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Report of persistent left superior vena cava associated with Ebstein's anomaly of tricuspid valve

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Case Report

Abstract

BACKGROUND: Ebstein's anomaly is one of the challenging congenital heart diseases (CHDs) that is presented with different anatomical and clinical symptoms. For this reason, patients with this complication require exact diagnostic methods and appropriate treatment approaches. In addition, multiple cardiac defects accompany this anomaly.

CASE REPORT: In this study, we describe a rare associated defect in an adult patient with Ebstein's anomaly and proper surgical methods performed for her.

CONCLUSION: The most important issue in adult patients with Ebstein's anomaly is to pay close attention to the presence of associated anomalies, in which careful examination and use of para-clinical methods is very helpful. Furthermore, accurate diagnosis of the associated defects determines the treatment and surgery of patients.

Keywords: Ebstein Anomaly; Heart Defects; Congenital; Left Superior Vena Cava

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Introduction

Ebstein's anomaly is a rare congenital anomaly of the heart which accounts for less than 1% of all patients with congenital heart defects.[1] This disease is present at birth, but depending on the severity of the disease and type of associated defects, the patients have different signs and symptoms that can appear in any ages.[2] Although all congenital heart disorders might associate with Ebstein's anomaly, mitral valve prolapse and left ventricular non-compaction (LVNC) are the two most common disorders which might be seen with Ebstein's anomaly.[3]

Additionally, in patients with Ebstein's anomaly, based on variation in anatomy and physiology, different medical and surgical treatments can be adopted. In this study, we described the symptoms and results of the para-clinical study of a rare association of persistent left superior vena cava (PLSVC) connection to left atrium (LA) in an adult patient with the Ebstein's anomaly for whom surgical management was performed.

Case Report

A 24-year-old woman who was a known case of Ebstein's anomaly referred to our clinic for better evaluation and management. She complained from exacerbation of dyspnea and cyanosis since two months ago. At the first visit, oxygen saturation (SO2) was 88% in upper and lower extremities and S3 and systolic ejection murmur was heard in heart auscultation. Central cyanosis and clubbing were evident. Mild cardiomegaly with increased pulmonary vascular marking was seen in chest X-ray (CXR) (Figure 1A). Right axis deviation (RAD), incomplete right bundle branch block (RBBB), and first-degree atrioventricular block (AVB) were electrocardiographic (EKG) findings (Figure 1B).

The EKG showed situs solitus, normal continuity of inferior vena cava (IVC) to right atrium (RA), D-loop ventricle, normal left ventricle (LV) size with mild systolic dysfunction (45%), abnormal septal motion, severe anatomical right ventricle (RV) enlargement with moderate systolic

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dysfunction, apical displacement of tricuspid valve (TV) leaflet (5.5 cm) with tethered and sail-like appearance of anterior TV leaflet suggestive of Ebstein anomaly resulting atrialization of RV and moderate to severe tricuspid regurgitation (TR), functional to anatomical RV ratio of about 40%, normally related great artery, moderate sized atrial septal defect (ASD) with right to left shunt, and PLSVC. In contrast echocardiography via left hand, we observed contrast agent in the LA immediately after injection and respectively via right hand, RA and then LA visualized after injection.

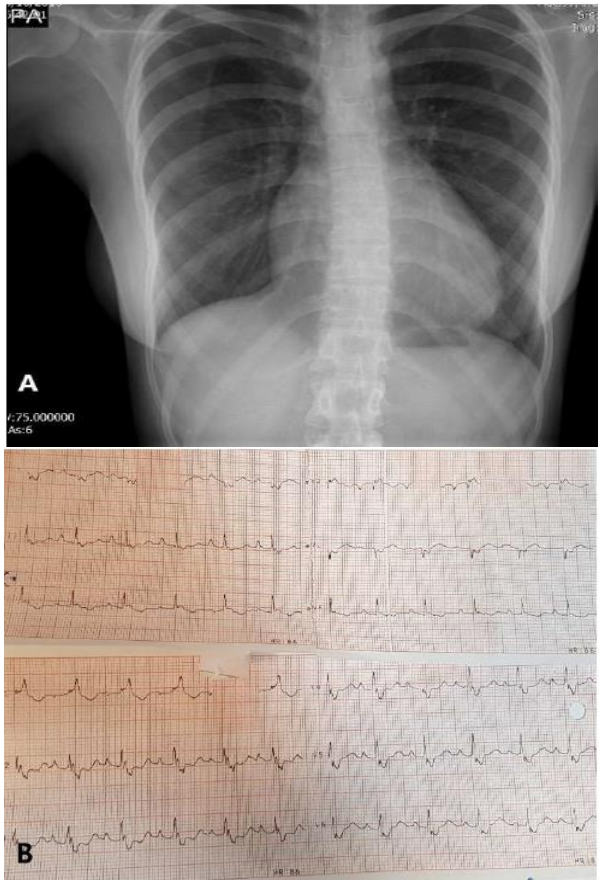

Figure 1. A) Chest X-Ray showed mild cardiomegaly and shunt vascularity; B) incomplete right bundle branch block (RBBB) and first degree atrioventricular block (AVB) in electrocardiography

With regard to inconsistency of symptoms and echocardiography findings, congenital computed tomography (CT) angiography was performed and showed PLSVC without bridging vein that connected to LA (Figure 2), right superior vena cava (RSVC), IVC, coronary sinus (CS) connected to RA, and unroofed CS (Figure 3).

