Two adult cases of anomalous left coronary artery from the pulmonary artery

Pulmoner arterden köken alan sol koroner arter anomalisi olan iki erişkin hasta

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Summary – Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly that presents as left-sided heart failure and mitral valve insufficiency during the first months of life. However, some cases may benefit from sufficient collateral blood supply from the right coronary artery, resulting in increased survival even to advanced ages. Herein, we report on two adult cases of ALCAPA, namely, a 52-year-old male patient that presented with angina and a 70-year-old female patient presenting with stroke, dyspnea, and pretibial edema. In both cases, ALCAPA was demonstrated by coronary angiography and multislice computed tomography angiography. The younger patient refused surgery and remained asymptomatic and event-free during a two-year follow-up with anti-ischemic medications, while the older with high surgical risk was considered for intracardiac defibrillator implantation along with medical therapy.

Özet – Sol ana koroner arterin pulmoner arterden köken alması (ALCAPA) çok nadir görülen bir doğuştan anomalidir. Hastalık genellikle yaşamın ilk aylarında sol kalp yetersizliği ve mitral kapak yetersizliği ile kendini gösterir. Ancak, bazı hastalarda sağ koroner arterden yeterli kolateral kan akımının sağlanması, sağkalımı ileri yaşlara kadar artırabilir. Bu yazıda, biri angina yakınmasıyla başvuran 50 yaşında erkek, diğeri inme, nefes darlığı ve bacaklarda şişlik ile başvuran 70 yaşında kadın olmak üzere, ileri yaşlarda ALCAPA tanısı konan iki hasta sunuldu. Her iki olguda da anomali koroner anjiyografi ve çokkesitli bilgisayarlı tomografi anjiyografi ile gösterildi. Daha genç yaştaki hasta ameliyatı kabul etmedi ve antiiskemik ilaçlarla sürdürülen iki yıllık takibi semptomsuz ve olaysız geçirdi. Daha yaşlı hastada ise, cerrahi riski yüksek olduğundan medikal tedavi yanı sıra intrakardiyak defibrilatör uygulaması planlandı.

Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome) is a rare congenital anomaly with an incidence of 1 in 300,000 births.[1,2] Both pulmonary vascular resistance and pulmonary arterial pressure decrease after birth, with corresponding decreases in oxygen content of the pulmonary artery, antegrade flow, and oxygen content of the anomalous left coronary artery, resulting in myocardial ischemia. Collateral circulation develops between the right and left coronary systems, whereby reversed left coronary artery flow enters the pulmonary trunk as a result of decreased pulmonary arterial pressure (coronary steal phenomenon); hence, inadequate perfusion of the myocardium. This sequence of events is associated with severe left-sided heart failure and significant mitral valve insufficiency, occurring within the first two months of life, with corresponding symptoms such as difficulty in feeding, irritability, diaphoresis, tachypnoea, and tachycardia. However, some cases may have relatively minor symptoms throughout childhood due to sufficient collateral supply from the right coronary artery and may lead further life with complications ranging from dyspnea, chest pain, and exercise intolerance to sudden car-

Abbreviations:

ALCAPA	*Anomalous origin of the left coronary artery from the pulmonary artery*
CT	*Computed tomography*
ICD	*Implantable cardioverter defibrillator*
LMCA	*Left main coronary artery*

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diac death due to acute ischemia or malignant ventricular arrhythmias.[3,4]

