Surgery of Anomalous Left Coronary Artery from the Pulmonary Artery in an Adult

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ABSTRACT

A 38-year-old man with no medical history presented with dyspnoea on exertion. Coronary angiography and coronary computerised tomography (CT) angiography revealed anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). The left anterior mammary artery and venous graft were used as a double bypass graft to the left anterior descending coronary and obtuse margin arteries, respectively. The patient was asymptomatic on follow-up.

Key Words: ALCAPA syndrome; congenital coronary artery anomaly

Erişkinde Sol Koroner Arterin Pulmoner Arterden Çıkış Anomalisi Cerrahisi

ÖZET

Otuz sekiz yaşındaki erkek hasta, hastanemize yeni başlayan eforla nefes darlığı ile başvurdu. Yapılan koroner anjiyografi ve bilgisayarlı tomografik anjiyografinin sonuçlarına göre sol koroner arterin pulmoner arterden çıkışı anomalisi saptandı. LİMA sol desendan koroner artere, safen ven grefti ise obtus marjine anastomoz yaparak aortokoroner baypas greft yapıldı. Postoperatif dönemde yakınması olmayan hasta şifa ile taburcu edildi.

Anahtar Kelimeler: ALCAPA sendromu; konjenital koroner arter anomalisi

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as Bland-White-Garland syndrome, is a rare congenital abnormality that affects 1 in every 300,000 live births [(1)]. There are 2 types of ALCAPA syndrome: the infant type and the adult type, each having different manifestations and outcomes. Approximately 90% of infants with ALCAPA syndrome die within the first year of life due to myocardial infarction (MI) and congestive heart failure. The adult type of ALCAPA syndrome may cause MI, left ventricular dysfunction and mitral regurgitation, or silent myocardial ischaemia, which can lead to sudden cardiac death. Here we introduce a 38-year-old male patient who had undergone coronary artery bypass grafting at our centre due to ALCAPA syndrome.

CASE REPORT

A 38-year-old male presented to our hospital with complaints of dyspnoea on exertion; his functional capacity was class II according to the New York Heart Association (NYHA) classification. Physical examination of the patient showed that his blood pressure was 110/70 mmHg, his heart rate was 85 beats/min and there was no murmur on auscultation or pre-tibial oedema. Echocardiography revealed that the ejection fraction was 40%, there was mild mitral regurgitation, the pulmonary artery pressure was 30 mmHg and the left atrial volume was 3.1 cm. Coronary angiography was performed and showed that the collateral circulation between the right and left coronary arteries was well developed. The origin of the left main coronary artery (LMCA) was not observed properly; therefore, we decided to perform coronary computerised tomography (CT) angiography. Thus, the diagnosis of ALCAPA syndrome was confirmed and LMCA was obviously observed to be originating from the main pulmonary artery (MPA), as shown in Figure 1 and Figure 2. Surgical treatment was planned for the patient.

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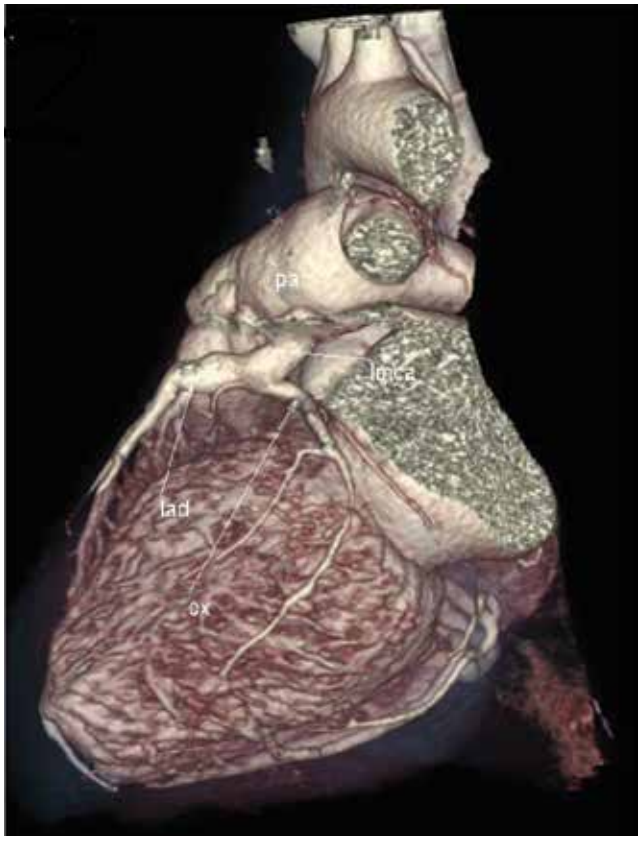

Figure 1. Lateral view of coronary of computerized tomography (CT) angiography shows that LMCA is originating from the main pulmonary artery (MPA).

