CASE REPORTS

AORTICO-LEFT VENTRICULAR TUNNEL ASSOCIATED WITH SINGLE CORONARY ARTERY IN AN INFANT

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Abstract: Aortico-left ventricular tunnel (ALVT) is a rare congenital malformation in which an abnormal communication between the aorta and the left ventricle (LV) bypasses the aortic valve. In infants, it usually presents with congestive heart failure. The clinical presentations mimic aortic regurgitation or ruptured sinus of Valsalva aneurysm into the LV. Progressive aortic regurgitation is a common problem in patients with ALVT. ALVT associated with a single coronary artery is extremely rare. We report the case of an infant who had congestive heart failure caused by ALVT. The diagnosis was made by echocardiography and angiography. In addition, a single coronary artery from the noncoronary cusp was found during surgery. Congestive heart failure resolved after successful surgical repair and he was asymptomatic throughout 2 years of follow-up. This case illustrates that early diagnosis of ALVT and corrective surgery can prevent aortic valve incompetence and is associated with a good clinical outcome.

Aortico-left ventricular tunnel (ALVT) is a rare congenital malformation characterized by a tunnel between the aorta and left ventricle (LV). ALVT was first described by Levy et al in 1963 [1], but there have been few reports from Asian countries [2, 3]. ALVT usually presents with congestive heart failure and occurs in infancy. Most patients will die without surgery [4–6]. Associated cardiac anomalies are common [5–7]. However, association with a single coronary artery has not been reported. We report the case of a 3-month-old infant who had ALVT with aortic regurgitation (AR) associated with a single coronary artery. He was successfully treated with surgery.

Case Report

A 1-day-old male newborn was referred for assessment of a heart murmur that had been noted immediately after birth. He was a full-term baby with normal pregnancy and delivery who weighed 3,000 g at birth. Physical examination revealed a non-cyanotic baby without distress. A unique grade III/VI to-and-fro murmur was heard at the left upper sternal border. There was no jugular vein engorgement, no hepatomegaly, and no leg edema. The rest of the physical examination was unremarkable.

Chest roentgenography demonstrated mild cardiomegaly (cardiothoracic ratio, 0.55). Lung markings were normal without pulmonary congestion. Electrocardiography revealed nonspecific ST-T changes. Two-dimensional echocardiography and color Doppler showed a small atrial septal defect (ASD), mild AR, and prominent sinus of Valsalva. There was a defect at the upper interventricular septum, but no flow communication between the LV and right ventricle (RV). Flow was noted from the aorta into the LV via this defect, but not through the aortic valve. ALVT or ruptured sinus of Valsalva aneurysm into the LV was suspected. He was treated with digoxin and a diuretic, and discharged in stable condition.
He was readmitted 3 months later due to congestive heart failure. Follow-up chest roentgenography revealed cardiomegaly (cardiothoracic ratio, 0.65). Repeat echocardiography in the parasternal long axis view demonstrated a tunnel with aneurysm dilatation between the LV and aorta. The tunnel arose from the region of the right aortic sinus and passed between the aortic valve and the outlet portion of the ventricular septum to terminate in the LV (Fig. 1). In addition, severe AR was noted. Cardiac catheterization and angiography were performed after informed consent was obtained from his parents. The findings on angiography (Fig. 2) were consistent with echocardiography. Because heart failure persisted despite inotropic support and diuretic therapy, surgical intervention was suggested.

Surgical repair of the lesion was performed under cardiopulmonary bypass. During surgery, a small ASD, dysplastic aortic valves with severe AR, and a tunnel from the aorta to the LV were confirmed (Fig. 3). A single coronary artery from the noncoronary cusp was also noted (Fig. 4). The ASD was closed. The tunnel was closed with a double pledget-buttressed suture and an aortoplasty was done to prevent distortion of the aortic valve. The postoperative course was uneventful and the patient was discharged 7 days after surgery. One year after surgery, electrocardiography showed normal sinus rhythm and chest roentgenography revealed normal heart size. Echocardiography demonstrated mild AR, and no flow through the tunnel was found. At this time, New York Heart Association functional status was improved from class IV to I. He remained asymptomatic without medication during 2 years of follow-up.

