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Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery Detected by Echocardiography in an Asymptomatic Young Athlete

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ABSTRACT

The anomalous origin of the coronary artery from the pulmonary artery (ALCAPA) is the most common congenital coronary artery anomaly. Up to 90% of patients die during the first year of life. It is unusual for an ALCAPA patient to survive to adulthood. We present a case of an asymptomatic young athlete with ALCAPA, in which the diagnosis was established by echocardiography during pre-participation physical evaluation. The patient underwent surgical closure of the left main coronary artery ostium through the inside of the main pulmonary artery and coronary artery bypass grafting. He was discharged after 6 days and remained well during follow-up visits. We emphasize the importance of echocardiographic examination during pre-participation cardiovascular screening in young athletic populations to avoid sudden death related to ALCAPA.

INTRODUCTION

The anomalous origin of the coronary artery from the pulmonary artery (ALCAPA), or Bland-White-Garland syndrome, is the most common congenital coronary artery anomaly, occurring in 0.26% of patients with congenital heart disease. It is unusual for an ALCAPA patient to survive to adulthood [Tian 2013]. Overall, the life expectancy of patients with untreated ALCAPA is very poor. Surgical correction is recommended as soon as the diagnosis is made, regardless of age or the degree of intercoronary collateralization [Kottayil 2011].

Here we describe an asymptomatic young athlete in which ALCAPA was diagnosed by echocardiography during pre-participation physical evaluation.

CASE REPORT

A 23-year-old male water polo athlete was referred to our hospital for the pre-participation physical evaluation before the World Cup. A mild holosystolic murmur (grade 2/6) was heard along the left sternal border on auscultation. Transthoracic echocardiography revealed a huge tortuous right

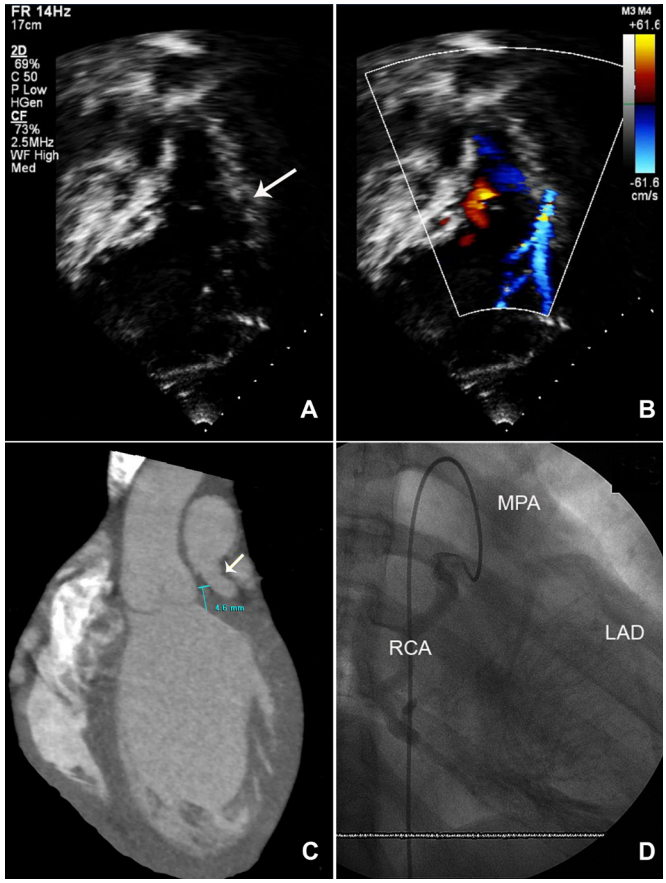

Figure 1. A, Transthoracic echocardiography showing the left main coronary artery (LMCA) originating from the pulmonary artery (PA) (arrow). B, Color Doppler reveals the retrograde flow from the LMCA to the PA. C, Multislice computed tomography coronary angiography showing the left main coronary artery arising from the main pulmonary artery (arrow). D, Coronary angiography demonstrating a dilated right coronary artery (RCA) giving rise to multiple dominant septal collaterals to the left anterior descending artery (LAD), which arose from the main pulmonary artery (MPA).

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coronary artery originating from the right sinus of Valsalva and anterolateral hypokinesia, and minimal mitral regurgitation with a left ventricular ejection fraction of 55%. The origin of the left main coronary artery (LMCA) was absent from the left sinus of Valsalva (Figure 1, A and B). Doppler color flow mapping demonstrated an abnormal jet from the left coronary system to the pulmonary trunk and retrograde blood flow in the proximal segment of the LMCA. Multislice computed tomography coronary angiography showed the LMCA originating from the pulmonary artery (Figure 1, C). Coronary angiography revealed a huge tortuous right coronary artery with marked collateral arteries filling the left coronary system, and the LMCA filling from the main pulmonary artery (Figure 1, D). Right heart catheterization demonstrated the left main coronary artery originating from the posterior side of the pulmonary artery. The patient was diagnosed with ALCAPA and was scheduled for surgery.

