Levoatrial Cardinal Vein: In two siblings with normal heart morphology

Ayşe Şimşek[1], Tulay Demircan[2], Fatma Sarioğlu[3], Sedat Bağlı [4], Engin Gerçeker [5], Kaan Yıldız [6], cem karadeniz[7], and Nazmi Narin[8]

[1]İzmir Democracy University
[2]Turkiye Cumhuriyeti Saglik Bakanligi Izmir Tepecik Egitim ve Arastirma Hastanesi
[3]Izmir Health Sciences University, Tepecik Education and Research Hospital, Department of Radiology, Division of Pediatric Radiology, Izmir / TURKEY
[4]Izmir Health Sciences University, Tepecik Education and Research Hospital
[5]Buca Seyfi Demirsoy Training and Research Hospital
[6]Izmir Health Sciences University, Tepecik Education and Research Hospital,
[7]Izmir Katip Celebi University
[8]Izmir Katip Celebi University, Department of Pediatric Cardiology

September 25, 2021

Abstract

Levoatrial cardinal vein (LACV) is anomalous connection between the left atrium or pulmonary veins and any systemic vein which is derived from cardinal venous system. Presence of the levoatrial cardinal vein without a cardiac anomaly is a very rare congenital anomaly of the systemic venous return. In the literature, no LACV anomaly was found in two siblings who were asymptomatic and did not have an additional cardiac anomaly. Therefore, we present two cases (two siblings) the symptoms, diagnosis (the echocardiographic finding, computed tomography (CT) and aniographic images) and treatment modalities of isolated levoatrial cardinal vein.

INTRODUCTION

Levoatrial cardinal vein (LACV) is an abnormal connection between the left atrium or pulmonary veins and any systemic vein which is derived from cardinal venous system (1). Most often, this anomaly has been described in the context of left-sided obstructive cardiac lesions such as mitral atresia or hypoplastic left heart (2). It has been reported to be seen together with left heart pathologies and additionally anomalies such as double –outlet right ventricle, tetralogy of Fallot (3), ventricular septal defect and atrioventricular septal defect. Presence of the levoatrial cardinal vein without a cardiac anomaly is a very rare congenital anomaly of the systemic venous return.

In the literature, no LACV anomaly was found in two siblings who were asymptomatic and did not have an additional cardiac anomaly. Therefore, we present two cases (two siblings) the echocardiographic finding, computed tomography (CT) and aniographic images who were diagnosed with levoatrial cardinal vein and did not have any additional cardiac pathology. At the same time, we wanted to share the information that genetic tests are planned due to this rare anomaly in two siblings.

CASE 1

An 11-year-old male adolescent He applied to the pediatric cardiology outpatient clinic in order to obtain

a health report for participation in sports activities. No cardiac pathological finding was detected during the physical examination of the patient. Transthoracic echocardiography showed (TTE) a pattern of flow towards innominate vein, that was not appropriate for the pulmonary vein and other cardiac structural structures were normal. Computed tomography (CT) was performed for further evaluation of the anomalous communication between innominate vein and left atrium. Computed tomography clearly demonsrated an anomalous vessel joining the innominate vein and the left atrium**(Fig 1**). Catheter anjıography was performed. During angiopraphy when a radiopaque contrast agent was administered to the innominate vein, it was observed that the solution filled the LACV first and then the left atrium **(Fig 2).** Initially, closure of the patient was attempted with a vascular plate, but the closure procedure was unsuccessful. Therefore, LACV ligation was applied to the patient surgically. The patient is currently being followed up without any problem.

