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Case Report / Olgu Sunumu

Anomalous origin of the left coronary artery from the pulmonary artery syndrome in an adult and his eight-year follow-up results

Erişkin bir hastada sol koroner arterin pulmoner arterden çıkış anomalisi sendromu ve sekiz yıllık takip sonuçları

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) which is also known as Bland-White-Garland syndrome, is a rare congenital anomaly. If it is left untreated, ALCAPA syndrome has a high mortality in the first year of life due to mostly myocardial ischemia and heart failure. Most adult patients may be asymptomatic or present with angina, dyspnea, syncope, myocardial infarction, arrhythmia or sudden cardiac death. Sudden cardiac death secondary to malign ventricular arrhythmias is the most common presentation in this patient population. In this article, we report a 36-year-old male case who was an active sportsman and presented with atypical complaints. Transthoracic echocardiography revealed an indefinite turbulence in pulmonary artery and coronary angiography showed ALCAPA syndrome.

Keywords: Adult; coronary angiography; coronary vessel anomaly.

Bland-White-Garland sendromu olarak da bilinen sol koroner arterin pulmoner arterden çıkış anomalisi (ALCAPA), çok nadir görülen bir doğuştan anomalidir. Tedavi edilmediği takdirde, ALCAPA sendromu çoğunlukla miyokard iskemisi ve kalp yetmezliği nedeniyle yaşamın ilk yılında yüksek mortalite ile seyreder. Erişkin hastaların birçoğu asemptomatik olabilir ya da bu hastalarda anjina, nefes darlığı, baygınlık, miyokard enfarktüsü, aritmi veya ani kardiyak ölüm görülebilir. Malign ventriküler aritmilere sekonder ani kardiyak ölüm, bu hasta grubunda en sık görülen durumdur. Bu yazıda, aktif bir sporcu olan ve atipik yakınmaları olan 36 yaşında bir erkek olgu sunuldu. Transtorasik ekokardiyografide pulmoner arterde tanımlanamayan türbülans tespit edilmesi üzerine yapılan koroner anjiyografide ALCAPA sendromu saptandı.

Anahtar sözcükler: Erişkin; koroner anjiyografi; koroner arter anomalisi.

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), or Bland-White-Garland syndrome, is a rare congenital anomaly.[1] If left untreated, ALCAPA has a 90% mortality rate in the first year of life primarily due to myocardial ischemia and heart failure.[1] Herein, we present the case of a patient who was an active sportsman and had atypical complaints. Since he had an indefinite turbulence in the pulmonary artery during transthoracic echocardiography TTE, we performed coronary angiography and detected ALCAPA.

CASE REPORT

A 36-year-old male patient was admitted to the Sanko Hospital Cardiology Clinic with a complaint of left-sided atypical chest pain in 2005. He was an active sportsman (professional football player) without any known diseases or drug use, and his

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physical examination revealed normal vital signs with a pansystolic murmur at the mesocardiac and pulmonary areas. Electrocardiography (ECG) showed sinus rhythm without any pathological finding, and the TTE was suboptimal due to poor echogenicity and revealed normal systolic functions with normal-sized heart chambers, mild mitral and tricuspid regurgitation, and a pulmonary artery pressure (PAP) of 40/18 mmHg. In the parasternal short-axis view, an indefinite turbulent flow was detected in the pulmonary artery by color and continuous wave Doppler. In addition, there were no ischemic ST-T changes in the exercise stress test. Soon afterwards, the patient underwent cardiac catheterization, and since the left coronary artery (LCA) could not be seated by left Judkins and Amplatz catheters (Medtronic Inc., Minneapolis, MN, USA) in the first step, the operator shifted to right coronary angiography in which the right coronary artery (RCA) was dilated. This detected well developed coronary collaterals filling the LCA leading to pulmonary artery washing by a reverse flow (Figures 1a and 1b). In coronary computed tomography (CT), the LCA was shown arising from the pulmonary artery and perfused by collaterals directly from the aorta and RCA (Figures 1c and 1d). Surgical correction was planned, but the patient refused this option. Since that time we have been performing TTE at six-month intervals along with an annual 24-hour ECG Holter analysis and detected no changes nor any ventricular or supraventricular tachycardia, bradycardia, or ventricular couplets and triplets. The patient has been under follow-up since 2005 and remains asymptomatic.

