Anomalous origins of a coronary artery from the pulmonary artery: A series of three case reports undetected into adulthood

Katie L. Mastoris, Ataul Qureshi, Navin K. Subrayappa, Matthew W. Martinez, James Wu

ABSTRACT

Introduction: An anomalous origin of the coronary artery from the pulmonary artery (ACAPA) is a rare congenital anomaly resulting in sudden death in 90% of infants during their first year of life. Diagnosis in living adults is particularly unusual, especially for left coronary artery arising from the pulmonary artery (ALCAPA) given the large perfusion distribution that the left system provides to the myocardium. Surgery to correct and restore a bi-coronary arterial circulatory system has historically been the standard of treatment given the lethality of the diagnosis. However, advances in coronary imaging have led to an increased incidence of diagnosis in adulthood and challenges the true association between sudden death and ACAPA. This makes the decision for surgical correction more complex.

Case Series: Herein, we presented three cases of coronary anomalies arising from the pulmonary artery that remained undetected until adulthood, only one of which was managed surgically.

Conclusion: Currently, the prevalence of ACAPA in adults is low. Hence, there is no strong evidence based recommendations for management of adult patients. With advances in diagnostic modalities and potentially an increase in reported ACAPA cases, more data will become available to help guide the decision of surgical versus medical management.
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Keywords: Anomalies, Cardiac surgery, Chest pain, Coronary vessel, Pulmonary artery

INTRODUCTION

Anomalous origins of the left or right coronary artery from the pulmonary artery (ALCAPA or ARCAPA) are rare congenital anomalies. Sudden death from myocardial infarction and congestive heart failure occurs in 90% of infants with this anomaly within the first year of life [1, 2]. For the few that survive into adulthood by way of collateral systems, ALCAPA/ARCAPA may be an important cause of sudden cardiac death. The pathophysiology between the two diseases is similar [2]. However, ARCAPA in adults has been reported more commonly than ALCAPA [3]. This distinction is likely due to the large perfusion distribution that the left system provides to the myocardium. Regardless, both diseases have remained rare diagnoses in adults [3, 4]. We present three cases of coronary anomalies arising from the pulmonary artery that remained undetected until adulthood, which may alter the current correlation...
between coronary anomalies, sudden cardiac death and the immediate need for surgical correction.

Case Series

**Case 1:** A 48-year-old Caribbean male was admitted to the hospital after experiencing chest pain and progressive shortness of breath over a three-month period. Initial ECG illustrated left ventricular hypertrophy with ST-T wave changes compatible with repolarization abnormality. This patient had an abnormal adenosine myocardial perfusion imaging study demonstrating a moderate perfusion abnormality in the inferobasal wall, which prompted a cardiac catheterization. Catheterization verified a dilated left coronary system in the absence of the right coronary artery ostium. Subsequent imaging with a cardiac MRI confirmed ARCAPA (Figure 1) along with a hyperdynamic left ventricle and severe left ventricular hypertrophy. The right ventricle was within normal limits for contractility and no ischemic scars were noted. Three months later, the patient underwent surgical correction to successfully restore a bi-coronary arterial circulatory system (Figure 2). Intraoperative findings were consistent with ARCAPA however significant right ventricular dysfunction and distension, particularly at the intersection of the pulmonary artery, was now noted. The right coronary artery measured 8 mm in diameter with a great deal of collaterals surrounding the heart on the epicardial surface. The surgery consisted of dissecting the right coronary artery from the pulmonary artery and re-implanting onto the aorta. The patient was discharged home after a normal postoperative course. His anginal chest pain completely resolved however some residual shortness of breath continued for the first few months postoperatively.

**Case 2:** A 49-year-old female, suffering from recurrent bilateral breast cancer and awaiting bilateral mastectomies, experienced right-sided chest pain that radiated to her axilla and right arm. Nuclear stress test demonstrated normal diaphragmatic attenuation artifact with no obvious signs of ischemia. Given the magnitude of the surgery planned, a cardiac catheterization was performed and displayed a very large, right coronary artery and a left main artery, which could not be cannulated. A cardiac CTA identified an ALCAPA and a diffuse ectatic coronary vessel with extensive collateral vessels throughout the myocardium and communicate between the right coronary artery and the left coronary artery (Figure 3). Echocardiogram evaluated normal regional wall motion of both the left and right ventricles with normal systolic function. Given the extensive collateral vasculature it was felt that the patient was not at an increased risk for surgery. She underwent the planned bilateral mastectomies successfully without any cardiac complications. Currently, the patient is being managed by a cardiologist; no surgical correction is planned at this time.

