Korean Journal of Pediatrics Vol. 51, No. 10, 2008
DOI : 10.3345/kjp.2008.51.10.1118
Case report

Coronary artery fistula associated with single coronary artery

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= Abstract =

A case of a single coronary artery complicated with a coronary artery fistula (CAF) to the right ventricle is extremely rare, and its management strategy and prognosis are not clear. A 5-year-old boy was hospitalized for evaluation of a continuous heart murmur. Transthoracic echocardiography suggested a CAF to the right ventricle, with an enlarged left coronary artery. Cardiac catheterization confirmed the CAF terminating at the right ventricle and the absence of a right coronary artery. The fistula was ligated at the right ventricular side under cardiopulmonary bypass. At follow-up 18 months later, the child was clinically asymptomatic, and coronary angiogram showed no recurrence of the fistula. **(Korean J Pediatr 2008;51:1118-1122)**

Key Words : Coronary vessel anomalies, Arteriovenous fistula, Child, Cardiac surgery

INTRODUCTION

Congenital coronary abnormalities are found in 0.2-1.6% of the population. Many of these abnormalities are incidental and benign findings, while others may have serious implications[1-3]. The anomalous originating of the left circumflex coronary artery (LCx) from the right coronary sinus is the most common congenital coronary abnormality. The absence of one coronary artery ostium (single coronary artery) is a rare finding. The congenital absence of the ostium of the right coronary artery, with the origin of the right coronary artery as a continuation of the distal LCx and left anterior descending artery (LAD) is extremely rare[3-5]. We report a case of this unusual single coronary artery abnormality complicated with a coronary artery fistula to the right ventricle.

CASE REPORT

A clinically asymptomatic 5-year-old boy with a continuous heart murmur was referred to our hospital. His body weight on admission was 19 kg, and his heart rate and blood pressure were 91 bpm and 100/60 mmHg, respectively. The lungs were clear to auscultation. Cardiovascular examination showed a symmetric chest wall and a normally located apical impulse. Auscultation of the chest disclosed a grade 2/6 continuous murmur along the left sternal border. There was no cyanosis, clubbing, or peripheral edema. The remainder of the examination was unremarkable.

A chest X-ray demonstrated no cardiomegaly. An electrocardiogram showed sinus rhythm, normal axis and intervals, and no ST-segment or T wave changes except an incomplete right bundle branch block. To evaluate the cardiac murmur, an echocardiogram was performed which showed marked distension of the left main coronary artery and color-flow imaging showed an abnormal jet arising from the anterior wall of the right ventricle (Fig. 1).

A selective coronary angiogram demonstrated a left-dominant system with dilated left main coronary artery, LAD, and LCx, with aneurysmal changes in the distal LCx terminating with an aneurysmal sac (Fig. 2A). An abnormal artery connected the mid LAD with the distal LCx. The remaining LAD beyond the abnormal branch was of diminished size (Fig. 2B). The anterior wall of the right ventricle was supplied by this abnormal artery, and the lateral wall by the distended distal LCx. The CAF originated from the aneurysmal sac of the distal LCx (Fig. 2C). The right coronary artery ostium could not be cannulated despite use of multiple catheters. A 45° left anterior oblique arteriogram confirmed the absence of the ostium (Fig. 2D). It was considered that the boy required surgery because of the risk of further enlargement of the LCx dilatation, further growth in the size of the fistula and aneurysmal sac, and the potential risk of bacterial endocarditis. Additionally, the pulmonary to

Received : 6 June 2008, Revised : 25 August 2008,
Accepted : 1 September 2008
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systemic flow ratio (Qp/Qs) value was 2.2.

In order to confirm the exact location of the CAF in the right ventricle, it was decided to feed a coronary angioplasty guide wire through the LCx to the right ventricle before surgery (Fig. 3A). With the guidance of the wire, surgeons could reduce both the total operation time, and the cardiopulmonary bypass time, reducing the likelihood of fatal complications. Surgery was performed through a median sternotomy incision. The right atrial wall was opened, and the orifice of the CAF closed via the tricuspid valve. Weaning the patient from cardiopulmonary bypass and closure of the sternotomy were performed in the usual manner.

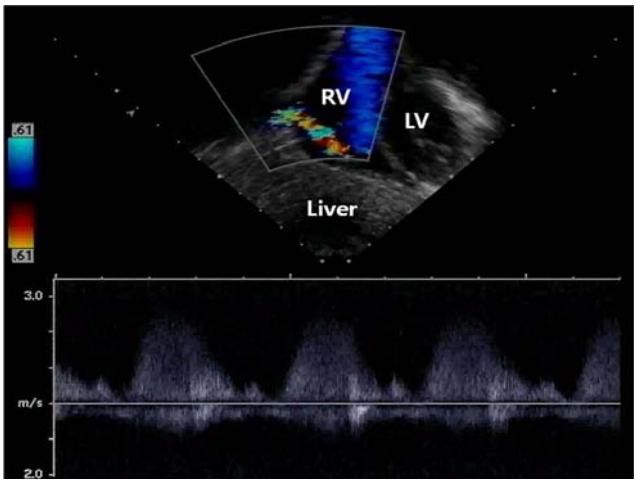

Fig. 1. The subcostal view of color-flow imaging and the corresponding Doppler recording by echocardiography. The imaging shows an abnormal jet from the anterior wall of the right ventricle. The jet can be traced into the right ventricle.

