Case Report /Olgu Sunumu
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Anomalous Origin of Left Coronary Artery from Pulmonary Artery (ALCAPA) in an Infant with Bronchiolitis and Dilated Cardiomyopathy

Bronşiolit ve Dilate Kardiyomyopatisi Olan Bir Bebekte Sol Koroner Arterin Pulmoner Arterden Çıkış Anomalisi (ALCAPA)

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SUMMARY

The most common cause of dilated cardiomyopathy (CMP) is viral myocarditis. However, anomalous origin of the left coronary artery is a rare condition that should be evaluated with high suspicion in the differential diagnosis of an infant with dilated CMP. With early surgical correction, an anomalous coronary artery origin has a good prognosis. Awareness of this condition is essential for prompt diagnosis. This article reports a 5 months-old infant with dilated cardiomyopathy in the setting of RSV bronchiolitis. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) was the final diagnosis in the infant, who presented as viral myocarditis with RSV infection. She was operated successfully with the direct reimplantation of the coronary artery to the aorta. (Journal of Current Pediatrics 2013; 11: 142-5)

Key words: ALCAPA, cardiomyopathy, infant, myocarditis, RSV bronchiolitis

ÖZET

Dilate kardiyomyopatinin en yaygın nedeni viral myokarditтir. Bununla birlikte sol koroner arterin pulmoner arterden çıkış anomalisi nadir bir durum olup dilate kardiyomyopatinin etiyolojisinde yüksek şüphe ile irdelenmelidir. Erken cerrahi düzeltme ile koroner arter çıkış anomalisinin prognozu iyidir. Bu bozukluğun farkındalığında olmak hızlı teşhis için önemlidir. Makalede 5 aylık bir süt çocuğunda RSV bronşioliti zemininde gelişen dilate kardiyomopati olgusu bildirilmektedir. RSV infeksiyonu ile viral myokardit geliştiği düşünülen bebekte kesin tanı sol koroner arterin pulmoner arterden çıkış anomalisi (ALCAPA) idi. Sol koroner arterin aortaya doğrudan reimplantasyon ameliyatı başarı ile sağlandı. (Güncel Pediatri 2013; 11: 142-5)

Anahtar kelimeler: ALCAPA, kardiyomyopati, infant, myokardit, RSV bronşioliti

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Introduction

Cardiomyopathy (CMP) is the leading cause of transplantation in children over 1 year old in developed countries. The incidence of dilated CMP is 51% to 58% among all types of CMP (1,2). Congestive heart failure (CHF) is the presenting symptom in most cases. Chest radiographs of dilated CMP show evidence of cardiomegaly and increased pulmonary vascular markings. Almost half of all patients with dilated CMP present within the first year of life, with a median age at presentation of 7.5 months.

Only one-third of cases with dilated CMP have a known cause, with lymphocytic myocarditis accounting for 27% to 40% of these cases. Enterovirus and adenovirus are frequently associated with dilated CMP (2,3), and respiratory syncytial virus (RSV) is a causative agent for myocardial dysfunction and dysrhythmias. Bacterial infections, structural heart malformations, tachyarrhythmias, idiopathic dilated CMP, inborn errors of metabolism, connective tissue disorders, and drug toxicity should also be evaluated in the differential diagnosis of viral myocarditis in infants (3).

In this paper, we report a case of severe heart failure in a 5-month-old female in the setting of RSV bronchiolitis. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), a very rare congenital heart anomaly, was the final diagnosis.

Case Report

In the present case study, the patient was a 5-month-old female, born at term with no history of complications during labor. She was admitted to another pediatric clinic with symptoms of rhinitis, coughing, and tachypnea. The runny nose had started 5 days prior to presenting at the clinic. A nasal swab rapid antigen test was positive for RSV infection. Cardiomegaly was prominent in the chest radiography, and echocardiography revealed LV dysfunction. She was referred to the pediatric cardiology department with initial diagnoses of RSV bronchiolitis and myocarditis.

