A RARE BUT TREATABLE CAUSE OF DILATED CARDIOMYOPATHY: ALCAPA

Case Report

DİLATE KARDİYOMİYOPATİ'NİN NADİR ANCAK TEDAVİ EDİLEBİLİR BİR SEBEBİ: ALCAPA

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ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart disease (CHD) which accounts for 0.023% of all congenital heart diseases. It is asymptomatic in many cases and is mostly diagnosed in the first few months of life. If untreated, the mortality rate in the first year of life will be more than 90%. We report a 6-month-old infant admitted to out clinic because of heart failure. Echocardiography revealed a dilated cardiomyopathy and left main coronary artery originates from pulmonary artery. Diagnosis was confirmed by coronary angiography and treated succesfully by surgical procedure.

Key words: *Anomaous origin of the left coronary artery from the pulmonary artery; cardiac insufficiency; dilated cardiomyopathy;newborn.*

ÖZET

Pulmoner arter'den köken alan sol koroner arter anomalisi, oldukça nadir görülen (% 0.023) bir konjenital kalp hastalığıdır. Genellikle yaşamın ilk birkaç ayında teşhis edilir, vakaların çoğu asemptomatiktir. Tedavi edilmezse, bir yıl içinde ölüm oranı % 90'dan fazladır. Burada kalp yetersizliği nedeniyle önce dış merkeze başvuran 6 aylık bir bebek rapor edilmiştir. Kliniğimizde yapılan ekokardiyografi'sinde dilate kardiyomiyopati ve sol ana koroner arter'in pulmoner arter'den kaynaklandığı saptanmış, tanı koroner anjiyografi ile teyit edilerek başarılı bir cerrahi tedavi uygulanmıştır.

Anahtar kelimeler: *Dilate kardiyomiyopati; kalp yetersizliği;sol koroner arterin pulmoner arterden çıkış anomalisi; yenidoğan.*

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart disease (CHD) which accounts for 0.5% of all CHD and occurs in approximately 1 per 300,000 live births (1). Bland,White and Garland were first described this pathology in 1933. 87% of patients with this anomaly become symptomatic during infancy and 65% to 85% of them die before 1 year old if untreated (2). Anomally originated coronary artery cannot supply the myocard due to unoxigenated blood and low pressure in pulmonary artery. Althoug the the colleterals from right coronary artery try to supply the myocardium, congestive failure is inevitable. In this case report, we present an infant with ALCAPA who admitted to our clinic with heart failure The electrocardiography showed deep wide Q waves in D1 and aVL, ST elevation in leads V1-V6. Echocardiography revealed a dilated cardiomyopathy and left main coronary artery originates from pulmonary artery. Diagnosis was confirmed by coronary angiography and treated succesfully.

CASE REPORT

A six month old infant was admitted to our emergency department because of fast breathing, mild cyanosis and extreme sweating. She was 5 kg (10-25p), 60 cm (50-75p). Heart rate was 162 beat/minute and respiration rate was 62/minute. Subcostal and intercostal retractions were evident. Blood pressure was in normal limits. But a 3/6 systolic murmur and a 5 cm hepatomegaly was detected.on physical examination. The cardiothoracic ratio was 0.61 on telecardiography. The electrocardiography showed deep wide Q waves in D1 and aVL, ST elevation in leads V1-V6. Echocardiography revealed mild to moderate mitral regurgitaion with dilated left ventricule (**Figure 1A**).

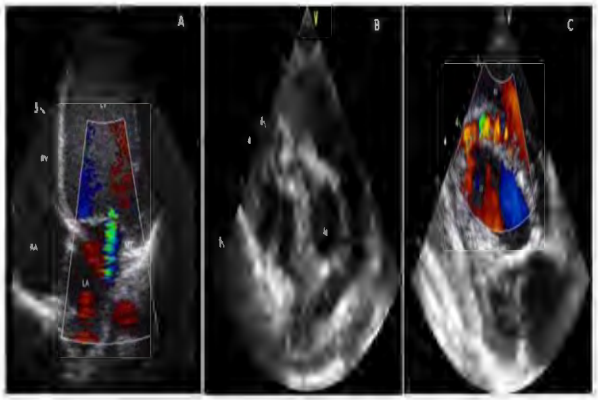

Figure 1: A. *Mild to moderate mitral regurgitaion with dilated left ventricule, B. The right coronary artery was shown to be originated from aorta and it was larger than normal. But the origin of the left coronary artery was not clear, C. Color doppler echocardiography showing the colletaral circulation through the interventricular septum.*

The right coronary artery was shown to be originated from aorta and it was larger than normal. But the origin of the left coronary artery was not clear (**Figure 1B**).

