Adult Type Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) with Left Main Coronary Artery Osteal Stenosis: Case Report

Erişkin Tipi Sol Koroner Arterin Pulmoner Arterden Çıkması (ALCAPA) ve Sol Ana Koroner Arter Osteal Stenozu

Gökhan İPEK,[a]
Deniz GÖKSEDEF,[a]
Ozan Onur BALKANAY,[a]
Zeki TALAS,[b]
Suat Nail ÖMEROĞLU,[a]
Nevzat Cem SAYILGAN,[c]
İrfan Levent SALTIK[d]

Departments of
[a]Cardiovascular Surgery,
[c]Anesthesiology and Reanimation,
[d]Pediatrics,
İstanbul University
Cerrahpaşa Faculty of Medicine,
İstanbul
[b]Clinic of Cardiovascular Surgery,
Sivas Numune Hospital, Sivas

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Yazışma Adresi/*Correspondence:*
Ozan Onur BALKANAY
İstanbul University
Cerrahpaşa Faculty of Medicine,
Department of Cardiovascular Surgery,
İstanbul,
TÜRKİYE/TURKEY
balkanay@doctor.com

ABSTRACT Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rarely seen congenital anomaly. Patients with ALCAPA usually become symptomatic until the second month of life. Left main coronary artery osteal stenosis can lead to late clinical presentation of ALCAPA. Herein, we described a patient with adult type ALCAPA and left main coronary artery osteal stenosis who underwent coronary artery bypass grafting successfully. An eleven-year-old male patient with fatigue was admitted to our hospital. Physical examination revealed no abnormality. Coronary angiogram showed ALCAPA and left main coronary artery osteal stenosis. Bypass grafting of the left anterior descending artery by left internal mammarian artery graft was performed. Then the origin of left main coronary artery from inside of the main pulmonary artery was primarily sutured. Early and late postoperative periods were uneventful. Regardless of the age and clinical symptoms, treatment is recommended soon after the diagnosis of ALCAPA.

Key Words: Coronary vessel anomalies; pulmonary artery; congenital abnormalities; coronary stenosis; coronary artery bypass; coronary angiography

ÖZET Sol koroner arterin pulmoner arterden çıkması (ALCAPA) nadir görülen bir doğumsal anomalidir. ALCAPA bulunan hastalar genellikle yaşamlarının ilk 2 ayı içinde semptomatik hale gelirler. Eş zamanlı sol ana koroner arter osteal darlığı bulunması, ALCAPA'nın geç klinik bulgu vermesine neden olabilir. Burada erişkin tipi ALCAPA ve eş zamanlı sol ana koroner arter osteal darlığı nedeniyle koroner arter baypas greftleme operasyonunun başarıyla uygulandığı olgumuz ele alındı. On bir yaşında erkek hasta yorgunluk şikayetiyle hastanemize başvurdu. Yapılan fizik muayene bulguları olağandı. Çekilen koroner anjiografide ALCAPA ve sol ana koroner arter osteal stenozu teşhisi konuldu. Sol ön inen artere sol internal mammarian arter ile baypas greftleme yapıldı. Daha sonra sol ana koroner arterin ana pulmoner arter içyüzündeki çıkış yeri primer olarak sütüre edilerek kapatıldı. Erken ve geç postoperatif dönemde sorun yaşanmadı. Klinik başvuru yaşından ve semptomlardan bağımsız olarak, ALCAPA tanısı konulur konulmaz tedavi uygulanması önerilmektedir.

Anahtar Kelimeler: Koroner damar anomalileri; pulmoner arter; doğumsal anomaliler; koroner darlığı; koroner arter baypas; koroner anjiyografi

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The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rarely seen congenital anomaly.[1] Patients with ALCAPA usually become symptomatic until the second month of life.[2] Left main coronary artery (LMC) osteal stenosis which is a very rarely seen condition can lead to late clinical presentation of ALCAPA. Here we described a patient with adult type ALCAPA and LMC osteal stenosis who underwent coronary artery bypass grafting (CABG) successfully.

CASE REPORT

An eleven-year-old male patient with fatigue was admitted to our hospital. Physical examination revealed no abnormality. Trans-thoracic echocardiography (TTE) showed right coronary artery dilatation. Coronary angiogram revealed dilated right coronary artery, well-developed collateral interseptal arteries, retrograde blood flow in the left coronary artery system, coronary steal from the LMC to the main pulmonary artery and LMC osteal stenosis (Figure 1). The informed consent was taken and operation was decided.

