Case of anomalous origin of right coronary artery from pulmonary artery associated with interrupted aortic arch type A, diagnosed by multidetector computed tomography angiography

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ABSTRACT

Anomalous origin of the right coronary artery from pulmonary artery (ARCAPA) is a rare congenital anomaly of the coronary circulation, which can be easily missed by echocardiography. Interrupted aortic arch (IAA) is another rare congenital cardiac abnormality that typically presents in the first few weeks of life. We present a case of ARCAPA associated with IAA diagnosed with the help of multidetector computed tomography angiography, in a 7-year-old boy.

Keywords: Anomalous origin of the right coronary artery from pulmonary artery, interrupted aortic arch, multidetector computed tomography angiography

INTRODUCTION

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly of the coronary circulation, which can be easily missed by echocardiography and may remain asymptomatic. We describe a case of ARCAPA with interrupted aortic arch (type A), discovered with 64-slice multidetector computed tomography (MDCT) angiography. ARCAPA and interrupted aortic arch (IAA) are both very rare congenital heart anomalies. To the best of our knowledge, no case of ARCAPA associated with IAA has been described earlier.

CASE

A 7-year-old boy presented with breathlessness and recurrent respiratory tract infection for 3 months. His physical examination revealed tachycardia (heart rate – 122 beats per min), tachypnea (respiratory rate – 36 breaths per min), and Grade II/VI early systolic murmur at the left upper sternal border. Echo revealed patent ductus arteriosus (PDA), with a left-to-right shunt. Coarctation of the aorta was also suspected on echocardiography, and therefore, cardiac computed tomographic (CT) angiography was planned. MDCT angiogram showed ARCAPA with IAA type A and PDA [Figure 1], dominant left coronary system with single left coronary artery arising from the left anterior coronary sinus [Figure 1]. The pulmonary artery was markedly dilated and connected to descending aorta via large PDA, giving the appearance of a low aortic arch [Figure 2]. The patient was referred to a higher center for cardiothoracic surgery evaluation and unfortunately lost to follow-up.

DISCUSSION

Anomalous origin of the coronary artery from the pulmonary artery is a rare congenital heart anomaly.

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Four subtypes were described by Soloff in – anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), ARCAPA, both coronary arteries from the pulmonary artery, and an accessory coronary artery from the pulmonary artery. ALCAPA is most common anomaly of these with an incidence of 0.008% in the general population compared to 0.002% for ARCAPA. Unlike ALCAPA, isolated ARCAPA may remain asymptomatic and usually gets diagnosed early in life when associated with other structural heart defects (reported in 25%–30% cases of ARCAPA).[1] To the best of our knowledge, this is the first case report where ARCAPA was associated with IAA. There is a case report published about anomalous origin of the left anterior descending artery from pulmonary trunk associated with type B interrupted arch.[2] The pathophysiology of ARCAPA depends on the coronary artery blood flow direction and collateralization between the normal and anomalous vessel to maintain adequate myocardial perfusion in the presence of coronary steal.[3] In ARCAPA, the most common presenting symptom is angina (18%), followed by dyspnea on exertion (13%), congestive heart failure (11%), sudden cardiac arrest (6%), cyanosis, acute myocardial infarction, and palpitations.[4,5] IAA is a rare congenital anomaly due to complete luminal, and anatomic discontinuity between the ascending and descending thoracic aorta.[6] IAA is very rare with an incidence of about 2 per 100,000 live births.[7] It is almost always associated with PDA, ventricular septal defect, or both. It typically presents in the first few days to first few weeks of life. Survival to late childhood or adolescence is very rare with IAA. Very few cases of late presentation have been reported.[8] According to the Celoria–Patton classification of IAA, type A is defined as interruption distal to the left subclavian artery (42%); type B, interruption is between the left subclavian artery and left common carotid artery (53%); and type C, interrupted segment is between the innominate artery and left common carotid artery (4%). Each type is further divided into three subtypes depending on the normal or aberrant subclavian artery origin.[9] With the advent of fast MDCT and dual slice CT scanners, CT has become a single-stop shop for evaluating various cardiac anomalies.[10] Definitive management for ARCAPA is surgery even for asymptomatic patients as it decreases the risk of sudden cardiac death and also provides relief from coronary steal.[11] Definitive management of IAA is also surgical repair.

