Case Report

Abnormal origin of the left coronary artery from the pulmonary artery (ALCAPA): A rare case report

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Abstract. Abnormal origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome is a rare congenital coronary anomaly (1/300000 live births). Generally most of the patients with the anomaly die in childhood because of several reasons. It is seen that only a few patients survive up to adulthood in the literature. We report a 39 years old man with the anomaly which reached adulthood and successfully treated surgically.

Key words: ALCAPA, adult, congenital, coronary, abnormality

1. Introduction

Abnormal origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome which is abnormal left coronary artery stemming from the pulmonary artery, is a rare congenital cardiovascular defect. The anomaly is anticipated to appear 1/300 000 live births, representing between 0.24% and 0.46% of all congenital cardiac anomalies (1).

It was first described in 1866. The first clinical definition on the ALCAPA syndrome was reported with autopsy findings by Bland and colleagues in 1933, hence the congenital defect is also known the Bland-White- Garland syndrome (2).

We report one of the few patients who can live for up to an advanced age.

2. Case report

A 39-years-old white male presented to cardiology clinic with shortness of breath and atypical angina during the last two months. There were no cardiovascular risk factors. The patient did not use cigarettes, alcohol and any harmful substance. The patient's physical examination was normal.

The echocardiographic result was normal, but the electrocardiogram (ECG) showed incomplete right bundle branch block (RBBB) and anterolateral ST depressions. The stress scintigraphy using single-photon emission computed tomography (SPECT) pointed to a reversible perfusion defect in the left ventricular anterior wall; therefore, we had to perform coronary angiography.

It was performed under local anesthesia with the rules of asepsis and antisepsis. And then '6 F Sheath' was placed into the right femoral artery with the standard Seldinger technique. Afterward the left coronary artery was not found with 6 F left judkins catheter. Aortography also didn't show it, but we saw a large right coronary ostium with the opaque material, and then displayed the right coronary artery with 6 F left Amplatz catheter (Figure 1a). The right coronary artery (RCA) was seen to be tortuous and very ectatic, with a well-developed collateral flow toward the left coronary artery (LCA) which drained into the mean pulmonary artery trunk (Figure 1b).

At the same time pulmonary angiography was performed through the femoral vein and shown in a pose silhouette of the left main coronary ostium and systolic pulmonary artery pressure was 28 mmHg. Moreover, the present anomaly was displayed using 128-slice multi detector computed tomography. It clearly showed that the RCA originated from the right coronary sinus and the left mean coronary artery (LMCA) stemmed from the inferolateral mean pulmonary artery (Figure 2a, 2b). Surgical corrections were planned due to the anterior wall

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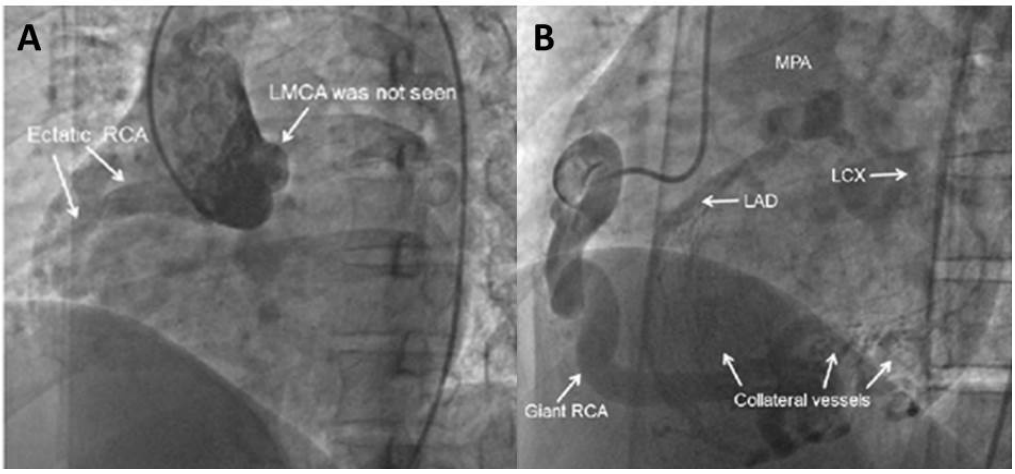

Fig. 1. (A) Aortography shows that here was no LMCA arising from aorta, (B) right coronary angiography displays the ectatic RCA and the left coronary arterial system which was filled from the RCA via collateral vessels. LAD: Left anterior descending artery, LCX: Left circumflex artery, LMCA: Left mean coronary artery, MPA: Mean pulmonary artery, RCA: Right coronary artery.

