ORIGINAL ARTICLE

A rare pathology: Levoatriocardinal vein

Nadir bir patoloji: Levoatriyokardinal ven

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ABSTRACT

Objective: Levoatriocardinal vein (LACV) is a rare cardiac pathology that represents a connection between the pulmonary venous and cardinal systems. The aim of the present study was to discuss morphological and clinical characteristics, as well as diagnostic methods, of experience with LACV.

Methods: Records of 11 patients (4 male, 7 female; mean age 79±1.83 days; range 1-390 days) diagnosed with LACV between 2010 and 2014 were retrospectively reviewed. Presence of LACV was confirmed with echocardiography. The primary obstructive lesion associated with cardiac defects and the integrity of the interatrial septum was identified in each patient with left-sided obstruction.

Results: Mean weight was 4.4±0.4 kg (range: 2–8). Age at presentation was under 1 year in 82% of patients. Nine patients had left-sided obstruction, and 2 had normal intracardiac anatomy and pulmonary venous return. In patients with left-sided obstruction, LACV was initially demonstrated with echocardiographic evaluation, performed in apical 4-chamber, high parasternal, and subcostal views. Atrial septum was restrictive or intact in patients with left-sided obstructions. LACV originated directly from the left atrium in all patients.

Conclusion: Levoatriocardinal vein is an extremely rare cardiac pathology, presenting almost exclusively in patients with left-sided obstructive lesions. In patients with left-sided obstructions, LACV must be kept in mind. It may also present in patients with normal intracardiac anatomy and pulmonary venous return.

ÖZET

Amaç: Levoatriyokardinal ven, pulmoner venöz sistem ile kardinal sistem arasındaki bağlantıyı gösteren nadir bir kardiyak malformasyondur. Bu çalışmada, levoatriyokardinal veni olan olguların morfolojik, klinik özellikleri ve tanı yöntemleri tartışıldı.

Yöntemler: 2010-2014 yılları arasında levoatriyokardinal ven tanısı alan 11 hastanın (4 erkek, 7 kız; ortalama yaş 79±1.83 gün; dağılım 1-390 gün) kayıtları geriye dönük olarak incelendi. Levoatriyokardinal ven tanısı tüm hastalarda ekokardiyografik inceleme ile kondu. Primer obstrüktif lezyon, ilişkili kardiyak defektler ve interatriyal septumun durumu, sol taraf obstrüksiyonu olan her bir hasta için tanımlandı.

Bulgular: Hastaların, ortalama vücut ağırlığı 4.4±0.4 kg (dağılım, 2-8) idi. Başvuru esnasında hastaların %82'sinin yaşı bir yaşın altında idi. Hastaların 9'unda sol taraf obstrüksiyonu, 2'sinde normal intrakardiyak anatomi ve pulmoner venöz dönüş vardı. Sol taraf obstrüksiyonu olan hastaların tümünde levoatriyokardinal ven, yüksek parasternal ve subkostal pencereler ile apikal 4 boşluk bakıdan yapılan ekokardiyografik değerlendirme ile gösterildi. Sol taraf obstrüksiyonu olan hastalarda atriyal septum restriktif ya da sağlamdı. Levoatriyokardinal ven tüm hastalarda direk olarak sol atriyumdan kaynaklanıyordu.

Sonuç: Levoatriyokardinal ven neredeyse sadece sol taraf obstrüksiyonu olan hastalarda görülen oldukça nadir bir kardiyak patolojidir. Bu patoloji sol taraflı obstrüksiyonu olan hastalarda akılda tutulmalıdır. Ancak levoatriyokardinal venin normal intrakardiyak anatomi ve pulmoner venöz dönüşe sahip olan hastalarda da görülebileceği unutulmamalıdır.

