

Correctable Cause of Dilated Cardiomyopathy in an Infant with Heart Failure: ALCAPA Syndrome

Süt Çocuğunda Kalp Yetersizliğı ile Başvuran Dilate Kardiyomiyopatinin Düzeltilebilir Bir Nedeni: ALCAPA Sendromu

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Abstract

Anomalous origin of the left coronary artery arising from pulmonary artery ALCAPA syndrome is a rare congenital heart disease seen in children. If untreated, it may lead to congestive heart failure, dilated cardiomyopathy (DCM), ischemic and arrhythmic complications may lead to patient's death. ALCAPA is diagnosed with echocardiography; in the patients of suspected diagnosis, computerized tomography, magnetic resonance imaging and cardiac catheterization are used for further testing. Surgically correctable ALCAPA syndrome must be considered as etiology of DCM in children. In this report, we presented the case of an infant that was referred to our center with the diagnosis of DCM, who was echocardiographically diagnosed with ALCAPA syndrome and successfully treated with surgery, as well as a review of recent literature.

Öz

Sol koroner arterin pulmoner arterden çıkış anomali olarak tanımlanan ALCAPA sendromu, çocuklarda nadir görülen bir konjenital kalp hastalığıdır. Tedavi edilmediği zaman konjestif kalp yetmezliği, dilate kardiyomiyopati (DKM), iskemik ve aritmik komplikasyonlarla hasta kaybedilebilir. Tam ekokardiyografi bulgularıyla koyulur, tanıdan şüphelenilen olgularda bilgisayarlı tomografi, manyetik rezonans görüntüleme ve kalp kateterizasyonu gibi ileri tetkiklerden faydalanılabilir. Çocukluk çağında DKM tanısı alan hastalarda etiolojide, cerrahi olarak düzeltme şansı olan ALCAPA sendromu mutlaka düşünülmelidir. Bu yazıda, merkezimize DKM tanısıyla sevk edilen, ekokardiyografi ile ALCAPA sendromu tanısı konulup, başarılı cerrahi tamir yapılan hasta olgu sunumu yapıldı ve son literatür gözden geçirildi.

Keywords

ALCAPA syndrome, dilated cardiomyopathy, echocardiography

Anahtar kelimeler

ALCAPA sendromu, dilate kardiyomiyopati, ekokardiyografi

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Introduction

Anomalous origin of the left coronary artery (ALCA) arising from the pulmonary artery (PA), also known as ALCAPA syndrome or Bland-White Garland syndrome, is one of the most common coronary artery origin anomalies. When left untreated, it is fatal within the first year of life in 90% of cases. It clinically presents as congestive heart failure, dilated cardiomyopathy (DCM), arrhythmia and ischemic events such as myocardial infarction. Early diagnosis and timely surgical intervention yield excellent results (1-4). In this article, we present a patient referred to us with DCM, who was echocardiographically diagnosed with ALCAPA and successfully treated with surgery.

Case Report

A 2.5-month-old female patient, who had no previous symptoms, presented with distress, constant crying, feeding problems, tachypnea and sweating for 3-4 days. Teleradiography performed at a different center showed cardiomegaly, echocardiographic evaluation showed DCM. Following inconclusive metabolic and genetic tests, the patient was referred to us for further examination. Her prenatal history was unremarkable. The patient was born on term to a 22-year-old mother, her birth weight was 3000 g. There was no parental consanguinity. Weight and height percentiles were normal for the patient's age. During physical examination, the patient's general condition was fair, she appeared to be in distress and had a body temperature of 36.8 °C, heartrate of 170 bpm, blood pressure of 80/50 mmHg, oxygen saturation of 96% in room air and respiratory rate of 62/min. Heart sounds were rhythmic and there were no murmurs. Her respiratory system was normal, her liver was palpated under the costal margin as sized at 2-3 cm. Teleradiography showed a cardiothoracic ratio of 0.65 (Figure 1). Electrocardiography (ECG) showed a 176 bpm sinus tachycardia, deep Q waves and inverted T waves in leads D1, aVL and V4-6 as well as left ventricular (LV) hypertrophy findings (Figure 2). Laboratory tests results were as follows; hemoglobin: 11.6 g/dL, white blood cells: 6200/mm³, thrombocytes: 318000/mm³, normal renal and liver function, electrolytes and acute phase reactants, elevated