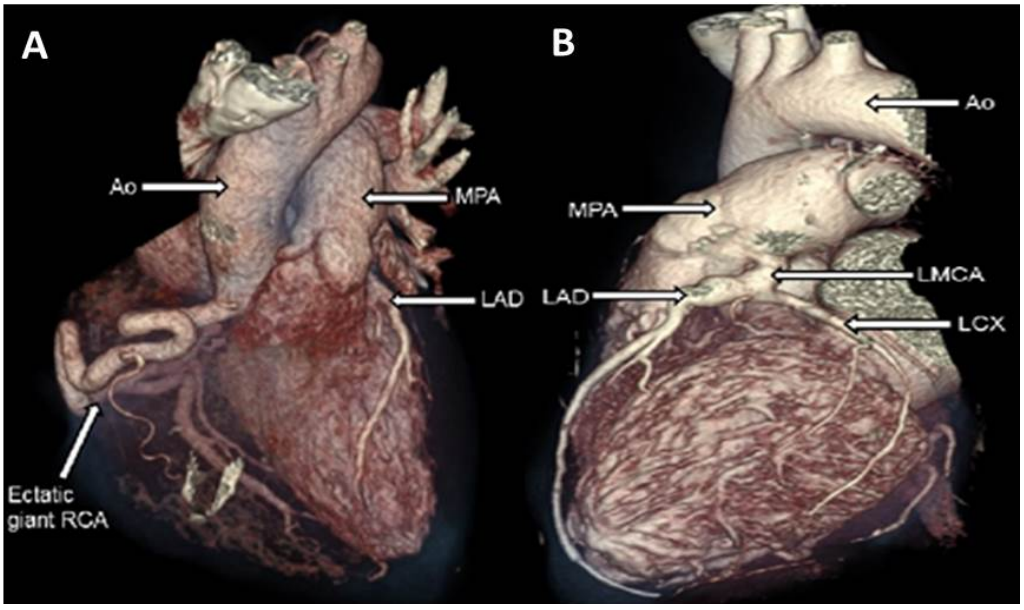

Fig. 2. (A) Multislice tomographic angiography displays RCA arising from the aortic root (anterior-superior position), (B) and demonstrates LMCA arising from the MPA (left-lateral position). Ao: Aorta, LAD: Left anterior descending artery, LCX: Left circumflex artery, LMCA: Left mean coronary artery, MPA: Mean pulmonary artery, RCA: Right coronary artery.

ischemia. And then the patient was successfully underwent the surgery consisting of the LMCA ostium ligation and aortocoronary bypass grafting which includes the left internal mammary artery (LIMA)-left anterior descending artery (LAD) and the obtuse-saphenous anastomoses.

3. Discussion

The ALCAPA syndrome is an infrequent but severe congenital anomaly. This anomaly in children is one of the most common reasons for myocardial infarction (3). There are two types of the syndrome: the first type is called as the 'adult type' and have well-established collaterals. The other type is also called as the 'infantile type' and has no collaterals (4). The case was also the first type.

In the ALCAPA syndrome, clinical findings are significantly owing to the pulmonary circulation steal from left coronary artery (5). In this way, it

may be myocardial ischemia, left ventricular dilatation and ischemic mitral regurgitation.

If the anomaly is not corrected surgically in the early years of lifetime, a large myocardial infarction or ventricular arrhythmias may be lethal for patients with the ALCAPA syndrome. The patients who survive adulthood frequently have several symptoms of left ventricular failure or myocardial ischemia, on the basis of the development of collateral flow from right to left, and almost 90 percent die suddenly at average of 35 years old (6, 7). Therefore, surgical treatment is recommended even in asymptomatic patients. In our case, there were no symptoms until two months ago. However, shortness of breath and atypical angina had begun in the last two months. Left ventricular diameters, systolic and diastolic functions were echocardiographically normal. The well-developed collateral may have provided sufficient flow from the RCA to the LCA at least during resting state. Therefore, they may have been preserved and he remained asymptomatic until recently.

The recent literature suggests that only if the patient is asymptomatic and has mild chronic ischemia, survival without surgical treatment is probable (4). If surgical correction is carried out for these patients, collateral flow is interrupted due to the normalization of antegrade flow in the LAD, and the flow rate is reduced in the ectatic RCA. Thus, the risk of thrombosis may increase in the RCA. In our case; the LMCA ostium ligation and aortocoronary bypass grafting (the LİMA-LAD and the obtuse-saphenous anastomoses) were performed, because there was large anterior wall ischemia in myocardial perfusion scintigraphy.

There are several surgical methods for these patients. However, the most popular surgical methods are to constitute two coronary artery systems via the LMCA ligation and aortocoronary bypass, Takeuchi operation (intrapulmonary re-rooting) and direct reimplantation of the LMCA to the aorta (8). We performed the LMCA ostium ligation and aortocoronary bypass grafting to the patient. After the surgery, although the patient has no atherosclerosis, we proposed to use 100 mg acetylsalicylic acid per day in a lifetime for him on account of the risk of thrombosis in the RCA. However, there is no evidence for lifelong aspirin use in literature. We consider that aspirin prevent potential acute coronary events for this anomaly after surgery.

Although the ECG changes in the ALCAPA may include in anterolateral ischemia or infarction, left electrical axis deviation, left ventricular hypertrophy and normal limits (9). There was incomplete right bundle branch block in our case. But, the right heart chambers were within normal limits by echocardiography. Furthermore, systolic pulmonary artery pressure was measured within normal limits with echocardiography and right heart catheterization. It was 28 mmHg. And this does not explain the cause of incomplete RBBB. We think that this event is incidental but anterolateral ST depression was because of the anterior wall ischemia. Already, the SPECT also showed this.

As in our case; the patients who reached adulthood usually show clinical trips associated with left ventricular ischemia. For this reason, physicians must be careful and implement the necessary tests for the diagnosis. Because, the patients with ALCAPA syndrome generally may have a very good prognosis if early diagnosis and treatment is established.

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