CASE REPORT

Case 1– A 52-year-old man was admitted to our outpatient clinic with worsening exercise capacity for the past six months. He complained of an exertional backache relieved by rest. He had an uneventful childhood. Physical examination was normal except for a holosystolic murmur at the mitral point. He was hemodynamically stable. The electrocardiogram showed ST/T-wave changes in leads I, AVL, and V6, suggesting lateral ischemia. Telecardiography was normal. Transthoracic echocardiography showed mildly decreased left ventricular systolic functions (ejection fraction 45%), mild mitral regurgitation, and reversed flow in the pulmonary artery. A single-photon emission computed tomography scan revealed an area of infarction at the apex and hypoperfusion in the mid and basal anterolateral wall. Coronary angiography showed ALCAPA with a retrograde filling through collaterals from an enlarged right coronary artery. Negative contrast was demonstrated on pulmonary angiography. For better visualization, multislice coronary CT angiography was undertaken, which also showed an anomalous left main coronary artery originating from the left pulmonary artery. A retrograde flow from the left anterior descending artery and left main coronary artery was filling the pulmonary artery (Fig. 1). Operation was proposed to the patient, but he refused surgery. He was discharged with medical therapy. During a two-year follow-up with anti-ischemic medications, the patient remained asymptomatic and event-free.

Case 2– A 70-year-old woman was referred to our clinic with ischemic stroke and pretibial edema. The patient had an uneventful childhood. On physical examination, she had dyspnea, 2+ pretibial edema, a 3/6 holosystolic murmur, and basilar crepitant rales. The electrocardiogram showed ST/T-wave changes in precordial leads, suggesting anterior ischemia. Telecardiography showed cardiomegaly. Transthoracic echocardiography showed decreased cardiac systolic functions (ejection fraction 35%), ventricular dilation with an anteroapical aneurysm containing a mural thrombus, moderate mitral regurgitation, and a reversed flow in the pulmonary artery. Single-photon emission CT demonstrated infarct areas in the anterior and apical segments and hypoperfusion in the mid anterolateral wall and basal segment. Coronary angiography showed ALCAPA with a retrograde filling through collaterals from an enlarged right coronary