Figure 1. Cardiomegaly as seen on teleradiogram

troponin-I at 27.3 ng/mL (normal range: 0.02-0.06 ng/mL) and elevated creatine kinase-MB at 12.6 ng/mL (normal range: 0-5 ng/mL). Echocardiography showed dilated left heart cavities and impaired LV systolic function (13% shortening fraction) (Figure 3). The right coronary artery (RCA)/aortic annulus ratio was 0.2. The patient had first degree mitral valve regurgitation (MVR); the chordae had the appearance of mild fibroelastosis. Her RCA outlet was dilated and LCA arose from the PA, resulting in a diagnosis of ALCAPA syndrome and secondary DCM (Figure 4). The patient was started on anticongestive treatment and underwent

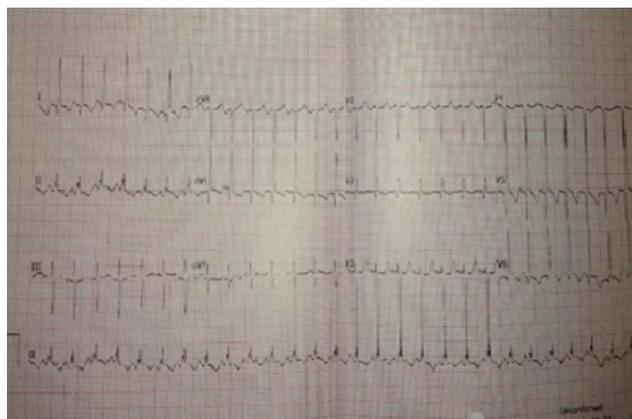


Figure 2. Broad Q waves and left ventricular hypertrophy findings can be observed on electrocardiogram

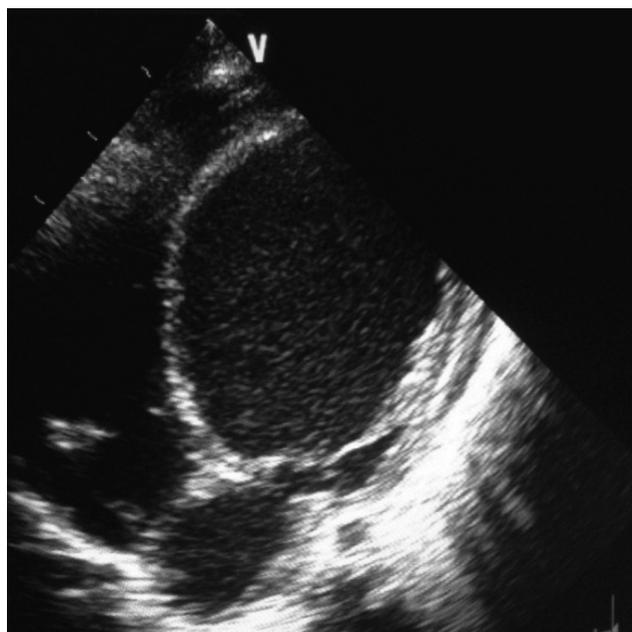


Figure 3. Echocardiographic image in the parasternal long-axis view showing severe dilation of left ventricle

reparative surgery using the direct reimplantation technique. The control echocardiography sessions showed the origin of ALCA at the aorta in addition to the presence of flow and progressive improvement of systolic function. She recovered as confirmed by clinical and echocardiographical assessments and she was discharged.

Discussion

ALCAPA syndrome accounts for 0.25-0.5% of all congenital heart diseases and is more prevalent in males. It was first described in 1933 by Bland, White and Garland. In ALCAPA syndrome, the ALCA arises from the PA or its branches. Anomalous origin of RCA at PA is infrequent, and the origin of both coronary arteries at PA is extremely infrequent (1-7).

ALCAPA syndrome generally occurs as an isolated cardiac anomaly but can also be concomitant with congenital heart defects such as patent ductus arteriosus, ventricular septal defect, pulmonary stenosis, Tetralogy of Fallot and aortic coarctation (5,8-10).