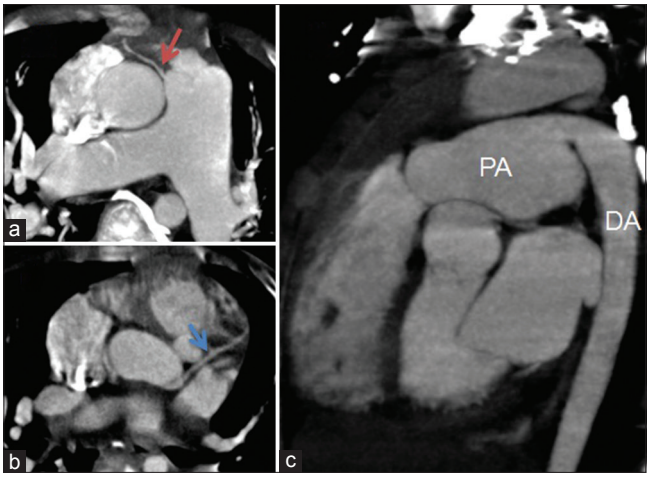

Figure 1: (a and b) The axial section of computed tomographic angiogram (maximum intensity projection) images showing anomalous origin of right coronary artery from pulmonary artery (red arrow) and normal left coronary artery origin (blue arrow). (c) Sagittal section of computed tomographic angiogram (maximum intensity projection) image showing dilated PA connected to DA. PA: Pulmonary artery, DA: Descending aorta

CONCLUSION

ARCAPA is a rare congenital coronary anomaly, and it is even rarer to have an association with IAA. Clinical symptoms depend on the degree of collateralization between the right and the left coronary system. As illustrated by this case, MDCT is well suited for evaluating

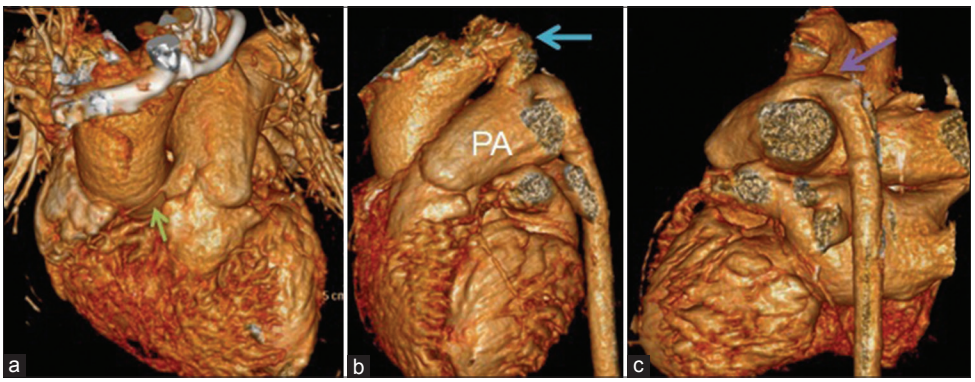

Figure 2: (a-c) Three-dimensional reconstructed volume-rendered computed tomographic images showing anomalous origin of right coronary artery (green arrow) from PA (a) further coursing in right atrioventricular groove: interrupted aortic arch (blue arrow), dilated PA connected to descending aorta via large patent ductus arteriosus (purple arrow) giving the appearance of a low aortic arch. PA: Pulmonary artery

ARCAPA because of its high spatial and temporal resolution and its flexible postprocessing techniques for distinguishing complex coronary anatomy and for assessing other associated cardiac anomalies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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