Rare Etiology of Cardiogenic Shock in Children: Anomalous Origin of the Left Coronary Artery from the Opposite Sinus with an Intramural Course

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J Pediatr Intensive Care 2015;4:166–167.

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Abstract

Keywords

- ▶ congenital coronary artery anomaly
- ▶ presentation
- ▶ diagnosis

Anomalous origin of the left coronary artery from the right sinus of Valsalva with an intramural course is a rare but potentially lethal congenital defect. While anomalous origin of the left coronary artery is a well-known cause of sudden cardiac death, it is less well appreciated that this anomaly may manifest in a variety of other clinical presentations, thereby leaving these patients at risk of missed or delayed diagnosis and subsequent poor outcomes. In this report, we describe an adolescent male with an unusual etiology of cardiogenic shock.

Introduction

Anomalous origin of the left coronary artery (AOLCA) from the right sinus of Valsalva (RSOV) with an intramural course is a rare but important congenital defect that is associated with increased risk of cardiac morbidity and sudden cardiac death (SCD).¹ The true prevalence of this anomaly is impossible to ascertain, as most patients remain asymptomatic. While exercise-induced SCD is often the first and only presentation,¹ left AOLCA from RSOV has been shown to manifest as a variety of other presentations in children, including syncope, angina, arrhythmia, and acute cardiac decompensation.² For children presenting with acute cardiac decompensation, timely diagnosis of left AOLCA from RSOV is crucial to preventing the detrimental clinical sequelae seen in these patients, given this anomaly has proven highly amenable to successful treatment with simple surgical unroofing of the intramural segment.³ Here we report the case of an adolescent male with an unusual etiology of cardiogenic shock.

Case Report

A 14-year-old boy with history of recent viral illness presented to a regional center in acute cardiac failure after developing exertional syncope. Initial electrocardiography

showed left ventricular hypertrophy and inferior lead ST depression. Transthoracic echocardiography (TTE) demonstrated subtle hypertrophy of the interventricular septum, apical and anterior segments, and ejection fraction (EF) of 42%. The coronary arteries were not visualized. His clinical condition subsequently deteriorated after developing wide-complex tachycardia and hypotension. Elective intubation and intra-aortic balloon pump insertion were undertaken and he was emergently transported to our institution for transition to extracorporeal membrane oxygenation (ECMO) support with the presumed diagnosis of acute myocarditis.

Upon arrival, he deteriorated into cardiac arrest and underwent emergent cannulation for venoarterial ECMO. Cardiac catheterization with atrial septostomy was performed. Myocardial biopsy demonstrated myocyte hypertrophy and focal interstitial fibrosis, with no evidence of inflammation. He was maintained on multiple inotropes and volume resuscitation, prompting evaluation for transplantation. By day 5, myocardial function had somewhat recovered and ECMO decannulation was attempted. However, electrocardiography showed ischemic ST-segment changes and assessment of coronary artery perfusion with focused echocardiogram demonstrated an intramural left AOLCA from RSOV. Given the clinical picture and myocardial

received

September 8, 2014

accepted after revision

November 30, 2014

published online

August 20, 2015

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Verlag KG, Stuttgart · New York

DOI <http://dx.doi.org/10.1055/s-0035-1559823>.
ISSN 2146-4618.

Fig. 1 Anomalous left coronary artery arising from a separate ostium in the right sinus of Valsalva.

biopsy findings, it was felt this anatomical finding was the cause of his presentation.

He was taken emergently to the operating room for surgical correction. Transition from ECMO to cardiopulmonary bypass was instituted. Transverse aortotomy was performed and examination of the aortic root demonstrated an anomalous left coronary artery arising from a separate ostium in the right sinus (► **Fig. 1**), with an intramural course at the level of the most superior portion of the intracoronary commissure. Unroofing of the left coronary artery was subsequently performed. He was successfully weaned off cardiopulmonary bypass and transesophageal echocardiogram demonstrated mild improvement in ventricular function. He remained hemodynamically stable and the sternum was closed. His postoperative course was complicated by poor ventricular function, vasopressor dependence, flash pulmonary edema, and a pericardial effusion. Cardiac magnetic resonance imaging at discharge showed moderate to severely depressed global systolic dysfunction with an EF of 29%. Follow-up magnetic resonance imaging at 5 months demonstrated persistent left ventricular hypokinesia and only modest recovery in systolic function with EF of 36%. At 1-year follow-up, he is on maximal medical therapy and is participating in school activities.

Discussion

We present the case of a previously healthy adolescent male who developed acute myocardial failure after an episode of exertional syncope. The history of recent viral illness and rapid clinical deterioration led us to assume a diagnosis of acute myocarditis. This case demonstrates that congenital coronary artery anomalies must always be considered as potential diagnoses in any young patient presenting with exertional syncope, acute cardiac failure, or SCD. Missed or delayed diagnosis of this anomaly in the setting of severe cardiac decompensation leaves patients at high risk of

misdirected clinical management and subsequent suboptimal outcomes. As this report and others illustrate, TTE is an accurate tool to identify life-threatening coronary artery abnormalities.⁴ Coronary angiography is also effective in delineating coronary anatomy. Based on this experience, we recommend that TTE be routinely included in the work-up of all young patients presenting with acute cardiac decompensation. However, in cases where the coronary arteries cannot be accurately identified on TTE, additional imaging may be required to confirm/exclude a diagnosis of anomalous coronary artery anatomy. In such cases, it is appropriate to utilize coronary angiography or high-resolution cardiac computed tomography to elicit confirmatory images of coronary anatomy.

Patients with left AOLCA from RSOV may develop chronic myocardial ischemia and be at risk of sudden infarction/decompensation or cardiac arrest. The exact mechanisms leading to myocardial ischemia in left AOLCA from RSOV with intramural course remain unclear,⁵ and symptoms alone do not enable appropriate risk stratification.^{3,5} Surgical management of left AOLCA from RSOV has traditionally been limited to symptomatic patients with known myocardial ischemia. Recently, surgical unroofing of the intramural segment of the anomalous coronary has emerged as a safe and effective strategy to create a large neo-orifice in the appropriate sinus,³ without the need for coronary revascularization. Currently, it is not possible to accurately evaluate the risk of cardiac decompensation or SCD in patients with intramural left AOLCA from RSOV. We therefore agree with the recommendation to undertake surgical intervention in all patients identified with this anomaly.⁶ Surgical unroofing repair, coupled with aggressive heart failure therapy, allows for some recovery of myocardial function in patients with chronic myocardial ischemia secondary to intramural left AOLCA from RSOV.

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