DISCUSSION

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital malformation

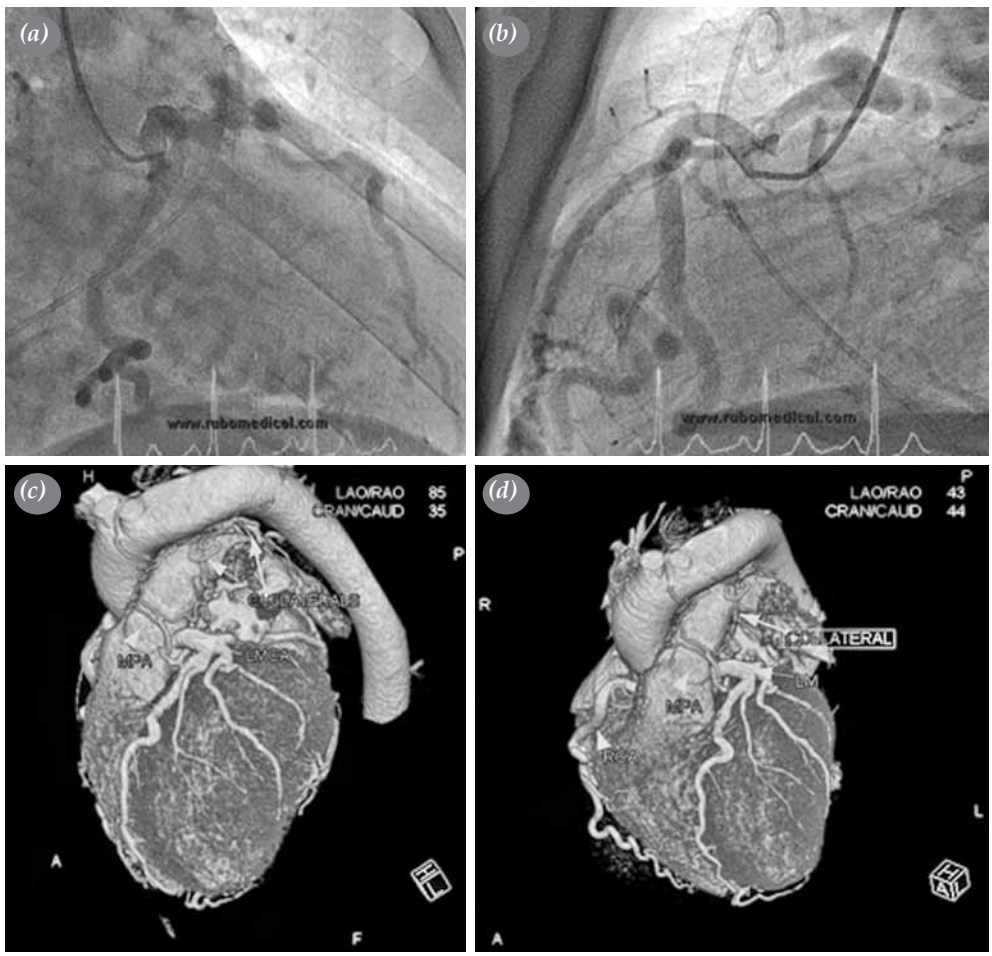

Figure 1. (a, b) Right coronary angiograpy showing the well developed coronary collaterals filling the left coronary artery leading to pulmonary artery washing by a reverse flow. **(c, d)** Coronary computed tomography showing the left coronary artery arising from the pulmonary artery and perfused by collaterals directly from the aorta and right coronary artery.

which occurs in 1/300,000 live births and constitutes about 0.25-0.5% of all congenital heart defects.[1]

In ALCAPA, the systemic arterial pressure (SAP) and PAP as well as the oxygen saturation (OS) levels are equal during the fetal period.[2] The pulmonary pressure falls and the ductus arteriosus closes in the neonatal period. The flow of the LCA reverses, and the perfusion pressure of this artery falls after closure of the ductus.[3] The major factor for determining the symptoms in patients with this condition during this period is the development of collaterals, and myocardial ischemia, and the degree of symptoms due to ischemia depends on the development of collaterals between the RCA and LCA.[3] Patients with well developed collaterals are defined as having adult-type ALCAPA while those without collaterals have the infantile-type of this syndrome.[3] Unless the latter type is diagnosed and treated, 90% of children die in their first year of life because of mitral regurgitation due to widespread myocardial ischemia and heart failure.[1,4]

Adult patients can be completely asymptomatic or present with angina, dyspnea, syncope, myocardial infarction (MI), arrhythmias, or sudden cardiac death,[5] with sudden cardiac death secondary to malignant ventricular arrhythmias being the most common presentation.[5] Today, an increase in the number of diagnostic procedures helps us to detect ALCAPA in older patients; however, well developed collaterals, symptoms, and life-threatening arrhythmias occur less frequently in older patients.[5] Tian et al.,[2] conducted a study that focused on the short-term follow-up of asymptomatic patients who refused surgical therapy and found that all were asymptomatic.

Echocardiography can show the abnormal origin of the LCA from the pulmonary artery, the left coronary artery flow via Doppler imaging, the dilated RCA, severe left ventricular dysfunction, and mitral regurgitation.[1,5] Since our patient had poor echogenicity, only an indefinite turbulent flow in the pulmonary artery could be detected by color and continuous wave Doppler.

Coronary angiography usually aids in the diagnosis by showing a dilated and tortuous RCA with collaterals to the LCA system along with variable degrees of shunting to the pulmonary artery.[6] In our case, the coronary angiography revealed that the LCA arose from the pulmonary artery, and aortography detected that that artery had collaterals arising directly from the aorta.

Coronary CT angiography is an important, noninvasive diagnostic tool used to delineate the abnormal origin, projection, and collaterals of coronary arteries.[2] In our case, we performed coronary CT in which we detected the abnormal origin of the LCA from the pulmonary artery and visualized the course of the LCA with its collaterals arising directly from the aorta.

The most important reason for symptoms associated with ALCAPA is deficient perfusion in the LCA region. The better the collaterals are, the better the myocardium perfuses. In our patient, the collaterals arose directly from the aorta and the ones from the RCA provided better perfusion in the LCA region, which contributed to the patient remaining asymptomatic.

Surgical correction is recommended in adult patients, especially those with symptoms and large left-to-right shunts; however, many authors have reported that surgery should also be performed on asymptomatic individuals in order to prevent ventricular arrhythmias, myocardial ischemia, and sudden death.[3] We recommended surgical correction to our patient, but he refused this option. He has been coming for regular controls since he was diagnosed in 2005 and has been taking part in sports activities with lower intensity without experiencing any symptoms.

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital anomaly and in patients with this condition direct branches from aorta to LCA which provides good coronary perfusion should taken into consideration and aorta should be evaluated thoroughly.

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