Embryologically, levoatriocardinal vein (LACV) is a result of abnormal persistence of the splenic plexus. It was first described by McIntosh in 1926.[1] LACV is commonly associated with left-sided obstructive conditions such as mitral stenosis or atresia, aortic atresia, cor triatriatum, hypoplastic left heart syndrome (HLHS), and coarctation.[2] It provides an alternative exit for pulmonary venous blood. How-

Received: September 14, 2015 *Accepted:* November 25, 2015

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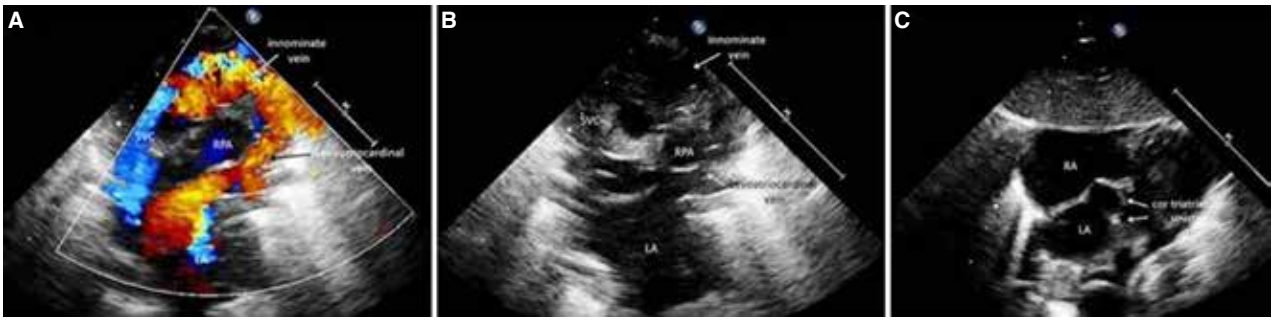

Figure 1. (A-C) Transthoracic echocardiographic image of patient with cor triatriatum sinister and levoatriocardinal vein.

ever, LACV can also present with normal intracardiac anatomy and pulmonary venous return.[3,4]

Abbreviations:

HLHS	*Hypoplastic left heart syndrome*
LACV	*Levoatriocardinal vein*
TAPVR	*Total anomalous pulmonary venous return*

Early infants may present with symptoms of pulmonary venous obstruction and low cardiac output.[2] In the present retrospective study, 11 cases with LACV were evaluated. Primary obstructive lesions and associated cardiac defects were examined, as was the role of atrial septal integrity in the formation of LACV.

METHODS

Records of 11 patients that had presented with LACV between 2010 and 2014 were retrospectively examined. Clinical presentation, physical examination, and demographic features were recorded in detail. Presence of LACV was diagnosed with echocardiography (Figure 1). In order to detail anatomy prior to surgery, 128-slice dual-source computerized tomography angiography was performed in 2 patients (Figure 2), and catheter angiography was performed in 5. Anatomic information was obtained from echocardiographic, angiographic, and tomographic records. Medical records were reviewed for details of the clinical presentations of these patients. Primary obstructive lesion, associated cardiac defects, and integrity of the interatrial septum was determined for each patient with left-sided obstruction. Clinical and anatomic features of the patient population are shown in Table 1.

As the present study was retrospective, consent was only obtained from the parents of each patient for

