Collateral Deception: A Unique Presentation of an Anomalous Coronary Artery

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INTRODUCTION

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a very rare congenital cardiac defect, with a reported incidence of 0.002%.[1] ARCAPA is typically an incidental, isolated finding in childhood or adulthood, and diagnosis remains challenging because of the variability in presenting age and the lack of symptoms. ARCAPA is much less common than anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), which has an incidence of 0.008% and usually presents in infancy with symptoms of heart failure.[1,2] Transthoracic echocardiography is the initial modality for diagnosing coronary artery anomalies, although additional advanced imaging can be complementary and helpful for management.[3]

CASE PRESENTATION

A 6-year-old boy presented to the emergency department with intermittent vague exertional chest pain. Review of systems was negative for shortness of breath, palpitations, dizziness, or syncope. There was no family history of congenital heart disease, sudden death, or cardiomyopathy. Results of physical examination were normal, with no appreciable murmur, crackles, or hepatomegaly. Electrocardiography showed possible biventricular hypertrophy and mild intra-atrial conduction delay, which prompted referral to pediatric cardiology.

Initial echocardiography suggested an anomalous origin of the right coronary artery (RCA) from the left main coronary artery (LMCA; Figure 1); the patient was therefore referred for cardiac computed tomographic angiography (CTA) to further delineate the coronary artery ostium and course. Cardiac CTA showed a retrograde blush of contrast at the origin of the RCA into the main pulmonary artery, suggesting the alternative diagnosis of ARCAPA (Figure 2). Repeat echocardiography demonstrated the ARCAPA with diastolic retrograde flow into the main pulmonary artery, as well as the initial finding, which was now consistent with a prominent collateral coronary vessel from the LMCA (Figure 3, Video 1). This collateral vessel traveled anteriorly between the great arteries toward the RCA distribution. Cardiac catheterization with coronary angiography confirmed a rightward, inferior-aspect ARCAPA, with additional RCA distribution supplied by collateral vessels from the left coronary arterial system (Figure 4). The patient successfully underwent surgical reimplantation of the anomalous RCA.

DISCUSSION

The true incidence of coronary artery anomalies is unknown, as some may remain undetected in life, but is estimated to be up to 1% of the general population.[4] Coronary artery anomalies can be classified into anomalies of origin, course, anastomosis, and termination.[5] Management and treatment depend on the type of coronary artery anomaly and the presence of coronary insufficiency.[1]

We describe an interesting case of ARCAPA, in which a prominent collateral vessel on initial echocardiography deceptively suggested the alternative diagnosis of an anomalous RCA from the LMCA. The management of these two types of coronary artery anomalies can be on opposite ends of the spectrum; some cases of anomalous RCA from the LMCA may be observed and managed conservatively whereas ARCAPA needs surgical repair. In a recent review of ARCAPA cases, only 70 patients were reported between 1885 and 2006. Most cases were diagnosed incidentally during childhood, with heart murmur being the most common indication for initial cardiac evaluation. Symptomatic patients presented with chest pain, shortness of breath, cyanosis, palpitations, bradycardia, and/or congestive heart failure. Rare but serious presentations include cardiac arrest, acute myocardial infarction, and/or sudden death. ARCAPA is typically isolated, but in approximately one third of reported cases, it was associated with other congenital heart anomalies such as atrial septal defect, ventricular septal defect, patent ductus arteriosus, pulmonary or aortic stenosis, double-outlet right ventricle, aortopulmonary window, tetralogy of Fallot, or coarctation of the aorta.[1]

The pathophysiology of ARCAPA and ALCAPA depends on the direction of coronary blood flow and the adequacy of oxygen delivery to the myocardium. As the pulmonary artery pressure decreases in early infancy, a coronary steal phenomenon may occur when diastolic flow from the coronary artery is directed into the pulmonary artery instead of into the myocardium, resulting in ischemia. Unlike ALCAPA, ARCAPA is less frequently associated with ischemia or sudden cardiac death; some hypotheses include the smaller area of myocardium supplied by the RCA as well as the lower oxygen demand of the right ventricle. Ostial obstruction of the anomalous RCA origin may also be protective. Collateral vessels from the left coronary system frequently develop to supply the RCA distribution, which may also decrease the risk for myocardial ischemia.[6]

Initial screening diagnostic test results of patients with the eventual diagnosis of ARCAPA range from normal to nonspecific, as no characteristic features exist. Electrocardiography may show right-axis deviation, ventricular hypertrophy, ST-segment and T-wave abnormalities, inferior-lead Q waves, bundle branch block, premature atrial