We performed median sternotomy. After pericardiotomy we observed that right coronary artery and its branches were well developed (Figure 2A). The operation was performed on cardiopulmonary bypass. After the aortic and pulmonary cross-clamping we used potassium-enriched blood cardioplegia via antegrade way to arrest the heart and protect the myocardium. Bypass grafting of the left anterior descending artery by left internal mammarian artery graft was performed by using separate 8/0 polypropylene sutures (Figure 2B). Then we primarily sutured the origin of LMC from inside of the main pulmonary artery. Early and late postoperative periods were uneventful.

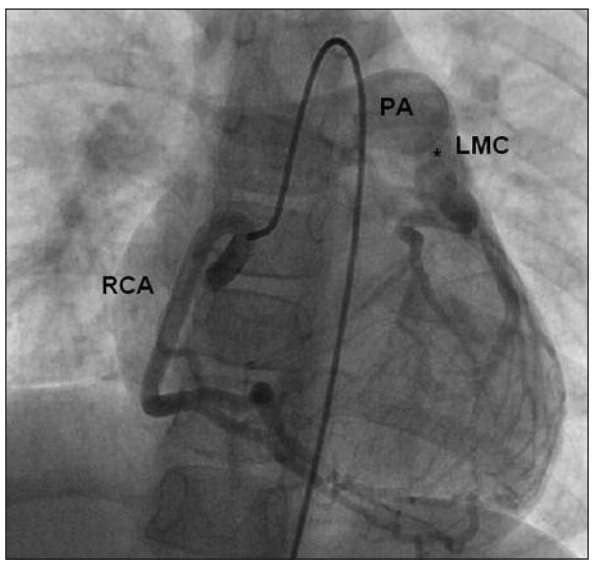

FIGURE 1: Coronary angiogram of left main coronary artery originated from the main pulmonary artery and left main coronary artery stenosis.
RCA: Right coronary artery; PA: Pulmonary artery; LMC: Left main coronary artery; * Stenosis of the origin of the left main coronary artery.

