Different Presentation of Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery in Adults: Case Reports

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac malformation. We report three cases of ALCAPA who survived to adulthood. The first case was a 51-year-old woman who complained of typical chest pain that was diagnosed with ALCAPA using cardiac catheterization and coronary computed tomographic angiography (CTA). The second case was a 30-year-old woman with a history of surgery for atrial septal defect at 10 years old who presented with progressive exertional dyspnea. Cardiac catheterization confirmed the diagnosis of ALCAPA. The third case was a 19-year-old man who was brought to our clinic due to aborted sudden cardiac death on the previous day. Cardiac catheterization and coronary CTA confirmed the diagnosis. They underwent the closure of orifice of the anomalous left coronary artery and grafting the left anterior descending artery concomitantly with mitral valve repair. All patients were followed up during a mean of 8.7 months and they were asymptomatic.

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Keywords

● Coronary vessel anomalies
● Heart defects
● Congenital

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac malformation.¹ ALCAPA was first described in 1933 by Bland, Garland, and White during an autopsy of a 3-month-old infant presented with progressive feeding problems, cardiomegaly, and left ventricular damage.² The first reported case of ALCAPA in an adult was based on postmortem findings of a 60-year-old asymptomatic female patient who had died accidentally.³ The earlier a patient undergoes surgery, the better the recovery of the cardiac function. Patients with ALCAPA who survive to adulthood may suffer from sudden cardiac death in the third decade of life.⁴ Adult survivors are either asymptomatic despite ongoing subclinical myocardial ischemia⁵ or presenting with malignant arrhythmias.⁴,⁵ Herein, we report three cases of adult ALCAPA with different presentations.

Case Presentation

The first case was a 51-year-old woman with a history of myocardial infarction who complained of typical chest pain.
The electrocardiogram (ECG) showed ST-T segment changes in I and aVL leads. Transthoracic echocardiography (TTE) showed left ventricular ejection fraction (LVEF) of 40%, moderate mitral valve regurgitation (MR), and suspicion of ALCAPA that was confirmed by cardiac catheterization and coronary computed tomographic angiography (CTA) (figure 1).

On operation, the orifice of the anomalous left coronary artery located in the left posterolateral aspect of the pulmonary artery (PA) was closed with a fresh autologous pericardial patch and then the proximal left anterior descending (LAD) artery was grafted by a left internal mammary artery (LIMA) graft. The mitral valve leaflets were thickened with flail anterior segments (A1 and A2), hence neochordae were constructed for these segments in addition to ring annuloplasty using Sorin Memo 3D annuloplasty ring No. 30 (Sorin Biomedica Cardio S.r.l., Saluggia, Italy). TEE after cardiopulmonary bypass (CPB) revealed a LVEF of 50% and mild MR. At 10 months follow-up, the patient was asymptomatic with normal findings on echocardiographic examination. The patient’s consent was obtained.

The second case was a 30-year-old woman who presented with progressive exertional dyspnea. She underwent surgery for atrial septal defect at 10 years of age and the repair of suspected right coronary artery (RCA) fistula into the right ventricle according to cardiac catheterization, while the ALCAPA was not detected. During surgery, the coronary fistula was not found and the surgery ended without complete repair. Twenty years later, echocardiographic examination revealed a LVEF of 35%, moderate to severe MR, and severe free tricuspid regurgitation (TR). Cardiac catheterization confirmed the diagnosis of ALCAPA. During surgery, there was no possibility for reimplantation of the orifice of the anomalous left coronary artery originating from the posterolateral sinus of valsalva of the PA was closed with a fresh autologous pericardial patch, and the PA arteriotomy was closed. Next, the proximal LAD was grafted by the LIMA. Mitral valve repair was performed by saddle ring annuloplasty No. 28 (St. Jude Medical Inc., MN, USA). TEE after CPB revealed a LVEF of 50% and trivial MR. At 6 months follow-up, the patient was asymptomatic with a LVEF of 50% and mild MR. The patient gave consent for this case report.

**Discussion**

The diagnosis of ALCAPA in adults is very rare; however, the availability of less invasive diagnostic modalities has resulted in increased diagnosis at older ages. To survive beyond childhood, patients develop collateral circulation

![Figure 1: Computed tomographic angiography (A) and three-dimensional computed tomographic angiography (B) show anomalous left main coronary artery arising from the main pulmonary artery.](image_url)
from the RCA to the left coronary artery. However, it is not sufficient to supply the left ventricle, especially in the subendocardial region, which can attribute to chronic left ventricular subendocardial ischemia. Consequently, patients may develop malignant ventricular rhythm disturbances increasing the incidence of sudden cardiac death, which occurs in 80-90% of patients. Although arrhythmic events are of great paramount in adult ALCAPA, but a wide range of symptoms from an asymptomatic one (14% of 151 reported cases) to life-threatening presentations (18% of 151 reported cases) has been reported in the literature. Early diagnosis and prompt surgical intervention with the aim of restoring a two-coronary-artery circulatory system have excellent results and lead to gradual myocardial recovery.6,7

Regarding the management of ALCAPA, the guidelines suggest surgical correction with a reestablishment of two-vessel anatomy.4 However, adult ALCAPA has less elastic vessels originated distant from the implant target, thus there is a little possibility to directly reimplant on the aorta.1 Alternatively, bypass grafting to the LAD with venous or arterial grafts along with a closure of ostium of left coronary artery is also advised.4 In our cases, all patients were treated with a closure of the ALCAPA orifice using fresh autologous pericardium along with coronary artery bypass graft (CABG) surgery and mitral valve repair. The implementation of CABG is associated with a risk of graft stenosis and a greater risk of late obliterative change in venous graft compared with an arterial graft.8 In a study by Moodie et al.,7 they showed a patency rate of 80% in SVGs after a mean follow-up of 5.8 years. In our cases, we aimed at using an arterial graft of LIMA for grafting the LAD; however, in one patient we used a SVG because of adhesions. After six months follow-up, a significant graft stenosis was detected. This stenosis could not be attributed to obliterative changes in the venous graft, but it may be caused by technical pitfall. However, this case had good outcomes at late follow-up.

There has been a great deal of debate regarding concomitant mitral valve repair at the time of ALCAPA repair for children with MR.5 The majority of MR cases in children is functional and will improve with reperfusion, but structural mitral valve abnormalities should be repaired at the time of surgery.10 The previous reports dealing with MR at the initial management of ALCAPA are certainly not for adults and whether this approach can be adopted for adults is not clear.4 In adults; however, structural mitral valve abnormalities develop due to chronic MR and valve repair is recommended at the initial operation as the left ventricular function recovery is not expected to correct the preoperative MR. In our cases, the mitral valve was repaired with ring annuloplasty in two patients and concomitant neochordae construction in the other patient.

**Conclusion**

The surgical repair of ALCAPA in adult cases is indicated at the time of diagnosis, regardless of its presentation and patient’s symptoms. Moreover, the surgical correction of malformation associated with concomitant CABG and mitral valve repair is associated with excellent exercise capacity and quality of life at follow-up period.

**Conflict of Interest:** None declared.

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