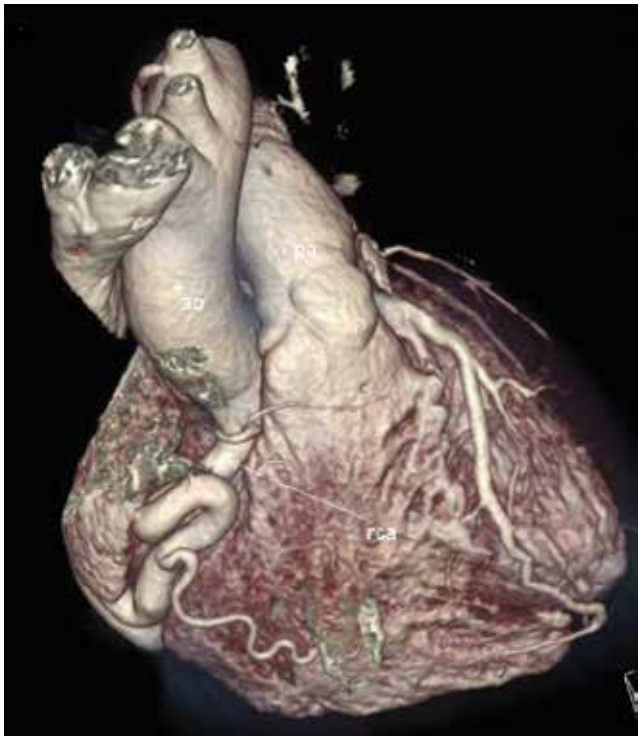

Figure 2. Anterior view of coronary computerized tomography (CT) angiography shows that LMCA is originating from the main pulmonary artery (MPA).

Surgical Technique

After median sternotomy aorto-bicaval cannulation was performed, cardiopulmonary bypass was established and the ascending aorta was cross-clamped. Cardiac arrest was induced by administering antegrade and retrograde crystalloid cardioplegic solutions. While introducing the cardioplegic solution the MPA was snared in order to prevent damaging run-off and minimizing the coronary steal from the decompressed right heart into the pulmonary artery. MPA was opened, and the origin of LMCA was identified and closed by sutures. The harvested left internal mammary artery (LIMA) was anastomosed end-to-side to the left anterior descending (LAD) artery. The saphenous vein graft was implanted end-to-side of the circumflex coronary artery.

The patient was extubated on the same day and discharged from hospital on the 6th postoperative day. Post-operative echocardiography revealed mild mitral regurgitation, and the ejection fraction was 55%.

DISCUSSION

ALCAPA syndrome is a rare but serious congenital cardiac anomaly. In foetal and early neonatal life, the origin of LMCA from the pulmonary artery is well tolerated because pulmonary arterial pressure is equal to systemic pressure, which leads to antegrade flow in both anomalous LMCA and the normal right coronary artery (RCA)[2]. Soon after birth, when pulmonary arterial pressure decreases, the flow in LMCA decreases and then reverses, which leads to myocardial ischaemia and infarction and congestive heart failure, and approximately 90% of patients die within the first year of life.

ALCAPA syndrome can be very rarely seen in adults and can lead to sudden cardiac death. In the adult type, as in our case, inter-arterial collateral anastomosis develops between the right and left coronary arteries. Blood flow in the left coronary artery is then reversed, and it empties into the pulmonary artery, a condition known as myocardial steal syndrome. Mitral insufficiency is a frequent complication secondary to a dilated valve ring or infarction of a papillary muscle. In our case, echocardiography did not reveal significant mitral insufficiency, and this may be due to the presence of well-developed collaterals between the right and left coronary arteries. Adult patients may complain of symptoms due to myocardial ischaemia because the development of extensive collaterals between the right and left coronary arteries reaches only one-third of that in case of the native artery. The anastomosis resulted in arterial blood into the left coronary artery, which emptied into the pulmonary artery, causing coronary insufficiency. In ALCAPA syndrome, left-to-right shunting causes continuous hypoperfusion of the left ventricle and subsequent chronic ischemic dysfunction. Askenazi and Nadas, in their report of 15 patients with ALACAPA syndrome, showed that only those with an appreciable left-to-right shunt survived[3].

Historically, ALACAPA syndrome was diagnosed by conventional coronary angiography. However, the development of electrocardiographically gated multi-detector CT angiography and magnetic resonance imaging enables accurate non-invasive imaging[4]. CT angiography was used to confirm the diagnosis of ALCAPA in our patient.

Surgery is the main treatment of ALACAPA syndrome: ligation of LMCA in patients with adequate collateral circulation and aorto-coronary connection with the use of a subclavian artery or a graft or creation of an aorto-pulmonary window and intrapulmonary tunnel. In adults, the preferred method is ligation of LMCA at its origin from the main pulmonary artery to stop competitive flow combined with CABG placement using LIMA and the saphenous vein. It is very important to vent or open the pulmonary artery during cardioplegic arrest. The heart is decompressed by an aortic sump before opening the pulmonary artery. Bypass grafting of the LAD artery may be sufficient in such cases; however, because of the increased need of cardiac muscle for blood supply in the acute phase and probability of the presence of atherosclerotic lesions in coronary arteries, we preferred performing bypass grafting of LAD and obtuse margin (OM) arteries.

We prescribed β-blocker and aspirin because RCA was tortuous and with an aim of decreasing the risk of long-term saphenous vein graft occlusion.

At the 3rd-month follow up, the patient's ejection fraction was 50% and functional capacity was class I according to the NYHA classification.

In conclusion, patients with ALCAPA syndrome must be treated surgically as soon as the diagnosis is confirmed. MPA must be vented in order to prevent damaging run-off and minimizing the coronary steal from the decompressed right heart into the pulmonary artery. Both the LAD and circumflex coronary arteries must be subjected to bypass grafting in such patients.

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