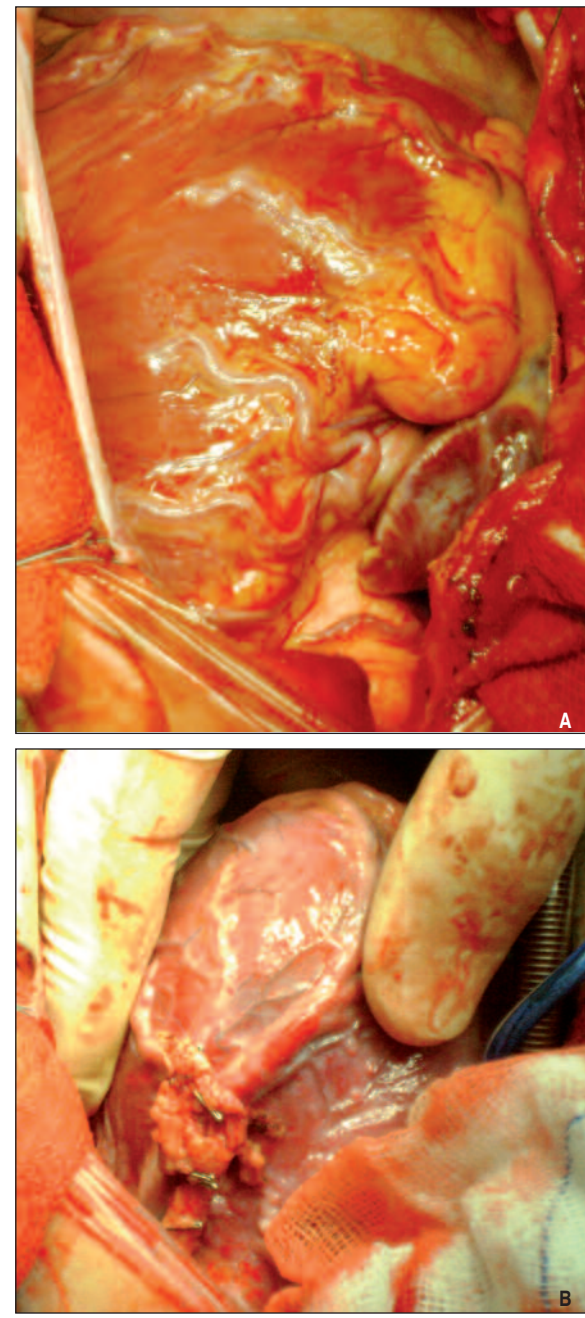

FIGURE 2: Intra-operative view **A)** Intra-operative view of enlarged right coronary artery and its branches. **B)** After LIMA-LAD bypass.
LIMA: Left internal mammary artery; LAD: Left anterior descending artery.
(See color figure at
http://www.turkiyeklinikleri.com/journal/cardiovascular-sciences/1306-7656/)

DISCUSSION

ALCAPA or Bland-White-Garland syndrome is a rarely seen congenital anomaly with an incidence rate of 1 in 300 000 live births, accounting for 0,25-0,5% of all congenital heart anomalies.[1-3]

Decrease in pulmonary vascular resistance after birth causes an increase in coronary steal that usually makes the patients symptomatic until the second month of life.[2] Previous reports used the term "adult type ALCAPA" for the patients who were asymptomatic during infanthood and early childhood periods. Common symptoms of adult type ALCAPA are fatigue, myocardial ischemia and infarction in childhood; fatal arrhythmias and sudden death in adulthood.[4,5]

TTE is one of the most valuable non-invasive diagnostic tools for ALCAPA.[1,4] In patients with adult type ALCAPA, TTE usually shows dilated right coronary artery system that compensates the left coronary artery system with well-developed collateral interseptal arteries. However, TTE can only diagnose 50% of adult type ALCAPA.[4]

Coronary angiography is the most valuable diagnostic tool for ALCAPA.[1] The incidence rate of adult type ALCAPA among patients who underwent coronary angiography is 1 in 10 000 patients.[6] Coronary angiogram usually reveals no orifice in the left valsalva sinus, dilated right coronary artery with well-developed interseptal arteries and retrograde blood flow in the left coronary artery system. Contrast medium passage from LMC to pulmonary artery is diagnostic for ALCAPA.[2,5]

Nearly 90% of untreated patients do not survive more than 1 year after birth.[4,7] Although the surviving patients with adult type ALCAPA can be asymptomatic until childhood or adulthood, majority of them will be lost until their thirties due to fatal arrhythmia or sudden death.[2,4,7] Regardless of clinical symptoms, treatment is recommended soon after the diagnosis of ALCAPA because of high rates of mortality and morbidity.[2]

Closure of the origin of LMC alone by percutaneous vascular plug device or surgery was described in patients with well-developed collateral interseptal arteries and adequate right coronary blood flow.[4,6,8] However, maintaining double coronary artery system is also recommended especially in patients with adult type ALCAPA.[4,6,8] In this way, whole blood supply of the heart is not left dependent only on the right coronary artery system. Direct or transpulmonary re-implantation can be used in suitable patients.[4,8,9] In patients with adult type ALCAPA, concomitant osteal stenosis of the left main coronary artery should be suspected before deciding on the method of the surgical treatment. In patients with osteal stenosis of the left main coronary artery, coronary artery bypass grafting should be preferred instead of direct re-implantation technique as we did in our case.[4]

Myocardial protection can be maintained by using aortic and pulmonary cross-clamping at the same time to minimize pulmonary steal of the cardioplegic solution. Concomitant direct closure of the ostium of the left main coronary artery via pulmonary arteriotomy can be preferred as well. We used aortic and pulmonary cross-clamping at the same time to minimize pulmonary steal of the potassium-enriched blood cardioplegia given via antegrade way. Direct closure of the ostium of the left main coronary artery was not found necessary in our case because of its severe osteal stenosis.

Although patients with ALCAPA usually become symptomatic in their very beginning of their lives, they can be asymptomatic until advanced ages.[10] Regardless of the age and clinical symptoms, treatment is recommended soon after the diagnosis of ALCAPA.[2,10] CABG using left internal mammarian artery graft with the closure of anomalous origin of LMC can be successfully used in patients with adult type ALCAPA and LMC osteal stenosis.[4,5,11]

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