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Anomalous left coronary artery from pulmonary artery (ALCAPA) as a silent cause of mitral regurgitation in children

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Abstract

Anomalous left coronary artery from pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome, is a rare cardiac disease. This condition may present with complications such as myocardial infarction, left ventricular dilatation, mitral regurgitation, and left heart failure in children. We report a case of a four-year-old boy who presented with shortness of breath, palpitations, and recurrent upper respiratory tract infections. He was diagnosed with mitral regurgitation. During the surgery, left coronary artery (LCA) was not present in its anatomical position and ALCAPA was identified. One should keep in mind the possibility of ALCAPA in presentation of mitral regurgitation in children despite not being reported in echocardiography.

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Introduction

Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare cardiac disease with an incidence of 1 in 300,000 live births [1]. The aberrant artery leads to coronary steal phenomenon due to which the left myocardium undergoes ischemia, which may lead to myocardial infarction, left ventricular dilatation, mitral regurgitation, and left heart failure [2]. Untreated ALCAPA has a high mortality rate of up to 90% in the first year of life [3]. The sudden cardiac death can occur due to arrhythmias [4]. Therefore, early diagnosis and surgical correction are important in survival. There are risks of diastolic dysfunction, left ventricular (LV) torsion, LV longitudinal deformation, and arrhythmias despite successful surgical correction but these complications occur mainly in the adult population [5]. Due to the rarity of this disease, it often goes unnoticed. The most common diagnostic tool is echocardiography [6]. However, some cases may go undiagnosed, leading to complications later in life or during surgical correction of the mitral valve [7]. Here, we present a case in which echocardiography failed to identify anomalous coronary supply and it was discovered during surgery.

Case Report

A 4-year-old boy presented to the Outpatient Department with the complains of shortness of breath and palpitations at the age of 8 months and was diagnosed to have mitral regurgitation (MR). He had recurrent upper respiratory tract infections for which he had multiple hospitalizations. On examination he had left precordial bulge, palpable thrill, grade 4 systolic murmur that was radiating towards the axilla and intercostal recessions. Echocardiography showed hugely dilated aneurysmal left atrium, severely dilated left ventricle with mitral valve prolapse, and non-coapting thickened leaflets with an annulus size of 32 mm and severe MR with ejection fraction (EF) of 76% and fractional shortening of 45% (Figure 1).

Median sternotomy was performed to approach the heart. Left coronary artery (LCA) was not present in its anatomical location, so the main pulmonary artery (MPA) was inspected and an anomalous left coronary artery arising from the pulmonary artery (ALCAPA) was discovered (Figure 2). Aorto-bicaval cannulation was performed and cardiopulmonary by-pass (CPB) was established. Patient was cooled to moderate hypothermia. Right and left pulmonary arteries were dissected and snugged. Aortic cross clamp was applied and anterograde cold blood cardioplegia was given in the aorta and main pulmonary artery. Pulmonary artery (PA) was transected and leftward facing sinus wall along with ALCAPA was harvested as a button and suitable length was achieved by dissecting it free from the surrounding tissue for a tension free anastomosis. Partial occluding clamp was applied to the left lateral wall of the aorta which was opened and end-to-side anastomosis of the ALCAPA was performed. Posterior wall of the MPA was reconstructed using bovine pericardium patch to prevent any compression on ALCAPA and the anterior wall was closed primarily. Attempt was made to repair the mitral valve but due to persistent regurgitation, mitral valve was replaced with a 27 mm Medtronic mechanical valve. CPB was weaned off with minimum ionotropic support and patient made an uneventful recovery. Immediate postoperative EF was 41% and at discharge it was 45%.

Discussion

Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare cardiac disease which is also known as Bland-White-Garland syndrome. It occurs in 1 in 300,000 live births and comprises 0.24-0.46% of all congenital cardiac defects [1]. It occurs as an isolated defect in 95% of the cases [8]. It presents usually after the early neonatal period with symptoms of crying during feeding, dyspnea, pallor, excessive sweating, and failure to thrive. The neonate remains asymptomatic because of high pulmonary vascular resistance. With time, the resistance in the pulmonary artery drops and reversal of flow starts from left coronary artery. This causes coronary steal phenomenon. Later, the collaterals start to form between the two circulations i.e., left and right coronary arteries resulting in lesser ischemia. ALCAPA can cause myocardial ischemia, mitral regurgitation, and dilated cardiomyopathy. The reason behind mitral regurgitation in ALCAPA is ischemia induced left ventricular and papillary muscle dysfunction [2]. If left untreated, it has a high mortality rate of up to 90% within 1 year of life due to complications such as ventricular failure, myocardial infarction, and arrhythmias [3]. However, many untreated patients survive as adults and remain asymptomatic or present later in life as arrhythmias or sudden cardiac death [8].

