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CASE REPORT

Incidentally Detected Anomalous Origin of Right Coronary Artery from Pulmonary Artery Associated with Atrial Septal Defect and PDA

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Abstract
Anomalous origin of right coronary artery (RCA) from pulmonary artery (ARCAPA) with atrial septal defect (ASD) and patent ductus arteriosus (PDA) is a rare presentation. Anatomic abnormalities of coronary arteries may be subtle, and echocardiographic evaluation is difficult in many cases especially in absence of significant myocardial damage. We report a case of a four years old boy who was admitted for ostium secondum ASD closure and was incidentally detected to have ARCAPA intra-operatively. The right coronary artery was re-implanted to the aorta with closure of ASD and PDA ligation. The association of ARCAPA with ostium secondum ASD and PDA is relatively rare. The presence of left to right shunt affects the pathophysiology of ARCAPA and makes the preoperative diagnosis difficult. Surgical correction is mandatory to prevent further myocardial dysfunction.

Keywords ARCAPA · Coronary arteries · Ostium secondum ASD · PDA · Myocardial damage · Case report

Background

ARCAPA is a relatively rare congenital coronary artery anomaly affecting 0.002% of the population [1]. There is a paucity of literature regarding its actual incidence, natural history, clinical course, and management guidelines. In most cases, the diagnosis was made during evaluation of other cardiac anomalies. We discuss an incidentally detected case of ARCAPA in a 4-year-old male child who was otherwise diagnosed as a case of ostium secondum ASD and small PDA without any evidence of cardiac dysfunction.

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Case Report

A 4-year-old boy presented with history of recurrent lower respiratory tract infections having been diagnosed as a case of ostium secondum ASD at the age of one and half years. He was asymptomatic at presentation. On echocardiographic evaluation, there was normal atrio-ventricular and ventriculo-arterial concordance, laminar inflow, and outflow with normal functioning cardiac valves. There was a large ostium secondum ASD of about 18 mm × 20 mm size with left to right shunt. Right atrium and ventricle was dilated, and biventricular function was normal. Arch was left sided with normal branching pattern without any coarctation. There was a small patent ductus arteriosus (PDA) with left to right shunt. No coronary abnormality was reported in pre-operative echocardiography. Cardiomegaly was present on chest X-ray. Electrocardiogram (ECG) showed right axis deviation with right ventricular hypertrophy. On the basis of pre-operative evaluation, the child was taken up for ASD closure and PDA ligation under cardiopulmonary bypass. At the preoperative evaluation, his body weight was 12 kg, height was 94 cm, and body surface area (BSA) was 0.56 m^2.

Intra-operatively, right coronary artery (RCA) was found to take its origin from main pulmonary about 2.5 cm above the pulmonary annulus with a long intramural

course in the aorta. The main pulmonary artery (MPA) was dilated and soft. Right atrium and ventricle were dilated. After evaluating the anatomy of ARCAPA, cardiopulmonary bypass (CPB) was initiated by aorto-bicaval cannulation. The PDA was dissected, looped and ligated. Aortic cross-clamp was applied and cardioplegic arrest achieved with del Nido cardioplegia at 20 ml/kg. Aorta was opened obliquely and left coronary ostium was detected in its usual position in left coronary sinus. No other coronary ostium was detected on other sinuses. The MPA was opened longitudinally in between stay sutures, and right coronary ostium was localized. Direct osteal cardioplegia was given to RCA. RCA was dissected and coronary button harvested with adequate pulmonary wall tissue around. The defect in the MPA was repaired with fresh autologous pericardium. The RCA was dissected downward from its origin and made free from its attachment from the aortic wall. There was some bleeding from the aortic wall which was managed by buttressing a small piece of pericardium over it. The coronary button was then rotated and implanted to the ascending aorta (Fig. 1). The aortotomy was closed and a small dose cardioplegia was given through the aortic root to assess coronary filling. The ASD was closed with autologous fresh pericardial patch with continuous prolene suture. Weaning from CPB was smooth. The cardiopulmonary bypass time was 157 min, aortic cross clamp time was 88 min, and the temperature was reduced to 32° centigrade at the time of the procedure. There was no hemodynamic or electrocardiographic changes after termination of CPB. On epicardial echocardiography, ventricular function was normal without any obvious regional wall motion abnormality. The 12 lead ECG done on 1st post-operative day showed sinus rhythm with 1:1 conduction, T wave inversion in V1 to V2, and features of right ventricular hypertrophy. The cardiac enzymes were elevated in immediate post-operative blood sample (creatine kinase, CK, 1545 U/L; creatine kinase, MB, 44.8 ng/L). The enzyme level however normalized in subsequent blood samples at 2nd and 4th post-operative day (CPK < 300 U/L and CPK-MB < 6 ng/L). The patient had smooth post-operative recovery and was discharged from the hospital on the 5th postoperative day. The serial postoperative echocardiography showed normal ventricular function.