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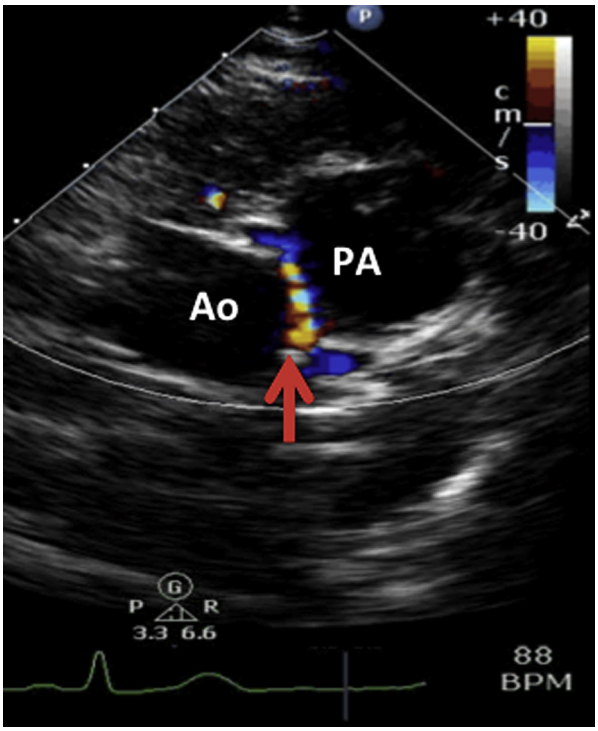

Figure 1 Parasternal short-axis view with color flow Doppler showing a vessel (*red arrow*) coursing between the aorta (Ao) and pulmonary artery (PA), suggesting anomalous RCA from the LMCA.

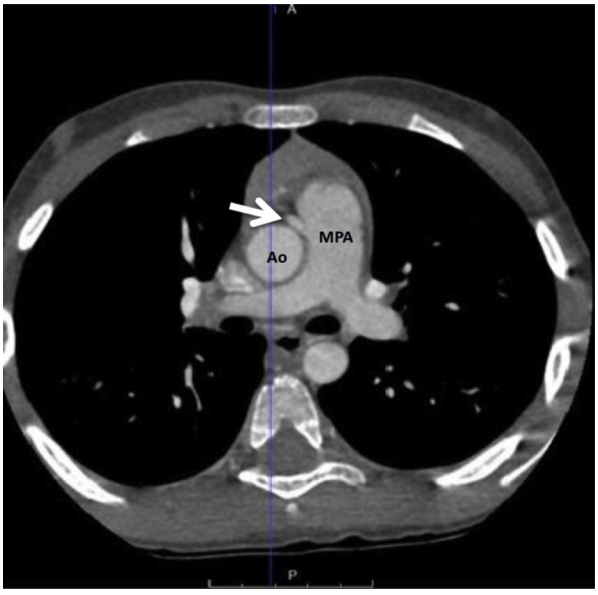

Figure 2 Cardiac computed tomography shows a retrograde blush of contrast from the origin of the RCA (*white arrow*) into the main pulmonary artery (MPA). *Ao*, Aorta.

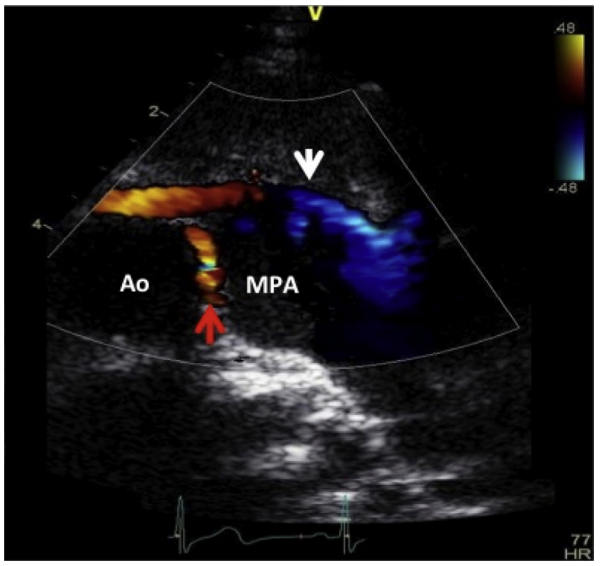

Figure 3 Modified parasternal short-axis view with color flow Doppler demonstrating the ARCAPA (*white arrow*) with diastolic retrograde flow into the main pulmonary artery (MPA) as well as the prominent collateral coronary vessel (*red arrow*). *Ao*, Aorta.