**Case 3:** A 63-year-old female with an extensive cardiac history including childhood myocardial infarctions, congestive heart failure, and a presumed diagnosis of endocardial fibroelastosis presented with left shoulder pain radiating to her upper back. She underwent a nuclear stress test, which was abnormal, and an echocardiogram, which showed decreased left ventricular systolic function with heavy calcification of the endocardium in the apical lateral, anterolateral and inferolateral segments. The patient had subsequent cardiac CTA, which showed the presence of ALCAPA with a scarred anterior and anterolateral wall resulting in hypokinesis (Figure 4). Moderate central mitral regurgitation was also identified. Although surgical intervention was initially recommended, the patient declined at the time of diagnosis. She is currently being managed in collaboration with an adult congenital heart disease specialist who also recommended closure of the anomalous left coronary artery through catheterization to prevent the steal phenomenon. The patient however remains satisfied with her overall level of functioning and symptoms and has declined interventions. She continues to be monitored for cardiac arrhythmias.

Discussion

An anomalous origin of the coronary artery from the pulmonary artery (ACAPA) presenting into adulthood remains a scarce diagnosis as over 90% of patients die within their first year of life [1, 2]. During fetal life, the pulmonary arterial pressure equals the systemic pressure allowing for antegrade flow into the anomalous coronary artery and perfusion of the myocardium. After birth,
however, the pressure in the pulmonary artery decreases and retrograde flow into the anomalous coronary artery ensues. This is known as coronary steal phenomenon and results in myocardial ischemia, infarction and sudden cardiac death [2].

The proposed mechanism of both ARCAPA and ALCAPA survival into adulthood involves significant collateral circulation to the myocardium originally supplied by the anomalous vessel. ARCAPA has more commonly been reported in adults and thought to be less fatal in infancy than ALCAPA due to the smaller perfusion distribution supplied by the right coronary artery compared to the left [3, 4]. ARCAPA’s overall incidence is estimated to be 0.002% compared to 0.008% for ALCAPA [3].

With advances in technology, the modality of diagnosis has changed over time from autopsy to surgery to catheterization and now angiography [5, 6]. These modalities have led to an increased number of reported adult cases of ACAPA. Many have postulated that the actual prevalence in adult population may be higher than reported and therefore the true association between sudden death and ALCAPA/ARCAPA is lower than described [2].

Historically, due to its association with myocardial infarct, heart failure, ventricular arrhythmias and sudden cardiac death, the standard treatment has been surgical correction [2, 7]. Limited data is available regarding the most effective surgical strategy, however, establishment of a dual coronary artery system with coronary transfer is preferred even if interposition graft is required [8]. Traditionally, plans for surgical correction before the onset of left ventricular function was favored. Owing to the possible lower association between sudden death and ACAPA in adults, the decision for a challenging surgical intervention becomes more complex. Recent literature has suggested that if mild to moderate chronic ischemia and limited necrosis is present, survival without surgical correction is possible [2]. There is even less evidence based recommendations for the medical management of ALCAPA and ARCAPA. Theoretically, optimizing risk factors for acute coronary syndrome and preserving collateral flow should delay the onset of the coronary steal phenomenon.
Data remains limited on adult manifestations of ALCAPA and ARCAPA and its true association between sudden cardiac death, progression to severe cardiac pathology, and treatment modalities. Our cases represent three symptomatic adult manifestations of ACAPA; only one of which underwent recommend surgical correction to restore a bi-coronary arterial circulatory system. The latter two preferred monitoring and referral to an adult congenital specialist for further guidance and management.

CONCLUSION

Currently, there are no strong evidence based recommendations for management of adult patient’s with anomalous origin of the coronary artery from the pulmonary artery (ACAPA) and strong collateral circulation due to the low prevalence of the anomaly. Our hope is that with advances in diagnostic modalities and an increase in reported ACAPA cases, more data will become available to support the decision of surgical or medical management.

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Katie L. Mastoris – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ataul Qureshi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Navin K. Subrayappa – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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