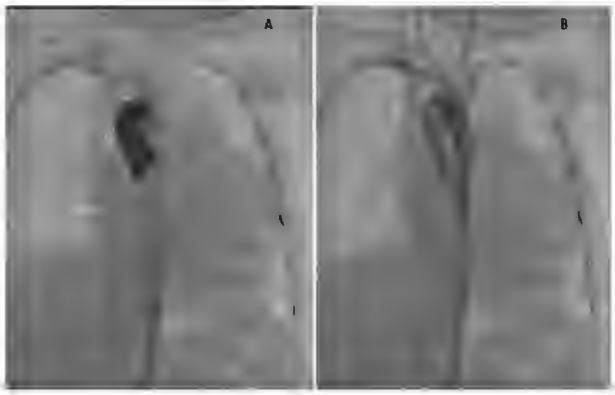

Figure 2: A and B. *Catheter angiography images of the retrograde filling of left coronary artery from right coronary artery and anomalous origin of the left coronary artery from pulmonary artery.*

A reverse flow was detected in the pulmonary artery by color doppler echocardiography. 2D echocardiography revelaed the left coronary artery and its branches originating from the pulmonary artery. Color doppler echocardiography also showed the colletaral circulation

through the interventriculer septum (**Figure 1C**).

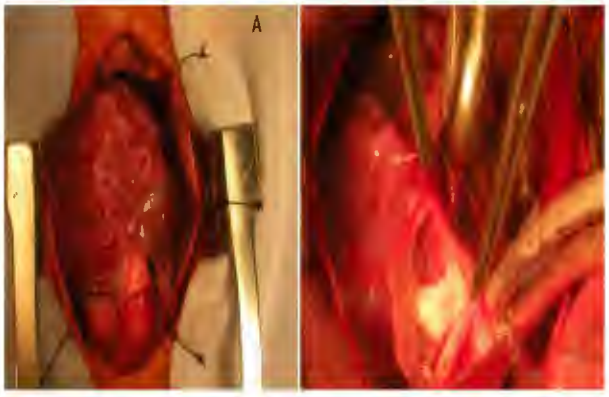

Figure 3: A. *Dilated right coronary artery (RCA), B. The creation of the tunnel between the A-P window and the left coronary artery from pulmonary artery using a Dacron patch (internal view of the pulmonary artery)*

The systolic functions of the left ventricule was decreased (ejection fraction was 44%). Catheter angiography confirmed the retrograde filling of left coronary artery from right coronary artery and anomalous origin of the left coronary artery from pulmonary artery (Figure 2A, B), and finally patient was sent for surgery.

The ostium of the left main coronary artery was not appropriate for buton transverse. So modified intrapulmoner tunnel (modified Tekauchi procedure) was performed. Pulmonary artery and aorta was drilled just above the sinotubuler junction by using 4 mm punch. These holes were joined by 6-0 prolen and a aortapulmonary window was built. Then 4 mm dacron patch was longitudianlly placed to make a tunnel between the ostium of the left coronary artery and the aorticopulmonary window (Figure 3A, B). So the ostium of the left coronary artery was transferred to the aorta just above the sinotubuler junction. Pulmonary artery was repaired by autologous pericard patch. The patient was discharged five days after the operation. At follow-up echocardiography confirmed normal the ejection fraction (61%).

DISCUSSION

Although ALCAPA is a rare disease it is fatal in the first years of life if not treated. The pathophysiologic and clinical features of this disease are directly related with the poor perfusion of the left ventricular myocardium. This anomaly is well tolerated during fetal life because the pressure in aorta and pulmonary artery is almost the same. After birth the pressure in pulmonary artery decreases and it cannot supply the left coronary artery well enough and the circulation of the left ventricular myocardium becomes depended to the collaterals from right coronary artery. The numbers of the collaterals and the leakage from left coronary artery to pulmonary artery determines the degree of ischemia (3). Because of this pathophysiologic process 85% of patients become symtomatic in the first 2-3 months of life (4). In 15% of the patients clinical symptoms can be delayed to adulthood (4,5). Florent et al reported the patients with ALCAPA between 3-54 months (6). Few patients with this anomaly are diagnosed during newborn period (7).

Poor feeding , extreme sweating and pallor are the initial symptoms. Developmental delay, tachycardia, dyspnea and hepatomegaly are other symptoms (8).

Determining the ALCAPA syndrome in patients with dilated cardiomyopathy (DCMP) is very important. Because ALCAPA is a treatable cause of DCMP. Most of the patients with DCMP undergo a group of long lasting laboratory tests . This situation is one of the most reason of the delay of the diagnosis.

There are some clues sugessting the diagnosis of ALCAPA in DCMP patients. These are; pathologic Q waves in DI, AVL and V4-V6. A dilated right coronary artery, and unable to visualization of the left coronary arter are echocardiographic indicators. But in literature the reported

rate of false negativitiy is ranged between 50 to 71% (9,10).

In our patient the right coronary artery was larger than normal and there were multiple collalerals on the interventricular septum which considered the ALCAPA syndrome.

Some authors suggest to use echocardiography for diagnosis only but most of them offer aortic root injection and selective right coronary artery angiography (1).

The essential treatment of ALCAPA is surgery. There are several surgical methods for ALCAPA. Original operation was ligation of the LCA at its origin from the PA to interrupt the shunt and improve myocardial perfusion. Although there were several case reports with good long-term prognosis (11). most studies demonstrated high mortality in children after LCA ligation surgery (12,13). Hence, it is generally advocated to create two-coronary-artery system (14) . After surgical procedure the left ventricule improves gradually. Michielon et al reported that this improvement is better in infants but the report of Ando et al showed the same improvement in adults (15,16). Our patient was discharged on the fifth day of the operation without any problem.

In conclusion ALCAPA syndrome must be recognized in patients with DCMP because it is a treatable reason of DCMP.

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