

Anomalous Left Coronary Artery from the Pulmonary Artery in an Asymptomatic Two-year-old after Patent Ductus Arteriosus Closure: A Near Miss Case Report

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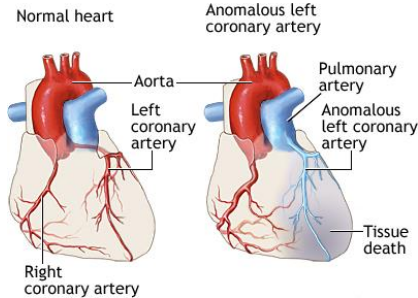
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Introduction

Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA) accounts for less than 0.5% of congenital heart anomalies¹.

ALCAPA usually results in congestive heart failure with left ventricular dysfunction early in life, but may be asymptomatic if a patent ductus arteriosus (PDA) is present.

In this case report, we present a patient who had a PDA with ALCAPA likely resulting in delayed diagnosis secondary to maintained perfusion in the anomalous coronary artery.



Case Description

A 2/6 continuous heart murmur was heard at 2 years of age in our previously healthy patient. An echocardiogram showed a moderate PDA, trivial mitral regurgitation, and dilated left ventricle with normal systolic function. She underwent successful transcatheter device closure of her PDA. Her coronary anomaly was not detected during the catheterization.

At her follow up visit, her echocardiogram had similar findings; however echobright mitral valve papillary muscles and a coronary artery fistula to the main pulmonary artery were noted.

The patient returned to the catheterization lab for a planned occlusion of the coronary artery fistula. Aortic root angiography demonstrated a single, dilated right coronary artery. Selective injection confirmed the diagnosis of ALCAPA.

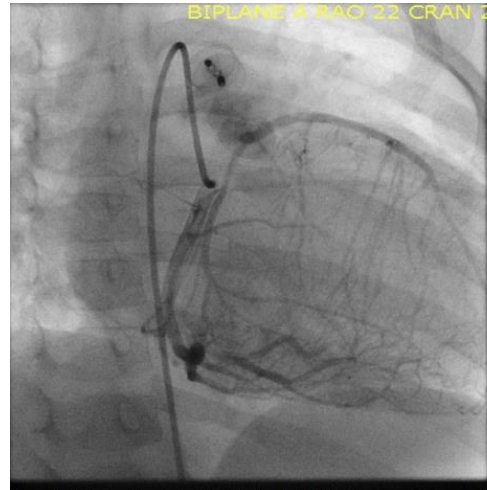


Figure 1: Selective injection in the RCA with the 4 Fr JR 3.0 catheter demonstrating a dilated RCA with normal origin, course and distribution. No areas of stenosis are appreciated. There is extensive collateralization via the PDA and septal perforators to the left coronary artery which clearly originates from the main pulmonary artery. A normal left main coronary artery arising from the posterior aspect of the MPA is noted and bifurcates normally into the LAD and circumflex arteries. A 10 mm Amplatzer vascular plug device is seen in the PDA from a previous catheterization.

The next day, the patient underwent successful ALCAPA repair by implantation of the left coronary artery into the aorta. The patient's recovery period was uneventful.

Discussion

In ALCAPA, prenatally through the first weeks of life, the persistently elevated PVR allows for adequate antegrade perfusion to the anomalous coronary. As PVR drops, coronary steal to the pulmonary circulation occurs, resulting in ischemia. The presence of a moderate PDA likely maintained adequately elevated pulmonary pressures to allow for antegrade flow through the left coronary artery.

Once closure of the PDA was performed, the pulmonary artery pressures likely lowered resulting in worsening ischemia. This was seen on echocardiogram in the form of echobright papillary muscles. By angiogram, the patient's coronary artery system was well collateralized consistent with chronic ischemia.

ALCAPA is an important diagnosis to make prior to any intervention. The anesthesiologist must adapt his/her anesthetic management with this finding. Measures to mildly increase PVR (i.e. mild hypercapnia, low FiO_2) can help minimize the coronary steal phenomenon and prevent further myocardial damage².

This case reminds us how important it is to systematically evaluate all patients with congenital heart disease on every encounter. Prior knowledge of compromised coronary flow would be essential to enable the appropriate anesthetic management, and avoid potentially life threatening complications.

This is the first reported case of ALCAPA status post PDA device closure who had successful surgical repair to our knowledge.

References

- 1) Korkmaz AA, Erkan H, Karadeniz A, Orem C. Missed Diagnosis and Treatment Dilemma: Large Patent Ductus Arteriosus Combined with Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery. *Iran J Radiol.* 2017 April; 14(2):e30713.doi: 10.5812/iranjradiol.30713.
- 2) Andropoulos DB, et al. "Anesthesia for specific lesions." *Anesthesia for Congenital Heart Disease*, 2nd ed., Wiley, 2010, pp. 485-486.