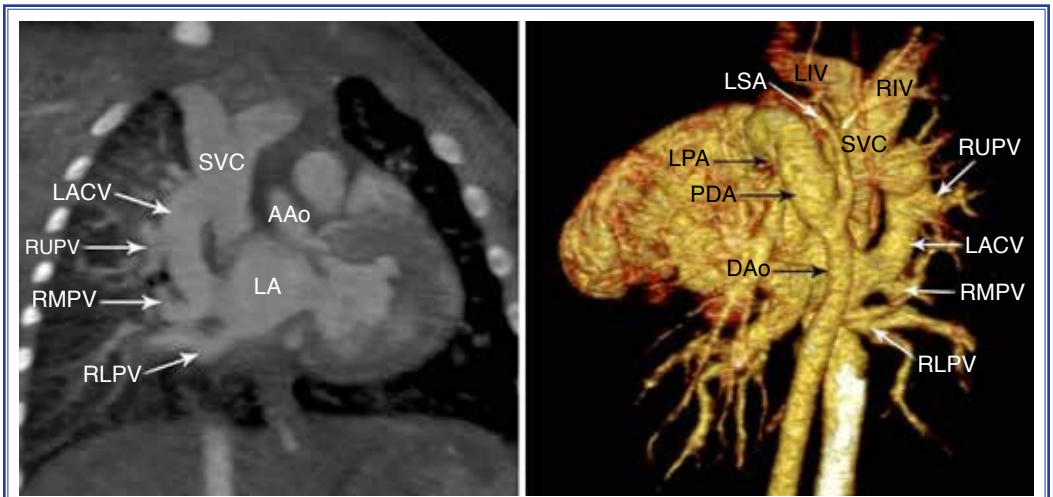

Figure 2. Computerized tomography angiographic image of patient with hypoplastic left heart syndrome and levoatriocardinal vein. LACV: Levoatriocardinal vein; SVC: Superior vena cava; AAo: Ascending aorta; LA: Left atrium; RLPV: Right lower pulmonary vein; RMPV: Right middle pulmonary vein; RUPV: Right upper pulmonary vein; LSA: Left subclavian artery; DAo: Descending aorta; PDA: Patent ductus arteriosus; LPA: Left pulmonary artery; RIV: Right innominate vein; LIV: Left innominate vein.

Table 1. Clinical and anatomic features of 11 patients with levoatriocardinal vein

Patient No	Age at presentation/ gender	Primary obstruction	Associated cardiac defects	Atrial septum	Origin of LACV	Drainage of LACV	Diagnosis	CT	Angio
1	4 months/M	None	None	Intact	LA	IV	Echo	No	No
2	13 months/F	None	None	Intact	LA	IV	Echo	No	No
3	4 months/F	Cortriatriatum	None	Intact	LA	RSVC	Echo	No	Yes
4	3 months/F	Cortriatriatum	VSD	Intact	LA	RSVC	Echo	No	Yes
5	2 months/M	Coarctation	PDA	PFO	LA	RSVC	Echo	Yes	Yes
6	2 days/M	AS, MA	PDA	PFO	LA	IV	Echo	No	No
7	1 days/F	AS, MA	PDA	PFO	LA	RSVC	Echo	No	No
8	4 days/M	AS, MA	PDA	Intact	LA	RSVC	Echo	No	No
9	23 days/F	AS, MA	VSD	Intact	LA	IV	Echo	No	No
10	1 month /F	TAPVR	VSD	PFO	LA	RSVC	Echo	No	Yes
11	5 month/F	Coarctation	CAVSD	PFO	LA	RSVC	Echo	Yes	Yes

AS: Aortic stenosis; CAVSD: Complete atrioventricular septal defect; F: Female; IV: Innominate vein; LA: Left atrium; M: Male; MA: Mitral atresia; PDA: Patent ductus arteriosus; PFO: Patent foramen ovale; RSVC: Right-sided superior vena cava; TAPVR: Total anomalous pulmonary venous return; VSD: Ventricular septal defect.

the conduction of general surgery during hospitalization and for each intervention and examination.

Statistical analysis

Distribution of variables were classified using SPSS software (version 11.5; SPSS Inc., Chicago, IL, USA). Descriptive results were also obtained using this program.

RESULTS

Patient characteristics

Clinical and anatomic findings are presented in Table 1. Mean age was 79±1.8 days (range: 1–390), mean weight was 4.4±0.4 kilograms (range: 2–8). Four patients were male; 7 were female. Age at presentation was under 1 year in 82% of patients. Nine had left-sided obstructions, and 2 had normal intracardiac anatomy and pulmonary venous return.

A 4-month-old and a 13-month-old asymptomatic patient were referred for innocent murmurs; they had normal intracardiac anatomy and pulmonary venous return. No important non-cardiac medical issues were present in these patients. LACV originated directly from the left atrium and drained to the innominate veins.