Cardiopulmonary bypass was established with ascending aortic and bicaval cannulation. The left atrium was vented through the right superior pulmonary vein. Cardioplegic arrest was achieved by injecting cold blood cardioplegic solution through the aortic root while simultaneously occluding the main pulmonary artery (MPA). The MPA was opened with transvers incision below the bifurcation. The ostium of the left main coronary artery, located at the left and posterior sinus of the MPA, was identified and closed with continuous 5/0 propylene suture primary through the inside of the MPA (Figure 2). The incision of the MPA was closed using 5/0 propylene suture. In order to provide perfusion of the left coronary artery system, coronary artery bypass grafting (CABG) to the left anterior descending and to the first marginal branch of the circumflex arteries was performed using the left internal thoracic artery and saphenous vein grafts respectively.

Postoperative course was uneventful. The patient recovered well and was discharged on the postoperative day 6. Six months after the procedure, he was asymptomatic and the treadmill test did not reveal evidence of myocardial ischemia.

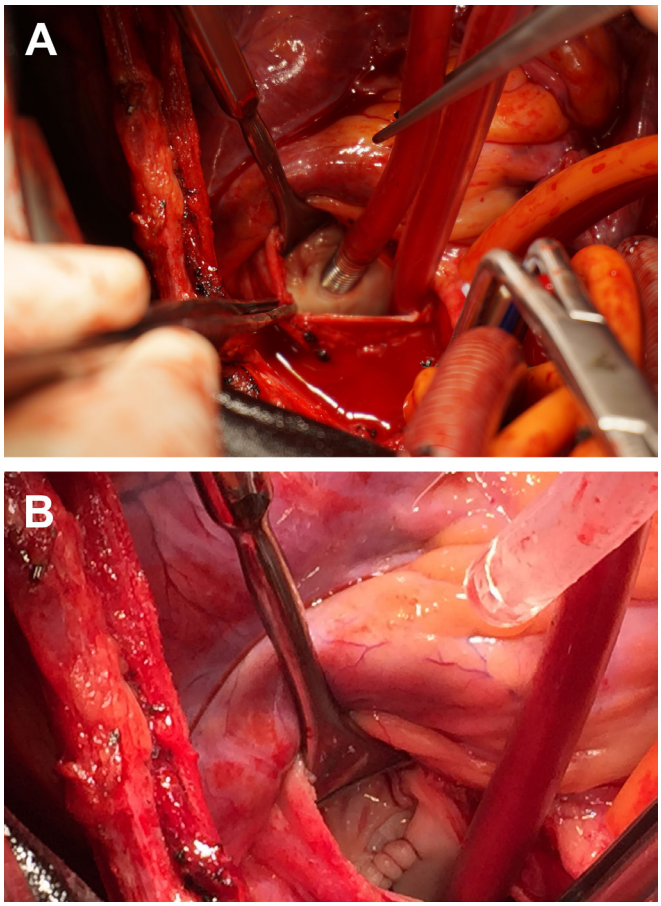

Figure 2. Intraoperative view of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). A, The suction cannula demonstrating the ostium of the left main coronary artery, located at the left and posterior sinus of the main pulmonary artery (MPA). B, The ostium is closed with continuous 5/0 propylene suture primary through the inside of the MPA.

DISCUSSION

ALCAPA is a rare anomaly and patients often present in early infancy with myocardial infarction and sudden death. Rarely, patients may present in adulthood [Tian 2013]. Atypical angina and systolic murmur can be the only sign detected during physical examination [Sahin 2012]. The asymptomatic patients can initially present with syncope or even sudden death [Yau 2011]. Therefore, in any young individual, easily identifiable echocardiographic markers of ALCAPA, including dilated right coronary arteries, well developed collateral vessels within the interventricular septum, and abnormal blood flow in the PA must raise suspicion to avoid missing a diagnosis of this anomaly [Tian 2013].

Whenever possible, direct implantation of the left coronary artery into the ascending aorta is performed. This technique has been most frequently used in the pediatric population with good long-term survival and freedom from reoperation. But in adult patients, this procedure can be technically challenging to mobilize the LMCA to reach the aorta. In these patients, ligation of the left coronary ostium combined with aortocoronary bypass grafting using the internal thoracic artery and venous grafts is an alternative surgical approach [Kreutzer 2000]. Associated mitral regurgitation is usually functional secondary to chronic ischemia, affecting the left ventricular geometry and function. Intervention is not recommended at the initial operation because most patients will recover to have normal mitral valve function after coronary repair [Kottayil 2011].

In conclusion, even though ALCAPA is a rare congenital anomaly, due to unfavorable prognosis it is necessary to diagnose the anomaly early and surgically correctly before cardiac problems arise. ALCAPA can be diagnosed with angiographic methods including invasive coronary and computed tomography angiographies. Though both have high diagnostic accuracy with superior imaging quality, there are still some inferiorities such as high doses of radiation and risk of contrast nephropathy. Recent echocardiography technology provides useful information about the anomaly without the risk of radiation and kidney failure, such as anatomic configuration,

including displaying the origin of coronary arteries with their courses, hemodynamic cardiac parameters and associated anomalies. This noninvasive and widely available diagnostic tool could be used as the initial method of diagnosis in patients with ALCAPA, especially in young athletic populations pre-participation exam.

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