Case report
Korean J Pediatr 2016;59(Suppl 1):S80-83
https://doi.org/10.3345/kjp.2016.59.11.S80
pISSN 1738-1061•eISSN 2092-7258

Korean J Pediatr

Anomalous right coronary artery from pulmonary artery discovered incidentally in an asymptomatic young infant

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Isolated anomalous right coronary artery originating from the pulmonary artery (ARCAPA) is a rare congenital coronary anomaly that is asymptomatic and discovered incidentally in most cases. ARCAPA is generally not considered a fatal defect in infancy or childhood, although cases of sudden death have been reported. Here, we report a 2-month-old female infant who presented with a prolonged fever that was determined to be caused by rhinovirus infection. Myocardial ischemia of the left ventricular posterior wall was already seen on echocardiography, and ARCAPA was discovered incidentally. The patient underwent successful surgical reimplantation of the right coronary artery to the aortic root to re-establish dual ostial circulation.

Key words: Coronary vessel anomalies, Infant, Asymptomatic disease

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Received: 15 September, 2014
Revised: 16 October, 2014
Accepted: 30 October, 2014

Introduction

Anomalous right coronary artery (RCA) from pulmonary artery is a rare congenital coronary anomaly. The prevalence is about 0.002% of general population[1)]. Anomaly such as aortopulmonary window, tetralogy of Fallot, and septal defects has been reported in one-third of anomalous RCA originating from the pulmonary artery (ARCAPA) patients. Whereas patients with anomalous pulmonary origin of the left coronary artery (ALCAPA) usually present with signs of left ventricular ischemia and congestive heart failure in infancy, most ARCAPA patients are diagnosed during childhood with cardiac murmur, chest pain, or congestive heart failure. The diagnosis of ARCAPA is missed because of asymptomatic status[2,3)]. However, ARCAPA may present with subclinical myocardial ischemia or is at the risk of sudden cardiac death even in early childhood.

Case report

A 2-month-old female presented with 2 days of fever. The fever persisted for more than 5 days after admission. Generalized erythematous macular rash was followed. She was born with 40 weeks of gestational age and 2.7 kg of body weight without significant perinatal illness. She has been completely well without irritability, dyspnea, tachypnea, or cyanosis prior to illness. The height was 57.7 cm (10th–25th percentile) and body weight was 5.4 kg (10th–25th percentile). The blood pressure and the pulse rate were 84/60 mmHg and of 148 beats per minute respectively. The cardiac examination was unremarkable except for sinus tachycardia and breathing sounds was equal bilaterally. The laboratory

results were as follows: white blood cell, 9,640/μL; hemoglobin, 9.3 g/dL; platelet, 571,000/μL, erythrocyte sedimentation rate, 61 mm/hr; C-reactive protein, 12.9 mg/dL; procalcitonin, 1.96 ng/mL; prohormone of brain natriuretic peptide, 2,077 pg/mL; creatinine phosphokinase, 19 IU/L; troponin I, 0.024 ng/mL. Electrocardiography showed sinus tachycardia without ST-T changes or abnormal Q wave (Fig. 1). The chest X-ray showed normal heart contour and size. Because of concern of Kawasaki disease and infective endocarditis for prolonged febrile exanthema, transthoracic echocardiogram was done. The origin of the RCA was from the main pulmonary artery with RCA to pulmonary artery steal, predominantly diastolic (Fig 2). It showed a dilated left coronary artery with normal origin and collaterals

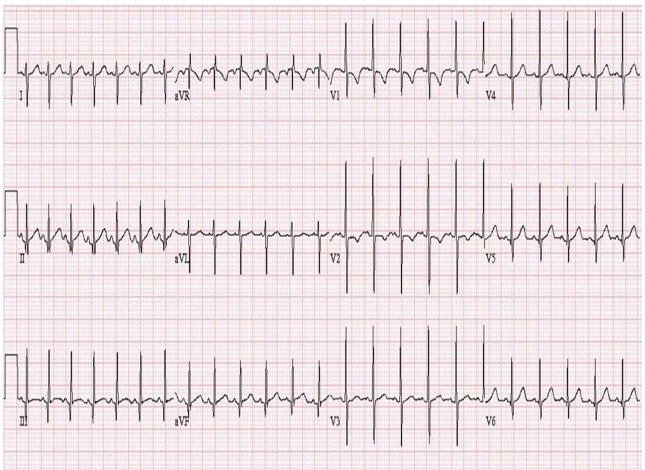

Fig. 1. Electrocardiogram shows nonspecific findings without an ST-T change or abnormal Q wave