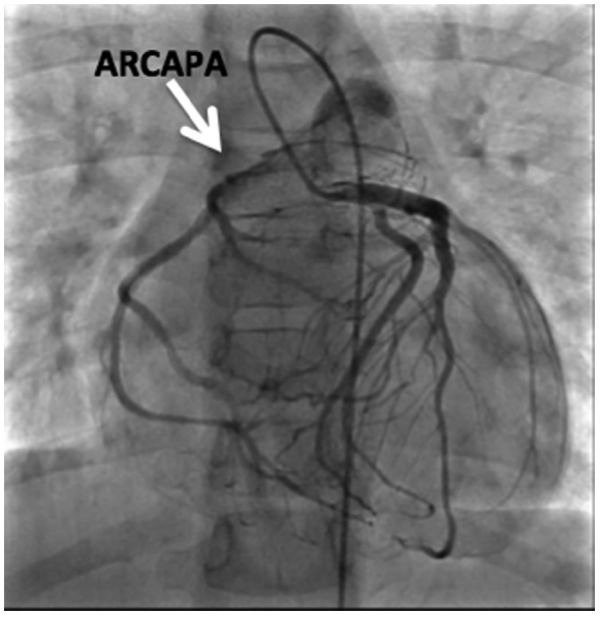

Figure 4 Coronary angiography injection into the LMCA, revealing collateral vessels supplying the right ventricular myocardium and retrograde flow into the ARCAPA (*white arrow*).

contractions, or atrial fibrillation. Chest radiography may demonstrate cardiomegaly and/or pulmonary edema caused by associated cardiac anomalies.

Historically, coronary artery anomalies were diagnosed by coronary angiography up until the mid-1980's. Since the 1990's, echocardiography has become the initial modality for diagnosing coronary artery anomalies. It is important to visualize coronary artery origins and proximal courses by two-dimensional and color flow imaging, which can be achieved by obtaining high-quality parasternal short-axis images. Focused imaging of the main and branch pulmonary arteries should be pursued when there is a high level of suspicion for anomalous origin of either the right or left coronary artery from the pulmonary artery. Echocardiographic findings in ARCAPA include retrograde flow from the RCA into the main pulmonary artery in

diastole, collateral vessels in the distribution of right and left coronary arteries, and dilated coronaries and left ventricle. Alternatively, color flow within the right ventricular myocardium may signal the existence of collateral vessels.[7]

Cardiac CTA and magnetic resonance imaging are excellent noninvasive, three-dimensional adjunct tools to aid in the evaluation of coronary artery abnormalities, because of their high spatial and temporal resolution.[3] Advanced cardiac imaging can provide additional information about coronary ostia and proximal and distal courses, which are important for surgical planning. Functional assessment with exercise stress test or myocardial perfusion imaging using positron emission tomography or single-photon emission computed tomography may show signs of ischemia and wall motion abnormalities. In this case, additional multimodal imaging with cardiac CTA and coronary angiography was valuable in confirming the diagnosis and guiding surgical planning.

Although many patients may be asymptomatic, surgical repair of ARCAPA is necessary to prevent coronary artery steal and subsequent myocardial ischemia and/or sudden cardiac death. Historically, some patients were treated with surgical ligation of the anomalous artery to avoid coronary steal.[1] Reimplantation of the RCA into the aorta is currently the treatment of choice to establish a normal, dual–coronary artery system.

CONCLUSIONS

Echocardiography remains the initial modality of choice for diagnosing coronary artery anomalies but requires a high index of suspicion with focused attention on coronary artery origins by both two-dimensional and color flow assessment. Adjunct imaging with higher resolution cardiac CTA or magnetic resonance imaging should be performed to further delineate detailed origin, course, and termination of coronary arteries for comprehensive presurgical evaluation.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.01.002.

REFERENCES

1. Williams IA, Gersony WM, Hellenbrand WE. Anomalous right coronary artery arising from the pulmonary artery: a report of 7 cases and a review of the literature. Am Heart J 2006;152:1004.e9-100417.
2. Kristensen T, Kofoed KF, Helqvist S, Helvind M, Sondergaard L. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) presenting with ventricular fibrillation in an adult: a case report. J Cardiothorac Surg 2008;3:33.
3. Lorber R, Srivastava S, Wilder TJ, McIntyre S, DeCampli WM, Williams WG, et al. Anomalous aortic origin of coronary arteries in the young: echocardiographic evaluation with surgical correlation. JACC Cardiovasc Imaging 2015;8:1239-49.
4. Davis JA, Cecchin F, Jones TK, Portman MA. Major coronary artery anomalies in a pediatric population: incidence and clinical importance. J Am Coll Cardiol 2001;37:593-7.
5. Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. Circulation 2002;105:2449-54.
6. Bansal M, Golden AB, Siwik E. Images in cardiovascular medicine. Anomalous origin of the right coronary artery from pulmonary artery with ostial stenosis. Circulation 2009;120:e282.
7. Afolabi-Brown O, Witzke C, Moldovan R, Pressman G. A different kind of Christmas tree: anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA). Echocardiography 2014;31:E52-4.