On examination, the patient looked pale. Her weight was 5.5 kg, axillary temperature was 36.5°C, room air oxygen saturation was 94%, respiratory rate was 52/min, heart rate was 140/min, and liver edge was 3 cm below the costal margin. Rhonchus in both lungs, subcostal retractions, and 2/6 pansystolic murmur at the lower left sternal border radiating toward the apex were noted in the chest examination. The peripheral leukocyte count was 8190 cells/mm^3, with 51% lymphocytes. Blood test results included hemoglobin of 11.2 g/dL, isoenzyme of creatinine kinase with muscle and brain subunits (CK-MB) of 29 u/L, alanine aminotransferase of 61 U/L, aspartate aminotransferase of 71 U/L, and troponin I of 0.161 ng/ml (normal range is <0.02 ng/ml). The history revealed poor feeding and a weight gain of only 300 g during the previous month.

Echocardiography revealed severe LV dysfunction, with an ejection fraction of 18% and mild mitral regurgitation. The left coronary orifice was not seen, and reverse blood flow within the left coronary artery into the pulmonary artery was present (Figure 1). The diagnosis of ALCAPA was made. Surgical treatment of ALCAPA was planned for as early as possible, with consideration given to the fact that the patient had RSV bronchiolitis.

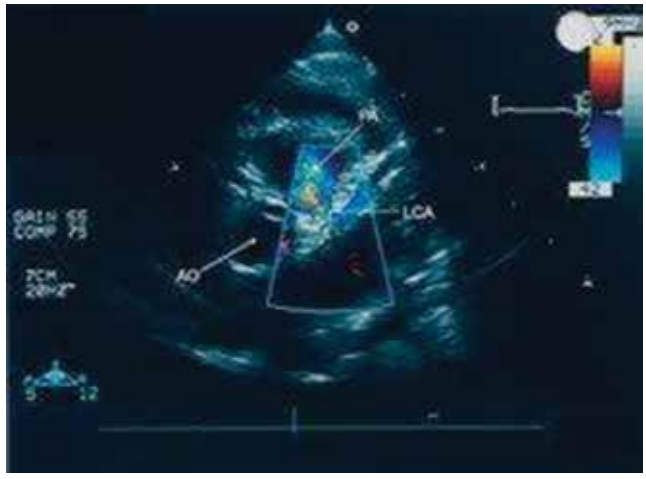

Figure 1. Color Doppler examination in the parasternal short-axis view demonstrating the left coronary artery (LCA) arising from the pulmonary artery (PA). AO: aorta

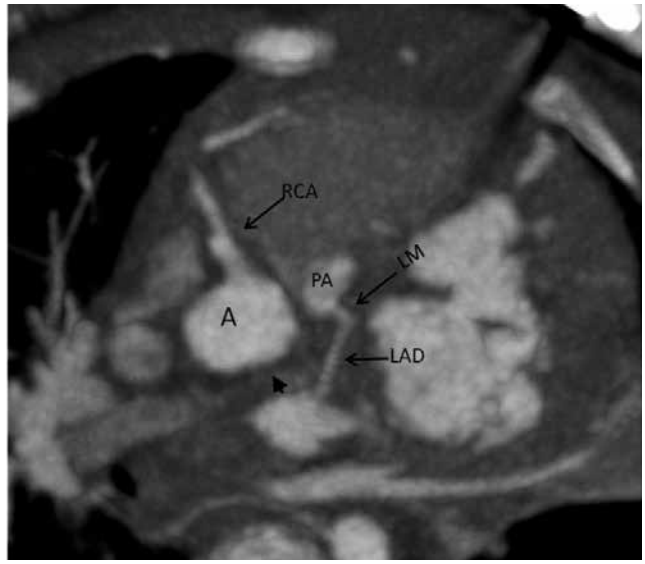

Figure 2. Maximum intensity projection (MIP) thick-slice image at the coronary artery level showing the left coronary artery (LCA) originating from the pulmonary artery (PA). The right coronary artery (RCA) originates from the right coronary sinus. No vessel originates from the left coronary sinus (arrow head). AO: aorta

Before the surgery, computed tomographic (CT) angiography was performed to determine which technique would be used for the operation (Figure 2). After 12 days, recovery from RSV infection was documented by a negative rapid antigen test. Direct aortic reimplantation of the left coronary artery was managed successfully on day 14. The cardiopulmonary bypass time was 168 minutes, and aortic clamp time was 96 minutes.

Severe inotropic support was needed postoperatively, which was decreased gradually and stopped on the 10th day. The patient was extubated on sixth postoperative day, and the total intensive care unit stay was 14 days. The ejection fraction by echocardiography increased and was 44% on the 20th postoperative day. The patient was discharged to home with digoxin, captopril, diuretic, and aspirin therapy, and will be followed-up in the pediatric outpatient clinic.

