Anomalous right coronary artery from the pulmonary artery in an infant with tetralogy of Fallot

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ABSTRACT

An anomalous right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly that does not have the typical presentation of the more common anomalous left coronary artery. We present an infant with tetralogy of Fallot with atypical findings on the preoperative echocardiogram. A cardiac computerized tomographic (CT) scan showed ARCAPA. This was confirmed intraoperatively and repaired successfully. Close attention to coronaries on echocardiography and a low threshold for additional imaging can successfully diagnose ARCAPA in the presence of additional congenital heart defects.

Keywords: Anomalous right coronary artery from the pulmonary artery, congenital heart defect, coronary artery anomaly, tetralogy of Fallot

INTRODUCTION

Anomalous right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly, occurring with a significantly lower incidence than the anomalous left coronary artery (LCA).[1] It is often discovered incidentally through imaging or may manifest as heart failure, sudden cardiac death, or syncope.[2-4] In addition, ARCAPA can be associated with other congenital heart defects (CHD) such as tetralogy of Fallot (TOF), aortopulmonary window, and simple shunts in a third of cases, emphasizing the importance of diligent preoperative assessment.

CASE REPORT

An 11-month-old infant (weight: 7.6 kg) was referred to our center for surgical correction of TOF. Clinical examination revealed a pulse oximetry reading of 94%, tachypnea, a harsh 3/6 systolic murmur at the upper sternal border, well-felt pulses, and firm 2 cm hepatomegaly. A 12-lead electrocardiogram (ECG) showed a strain pattern of the right ventricle (pure R wave in precordial lead V1 and ST segment depression in right-sided precordial leads) [Figure 1], while the chest radiograph displayed significant cardiomegaly instead of the typical boot-shaped heart [Figure 2]. The preoperative echocardiogram confirmed TOF with a dysplastic pulmonary valve. In addition, a dilated LCA appearing to give rise to the right coronary artery (RCA) that traversed the interatrial space [Figure 3a] was seen. There were prominent coronary artery branches in the anterior aortopulmonary groove and tiny but noticeable epicardial collaterals on color Doppler imaging of the ventricles in cross-section. No coronary ostium was seen in the right aortic cusp. There was moderate right ventricular systolic

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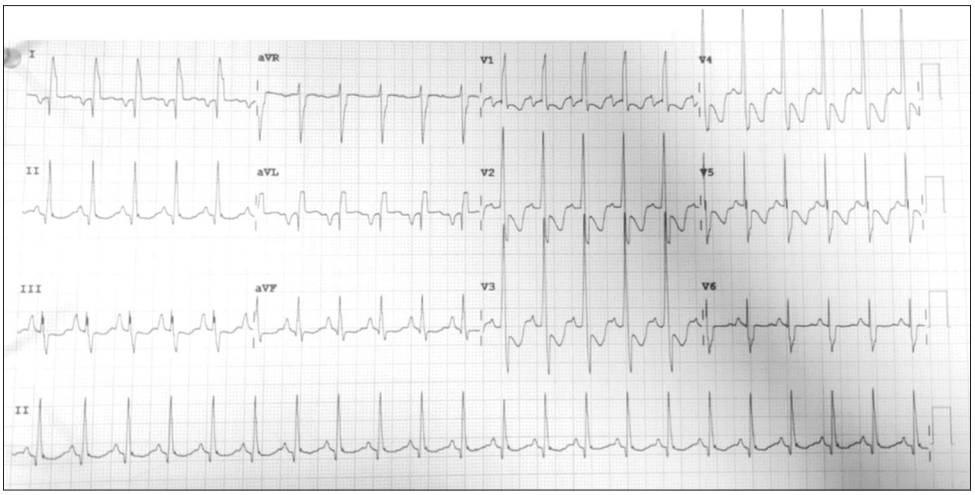

Figure 1: Electrocardiogram showing right ventricle strain pattern: ST depression and T wave inversion in the leads V1–V5, II, III, and arteriovenous fistula