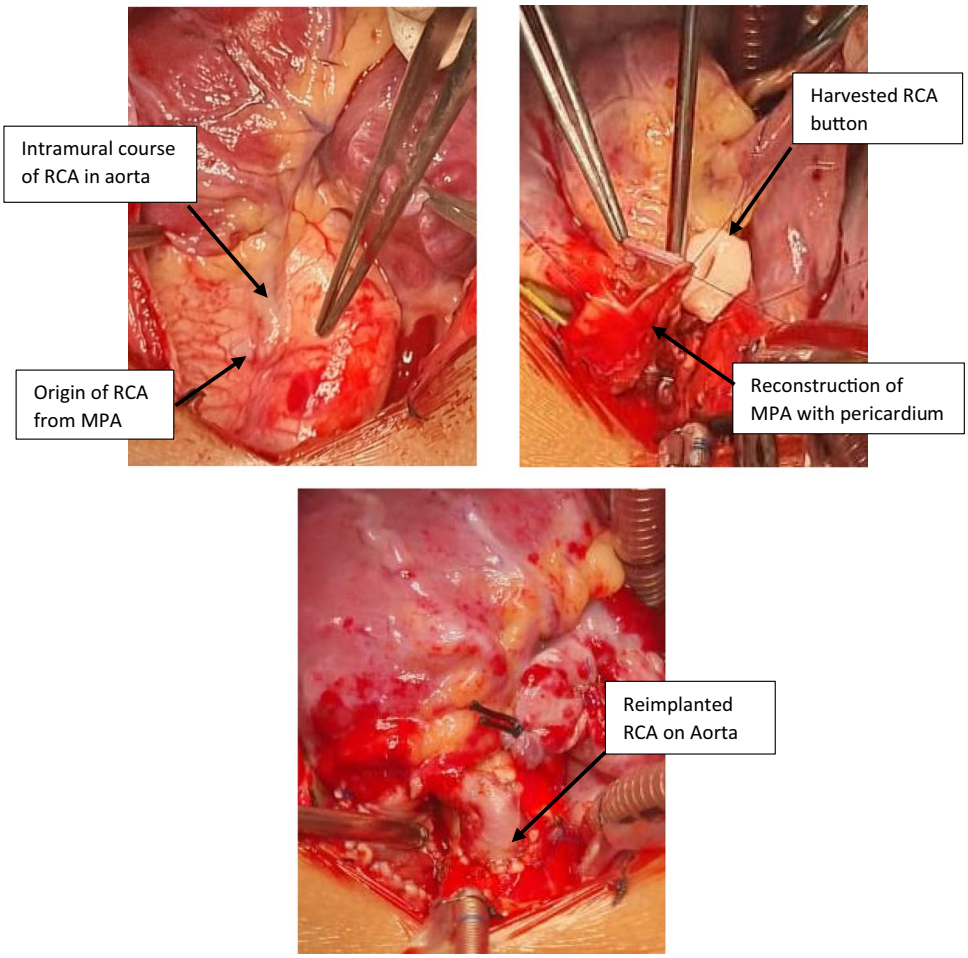

Fig. 1 Intraoperative images showing repair of ARCAPA by re-implantation of anomalous RCA into the ascending aorta

Discussion

Malrotation of the spiral septum dividing the truncus or malpositioning of the coronary buds is the primary cause of abnormal origin of coronary arteries. ALCAPA is the most common anomaly followed by relatively rare ARCAPA. Other forms of anomalies include origin of both the coronary arteries and accessory coronary artery from pulmonary trunk [1, 2]. In the fetal life and immediately after birth, due to increased pulmonary vascular resistance, blood flows from the pulmonary trunk to the anomalous coronary artery. The blood supply is sufficient in most of the cases to maintain normal ventricular function in spite of its low oxygen content. However gradually with fall of pulmonary vascular resistance, there is steal phenomenon resulting diastolic run off of blood from coronary artery to the pulmonary trunk [4].Collateral circulation gradually develops between the normal coronary and the abnormally originating coronary. If the collateralization is extensive, it can maintain the ventricular function initially. However, it may aggravate the coronary steal to pulmonary circulation due to relative difference in the diastolic pressure. Eventually, with the increase in ventricular demands cardiac function fails. If the collaterals are insufficient, the ischemic events and sudden cardiac deaths occur much earlier, and ventricular dysfunction is evident from the beginning [1, 3]. All these clinical features are more severe in anomalous origin of left coronary artery from pulmonary artery (ALCAPA) with earlier presentation. In majority of ARCAPA, there are no specific ECG changes or cardiac dysfunction in the early days of life. They are also less often associated with sudden coronary events than ALCAPA. This may be due to less right ventricular myocardial demand and smaller supply territory of the RCA [1, 3, 4]. However in right dominant coronary circulation, ARCAPA is less likely to be tolerated, and there are incidences of sudden cardiac deaths reported, especially in late presentation.