The 9 patients with left-sided obstructions were referred due to tachypnea, respiratory distress, and murmurs. HLHS with severe aortic stenosis and mitral hypoplasia accounted for the primary obstruction in 4 patients; cor triatriatum sinister (obstructive) was found in 2. Two patients had juxtaductal coarctation of the aorta, and 1 had a total anomalous pulmonary venous return (TAPVR). Of the patients with HLHS, atrial septal defect was restrictive in 2 patients, while 2 others had hemodynamically insignificant patent foramen ovale. The interatrial septum was intact in patients with cor triatriatum sinister. Patients with coarctation of the aorta and TAPVR had hemodynamically insignificant patent foramen ovale. This data is consistent with the hypothesis that LACV persists in the absence of non-restrictive interatrial communication in order to decompress pulmonary venous circulation.

Diagnostic imaging

In all patients with left-sided obstruction, LACV was echocardiographically demonstrated in apical 4-chamber, high parasternal, and subcostal views. LACV originated directly from the left atrium in all patients and drained directly to the right-sided superior vena cava in patients with coarctation of the aorta, TAPVR, and cor triatriatum sinister. Of the patients with HLHS, LACV drained to the left innominate vein in 2 patients and to the right-sided superior vena cava in 2 patients. Presence of LACV was also shown with echocardiography from the apical 4-chamber

and high parasternal views in patients with normal intracardiac anatomy.

Surgery and outcome

Norwood Stage I procedure and LACV ligation were performed in patients with HLHS. TAVPR and LACV repairs were performed in the patient with TAPVR. Left atrial membrane resection and LACV resection were performed in patients with cor triatriatum sinister. Coarctation repair and LACV ligation were performed in patients with coarctation of the aorta. Of the patients with HLHS, 3 underwent Norwood Stage II procedure, and 1 died on the tenth postoperative day due to sepsis. Patients with coarctation of the aorta, TAPVR, and cor triatriatum sinister are still being followed, without any complication. Patients with LACV and normal intracardiac anatomy remained asymptomatic in the follow-up period.

DISCUSSION

LACV is an extremely rare pathology that has only been described in case reports and case series.[1,2,5,6] The term was first used by Edwards and DuShane, who described LACV as the physiologic phenomenon that decompresses the left atrium through the cardinal venous system.[7] In most cases, left-sided obstructive lesions were observed, and LACV was the only form of communication between the left and right atriums. In cases of left-sided obstructive lesions, LACV was the only means of unloading the left heart. It was thought that high left atrial pressure caused patency of the splenic plexus, which is a natural connection between the cardinal system and the pulmonary veins in the early intrauterine period.[2] Paudel G et al. reported cases with HLHS, intact atrial septum, and LACV.[8] Only 2 cases without intracardiac pathology and normal pulmonary venous return have been reported.[3,4]

LACV should also be considered a distinct entity from left superior vena cava. These entities can be differentiated by examining the direction of blood flow using echocardiography or other imaging techniques. In cases of LACV, blood flows from the left atrium to the innominate vein, whereas left superior vena cava drains to the coronary sinus.[5]

A retrospective study conducted by Bernstein HS et al. in 1995[5] featured 12 cases of left-sided obstructions, including HLHS, coarctation, TAPVR, and cor triatriatum, and 1 case of normal intracardiac anatomy and pulmonary venous return, all of which had presented between 1980 and 1994. The authors also reviewed 12 patients with left-sided obstructive lesions. In the series, patients had proximal levels of left-sided obstructions, as in the present study. Interatrial septum was intact or restrictive in 21 of the patients with left-sided obstructive lesions, and LACV was the obligatory means of unloading the left heart. Four patients had hemodynamically significant atrial septal defect. The authors concluded that restrictive interatrial septum is not necessary for LACV formation, but that non-restrictive interatrial septum carries the patient until 2 years of age, maintaining asymptomatic status. Rauch et al. reported the case of a 10-week-old infant with Shone's complex, nonrestrictive atrial septum, and LACV.[9] Lee et al., Pinto et al., and Tůma et al. reported cases of patients with left-sided obstructive lesions and restrictive interatrial septums.[10–12] In these studies, all patients were symptomatic before 2 years of age. All patients with left-sided obstructions in the present study had restrictive or intact interatrial septums, and LACV was the only means of decompressing. These patients were also symptomatic before 2 years of age. In a study conducted by Lee et al., a 15-year-old patient with normal intracardiac anatomy and pulmonary venous return was incidentally found to have LACV on angiography when he presented for evaluation regarding left-to-right shunt.