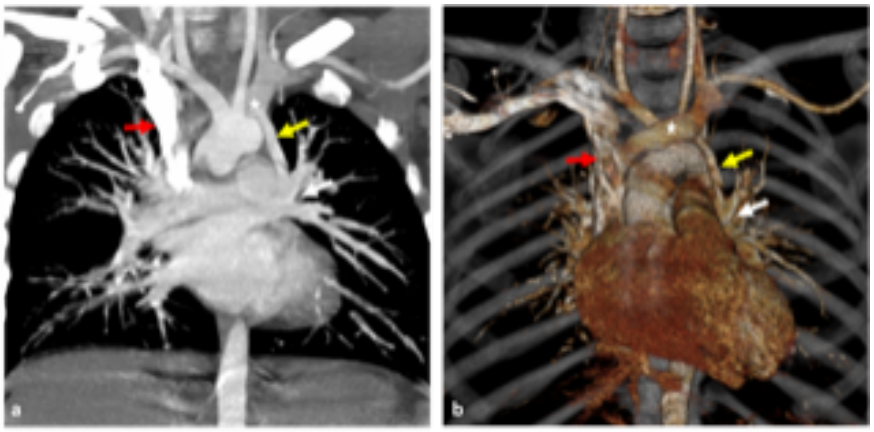

Fig. 1 . Coronal-reformatted CT image (a) and volume-rendered CT image (b) show levoatriocardinal vein (yellow arrow) connecting junction of left atrium and left superior pulmonary vein (white arrow) to innominate vein (white star). Superior vena cava is also seen (red arrow).

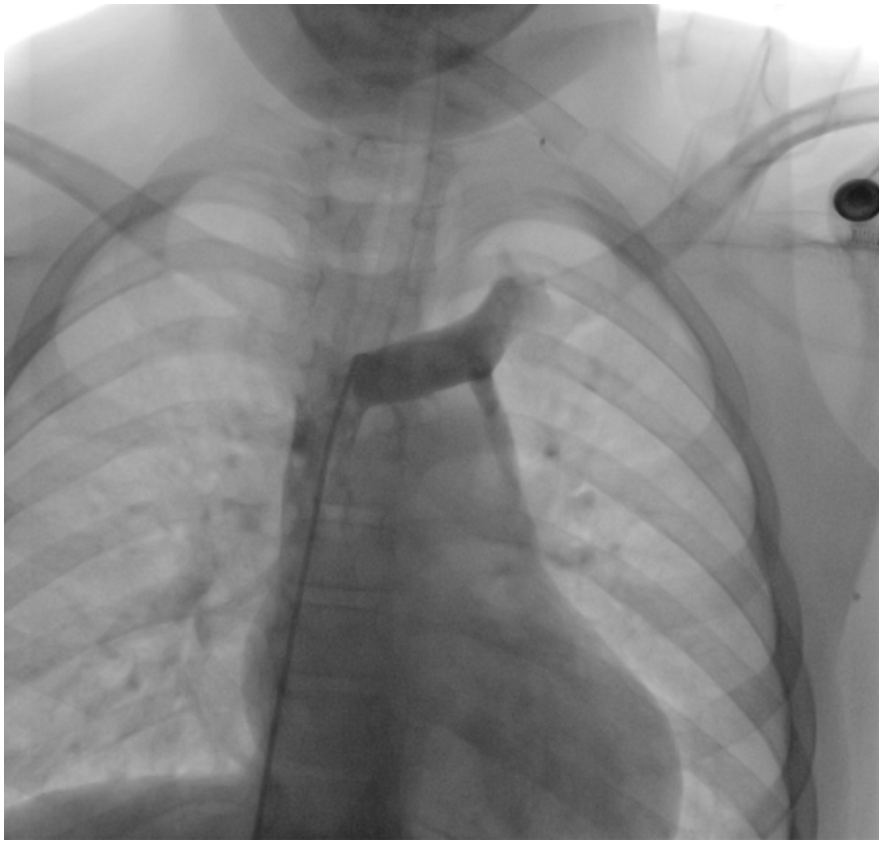

Fig.2. Catheter angiography view. Levoatrial cardinal vein (white arrow) ,innominate vein (red arrow)

