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Case Report / Olgu Sunumu

ALCAPA syndrome in an asymptomatic young soccer player

Asemptomatik genç bir futbol oyuncusunda ALCAPA sendromu

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ABSTRACT

Anomalous origin of left coronary artery from pulmonary artery syndrome is a rare, but severe congenital cardiac malformation. It is an important cause of dilated cardiomyopathy and left heart failure during infancy and, if left untreated, the prognosis is poor with an overall mortality rate over 90%. About 15% of patients can survive beyond the first year of life, depending on the development of collateral circulation and may present with angina, dyspnea, syncope, and arrhythmias. Myocardial infarction and sudden cardiac death may be the only and the first symptom in some cases. The treatment of choice for this syndrome is urgent surgical intervention with favorable long-term outcomes. Herein, we present an asymptomatic adolescent active sportsman who was diagnosed with anomalous origin of left coronary artery from pulmonary artery syndrome and underwent a successful surgery.

Keywords: ALCAPA, asymptomatic, coronary anomaly, left heart failure.

ÖZ

Pulmoner arterden kaynaklanan sol koroner arter anomalisi sendromu nadir, ama ciddi bir doğuştan kalp anomalisidir. Bebeklik döneminde dilate kardiyomiyopatinin ve sol kalp yetmezliğinin önemli bir nedeni olup, tedavi edilmez ise, prognozu kötüdür ve genel mortalitesi %90'ın üzerindedir. Hastaların yaklaşık %15'i kollateral dolaşım gelişimine bağlı olarak bir yaşından sonra hayatta kalabilir ve bu hastalarda göğüs ağrısı, nefes darlığı, senkop ve aritmiler görülebilir. Bazı olgularda miyokard enfarktüsü ve ani kardiyak ölüm ilk ve tek semptom olabilir. Bu sendromun tedavisi acil cerrahi düzeltme olup, uzun dönem sonuçları iyidir. Bu yazıda, pulmoner arterden kaynaklanan sol koroner arter anomalisi tanısı konulan ve başarılı bir cerrahi yapılan asemptomatik ergen aktif bir sporcu sunuldu.

Anahtar sözcükler: ALCAPA , asemptomatik, koroner anomali, sol kalp yetmezliği.

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a rare congenital coronary artery malformation. It is an important cause of left heart failure during infancy and, if left untreated, the overall mortality is over 90%.[1] Herein, we present an asymptomatic adolescent active sportsman who was diagnosed with ALCAPA and underwent a successful surgery to highlight the importance of meticulous evaluation of coronary arteries in each patient, considering the fact that ALCAPA syndrome may present in adulthood without any signs or symptoms.

CASE REPORT

An 11-year-old boy who was an active sportsman (licensed soccer player of a local soccer club academy) without any complaints, was admitted for cardiac evaluation upon his request. He had no history of familial cardiac disease or sudden death. Three years ago, he was evaluated in another institution and was diagnosed with multiple small ventricular septal defects (VSDs). Physical examination findings were normal. Electrocardiography (ECG) showed normal sinus rhythm and mild (<1 mm) ST changes in leads

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V4-V6 and lead III (Figure 1a). Echocardiography (ECHO) revealed normal sized cardiac chambers, an ejection fraction of 65%, and shortening fraction of 35%. The multiple intercoronary collateral flows, which were previously misinterpreted as multiple small VSDs, within the ventricular septum were detected by color Doppler (Figure 1b). The origin of the left coronary artery (LCA) was unable to be detected; however, the mosaic flow pattern in the pulmonary artery (PA) (Figure 1c) and the reverse flow in the LCA (Figure 1d) were detectable. In addition, the right coronary artery (RCA) was remarkably dilated (5.5 mm, Z-score: +5.23) (Figure 1e). Catheter angiography showed an anomalous origin of the LCA from PA and multiple intercoronary collateral arteries (Figure 1f). The patient was scheduled for surgery and a written informed consent was obtained from each parent.