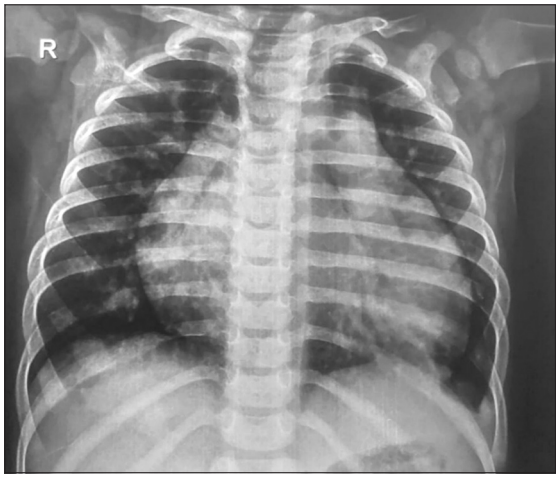

Figure 2: Chest radiograph showing cardiomegaly

dysfunction with a dilated inferior vena cava in the absence of severe cyanosis or acute respiratory illness. An ECG-gated computed tomography (CT) scan of the thorax with contrast confirmed the presence of ARCAPA, arising from the main pulmonary artery (MPA) well distal to the sinotubular junction [Figure 4]. Intravenous metoprolol was used to lower the heart rate.

Intraoperatively, ARCAPA was confirmed [Figure 5a and b] and the patient was placed on cardiopulmonary bypass. Aortic cross-clamping was performed, and the del Nido cardioplegic solution was delivered through the aortic root. Branch pulmonary arteries were snared to facilitate cardioplegia through the anomalous RCA. A right atriotomy was performed, and the heart was vented through a patent foramen ovale. The MPA was opened between stay sutures, and the RCA ostium was located. A partial aortotomy was done above the sinotubular junction, and ostial plegia was administered through ARCAPA. The RCA ostium was harvested from the pulmonary artery, mobilized, and implanted on the aorta using the open trap door technique with 7-0 Prolene continuous suturing [Figure 5c]. The defect in the pulmonary artery was reconstructed. The aortotomy was closed in layers, and the TOF component was corrected with a transannular patch and a monocusp valve. Cardiopulmonary bypass and cross-clamp times were 182 min and 161 min, respectively.

The patient had an uneventful postoperative course and was discharged on the 5th postoperative day. Follow-up echocardiography showed normal flow through the RCA ostium, improved right ventricular function, and satisfactory somatic growth.

DISCUSSION

ARCAPA is a rare congenital anomaly often associated with other CHDs, such as TOF. In such instances, the major structural heart defect gets highlighted in the preoperative work and the coronary anatomy may be relegated to a secondary "checklist" status with potentially disastrous consequences if the ARCAPA is undetected.[3] Although both coronary ostia can be seen clearly on the majority of echocardiograms done on infants, occasionally, artifacts may be mistaken for a coronary ostium.[2] The artifacts may be pericardial spaces or drop-out artifacts when the wall is parallel to the sonography beam [Figure 3a]. Another source of error is when the echocardiographer twists the probe to unconventional angles and images the coronary "satisfactorily" missing the fact that the coronary imaged in this manner actually arises from elsewhere. On retrospective review of the preoperative echo, the RCA was clearly seen arising off the MPA [Figure 3b] in an "off-angle" view. The dilated main LCA was also a pointer to compensatory dilatation due to the ARCAPA. The depressed right ventricular function seen preoperatively was due to reduced blood supply to it, secondary to adequate collateralization between the LCA and the RCA

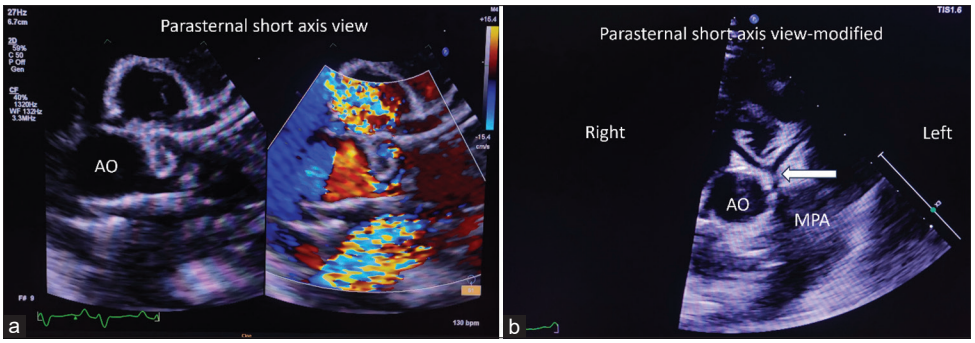

Figure 3: Echocardiogram showing the dilated left coronary artery (a) and origin of right coronary artery (RCA) from main pulmonary artery when retrospectively reviewed (b). Arrow shows anomalous RCA arising from the pulmonary artery. AO: Aorta, MPA: Main pulmonary artery