CASE 2

5 years old male patient. Since his brother was diagnosed with LACV anomaly, he was brought to the pediatric cardiology outpatient clinic for control purposes. Physical examination was normal and no cardiac pathology was detected. That's why transthoracic echocardiography (TTE) was done. Transthoracic echocardiogram showed an anomalous vein communicating between the innominate vein and the left atrium. The patient's pulmonary venous return was normal, and other cardiac morphological structures were also normal. Computed tomography (CT) was performed for further evaluation of the anomalous communication. This showed anomalous vessel between the pulmoner vein and the innominate vein, was compatible with LACV **(Fig 3).** Cardiac catheterization was performed and revealed a left to right shunt. Radiopaque contrast agent was administered to the innominate vein, it was observed that the solution filled the LACV first and then the left superior pulmonary vein **(Fig 4).**

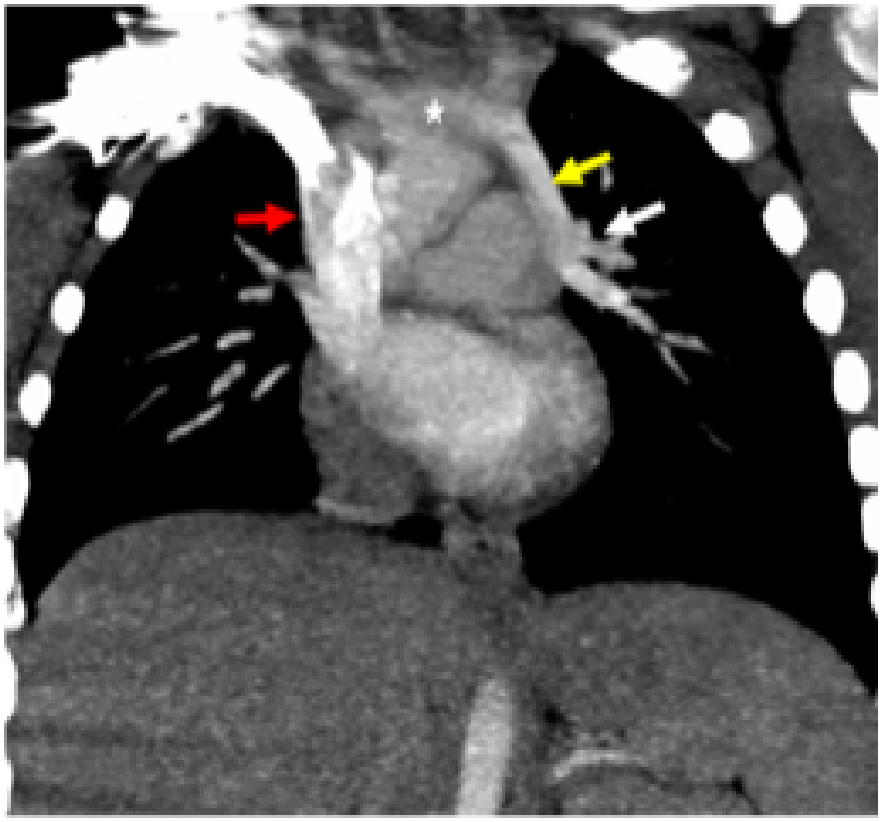

Fig. 3. Coronal-reformatted CT image (a) demostrates levoatriocardinal vein (yellow arrow) connecting left superior pulmonary vein (white arrow) to innominate vein (white star). Superior vena cava is also seen (red arrow).