This condition poses no threats during the fetal period as the pressure and oxygen saturation in the aorta and PA are equal (5). While patients are generally asymptomatic at birth, PA pressure eventually falls, the flow from PA to coronary artery gradually decreases, LV myocardial perfusion is disrupted, leading to progressive ischemia and LV dilation. As a compensatory mechanism, RCA dilates and collateral vessels are formed between RCA and

ALCA. Symptoms manifest themselves in the 2nd-3rd month of life in 80-90% of cases (3,8). Clinical signs include angina-related pain in the form of acute distress, feeding problems, weight loss, tachycardia, tachypnea and sudden death. Eventually, cardiomegaly and congestive heart failure develop. LV performance and onset of symptoms depend on PA pressure and collaterals (5,8-11). Our patient was asymptomatic at birth and was referred to us at 2.5 months of age with heart failure findings.

In cases with congestive failure or DCM, telerradiograms show pronounced cardiomegaly. Eventually, infarction develops in the LV anterolateral area with ECG showing deep Q and inverted T waves in D1, aVL and left precordial leads (2,3,5,12). We observed similar findings in our patient.

Echocardiographic findings with ALCAPA include enlarged left heart cavities and RCA, ALCA at PA instead of aorta, LV systolic dysfunction, MVR, hyperechogenic papillary muscles, RCA/aortic annulus diameter ratio of over 0.14 and left-to-right shunt through interventricular septum on color Doppler ultrasound image. MVR may be caused by mitral valve dilation or papillary muscle infarction. ALCAPA must be considered in the etiology of patients born with MVR. When coronary artery anomalies are suspected, but echocardiography is inconclusive, it is advisable to use computed tomography angiography, magnetic resonance imaging and coronary angiography (2-6,10-13). Our patient had typical echocardiographic findings and a correct diagnosis was reached without further studies.

In children with clinical signs of DCM, the treatable ALCAPA syndrome must be kept in mind as one of the possible causes. Most of these patients were examined using sophisticated tests for metabolic diseases and they were eventually classified as idiopathic in most of the cases. Diagnosis of ALCAPA is a definitive indication for surgery. Those who receive exclusively medical treatment are at high risk of mortality. While medical treatment supports the deteriorating myocardial function, early diagnosis and surgical intervention ensure a more rapid improvement. An important factor affecting postoperative mortality is severe LV dysfunction and MVR.

Previous studies have found that ALCAPA patients do not receive a correct diagnosis after the first



Figure 4. Echocardiographic image in the parasternal short-axis view showing the aorta and the left coronary artery (marked with arrows) arising from the pulmonary artery

echocardiographic evaluation in 66-78% of cases. In order to diagnose ALCAPA, one must look for indirect findings (14,15). If a neonate presents with bronchiolitis and heart failure, displays cardiomegaly on telecardiogram and pathologies on ECG, then DCM and ALCAPA must be considered in terms of differential diagnosis (2,5,8-10,13).

The most common surgical method for treating ALCAPA is intrapulmonary tunnel repair, also known as Takeuchi repair, or direct reimplantation of ALCA, which ensures dual coronary circulation. The direct transfer technique is preferred today on account of the coronary artery transfer experience built up during arterial switch operations. Long-term echocardiographic follow-up is necessary after the surgical correction of ALCAPA syndrome (10,12-14). In our patient, the coronary artery was directly re-implanted into the aorta, and the follow-up showed gradual improvement of systolic function and reduction of left heart size.

Conclusion

DCM is the most common cardiomyopathy among children, and is generally idiopathic or secondary to viral myocarditis. A more rare cause of DCM is ALCA from PA. The correctable ALCAPA syndrome must be taken into account for differential diagnosis of DCM in infants.

Ethics

Informed Consent: It was taken.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Osman Güvenç, Murat Saygı, Concept: Osman Güvenç, Alper Güzeltaş, Design: Murat Saygı, Alper Güzeltaş, Data Collection or Processing: Osman Güvenç, Murat Saygı, Analysis or Interpretation: Murat Saygı, Alper Güzeltaş, Literature Search: Erkut Öztürk, Murat Saygı, Writing: Osman Güvenç.

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