The current diagnostic method of choice is echocardiography [6]. The characteristic findings that can be seen are a dilated right coronary artery (RCA), retrograde Doppler flow from LCA to PA, and a septal flow due to collaterals [9]. However, there are reports of cases being missed on echocardiography just like our case. In some cases, mitral valve replacement has been done without correcting the ALCAPA. In extreme cases, even deaths have been reported because of missed diagnosis [7]. The echocardiography cannot pick significant findings in less developed collateral circulation. This also means that there are higher chances of diagnosis being missed in more critical patients. Cardiac catheterization and angiography are considered the gold standard for reaching the diagnosis. However, due to the invasive nature of the test, it is not commonly used. Noninvasive investigations such as CT angiogram and MRA can also be used as an alternative to catheterization in detecting suspected patients [4,8]. We strongly recommend using such investigations in case of strong suspicion such as

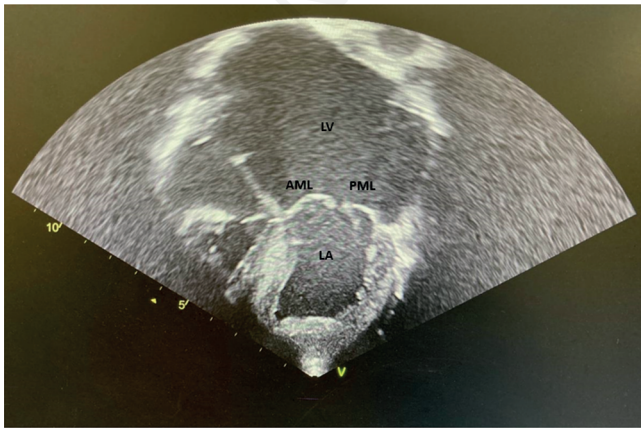

Figure 1. Echocardiography showing dilated left atrium and ventricle and non-coapting mitral valve leaflets. LV, left ventricle; AML, anterior mitral leaflet; PML, posterior mitral leaflet; LA, left atrium.

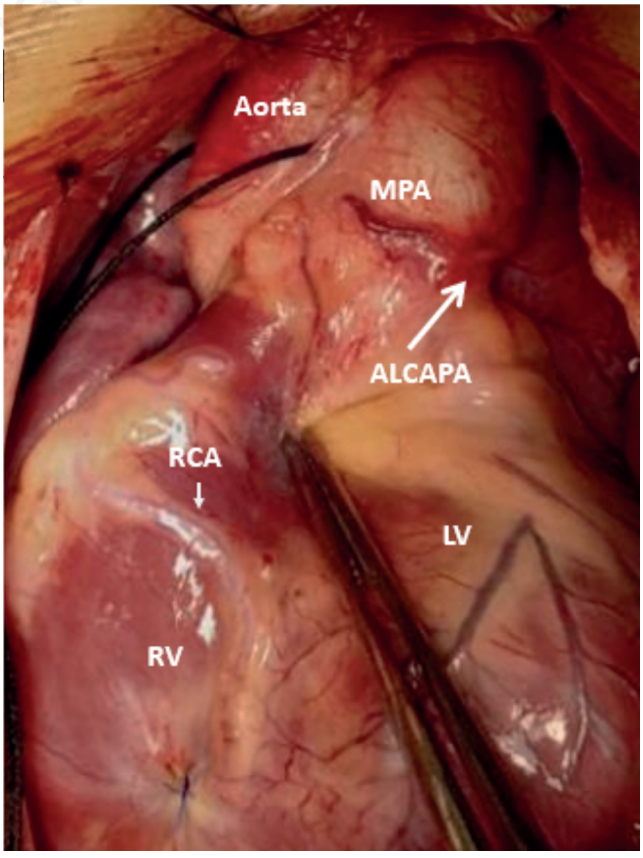

Figure 2. ALCAPA seen arising from leftward sinus of pulmonary artery. RV, right ventricle; RCA, right coronary artery; MPA, main pulmonary artery; ALCAPA, anomalous left coronary artery from pulmonary artery; LV, left ventricle.

unexplained mitral regurgitation or dilated cardiomyopathy in children to avoid unseen complications.

This rare cardiac anomaly can be definitively treated with surgery. The main objective of surgery is to restore coronary circulation. Various methods have been used among which coronary reimplantation method has been found superior [10]. It is generally not recommended to perform additional mitral valve surgery with the primary operation as mitral valve function improves after coronary vascularization. However, in cases with severe persistent MR, surgery with mitral valve repair or replacement can be considered [11]. Overall, there are good survival rates after surgery, and survival rate of 94.8% has been reported after 20 years of surgery [12]. However, asymptomatic patients who have been surgically corrected can have diastolic dysfunction, LV torsion, and LV longitudinal deformation and can present with reduced exercise performance and stress perfusion defects [5]. Similarly, patients who are surgically corrected in later stages of life or after an ischemic episode have a risk of life-threatening arrhythmias [4].

Conclusions

While echocardiography is the most effective diagnostic tool for detecting ALCAPA, one should entertain the possibility of ALCAPA in unexplained mitral regurgitation in children despite not being reported on echocardiography. We recommend using other diagnostic investigations in case of a strong suspicion of ALCAPA.

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