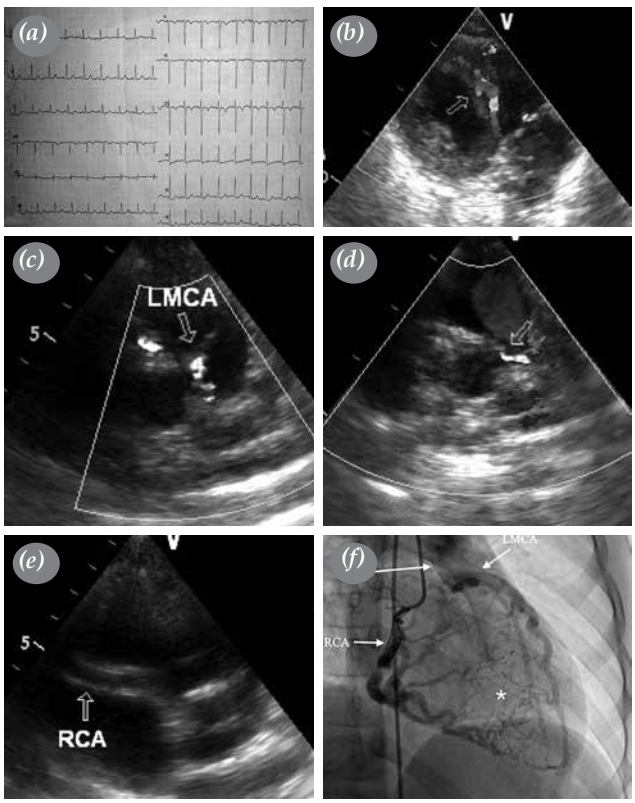

Figure 1. (a) ECG showing mild (<1 mm) ST changes in leads V4-V6 and lead III. **(b)** ECHO view showing multiple intercoronary collateral flows, which were previously misinterpreted as multiple small VSDs. **(c)** A parasternal short axis echocardiographic view with the mosaic flow pattern in pulmonary artery. **(d)** A parasternal short axis echocardiographic view showing reverse flow in LCA. **(e)** Remarkably dilated right coronary artery on parasternal short axis view. **(f)** Catheter angiography of the patient showing remarkably dilated right coronary artery, well-developed network of intercoronary collateral vessels, origin of left coronary artery from pulmonary artery.

LMCA: Left main coronary artery; RCA: Right coronary artery; PA: Pulmonary artery; ECG: Electrocardiography; ECHO: Echocardiography; VSD: Ventricular septal defect; LCA: Left coronary artery.

The patient underwent a successful coronary reimplantation surgery. After median sternotomy, standard aortic arterial and bicaval venous cannulation were utilized for cardiopulmonary bypass (CPB). After the onset of CBP, the right and left PA were snared. Myocardial protection with mild hypothermia was achieved via Del Nido antegrade cardioplegia. Pulmonary arteriotomy was applied horizontally and the slit-like ostium of the LCA, which was at the close sinus to aorta, was visualized. Coronary artery was removed from the pulmonary sinus and, then, anastomosed to the adjacent aortic sinus prepared by punch resection before the anastomosis. The defect in the pulmonic sinuses was reconstructed by using glutaraldehyde-fixed autologous pericardium. The patient was weaned from CPB lasting 62 min after removing cross-clamp lasting 41 min. Control ECG showed mild ST changes (Figure 2a) and control ECHO

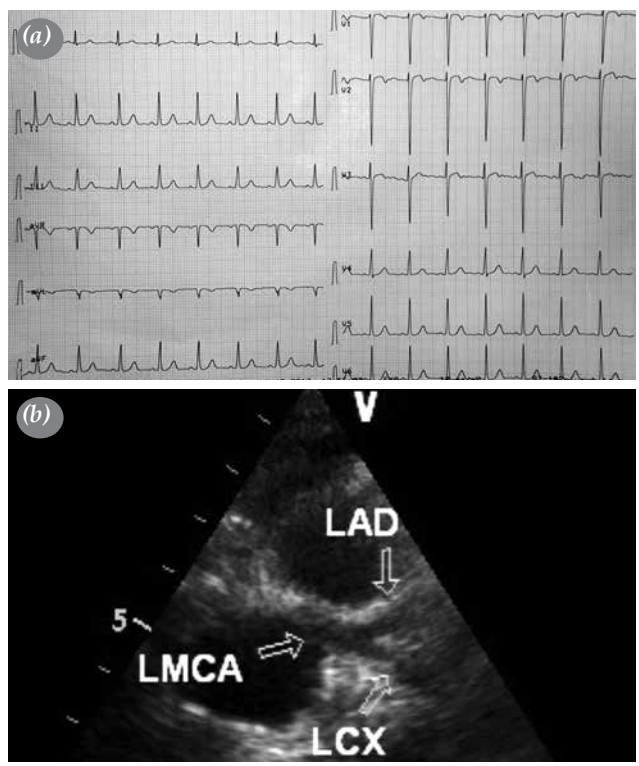

Figure 2. (a) ECG of the patient six months after surgery showing mild ST changes. **(b)** Parasternal short axis ECHO view of the patient after surgical correction showing left main coronary artery and its main branches.

LAD: Left anterior descending coronary artery; LMCA: Left main coronary artery; LCX: Left circumflex coronary artery; ECG: Electrocardiography; ECHO: Echocardiography.

revealed normal heart functions and no pathological findings (Figure 2b). The patient was discharged from the hospital on the postoperative Day 5 after one-day postoperative intensive care unit stay and is still under follow-up without any symptoms for nine months.

