Anomalous origin of the left coronary artery from the pulmonary artery presenting as dilated cardiomyopathy

Sol koroner arter pulmoner arterden köken aldığı dilate kardiyomiyopatiyi taklit eden bir olgu


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Summary– Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly and one of the causes of myocardial ischemia. It often presents with atypical signs and symptoms, especially in childhood. In this case report, an 11-year-old girl presented with dilated cardiomyopathy in our clinic and was followed for five years. Echocardiography showed multiple left-to-right shunts on the interventricular septum, the confirmation of which was done by multi-slice computed tomography and coronary angiography. Therefore, we suggest that ALCAPA should be suspected in young patients diagnosed with dilated cardiomyopathy.

CASE REPORT

An 11-year-old girl was admitted for follow-up examination of DCM, which had been diagnosed five years ago. Findings on physical examination were: weight: 28 kg (3-10 percentile), length: 141 cm (25-50 percentile), heart rate: 88/min, and blood pressure: 100/70 mmHg; a 2/6 systolic murmur was heard at the apex. Electrocardiography (ECG) showed inverted T waves in DI, AVL and left precordial leads. In telecardiography, cardiothoracic index was 62%. The left ventricle was larger than normal, there was systolic dysfunction (ejection fraction [EF] 50%), the right...
coronary artery was dilated (3.7 mm), and the right coronary artery/aortic annulus ratio was detected as 3.7/17=0.22 in ECHO. Color Doppler ECHO showed multiple left-to-right shunts on the interventricular septum (IVS). MSCT depicted that the left atrium and ventricle were larger than normal, the right coronary artery was normally located but hugely dilated and tortuous, and the left coronary artery (LCA) was abnormally traced to its origin from the pulmonary artery (Fig. 1a). A coronary angiography was performed before surgery. Contrast injection into the aorta demonstrated a gigantic right coronary artery which filled the LCA through extensive collaterals, and the origin of the LCA was from the pulmonary artery (Fig. 1b, Video 1*). After the diagnosis of ALCAPA syndrome, the case underwent surgical correction using the Takeuchi procedure (creation of an intrapulmonary tunnel). Follow-up ECHO revealed a mild systolic dysfunction one month after the surgery.

**DISCUSSION**

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital heart defect that affects approximately 1 in 300,000 live births and accounts for 0.5% of all congenital diseases.\(^2\) Due to low pressure in the pulmonary artery, the LCA cannot perfuse the myocardium. Collateral vessels develop between the right and left coronary arteries and may provide adequate perfusion of the left myocardium. As the pulmonary resistance decreases further, there is retrograde flow from the high-pressure coronary arteries to the pulmonary trunk. This is known as myocardial steal and further contributes to myocardial ischemia. Over time, there is anterolateral myocardial infarction, mitral valve dysfunction and congestive cardiac failure.\(^{[5]}\) During the first admission, the studied case was diagnosed as DCM with left ventricle dysfunction and dilatation. When the LCA arises from the pulmonary artery, this anomaly can cause myocardial ischemia that may be progressive and lead to left ventricular dysfunction, congestive heart failure and sudden cardiac death. Asymptomatic presentation after infancy and survival into adulthood by the collaterals is extremely rare.\(^{[6,7]}\) In the diagnosis of ALCAPA, clinic findings are insufficient. Chest radiography can reveal cardiomegaly, and ECG can show acute and chronic anterolateral myocardial infarction findings. There can be pathological Q waves, elevated ST segment and inverted T waves in leads DI, AVL and V4-6. In the studied case, there was cardiomegaly on X-ray, and the ECG showed inverted T waves in the DI and V5-6 leads. Transthoracic echocardiography (TTE) can reveal the origin of the coronary arteries.

![Figure 1.](image-url)
However, in some cases, false-negative echocardiographic diagnosis of this condition is also possible. Robinson et al.\textsuperscript{[8]} reported three false-negative cases that mimicked the appearance of normal LCA. ALCAPA needs to be suspected with the existence of significant enlargement of the right coronary artery and dilated left ventricle with global hypokinesia in ECHO. Additionally, in ALCAPA, all older patients have multiple unusual color flow Doppler signals within the ventricular septum, representing septal coronary collaterals. In these patients, retrograde-filling LCA connected to the main pulmonary artery can be diagnostic. Some authors have described systematic evaluation methods to distinguish between ALCAPA and DCM using a systematic approach. For example, Chang et al.\textsuperscript{[9]} detected the echocardiographic feature of right coronary artery diameter to aortic annulus ratio as ≥0.14 whereas Schmidt et al.\textsuperscript{[10]} reported the ratio of the diameters of the right coronary artery and the aortic root as 0.25 to 0.33 (normal: 0.12±0.02). In our case, ECHO showed left ventricle dilatation, dysfunction of the left ventricle, and enlarged right coronary artery (right coronary artery/aortic annulus=0.22). There were multiple color flow Doppler signals on the base of the IVS. Evaluation of ECG and ECHO are important in the diagnosis of ALCAPA. However, a precise diagnosis is made by MSCT and/or coronary artery angiography. In our case, the diagnosis was confirmed by MSCT, and a coronary angiography was performed before surgery. After the diagnosis via an intrapulmonary tunnel, the LCA was related to the aorta with surgical intervention (Takeuchi procedure).

In conclusion, in ALCAPA syndrome, left ventricle dilatation and dysfunction can be misdiagnosed as DCM. In cases of DCM, ALCAPA syndrome has to be kept in mind as a surgically correctable cause.

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*Supplementary video file associated with this article can be found in the online version of the journal.

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