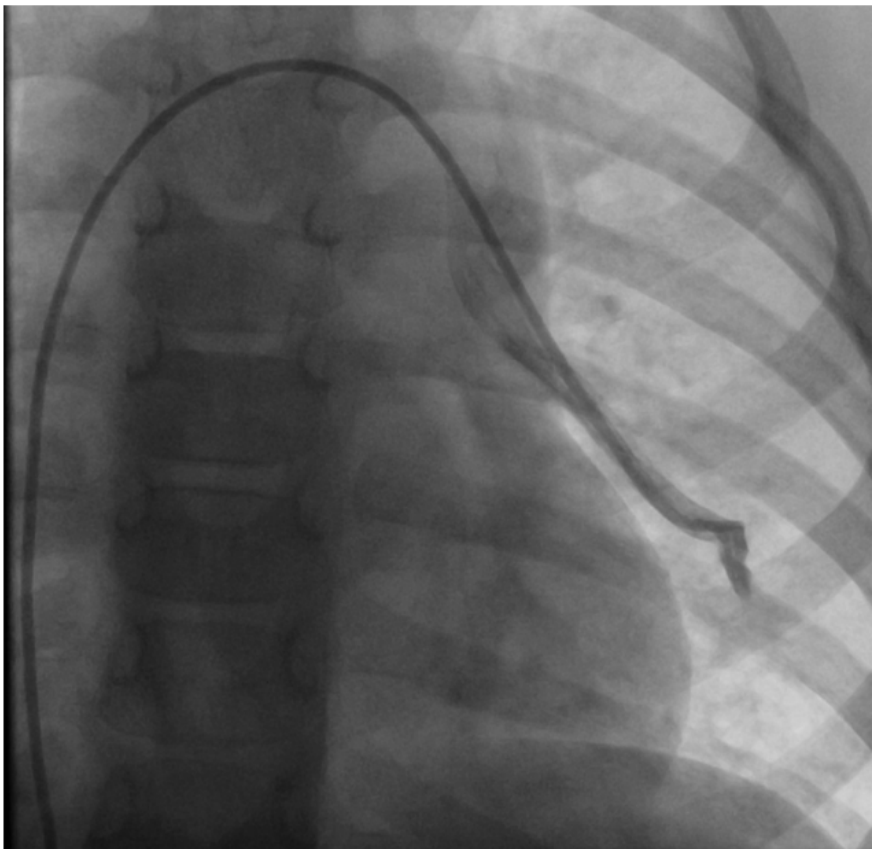

Fig.4. Catheter angiography view. İnnominate vein (red arrow), levoatrial cardinal vein (white arrow)

Since this rare anomaly was seen in two siblings, they were referred to the genetics department in terms of genetic anomalies.

DISCUSSION

There are many cases of LACV reported with left sided obstructive lesion. The largest study was done by Bernstein et al, who reported 25 patients of LACV with left sided obstructive lesions suc as mitral atresia, aortic stenosis, aortic coarctation and cor triatrium (4) In addition, LACV was reported as a concomitant anomaly in 3 patients with ventricular septal defect (VSD) ,atrial septal defect (ASD)-related shunts and pulmonary HT (5). Fujiwara et al reported a case of LACV in a boy being operated for tetralogy of Fallot. However, presence of LACV is very rare anomaly in the setting of normal heart morphology. Both of our cases did not have a concomitant cardiac pathology.

In addition to TTE and angiographic examinations, CT examination is also used in diagnosis. In the literature, the definitive diagnosis of venous return anomalies, which cannot be distinguished by TTE and angiographic examinations, has been clarified with CT scanning (6). In another case reported by Genç et al. LACV was detected incidentally during CT examination. (7). In each of our cases, CT was used to confirm the diagnosis.

Patients with LACV may present symptomatically. Symptoms may be dyspnea, tachypnea, pulmonary congestion findings, or systolic ejection murmur depending on the accompanying cardiac pathology and the amount of shunt. Rarely, it may be asymptomatic (7). In our cases LACV was detected incidentally and was asymptomatic.

LACV causes shunting between the pulmonary venous circulation and the systemic venous circulation, usually between the left atrium and the innominate vein, rarely between the pulmonary vein and the superior vena cava (SVC). In our first case, the direction of the shunt was from the innominate vein to the left atrium. In our second case, the flow direction was from the pulmonary vein to the innominate vein. In the literature, treatment modalities are selected depending on the amount and direction of the shunt flow. Transcatheter occlusion (8) and surgical correction treatments are applied (3),(5). In our first case, since the flow direction was from the innominate vein to the left atrium, surgical closure was preferred using the double ligation technique in terms of paradoxical embolism and infection risk. In our second case, it was decided to wait for treatment because the patient was young and asymptomatic..

In conclusion, **,** this report described a very rare case of LACV without any other cardiac abnormality. At the same time, the association of this rare anomaly in two siblings has not been reported in the literature. LACV may rarely be asymptomatic. If symptomatic, patients can also be treated noninvasively with device closure or surgical ligation.

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