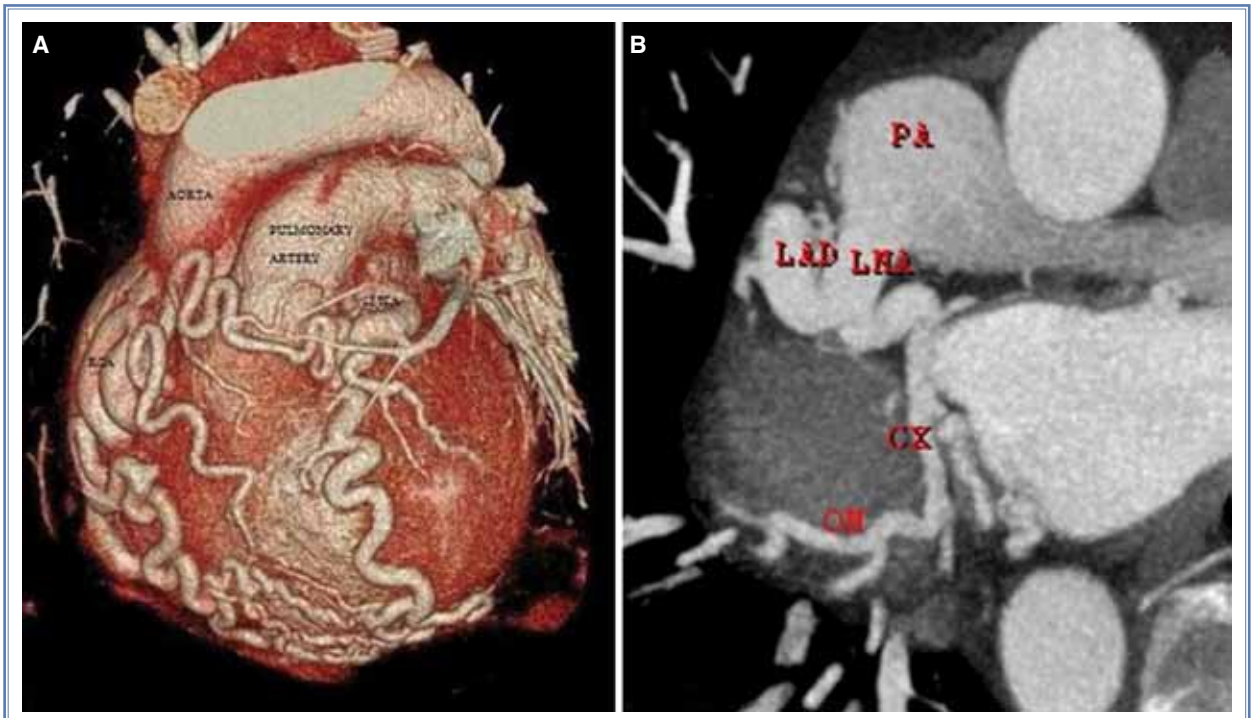

Figure 1. (A) Computed tomography angiography with three-dimensional reconstruction shows anomalous left coronary artery originating from the pulmonary artery. **(B)** A computed tomography angiography scan shows anomalous left coronary artery (LMA) originating from the pulmonary artery (PA), with its branches, the left anterior descending artery (LAD) and circumflex artery (Cx).

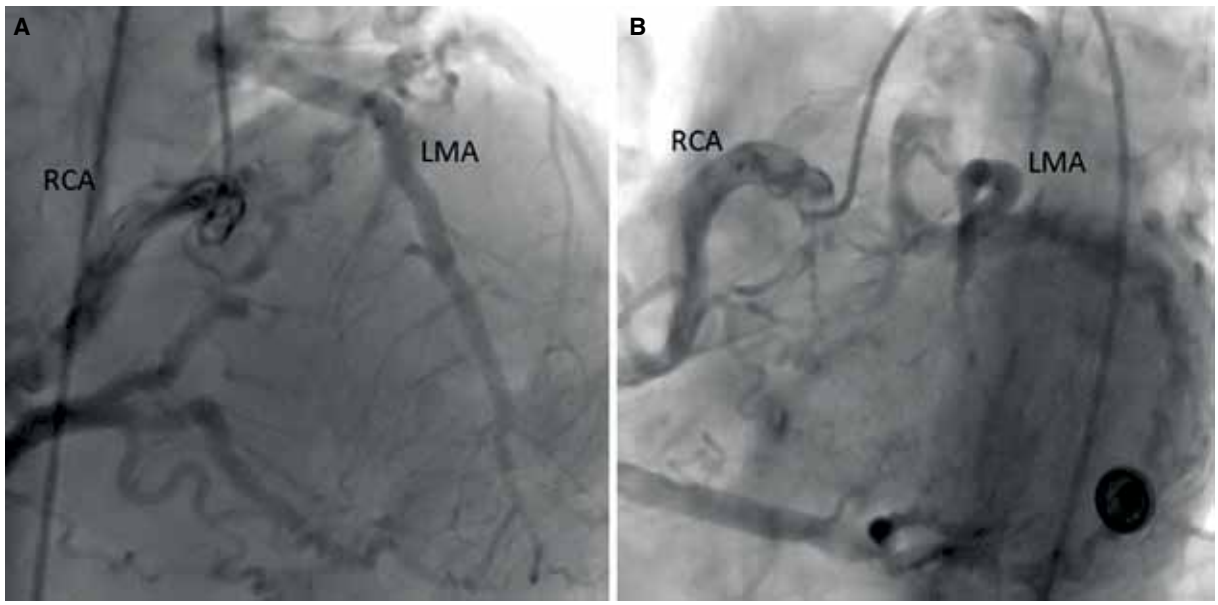

Figure 2. (A, B) Coronary angiograms show an ectatic right coronary artery (RCA) giving collaterals to the anomalous left coronary artery (LMA) originating from the pulmonary artery.

artery (Fig. 2). Negative contrast was demonstrated on pulmonary angiography. Multislice coronary CT angiography also showed an anomalous LMCA originating from the left pulmonary artery. The left anterior descending artery and LMCA were filling the pulmonary artery through a retrograde flow. Since the patient carried a high surgical risk and was reluctant to surgery, operation was not performed. Intracardiac defibrillator implantation was planned and the patient was discharged on optimal medical therapy.