DISCUSSION

Anomalous origin of left coronary artery from pulmonary artery syndrome, also known as Bland-White-Garland syndrome, is a rare, but severe congenital cardiac malformation with an incidence of 1 in 300,000 liveborn.[1] In this syndrome, as its name indicates, the left coronary artery branches of the PA, instead of the aortic sinus. At birth, there is an antegrade flow in the anomalous LCA due to the high pulmonary arterial pressure. Shortly after birth, when pulmonary arterial pressure decreases, antegrade flow in the LCA also decreases. As a result, most patients are usually asymptomatic, until the PA pressure decreases. With the development of collateral circulation between the right and left coronary systems, the flow in the LCA reverses and enters the pulmonic trunk due to the low pulmonary vascular resistance. Thus, there is a preferential blood flow into the low-pressure pulmonary circulation rather than into the high-resistance myocardial circulation. This left-to-right shunt is known as the steal phenomenon.[2,3]

Most patients present in infancy with varying symptoms of myocardial ischemia, left ventricular dysfunction, and progressive heart failure.[3] The prognosis of ALCAPA depends on the varying degree of collaterals developing between the LCA and RCA, when PA pressure gradually decreases. Patients with well-established collaterals have the adult type of the disease, and those without collaterals have the infant type.[2] In addition to the development of intercoronary collaterals, a slit-like ostium and/or restrictive opening between the LCA and PA are important anatomical factors leading to a better left coronary perfusion and longer survival by limiting coronary steal. Approximately 15% of patients can survive beyond the first year of life, depending on the development of collateral circulation. These patients may present with angina, dyspnea, syncope, myocardial infarction, arrhythmias, or sudden cardiac death.[4] Sudden death and myocardial infarction may be the first and the only symptom. Tachypnea, rapid and weak pulse, gallop rhythm, murmur of mitral incompetence and continuous murmur of intercoronary anastomosis and arrhythmias may be the signs of ALCAPA. Chest X-ray may show cardiomegaly. The ECG findings must be evaluated for signs of ischemia (pathological Q waves in leads I, AVL (augmented vector left) and V4-V6). Patients with a rich collateral circulation may have non-specific changes. Also, ECHO may show the abnormal origin of the LCA from the PA, the reverse flow in LCA, the dilated RCA, severe left ventricular dysfunction, and mitral regurgitation. Catheter angiography is performed to confirm the diagnosis. The treatment of choice for ALCAPA is urgent surgical intervention which mainly targets the correction of the coronary steal phenomenon. The simple ligation of ALCAPA, coronary artery bypass grafting, channel repair (Takeuchi surgery), and coronary artery reimplantation are the four surgical procedures recommended for the treatment. The reimplantation surgery, which is the ideal option to achieve a definitive two-coronary anatomy and physiology, has become the first-choice procedure for this anomaly with favorable short and long-term outcomes.[5,6]

Our patient was diagnosed with adult form of ALCAPA syndrome. There was no pathological Q wave or T wave inversion on ECG. Only mild ST changes (<1 mm) in leads V4-V6 and lead III were observed. The ECHO revealed normal sized cardiac chambers, an ejection fraction of 65%, no mitral regurgitation, collateral circulation that was previously misinterpreted as multiple small VSDs, dilated RCA, and reverse flow in LCA. Catheter angiography confirmed the diagnosis of ALCAPA. The reimplantation technique was preferred as the surgical treatment. It is the first-choice procedure for this anomaly in our institution, as we believe that, even in patients with sufficient collateral circulation, anatomical correction and the establishment of two-coronary system is important to avoid long-term morbidity and mortality. Besides the development of extensive intercoronary collaterals, slit-like ostium of the LCA was an important factor protecting our case from myocardial ischemia.

We present this case due to its rarity, as most patients with ALCAPA have congestive heart failure during infancy. However, very rarely, due to the development of extensive collaterals, some patients may survive beyond infancy. Although most patients with undiagnosed ALCAPA dies during infancy, it must be kept in mind that they may survive to adulthood without any signs and symptoms, although very rare. Therefore, careful evaluation of coronary arteries is of utmost importance. In case of any suspicion, even in asymptomatic patients, angiography must be performed to confirm the diagnosis, since the first symptom of ALCAPA may be sudden cardiac death. As it is a treatable cause of dilated cardiomyopathy, early and accurate diagnosis with an urgent surgical treatment is critical.

In conclusion, although most pediatric cardiologists pay attention to the evaluation of coronary arteries, particularly in patients with ventricular dysfunction and signs of myocardial ischemia, careful evaluation of coronary arteries in every patient is very important, as coronary artery anomalies such as anomalous origin of left coronary artery from pulmonary artery may present in adulthood without any sign and symptoms.

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