Case Reports

Perioperative Management of a Levoatrial Cardinal Vein

in the Absence of the Brachiocephalic Vein

Olcay Murat Disli, MD
Bektas Battaloglu, MD
Nevzat Erdil, MD
Cemsit Karakurt, MD
Ozlem Elkiran, MD

Levoatrial cardinal vein is a rare congenital anomaly of the systemic veins. It is commonly associated with left-sided obstructive conditions such as aortic atresia, mitral atresia, and cor triatriatum. We report the case of a 14-year-old boy who was undergoing surgery for correction of a secundum atrial septal defect. Intraoperatively, we discovered that he had a levoatrial cardinal vein and no brachiocephalic vein. However, collateral vessels provided adequate flow to the right atrium, and the patient's left-sided venous pressure was not excessive, so we ligated the levoatrial cardinal vein and directly repaired the septal defect. Postoperatively, the left venous drainage was satisfactory and the patient was asymptomatic. In addition to our patient's case, we discuss the embryology, diagnosis, and treatment of levoatrial cardinal vein. ***(Tex Heart Inst J 2013;40(2):201-3)***

Levoatrial cardinal vein, a rare congenital anomaly of the systemic veins, is often associated with left-sided obstructive conditions such as aortic atresia, mitral atresia, and cor triatriatum.[1] In this report of a patient who had a secundum atrial septal defect (ASD), we describe our intraoperative discovery and treatment of a levoatrial cardinal vein in the absence of a brachiocephalic vein. In addition, we discuss the embryology and diagnosis of this rare anomaly.

Case Report

In October 2011, a 14-year-old boy with a large secundum ASD was referred to our hospital. Chest radiographs showed cardiomegaly from right atrial and ventricular enlargement. Transthoracic echocardiograms showed a 32-mm secundum ASD without inferior and aortic rims. The location of the pulmonary venous opening and the size of the coronary sinus were normal. We decided to treat the ASD surgically.

At operation, the patient was placed on cardiopulmonary bypass. Intracardiac exploration through an oblique right atriotomy revealed that the venous blood from the upper left side was draining into the left atrium through a sizable vein connected to the left atrial appendage. This vein was confirmed to be a levoatrial cardinal vein. There was no brachiocephalic vein, and its absence had not been noticed at the time of cannulation, which had been completed in routine bicaval fashion.

The pulmonary veins and coronary sinus were in normal locations, and the superior vena cava (SVC) was of normal size. There was no left-sided obstructive condition or anomaly of the pulmonary veins. The vein connecting to the left atrial appendage was apparently draining the left subclavian vein.

We snared the levoatrial cardinal vein with a tape and temporarily obliterated its blood flow without consequent change in the patient's systemic oxygen saturation or blood pressure. His mean venous pressure increased from 8 to 14 mmHg. A perioperative cardiac angiogram showed an anomalous vein draining into the azygos vein, as well as 2 collateral connections between the levoatrial cardinal vein and the SVC. Accordingly, we double-ligated the levoatrial cardinal vein.

Our original surgical strategy was not affected by these anomalous connections: we closed the ASD directly without using a patch. At no time was cannulation affected by the anomalous systemic venous drainage.

The patient was weaned from cardiopulmonary bypass without inotropic support. His recovery was uneventful, with no cyanosis or edema of the left arm, and he

Key words: *Heart atria/abnormalities/surgery; heart defects, congenital/surgery; treatment outcome*

From: *Departments of Cardiovascular Surgery (Drs. Battaloglu, Disli, and Erdil) and Pediatric Cardiology (Drs. Elkiran and Karakurt), School of Medicine, Inonu University, 44280 Malatya, Turkey*

Address for reprints: *Olcay Murat Disli, MD, Department of Cardiovascular Surgery, Faculty of Medicine, Inonu University, Elazig Rd. (15 km), 44280 Malatya, Turkey*

E-mail: *olcaydisli@hotmail.com*

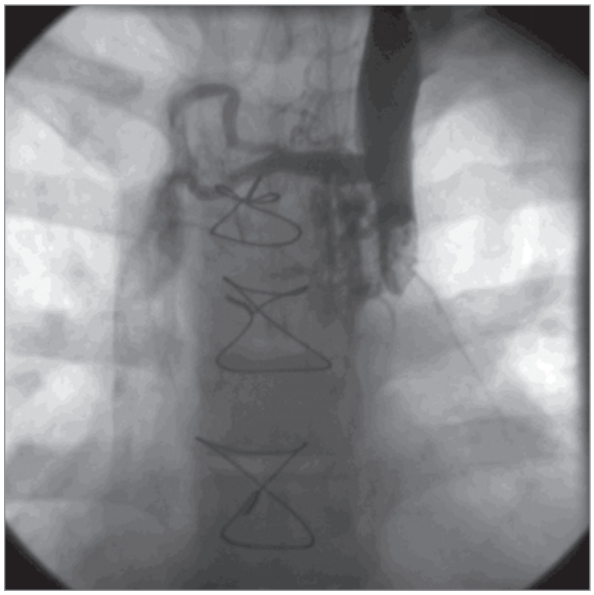

Fig. 1 *Angiogram after left subclavian vein injection shows a ligated levoatrial cardinal vein and 2 collateral venous connections between it and the azygos vein.*