DISCUSSION

The majority of cases (85%) with ALCAPA are diagnosed within the first months of life. However, some patients may have gone unnoticed because of misinterpretation, subtleness, or absence of symptoms, until the development of heart failure, mitral valve insufficiency, angina (as in our case), or arrhythmias in adult life. Both of our patients had dominant right coronary arteries that provided adequate collateral blood supply to the left coronary circulation, enabling the patients to survive till late adult ages. However, the maintenance of cardiac functions through the collaterals became insufficient, resulting in ischemic areas. Patients with ischemic cardiomyopathy are prone to develop malignant arrhythmias and sudden cardiac death due to arrhythmogenic potential of scar tissue. Implantable cardioverter defibrillator therapy is indicated in patients with ischemic cardiomyopathy and decreased left ventricular ejection fraction.[5] In the first case, ICD therapy was not considered because he refused surgery and his left ventricular ejection fraction was 45%. In the second case, however, there was a high surgical risk and her ejection fraction was 35%, so the patient was scheduled to ICD implantation.

Cardiomegaly on chest X-ray and an anterolateral infarct pattern on the electrocardiogram may be suggestive of ALCAPA. Although two-dimensional echocardiography provides direct visualization of the abnormal origin of the left coronary artery and retrograde flow into the pulmonary artery in pediatric patients, it may be difficult to visualize the origins of the coronary arteries in adults; thus, coronary angiography or CT angiography may be required. We performed multislice CT angiography in both patients and obtained three-dimensional construction of the anatomy with better delineation and spatial resolution compared to coronary angiography.[6-8]

Medical therapy is indicated in patients with ischemic heart disease to prevent deterioration in cardiac functions and fibrosis as well as to obtain symptomatic relief.[9] Since ALCAPA and ischemic heart disease have common features, patients with ALCAPA and worsening left ventricular functions may benefit from medical therapy. However, no data exist regarding the benefits of medical therapy in adult patients with ALCAPA syndrome. On the other hand, several surgical treatment options have been proposed for cas-

es of ALCAPA. Normalization of both mitral valve insufficiency and ventricular function was achieved in most infants with corrected ALCAPA,[10,11] yielding an estimated long-term survival rate of 95% at 20 years.[12] Considering the likelihood of spontaneous improvement in mitral valve function, conservative treatment is often recommended against surgical correction of mitral valve insufficiency. Corrective surgery in adult patients may well provide revitalization of ischemic areas, but long duration of ischemia may cause irreversible damage, resulting in scar tissue.[11] In patients unsuitable for or reluctant to surgery, ischemic areas may expand over time, but medical therapy in such cases may slow down the process and provide survival benefit and symptomatic relief, as in patients with ischemic heart disease.

In conclusion, medical therapy may be safe and feasible in adults under limited circumstances, especially in patients refusing or unsuitable for surgery. As long as the collateral blood supply from the right coronary artery is sufficient to maintain basal cardiac functions, and because the problem arises from ischemia due to inadequate vascular nourishment, patients may benefit from anti-ischemic medications, as seen in ischemic heart disease patients. Furthermore, just as in ischemic cardiomyopathy patients, ICD therapy may offer an extra survival benefit in patients with ALCAPA syndrome with decreased left ventricular systolic functions.

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Key words: Adult; coronary angiography; coronary vessel anomalies; echocardiography; pulmonary artery/abnormalities; syndrome; tomography, X-ray computed.

Anahtar sözcükler: Erişkin; koroner anjiyografi; koroner damar anomalisi; ekokardiyografi; pulmoner arter/anormallik; sendrom; bilgisayarlı tomografi.