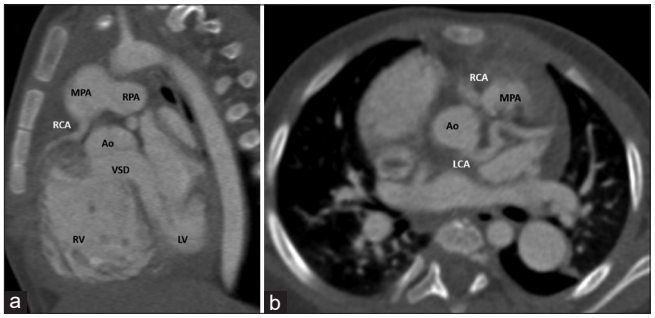

Figure 4: CT scan sagittal (a) and axial (b) views showing the left coronary artery and the anomalous right coronary artery from the pulmonary artery. AO: Aorta, MPA: Main pulmonary artery, RCA: Right coronary artery, LCA: Left coronary artery, RPA: Right pulmonary artery, RV: Right ventricle, LV: Left ventricle, VSD: Ventricular septal defect

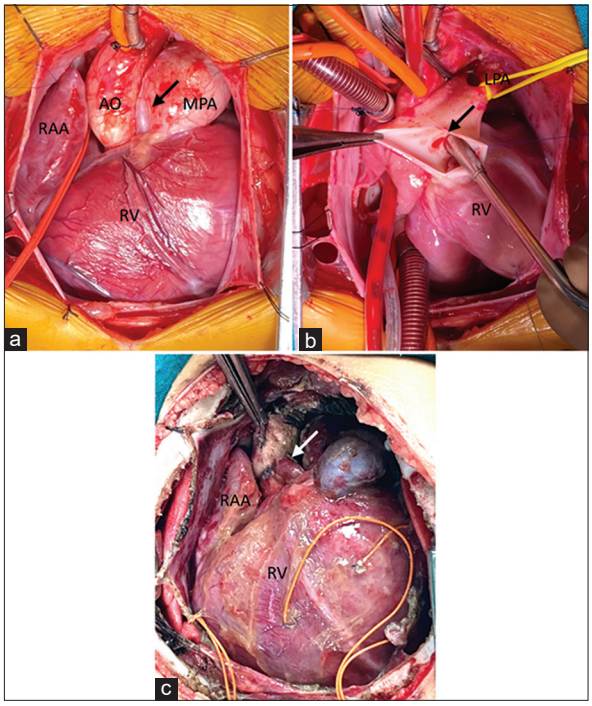

Figure 5: Intraoperative appearance of the right coronary artery (RCA) arising from the main pulmonary artery (a and b). RCA postimplantation (c). Arrows = RCA. RAA: Right atrial appendage, AO: Aorta, MPA: Main pulmonary artery, RV: Right ventricle

leading to coronary steal. Steps to minimize missing anomalous coronary origin include (i) color Doppler evaluation of the coronary with a low Nyquist limit, (ii) paying attention to the surrounding structures in "off-angle" unconventional views, and (iii) correlating presence of collaterals, dilated contralateral coronary with reduced ventricular function.

The investigative modality of choice beyond echocardiography in such a situation is angiography or a CT scan.[3] We chose to do a CT scan as it is less invasive. The surgical options for ARCAPA are translocation and reimplantation or ligating the RCA with or without a bypass graft.[4,5] The establishment of a dual coronary artery system is preferable.

CONCLUSIONS

This case underscores the importance of a thorough preoperative coronary artery evaluation, particularly in the presence of structural heart diseases, to ensure the detection of ARCAPA. The team should maintain a low threshold for obtaining additional imaging.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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