These finding were confirmed with congenital catheterization. (Figure 4).

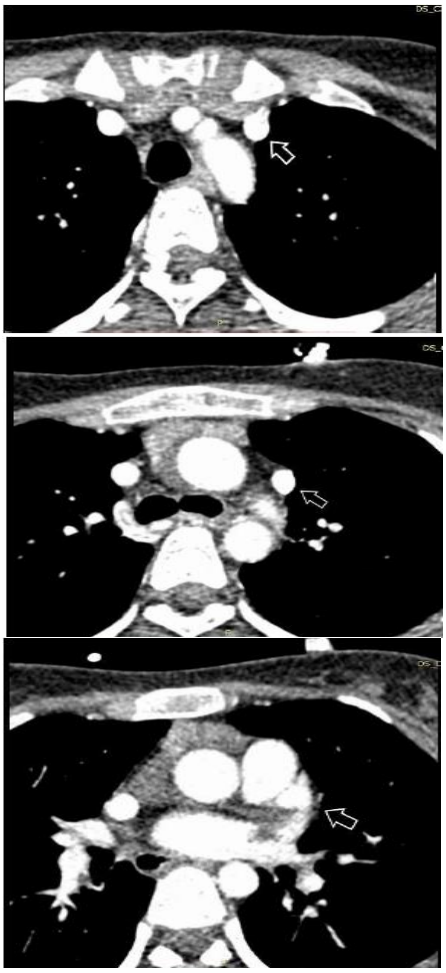

Figure 2. Persistent left superior vena cava (PLSVC) course that connected to left atrium (LA) (arrow)

Due to the special anatomy in this patient and the high volume of the right-to-left shunt and the fact that the patient's symptoms could not be controlled by medical treatment, the surgical method was considered for her.

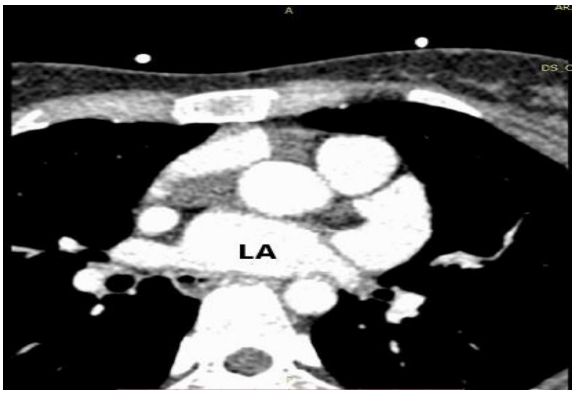

Figure 3. Left atrium (LA) seen in computed tomography (CT) angiography without unroofed coronary sinus (CS)

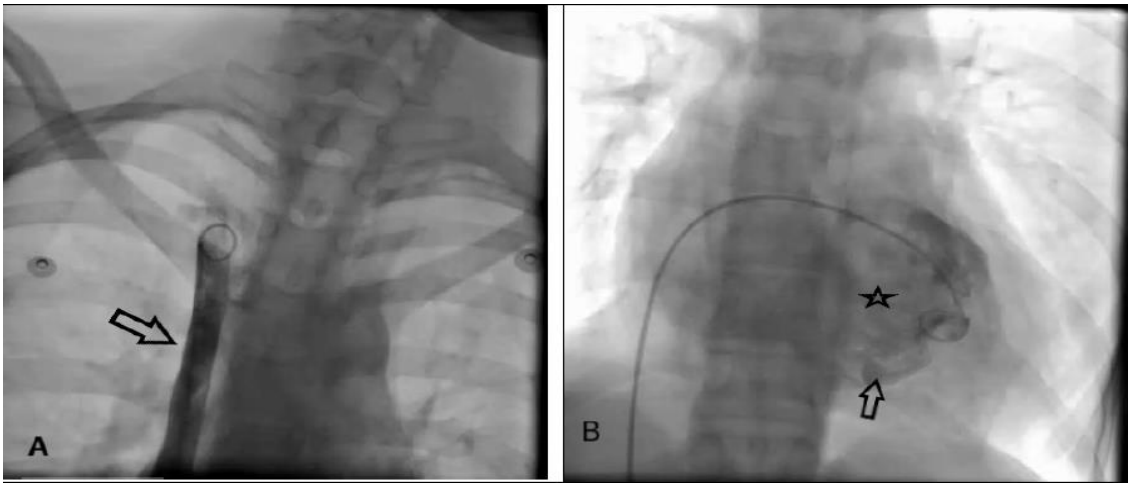

Figure 4. A) Persistent left superior vena cava (PLSVC), arrow, was seen without bridging vein; B): large atrialized right ventricle (RV), stars, and displacement of septal tricuspid valve (TV) leaflet, arrow

