapes. CS dilatation can be found in association with LPSVC in CT. As a result, it is important to use non-invasive cardiovascular examinations to make an optimal diagnosis of congenital cardiovascular variations and in order to avoid further interventional complications.

Keywords: Superior vena cava, Left persistent vena cava, Thorax radiology

Öz

Kalıcı sol superior vena kava (LPSVC) nadir ve önemli bir konjenital venöz anomalidir. Bu anomali, kardiyak gelişim sırasında sol anterior kardinal venin regresyonunda bir defektten kaynaklanır. LPSVC, % 90 oranında koroner sinüs (CS) yoluyla sağ atriyuma akar, % 10'unda sol atriyuma bağlanır. Kardiyak anomaliler mevcut olduğunda, LPSVC genellikle doğrudan sol atriyuma bağlanır. CS'ye drene olan LPSVC genellikle izole ve asemptomatiktir (4). Bizim olgumuzda, solunum testlerinin tümü normaldi ancak hasta herhangi bir tanı olmaksızın ağır bir solunum semptomatolojisi sundu. Bilgisayarlı tomografi (BT)'de LPSVC'nin, CS dilatasyonunu indülendiği saptanmıştır; CS literatürde belirtilen normal boyutun 3 kat üzerinde izlenmekteydi, herhangi bir konjenital anamali saptanmadı. LPSVC, farklı klinik ve anatomik şekillerle kompleks bir anatomik varyasyon gibi görünmektedir. CS dilatasyonu BT'de LPSVC ile ilişkili olarak bulunabilir. Sonuç olarak, konjenital kardiyovasküler varyasyonların optimal tanısını koymak ve girişimsel komplikasyonları önlemek için non-invaziv kardiyovasküler muayenelerin kullanılması önemlidir.

Anahtar kelimeler: Superior vena cava, Sol persistan vena kava, Toraks radyolojisi

Introduction

Persistent left superior vena cava (LPSVC) is a rare and important congenital venous anomaly and it is caused by a defect in the closure of the left anterior cardinal vein during cardiac development [1]. It can lead to coronary sinus (CS) dilatation when it drains to right atrium. In another perspective, this dilatation is very important to understand the way of drainage of LPSVC. Additionally, knowledge of LPSVC's physiopathology is also important before some interventional and surgery procedures [2,3]. We will put the light in our case report on LPSVC with a huge CS dilatation.

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Case presentation

We report the case of 56-years-old woman admitted to hospital with respiratory complains. Pulmonary functional tests were normal. There was a second line adjacent to the shadow of the descendant aorta in chest x-ray image. Bilateral lung parenchyma was normal. The ascending aorta was slightly tortoise and elongate. Trachea was in midline and borders were regular. No hilar pathology was noticed (Figure 1c). In computed tomography (CT) imaging, the patient had double superior vena cava. There was a normal formed right superior vena cava but LPSVC was detected at the anterior left side of the aorta. Left jugular and left subclavian veins merged and formed the LPSVC. Two of the vena cava had approximately the same diameter (Figure 1d). There was no contrast enhancement in left vena cava, because of right atrial communication (Figure 1a). In addition, long diameter of coronary sinus was 2.5 cm which was very large according to normal ranges (Figure 1b). In the report of echocardiography, no congenital anomaly was detected. Only they found a large coronary sinus connected with the right atrium and its roof was intact. In colored reconstruction images, LPSVC is connected with the coronary sinus. It goes between aortic arch and the vascular structures of the left pulmonary hilum and finally curves right and anteriorly to end in the right atrium. Atriums, ventricles and aorta were normal (Figure 2). The patient consent was taken before writing this case report.

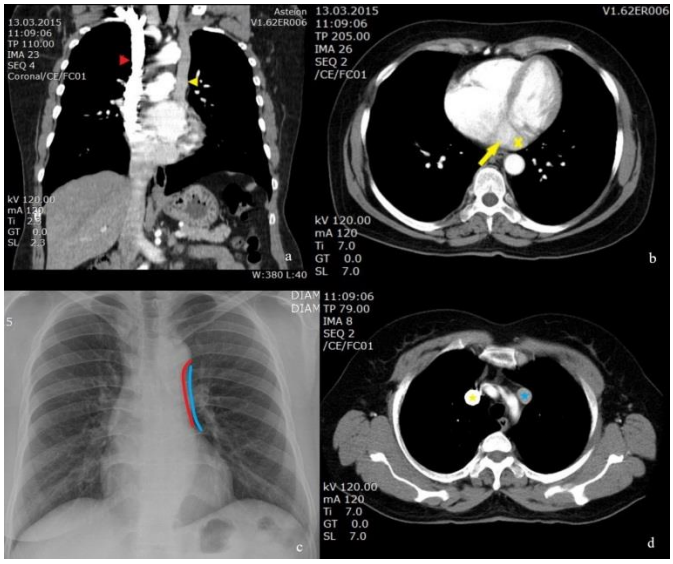

Figure 1: a: Right vena cava (red arrow head) LPVCS (yellow arrow head), b: Coronary sinus (yellow arrow) PLVCS connection with coronary sinus (yellow cross), c: Border of descendant aorta (red line) PLVCS (blue line), d: Enhanced contrast of normal Vena cava (yellow star) unenhanced contrast of PLVCS (blue star)