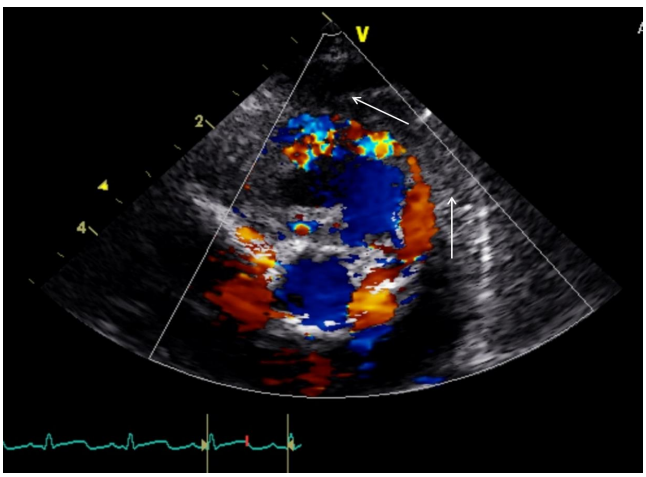

Fig. 3. Transthoracic echocardiography in the parasternal short-axis view shows a dilated left coronary artery with collaterals from the left anterior descending artery to the right coronary artery (arrows).

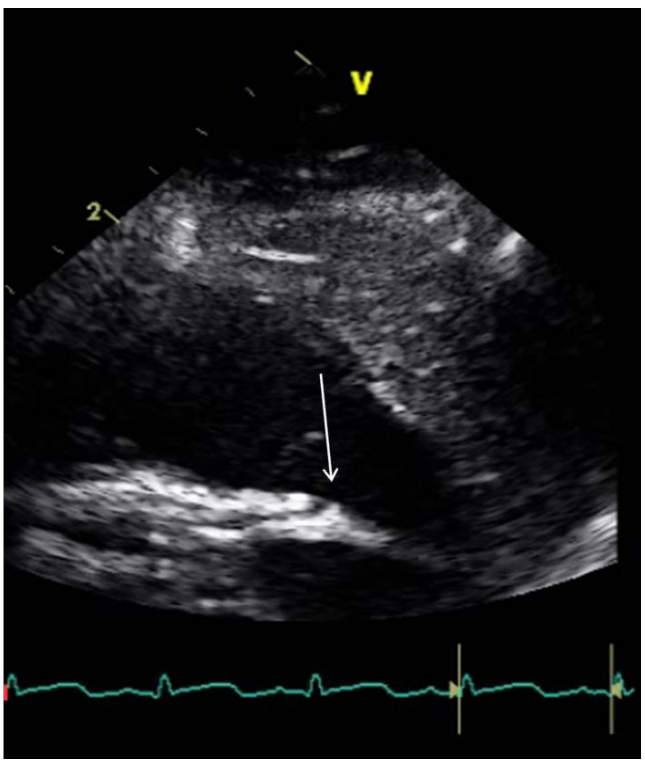

Fig. 2. Transthoracic echocardiography in the modified short-axis view shows the orifice of a normal-sized right coronary artery (arrow) arising from the main pulmonary artery.