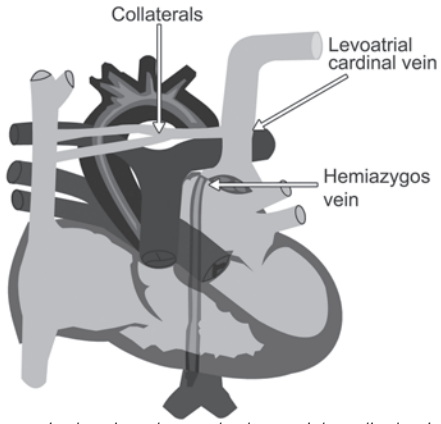

Fig. 2 *Schematic drawing shows the levoatrial cardinal vein connected to the left atrium without a brachiocephalic vein. The intercaval connection was provided by 2 collateral vessels.*

was discharged from the hospital 8 days after the operation. A postoperative angiogram showed that the left venous return drained into the SVC through the collateral veins (Figs. 1 and 2). Echocardiograms 2 months postoperatively showed regression of the right and left atrial dilation; also, the patient's effort capacity had increased.

Discussion

In the early development of the human embryo, the cardinal, vitelline, and umbilical veins exist collectively. Otherwise, the pulmonary vascular bed cannot drain directly into the heart. The pulmonary vascular bed is connected to the cardinal, vitelline, and umbilical venous systems via a splanchnic plexus, which surrounds the foregut. Almost all patterns of systemic or pulmonary venous embryologic abnormalities are explained by these venous connections. At this developmental stage, a brachiocephalic vein emerges and the left anterior cardinal vein, the so-called persistent left SVC, sequentially disappears. The levoatrial cardinal vein becomes permanent if the brachiocephalic vein is absent or small.[2,3] This abnormal vessel enables drainage of the pulmonary venous blood, especially in the presence of left-sided heart malformation. The clinical importance of this pathologic condition is well established: McIntosh,[4] and later Edwards and DuShane,[5] reported cases of levoatrial cardinal vein in patients with mitral atresia.

The surgical management of levoatrial cardinal vein involves simple ligation with or without intra- or extracardiac rerouting. After temporary occlusion, simple ligation is appropriate when left-side venous pressure does not exceed 30 mmHg and left-to-right venous communication is adequate.[6] Conversely, venous pressure over 30 mmHg indicates intra- or extracardiac rerouting. Our patient's pressure increased to 14 mmHg after temporary occlusion, so we performed simple ligation. In more typical instances of this anomaly, the levoatrial cardinal vein connects to the brachiocephalic vein and serves as a safety valve for left-sided venous drainage. Despite the absence of a brachiocephalic vein in our patient, simple ligation was still indicated, because his venous pressure rose only moderately. The postoperative angiogram showed satisfactory drainage, and the patient had no subsequent clinical symptoms.

Preoperatively, knowing the anatomic variations and determining the surgical approach are important. Therefore, when levoatrial cardinal vein is suspected, echocardiography should be supplemented with imaging methods such as multidetector computed tomography.[7]

In our patient, the conventional method of relying upon pressure measurements proved useful even in the presence of the variations that we found. Because of the normal SVC, the collateral vessels enabled flow to the right atrium after our use of simple ligation to correct the venous anomaly.

References

1. Sadler TW. Langman's medical embryology. Baltimore: Lippincott Williams & Wilkins; 1995. p. 219-24.
2. Raghib G, Ruttenberg HD, Anderson RC, Amplatz K, Adams P Jr, Edwards JE. Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus: a developmental complex. Circulation 1965;31:906-18.
3. Winter FS. Persistent left superior vena cava; survey of world literature and report of thirty additional cases. Angiology 1954;5(2):90-132.
4. McIntosh CA. Cor biatriatum triloculare. Am Heart J 1926; 1(6):735-44.

5. Edwards JE, DuShane JW. Thoracic venous anomalies. I. Vascular connection of the left atrium and the left innominate vein (levoatriocardinal vein) associated with mitral atresia and premature closure of the foramen ovale. II. Pulmonary veins draining wholly into the ductus arteriosus. Arch Pathol 1950; 49:517-37.
6. de Leval MR, Ritter DG, McGoon DC, Danielson GK. Anomalous systemic venous connection. Surgical considerations. Mayo Clin Proc 1975;50(10):599-610.
7. Chen K, Thng CH. Multislice computed tomography and two-dimensional echocardiographic images of cor triatriatum in a 46-year-old man. Circulation 2001;104(17):2117.