The association of other congenital cardiac anomaly also determines the clinical presentation in many cases. In our patient, the ARCAPA was associated with intracardiac left to right shunt through a large ostium secondum ASD and small PDA. Any left to right shunt increases the pulmonary blood flow and its oxygen content which maintains adequate perfusion pressure for antegrade coronary flow through the RCA with higher oxygen content. This in turn helps to preserve the ventricular function at normalcy for many years. Also, there is less development of collaterals in early years of life and less coronary run off to pulmonary artery. This poses difficulty in identifying the coronary artery origin anomaly by echocardiography [3, 4]. But it is important to recognize the defect and re-implant the RCA to the aorta to correct the coronary perfusion especially during closure of intracardiac shunt. Infants and young children present mostly with non-specific clinical features like cardiac murmurs, chest pain, arrhythmias, infections, and non-specific ECG changes [1, 3]. In adults, the presentation may be subtle with non-specific chest pains or effort intolerance. Patients with late presentation and ischemic symptoms or rhythm disturbances are at higher risk of developing sudden cardiac events [2]. In most of the cases however, the diagnosis was made during the evaluation for other congenital cardiac diseases. Many of the cases were detected preoperatively by echocardiography as well as by angiography [1]. Approximately 40% of the cases of ARCAPA are reported to be associated with other congenital cardiac anomalies. Tetralogy of Fallot, aorto-pulmonary window, bicuspid aortic valve, partial anomalous pulmonary venous drainage, septal defects, and aberrant right subclavian arteries are the congenital cardiac defects that have been reported to be associated with ARCAPA [5–7].

Echocardiography is the standard modality of investigation, and the origin of RCA from MPA can be demonstrated in parasternal long axis view in most of the cases with color showing antegrade or retrograde flow to the coronary artery. Color Doppler study also helps in detecting any intracoronary collaterals. The dimension of ventricles and right ventricular function can also be assessed. Sometimes the anomalous origin of RCA can be suspected from parasternal short axis view when (a) there is absence of coronary orifice in right coronary sinus, (b) the LCA is dilated with normal origin, and (c) the ratio of the diameter of the coronary artery originating from the normal sinus to the diameter of the aortic annulus is increased (normal value 0.10–0.28). However, misdiagnosis is also possible as mentioned by Wu et al. in their article where two cases has been misdiagnosed out of six with one of fistula between the pulmonary artery and the left coronary artery and another of ARCAPA diagnosed intraoperatively in one case of atrial septal defect [8]. Also, the size and the course of distal coronary arteries are often difficult to visualize in two-dimensional and color Doppler echocardiography because of poor image quality due to poor acoustic windows and their intramural or tortuous course [9].

Whether it is detected preoperatively or intra-operatively, the therapeutic recommendation is to re-implant the RCA into the aorta. Mishra in his article has mentioned four cases of ARCAPA. In all cases, the operative procedure included harvesting of RCA from MPA followed by its relocation to the aorta along with pericardial patch reconstruction of MPA. The additional defects were also addressed whenever present [10]. In some of the cases, the RCA was ligated when implantation was not possible [1]. During implantation, excessive tension to the anastomosis and bending of RCA due to redundancy should be avoided [3]. Thrombosis or stenosis is the possible late complication after coronary

transfer. Sometimes thrombosis occurs in dilated abnormal coronary artery due to late presentation. Antithrombotic medication should be started and continued to maintain luminal patency especially in cases where these kinds of complications are expected. All the patients require long-term cardiac follow up after surgery [3].

Abbreviations *RCA*: Right coronary artery; *ARCAPA*: Anomalous origin of right coronary artery from pulmonary artery; *ASD*: Atrial septal defect; *PDA*: Patent ductus arteriosus; *ECG*: Electrocardiogram; *BSA*: Body surface area; *CK*: Creatine kinase; *MPA*: Main pulmonary artery; *CPB*: Cardiopulmonary bypass; *ALCAPA*: Anomalous origin of left coronary artery from pulmonary artery

Author Contribution Both the authors are involved in preparing the manuscript. Both the authors have read and approved the final manuscript.

Data Availability Not applicable.

Declarations

Ethics Approval and Consent to Participate Ethics approval is not required as no new procedure is done. Consent to participate is not applicable.

Consent for Publication Obtained.

Competing Interests The authors declare no competing interests.

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