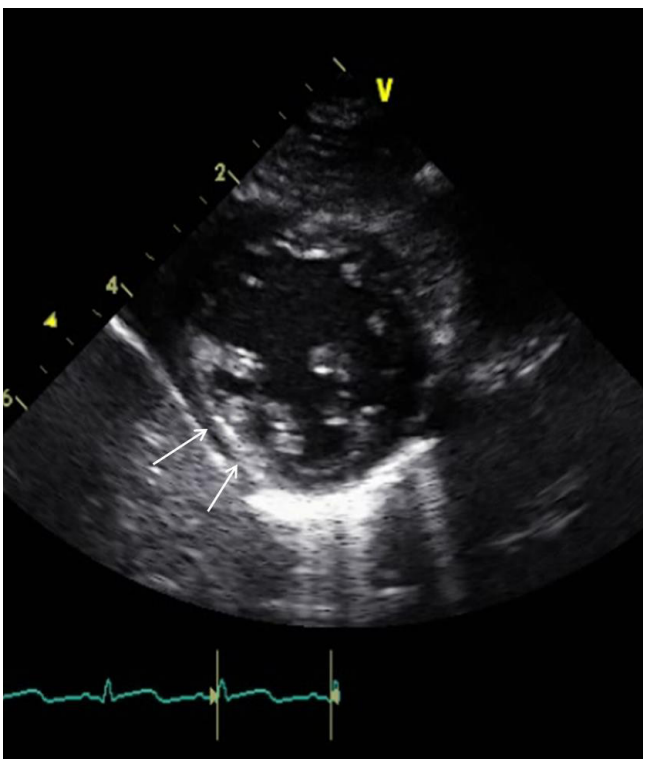

Fig. 4. Transthoracic echocardiography in the parasternal short-axis view shows bright echogenicity and thinning of the posterior wall of the left ventricle (arrows) suggestive of an ischemic change.

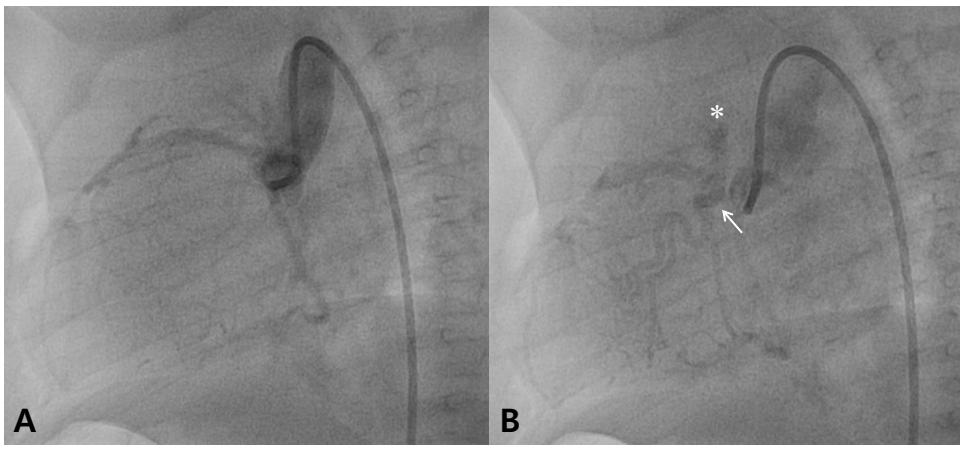

Fig. 5. Lateral aortography. (A) Aortogram clearly showing the left coronary system without filling of the right coronary system. (B) Aortogram showing delayed filling of the right coronary system (arrow) and pulmonary artery (asterisk) through collaterals in the late phase.

from left anterior descending coronary artery to RCA on short axis view (Fig. 3). The cardiac structure, function, and dimension were within normal range but ischemic change of posterior wall of left ventricle was noted (Fig. 4). The fever was subsided on 4th hospital day and the causative infectious agent was concluded as rhinovirus. Coronary angiography was arranged for further evaluation. Aortogram demonstrated prominent left main coronary artery, normal left anterior descending artery and circumflex artery. The left main coronary artery arose from the appropriate sinus without aneurysm or stenosis (Fig. 5A). After a slight delay, the posterior descending and RCA were filled up in a retrograde fashion through collaterals of the left coronary artery (Fig. 5B). On the 11th hospital day, the patient underwent direct reimplantation of anomalous RCA to aortic root establishing dual ostial circulation (Fig. 6). On the last follow-up, 5 months after the surgery, she was perfectly well on acethyl salicylic acid therapy. The echocardiographic study showed the normal biventricular function. The forward flow was well seen from aorta to RCA.