Eighteen months after surgery, the boy was readmitted to the hospital, and follow-up evaluation by coronary angiogram was performed. The selective coronary angiogram revealed no recurrence of the fistula, and complete occlusion of the aneurysmal sac of the distal LCx. The diameters of the dilated left main coronary artery and LCx had not reduced (Fig. 3B). However, that of the proximal LAD had markedly reduced (Fig. 3C). The blood flow between the distal LCx and LAD showed delayed filling, which was possibly due to competition between the arteries for blood flow (Fig. 3D).

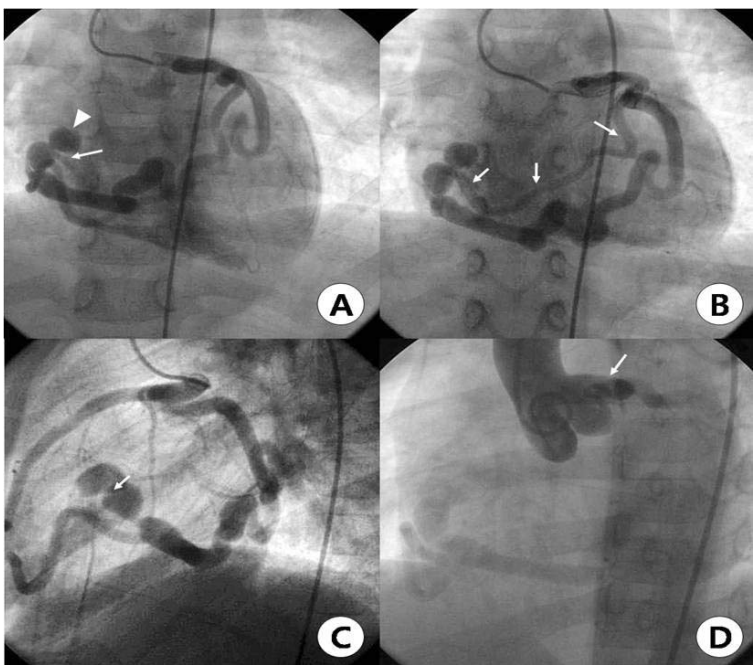

Fig. 2. (A) Anterior-posterior view showing aneurysmal changes in the distal left circumflex artery (LCx) terminating with an aneurysmal sac (arrow head) and the opening of the fistula (arrow). (B) The same view showing the abnormal branch (arrows) connecting the mid left anterior descending artery (LAD) with the distal LCx, and the diminished distal LAD blood flow beyond the branch. (C) Left lateral view showing how the terminal LCx connects with the abnormal branch and the opening of the fistula (arrow) (D) A 45° left anterior oblique aortogram showing the absence of the right coronary artery ostium. The dilated left main ostium is clearly seen (arrow).

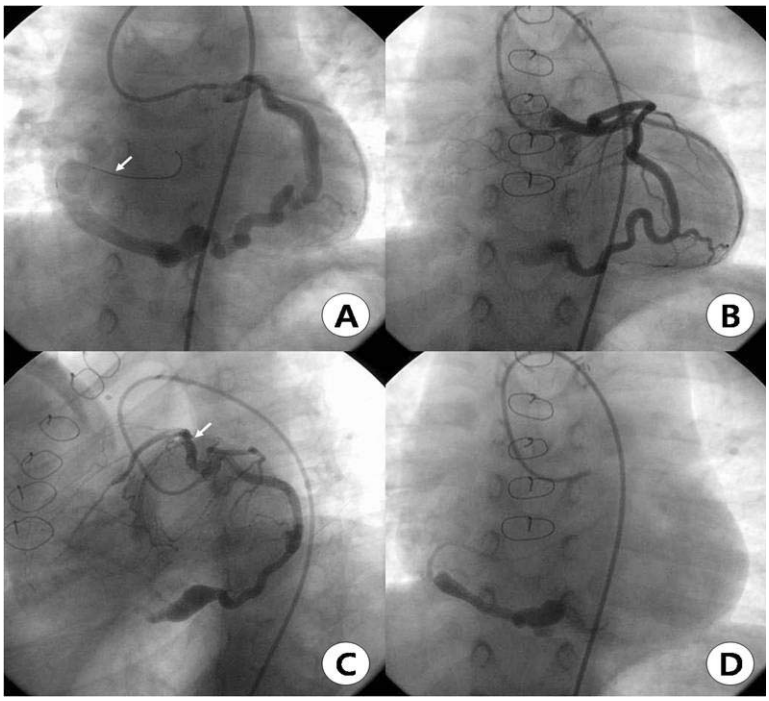

Fig. 3. (A) Anterior-posterior (AP) view showing the coronary angioplasty guide-wire (arrow) from left main coronary artery through the left circumflex (LCx) artery to the right ventricle. (B) Eighteen months after surgery, the dilated left main coronary and left circumflex arteries have not reduced in diameter. However, the diameters of the proximal and mid sections of the left anterior descending (LAD) artery have reduced (arrow), (C, arrow), and the distal LAD blood flow has improved. (D) Delayed filling between the distal LCx and the LAD branch, possibly due to competition for blood flow, with no recurrence of the fistula.