Odemis et al. reported the case of a 24-year-old patient suffering from fatigue. Through echocardiographic evaluation, intact interatrial septum, dilatation of the right atrium and ventricle, and an anomalous vein draining to the right-sided vena cava were revealed. Catheter angiography was performed with suspicion of abnormal partial pulmonary venous return. Normal pulmonary venous return and persistent LACV was shown with catheter angiography and magnetic resonance angiography, and surgery was successful. As the authors demonstrated, LACV can be isolated, and its clinical findings can mimic abnormal partial venous return and atrial septal defect. The present patients were asymptomatic because they were younger than 1 year. Patients with normal intracardiac anatomy and pulmonary venous return may show symptoms of low cardiac output and right heart volume overload during their lifetimes.[3] For this reason, the present patients are being followed with in-

termittent echocardiographic evaluation of right heart chamber dilatation and the other findings.

Conclusions

LACV is an extremely rare cardiac pathology, existing almost exclusively in patients with left-sided obstructive lesions that provide a mechanism for decompression of pulmonary venous circulation. Echocardiography provides a rapid, noninvasive, and inexpensive means of identifying LACV. LACV must be kept in mind in cases of left-sided obstructions, and may occur in patients with normal intracardiac anatomy and pulmonary venous return. Therefore, detailed examination must be performed in patients with low cardiac output and right heart volume overload in the absence of atrial septal defect, abnormal pulmonary venous return, or left-sided obstruction.

Conflict-of-interest issues regarding the authorship or article: None declared

REFERENCES

1. Eggleston C. The paroxysmal tachycardias. Med Clin North Am 1950;34:735–44.
2. Blieden LC, Schneeweiss A, Deutsch V, Neufeld HN. Anomalous venous connection from the left atrium to the cardinal venous system: "levoatriocardinal vein". AJR Am J Roentgenol 1977;129:937–8. Crossref
3. Odemis E, Akdeniz C, Saygili OB, Karaci AR. Levoatriocardinal vein with normal intracardiac anatomy and pulmonary venous return. Ann Pediatr Cardiol 2011;4:183–5. Crossref
4. Jaecklin T, Beghetti M, Didier D. Levoatriocardinal vein without cardiac malformation and normal pulmonary venous return. Heart 2003;89:1444. Crossref
5. Bernstein HS, Moore P, Stanger P, Silverman NH. The levoatriocardinal vein: morphology and echocardiographic identification of the pulmonary-systemic connection. J Am Coll Cardiol 1995;26:995–1001. Crossref
6. Amoretti F, Cerillo AG, Chiappino D. The levoatriocardinal vein. Pediatr Cardiol 2005;26:494–5. Crossref
7. Edwards JE, DuShane JW. Thoracic venous anomalies. Arch Pathol 1950;49:517–37.
8. Paudel G, Ng BY, Law IH. Hypoplastic left heart with intact atrial septum and levoatriocardinal vein: a challenge in identifying aortic arch branches. Cardiol Young 2015;25:171–3.
9. Rauch R, Grosse-Wortmann L, Jones J, Burrill E, Welisch E, Rosenberg HC, et al. Shone's complex and levoatriocardinal vein: a rare association. J Am Coll Cardiol 2012;59:698. Crossref
10. Lee ML, Wang JK, Lue HC. Levoatriocardinal vein in mitral atresia mimicking obstructive total anomalous pulmonary venous connection. Int J Cardiol 1994;47:1–4. Crossref
11. Pinto CA, Ho SY, Redington A, Shinebourne EA, Anderson RH. Morphological features of the levoatriocardinal (or pulmonary-to-systemic collateral) vein. Pediatr Pathol 1993;13:751–61. Crossref
12. Tůma S, Samánek M, Benesová D, Vorísková M. Premature closure of the foramen ovale with levoatriocardinal vein. Eur J Pediatr 1978;129:205–13. Crossref

Keywords: Child; echocardiography; levoatriocardinal vein.

Anahtar sözcükler: Çocuk; ekokardiyografi; levoatriyokardinal ven.