Discussion

Left coronary artery arising from the pulmonary artery instead of the aorta is a very rare congenital cardiac anomaly. Infants with ALCAPA appear normal at birth. After 2 or 3 months of life, when pulmonary arterial resistance decreases to an adult level, hypoperfusion of the LV with desaturated blood at a low pressure from the pulmonary artery predisposes the infant to myocardial ischemia. At that point, many patients present with symptoms of CHF, including tachypnea, tachycardia, sweating, poor feeding, and poor weight gain. The CHF deteriorates due to mitral incompetence, secondary to a dilated mitral ring or infarction of the papillary muscle. The anomaly is usually isolated, but is occasionally associated with patent ductus arteriosus or atrial and ventricular septal defects (4). Affected infants show marked cardiomegaly and evidence of pulmonary edema, a feature similar to CMP.

Anomalous coronary artery origin should be clinically suspected in an infant with poor feeding, irritability, dyspnea, and cardiomegaly. Doppler echocardiography is often diagnostic, identifying the abnormal attachment of the coronary artery from the pulmonary artery. ALCAPA has historically been evaluated by conventional angiography; however, the development of multidetector CT angiography and magnetic resonance imaging has enabled direct visualization of the left coronary artery arising from the main pulmonary artery.

ALCAPA is the main cause of ischemia and myocardial infarction in childhood. Mortality in the first year of life can reach 90% if the disease is not treated (4). Despite the commonly severe preoperative clinical status, the surgical mortality is low (10%) and long-term survival after surgery is high (5). The main incremental risk factor for mortality is the preoperative severity of LV dysfunction. However, early surgical correction promotes rapid recovery of LV.

The procedure of choice for ALCAPA treatment is the creation of a dual coronary system by the direct reimplantation of the anomalous coronary artery into the ascending aorta (5). Alternatively, left subclavian-left coronary artery anastomosis, intrapulmonary tunnel operation (Takeuchi procedure), or pulmonary artery flap can be used when the left main coronary artery is short or originates from the posterior-lateral position (5,6). In our patient, although the left coronary artery originated from the posterior wall of the pulmonary artery, direct reimplantation to the aorta was managed successfully.

Most centers do not recommend routine mitral valve repair in ALCAPA patients. Mitral insufficiency usually decreases with treatment of the anomalous coronary artery. However, some patients may need later mitral valve replacement or valvuloplasty (5). Stenosis or total occlusion of the left main coronary artery after surgery should be suspected in the case of persistent LV dysfunction or worsening mitral regurgitation.

The timing of surgery after recent respiratory tract infections is difficult. The decision for surgery in an infant who has not completely recovered from an episode of RSV infection may place the patient at higher risk for postoperative complications, especially postoperative pulmonary hypertension. Cardiopulmonary bypass may aggravate the inflammation and pulmonary hypertension that normally occur with RSV infection (7). The only similar case reported in the literature, a 4-month-old female who had respiratory failure after RSV infection with unrepaired ALCAPA, was surgically treated on the 27th hospital day. In that case, the troponin I and CK-MB levels and electrocardiograms were evaluated daily to assess for ongoing myocardial ischemia (8). We planned the surgery on 14th day after admission, and no postoperative complication related to the recent RSV infection was observed.

The incidence of ALCAPA may be greater than previously recognized, and it is possible that cases are being misdiagnosed or presenting as sudden infant death syndrome. Brotherton et al. reported the incidence of ALCAPA as 0.023 and emphasized awareness of the condition among pediatricians (9). Lipsett et al. reported an ALCAPA incidence of 0.2 in pediatric autopsy cases (10).

Dilated CMP due to viral myocarditis may mask the presence of ALCAPA. In our case, the clinical presentation was consistent with a diagnosis of CHF. If the underlying reason for severely decreased LV function had not been evaluated with such high suspicion and detailed examination, the patient likely would have been diagnosed with myocarditis after RSV bronchiolitis.

In conclusion, ALCAPA should be evaluated in the differential diagnosis of any infant with symptoms of unexplained severe CHF (with no obvious structural cause), mitral insufficiency, and cardiomegaly.

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