In the surgical procedure chosen for the patient, first the ASD was repaired and then the PLSVC was cut from the LA side and the left Glenn shunt was inserted on the same side to divert the draining of PLSVC to the left pulmonary artery. In this technique, SVC is disconnected from the heart and anastomosis to pulmonary artery unsaturated blood that comes from head and neck via SVC and directly drains to pulmonary artery and then lungs. By choosing this method for the patient, we diverted the poor oxygen blood in PLSVC that came from the head and neck to the left atrium directly to the left pulmonary artery and then to the lung, and this, along with closing the atrial septal defect, eliminated the patient's cyanosis and improved functional class.

On a follow up visit one month after surgery, the patient's dyspnea and cyanosis improved and SO2 was increased to 93%.

Discussion

Among CHDs, Ebstein's anomaly is a special type of TV involvement which manifests itself in the form of downward displacement of septal leaflet and atrialized RV.[4]

As mentioned earlier, this anomaly can be associated with any of the congenital diseases. In addition to the common defects mentioned earlier, ASD, patent foramen oval (PFO), ventricular septal defect (VSD), and pulmonary stenosis or atresia are other structural abnormalities often seen in Ebstein's anomaly. Furthermore, 39% of patients suffer from left heart myocardial or valvular involvement.[5,6]

Our recent patient who is described in this paper had very rare associated anomaly in which PLSVC was connected to LA which caused an extra shunt other than ASD. This anomaly was not diagnosed at birth and led to severe cyanosis and clubbing due to large amounts of right to left shunt.

The commonest symptoms of Ebstein's anomaly are cyanosis, arrhythmia, dyspnea due to right-sided heart failure and often sudden cardiac death. Nevertheless, the age at which patients become symptomatic and the type of symptoms depend on some factors including anatomical variation and degree of right-to-left interatrial shunting.[7]

In adult patients, the manifestations of the disease also depend on ventricular function and associated abnormalities, which makes the diagnosis of the disease in this age group challenging.[8] Several diagnostic methods such as echocardiography, CXR, and CT angiography are used to evaluate and determine the prognosis of these patients.[9] Likewise, due to the inconsistency of clinical symptoms (depressed level of o2 saturation) with echocardiographic findings (size of ASD), we used other diagnostic approaches to detect other associated anomalies. This demonstrated the connection of PLSVC to LA that can cause extra shunt on the atrial level and justify the patient's symptoms.

Another considerable point in these patients is deciding on therapeutic interventions which are of usually one time use for patients with regard to the symptoms and results of the diagnostic methods. For asymptomatic patients without right-to-left shunting or mild cardiomegaly, observation alone is recommended, however in symptomatic patients for whom evidence of worsening exists, surgery is preferred.

In spite of any symptoms in patients such as increased cardiomegaly, reduction of left ventricle

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systolic function, evidence of cyanosis or paradoxical emboli, the physician must decide based on a thorough evaluation to nominate the patient for surgery.[8]

Our patient was regularly followed up, but progressive symptoms led to another wide evaluation and we chose surgical treatment for her.

With regard to the different variations in anatomical and physiological defect in this anomaly, it is very important to choose the appropriate surgical method. Furthermore, surgical approaches are complex and different and must be decided on a case-by-case basis, and therefore it is necessary to perform a complete evaluation with different diagnostic methods before making any decision.

One of the surgical methods is biventricular repair in combination with the correction of all associated cardiac defects, which leads to good results in midterm.[10] Another surgical approach is a 1.5 ventricle repair which can be useful in patients with failing right ventricle and patients with severe biventricular dysfunction candidate for heart transplantation.[8] In our case, since the patient could not benefit from the closure of ASD alone because of the presence of another shunt from PLSVC to LA, we had to perform left Glenn shunt in addition to ASD closure.

Conclusion

The most important issue in dealing with these patients is to pay close attention to the presence of associated anomalies, in which careful examination and use of para-clinical methods is very helpful. Furthermore, accurate diagnosis of associated defects determines the treatment and surgery of patients.

Thus, these patients should be closely and regularly monitored by a cardiologist with expertise in CHDs to have a better quality of life (QOL) with timely diagnosis and appropriate medical treatment or surgery, and to find a suitable survival.

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Conflict of Interests

Authors have no conflict of interests.

Authors' Contribution

MA, ZK managed the patient, MA drafted the paper, ZK revised the draft. All authors read and approved the final version of the paper.

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