**Discussion**

ALVT is a rare congenital malformation in which an abnormal communication between the aorta and the LV bypasses the aortic valve. The tunnel usually arises...
Fig. 4. Schematic diagram showing a single coronary artery arising from the noncoronary cusp. ALVT = aortico-left ventricular tunnel; LCC = left coronary cusp; NCC = noncoronary cusp; RCC = right coronary cusp.

from the area of the right sinus of Valsalva and then passes anterior to the aorta, through the portion of interventricular septum, and into the LV immediately below the aortic valve. The typical presentation of ALVT is a to-and-fro murmur noted in the neonatal period. The natural course of ALVT depends on the degree of AR. In general, the age at presentation and the severity of the anomaly are related to the size of the tunnel and the severity of the regurgitation [8].

In the past, the standard diagnostic modality for ALVT was LV and ascending aorta angiography, which can demonstrate the course of the tunnel as well as the severity of AR. Sometimes, however, this method may not reveal the intracardiac portion of the tunnel, particularly when there is gross distortion of the LV outlet or association with severe AR [9]. With the aid of two-dimensional echocardiography and color Doppler, the diagnosis of ALVT can be made in the postnatal and prenatal periods [6, 8, 10–12]. The parasternal long axis view shows a septal dropout at the anterosuperior part of the interventricular septum, with a free communication area noted anteriorly in the RV outflow tract area ending just distal to the right coronary sinus of the aorta. In the parasternal short axis view, an echo dropout can be found at the level of the aortic valve. There is a crescent-shaped structure wrapping around the right coronary cusp anteriorly, clearly distinct from the aortic root [9]. Color flow mapping can confirm unobstructed to-and-fro flow through the structure [11].

The differential diagnosis of ALVT in infancy includes ventricular septal defect (VSD) with AR, ruptured sinus of Valsalva aneurysm into the LV, and coronary arteriovenous fistula [6, 8, 11]. Color Doppler shows a to-and-fro flow through the tunnel, which enables differentiation of ALVT from VSD with AR [11]. The aortic origin of the tunnel can be shown to be above the sinus and separate from the origins of both coronary arteries. This finding enables differentiation of the tunnel from a ruptured sinus of Valsalva aneurysm.

Associated anomalies have been described in several studies, including bicuspid aortic valve, aortic stenosis, patent ductus arteriosus, patent foramen ovale, pulmonary stenosis, absent right coronary artery, VSD and sinus of Valsalva aneurysm [5–7]. To our knowledge, association with a single coronary artery has not been described in the literature. In this case, selective coronary angiography was not performed; therefore, the anomaly of a single coronary artery was missed before surgery.

Surgical correction is the treatment of choice for ALVT. Various surgical techniques have been reported. Hovaguimian et al devised a surgical procedure that included direct closure of the aortic end of the tunnel, use of patch material to close the aortic end of the tunnel, or closure of both the aortic and the ventricular ends of the tunnel [5]. Surgical closure of the tunnel should not distort the aortic valve and should avoid the subpulmonary obstruction. Some authors have suggested that early surgery is associated with good outcome [7, 8]. However, it did not seem to prevent late deterioration of aortic competence in cases with preexisting aortic valve anomalies [13]. A follow-up study by the National Heart Institute in London demonstrated that surgical intervention in 50% of patients resulted in aortic valve incompetence with the necessity of valve replacement [13]. In this case, congestive heart failure subsided after surgery and mild AR was noted during follow-up. The growth and development of this patient were within normal limits after surgery, with only mild AR. However, long-term follow-up of aortic valve competence is mandatory in such patients.

References

Aortico-left Ventricular Tunnel with Single Coronary Artery