DISCUSSION

The great majority of cases of coronary artery fistula in the pediatric population are congenital in nature, possibly arising from persistence of sinusoidal coronary arterial connections. Acquired fistulas are often iatrogenic due to coronary surgery or the patient having undergone endomyocardial biopsies. It is difficult to estimate the true incidence of CAF because of its typically asymptomatic course in childhood. However, the use of echocardiography in the evaluation of heart murmurs has led to an increasing frequency of detection of the abnormality[6].

About 60% of CAF have been reported to arise from the right coronary artery[7], and 90% make their connection to the right ventricle[7]. Less frequently, the CAF empties into the right atrium, coronary sinus, or pulmonary artery[8]. The mean age at diagnosis of CAF was 7.2 years in 31 patients with an echocardiographic finding of a clinically silent CAF [9]. At the time of diagnosis, which is frequently made after the auscultation of a continuous murmur, children are usually asymptomatic.

A review of 187 patients has shown that complications related to CAF occured in 11% of patients under 20 years of age and in 35% of patients over 20 years of age[10]. The severity of the complications depends on the degree of altered myocardial blood flow. Complications include anginal chest pain, dyspnea and fatigue associated with congestive heart failure, arrhythmia, and myocardial infarction. The incidence of congestive heart failure and angina increases with age[10]. Thus, the likelihood of chest pain from CAF in young children is very low.

The natural history of CAF is of a progressive dilatation of the fistula accompanied by worsening of symptoms due to increased coronary steal. Late complications may include the formation of aneurysms, with possible rupture and tamponade, intimal ulceration, medial degeneration, atherosclerosis, calcification, and mural thrombosis formation within the CAF or proximal coronary artery[11].

The chest radiograph typically appears normal in patients

with CAF. The electrocardiogram usually appears normal or may show nonspecific abnormalities depending on the size and location of the CAF and the degree of ischemia due to coronary steal. Echocardiography has replaced coronary angiography as the method of choice for the diagnosis of CAF. The intra-cardiac anatomy is typically normal. The proximal coronary artery that supplies the fistula may appear dilated and tortuous due to increased blood flow through the vessel. The distal connection of the fistula may be defined by color Doppler imaging, but the proximal portion of the fistula is often difficult to image. Occasionally, cardiac catheterization with coronary angiography is required to make the final diagnosis.

The treatment of coronary artery fistula is dictated by both symptomatology and the size of the fistula. Spontaneous closure of small fistulae may occur in 23% of patients[9, 12]. While there is general agreement about the need to eliminate symptomatic fistulae, controversy surrounds treatment of the asymptomatic patient. Sherwood et al[9] concluded that silent CAF (those not associated with a heart murmur) in childhood and adolescence, especially those diagnosed incidentally by echocardiography, are not associated with adverse clinical outcomes. They recommended conservative management with continued follow-up. Others advocate closure of CAF associated with a continuous heart murmur, to eliminate the risk of bacterial endocarditis, and to avoid treating the same patient as an adult having developed a symptomatic, tortuous, aneurysmal CAF. The size of the proximal coronary artery at diagnosis may indicate the degree of left-to-right shunting and the potential risk of further dilatation as the child grows.

Trans-catheter coil embolization is the treatment of choice for the elimination of CAF. Embolization coils, or recently developed cardiac devices, can be delivered directly into the fistula during cardiac catheterization. After occlusion of the distal fistulous connection, complete thrombosis of the CAF occurs. Care must be taken to determine the precise anatomy of the normal coronary arteries to assure that the coil or device delivery will not adversely affect normal coronary blood flow.

In our case, surgical repair was performed. Firstly, because the coronary artery branch feeding the fistula could not be safely cannulated from the coronary ostium due to the extreme tortuosity of the LCx, and the very acute angle between the abnormal branch and the fistular opening (Fig. 2C). Secondly, because of the extensive draining network involved in the site. Thirdly and most importantly, because of the presence of a single coronary artery[13].

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단일관상동맥에서 우심실로 유출되는 동정맥루는 보고가 매우 드물며 이 경우 치료 방침이나 예후에 대해 명확하지 않다. 저자들은 지속성심잡음이 들려 시행한 심초음파 검사상 단일 좌관상동맥이 확장되어 있으며 우심실로 유출되는 동정맥루를 진단하였으며 관상동맥조영술로 확진하였다. 체외순환하에 우심실에서 동정맥루를 결찰하였다. 18개월 후 추적시 증상은 없었고 관상동맥조영술상 동정맥루의 재발도 없었다.

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