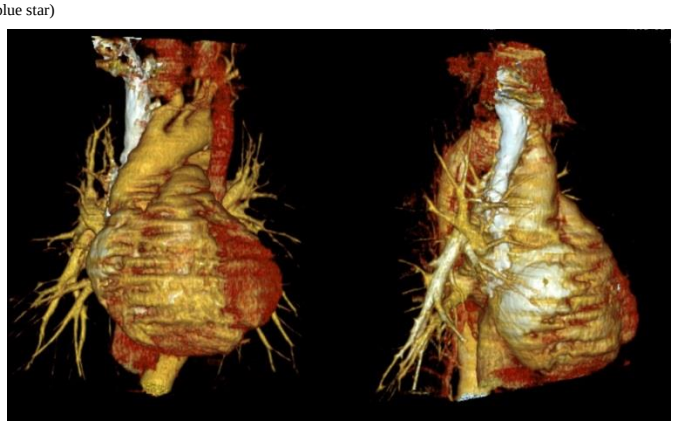

Figure 2: Double vena cava and normal configuration of heart, Right: Antero-posterior view of the heart, Left: Lateral view of heart

Discussion

LPSVC is a rare organogenesis abnormality [4]. The ratio of LPSVC is 0.3-0.5% in the general population [5]. It is due to the persistence of the terminal part of the left anterior cardinal vein, which normally involutes in the sixth month of uterine life [4]. The major venous drainage system of the embryo is constituted by the cardinal veins. The oblique anastomotic branch joins to the anterior cardinal veins and two of them forms left brachiocephalic vein. Normally the right anterior cardinal vein and the right common cardinal vein merge to form the SVC while the left anterior cardinal vein and the left common cardinal vein must undergo atrophy. Patency of embryonic veins lead to multiple variations for example double SVC, double inferior vena cava (IVC) or other venous connections which don't normally exist [6]. The PLSVC represents failure of obliteration of the left anterior cardinal vein in early embryological development [7].

In 90% of cases the persistent left-sided SVC connects to the right atrium via the coronary sinus, 10% the left SVC connects to the left atrium causing a right-to-left shunt. The variant linked with left atrium has potential hazard of systemic embolization of thrombus or air [7]. Cardiac anomalies accompanying LPSVC usually are a shunt type. The most commonly associated cardiac abnormality is an atrial septal defect (ASD). Others include Fallot's tetralogy, coarctation of the aorta, pulmonary stenosis and interventricular septal defect (VSD). If a double SVC is not associated with other congenital cardiac abnormalities, it is usually asymptomatic and hemodynamically insignificant [7]. LPSVC is the most common congenital venous anomaly of the thorax that causes a dilated CS [2]. As mentioned above, in the absence of congenital heart disease, LPSVC drains into the right atrium through the coronary sinus which expands because of hemodynamically overload [3].

Enlargement of the CS can be part of an anatomical variation like LPSVC or congenital anomaly like total anomalous pulmonary venous return, coronary atrioventricular fistula and anomalous hepatic venous drainage causing anomalous venous overload into the coronary sinus. It can also result from a wide spectrum of conditions causing right atrial dilatation, including tricuspid stenosis, tricuspid regurgitation, right ventricular dysfunction, and pulmonary hypertension [6,8]. Rare causes of dilated CS include postoperative obstruction, thrombosis and unroofing of the sinus [8].

In our paper we described the incidental discovery of a dilated CS due to a PLSVC in 56 years old female during CT screening for respiratory failure, in purpose to highlight the importance and usefulness of combining non-invasive cardiovascular examinations to make an optimal diagnosis and illustration of congenital cardiovascular variations.

PLSVC is often present in asymptomatic individuals and is discovered incidentally during cardiovascular imaging, device implantation, or surgery [3]. Although it is a benign condition, PLSVC has important clinical implications. From one hand, it can be associated with a variety of congenital malformations of the heart and great vessels, and in the other hand it may technically complicate some endovascular and surgery procedures.

Knowing the anatomy of the coronary sinus (CS) and cardiac venous drainage is important because of its relevance in electrophysiologic procedures and cardiac surgeries. Several procedures make use of the CS, such as left ventricular pacing, mapping and ablation of arrhythmias, retrograde cardioplegia for coronary artery bypass, targeted drug delivery, and stem cell therapy therefore it is primordial to detect the dilatation of CS before the intervention [6,8].

Otherwise, when the left subclavian vein is used as access, serious complications such as arrhythmias, shock and coronary sinus thrombosis may occur. Furthermore, pacemaker (PM) or implantable cardioverter defibrillator (ICD) placement can be difficult and dangerous [3]. Consequently, it is beneficial for physicians to interpret the results of cardiac CT examinations in order to identify normal variants and congenital anomalies and to understand their clinical importance [6].

In our case, the patient presented a heavy respiratory symptomatology without any diagnosis since all of the respiratory tests were normal. After realization of a CT, PLSCV had been discovered inducing a huge dilatation of CS. In the literature, using contrast-enhanced non ECG-gated chest CT, the mean diameter of the CS was reported as 7.05 mm ± 1.90 [6]. In our patient it was tree time more and this, in the absence of any congenital heart disease. It seems obvious that noninvasive investigation like CT make a considerable contribution for showing cardiovascular variations when other tests have failed to put a diagnosis.

Conclusion

PLSVC is a rare congenital variation. If not associated with congenital heart disease, it is benign condition without any clinical manifestation. The originality of our case resides in the fact that our patient presented with respiratory insufficiency when all of the tests were normal. The realization of a CT made the diagnosis of a PLSVC with a much dilated CS and in the absence of a congenital heart disease. This finding was highly important since it is necessary to take some precautions before risky interventions.

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