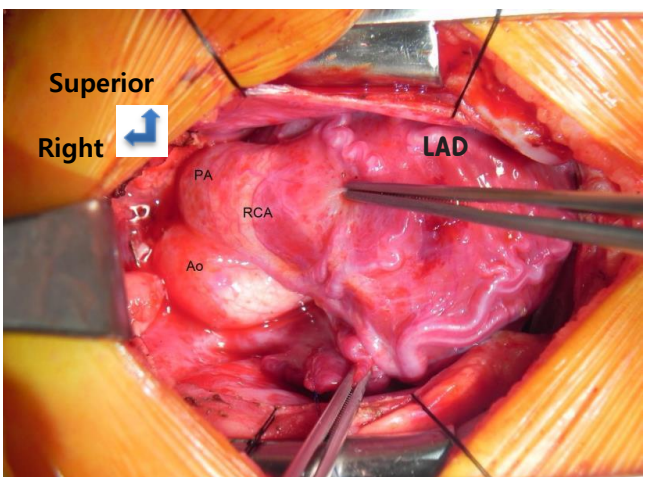

Fig. 6. Intraoperative view of the right coronary artery (RCA) arising from the pulmonary artery with normal RCA distribution and collaterals from the left coronary artery to the RCA. Ao, aorta; PA, pulmonary artery; LAD, left anterior descending artery.

Discussion

The prevalence of coronary anomaly is based on angiographic studies of adult from 0.6% to 1.3%[4]. ALCAPA, first described by Brooks[3], is more common than ARCAPA and is mostly fatal in infancy. The ARCAPA is rare, only about one-tenth as common as ALCAPA[1]. Unlike the ALCAPA presented with heart failure and myocardial ischemia in early infancy, many patients are asymptomatic[5]. This anomaly was usually diagnosed on the basis of heart murmur, mostly continuous in 50% of cases[1]. Other presenting symptoms and signs of ARCAPA are angina, dyspnea on exertion, congestive heart failure, and sudden cardiac arrest. Radke et al.[6] noted that 41% of patients of ARCAPA were asymptomatic, but 40% showed the ischemia. Our case was asymptomatic and diagnosed incidentally by echocardiography during the evaluation of prolonged fever. The timing and severity of symptoms depend on the type of anomaly, direction of blood flow in the anomalous vessels, and extent of collateralization[7]. Its small perfusion area results in the patient generally asymptomatic, compared with patients having ALCAPA. But some increase in oxygen demand leads to exhaustion of the coronary physiologic reserve, which triggers extensive cardiac ischemia and causes ventricular arrhythmia similar to the ALCAPA. Our patient showed the ischemia of left ventricular posterior wall even in asymptomatic 2 months old. The ARCAPA can be associated with other congenital anomaly like Aortopulmonary window (30% of associated defects)[8-10], Tetralogy of Fallot (19%), and

 https://doi.org/10.3345/kjp.2016.59.11.S80

isolated valvular anomalies[11,12]. The associated structural heart anomaly was not noted in the patient. Unlike ALCAPA, there are no characteristics of electrocardiogram findings associated with ARCAPA. Although the diagnosis of ARCAPA is made incidentally during heart surgery or on autopsy prior to 1965, it is diagnosed by angiography or echocardiography during evaluation of the cardiac murmur, chest pain or dyspnea. Both cardiac magnetic imaging and multislice computed tomography are becoming increasingly popular imaging modality for discovering coronary artery anomalies. Because of the risk of myocardial ischemia or sudden death and improvement in operative morbidity, surgical correction with reestablishment of dual coronary ostial circulation is recommended even in asymptomatic patients[13].

In conclusion, we treated a asymptomatic 2-month-old infant presenting with prolonged fever who was incidentally diagnosed with ARCAPA. The myocardial ischemia of left ventricular posterior wall was noted even in early and asymptomatic infant. We performed successfully the direct coronary reimplantation with reestablishment of dual coronary ostial circulation.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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