Rare case of anomalous right coronary artery from pulmonary artery (ARCAPA syndrome)

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ABSTRACT

Anomalous right coronary artery from pulmonary artery (ARCAPA) is a rare congenital anomaly of right coronary artery, most of the patient are asymptomatic, although myocardial infarction and sudden cardiac arrest has been reported. Herein we present a 5 month old infant presented with chest infection, during routine examination cardiac murmur detected, anomalous right coronary artery from pulmonary artery suspected by transthoracic echocardiography which showed dilated (4 mm) left main stem of coronary artery with mild left ventricular dilatation, but the origin of right coronary artery couldn’t be detected. Coronary angiography done which confirm the diagnosis and the child successfully operated on for re-implantation of right coronary artery to the aortic cusp.

Keywords: Anomalous right coronary artery from pulmonary artery, Right coronary artery, Anomalous coronary artery

INTRODUCTION

Anomalous right coronary artery from pulmonary artery is a rare congenital coronary anomaly with estimated prevalence of 0.002% [1]. It can be an isolated anomaly or be in association with other congenital cardiac defects [1]. The symptom and time of presentation may vary depending on the effective collateral branches to the myocardium, concurrent with decrease in pulmonary arterial pressure. Many patients are asymptomatic, but others may present with heart failure, myocardial infarction, valvular insufficiency and sudden cardiac death [2]. Diagnoses can be established by echocardiography, CT angiography, MR angiography and coronary angiography [1]. Surgical repair of the defect is recommended to prevent cardiac problems and sudden death [3], with good overall outcomes [4]. This is a rare case of anomalous origin of right coronary artery from pulmonary artery detected accidentally on routine checkup.

CASE REPORT

A 5-month-old boy, admitted to ICU for chest infection. During his routine physical examination, we could detect a cardiac murmur. After performing transthoracic echocardiography, mild left ventricular dilatation and 4 mm dilated left main stem of coronary artery with fistula to right ventricle detected (Figure 1).

Cardiac catheterization done which showed RCA arising aberrantly from the pulmonary artery from the right effacing cusp, with retrograde flow across multiple coronary fistulas draining into left and right ventricles and dilated left main coronary artery arising from the left coronary sinus, 4 mm in diameter, bifurcating normally into a large LAD and circumflex artery.
He was delivered normally at term, weighing 3 kg, no prenatal and natal abnormal event, he has no past medical history except for 2 times admission to hospital at the 1st year of life for chest infection (first admission was at 5th month of life, the 2nd one was at 9th month of age. Both admissions were for chest infection). He has no family history of coronary or cardiac disease, consanguinity is negative (Figures 2–8).

He was feeding well and gaining weight normally, at 3 year of age (weight 15.5 kg, height 96 cm) he underwent reimplantation of RCA in right aortic cusp. The early postoperative course was unremarkable; he needed inotropic support, extubated at postoperative day 1, with no respiratory complications. Discharged from hospital after 8 days, on Aspirin tab 80 mg daily (Figure 9).

Discharge echocardiography showed normal RCA at right coronary cusp with normal ante grade flow (Video 1 and Video 2). Discharge ECG showed normal sinus rhythm. Follow up echocardiography after 2 months (Figures 1–14) showed normal RCA.

**Video 1:** An anterioposterior view with catheter pigtail in the ascending aorta showing powered injection of dye through the left side which is normal, then backflushing of the dye to the pulmonary artery from the RCA. Additionally there is dilated left main steam.

**Video 2:** The lateral view of the same injection with biplane catheter lab in which shows the same power injection through the ascending aorta and dilation of left main steam. The flow comes back through collaterals to RCA, then drains directly to pulmonary artery.

**Videos URL**
http://www.ijcasereportsandimages.com/archive/article-full-text/100979Z01A82018

**DISCUSSION**

Anomalous origin of right coronary artery from pulmonary artery is a rare congenital lesion of the heart, it has been first described at 1885 [2]. With an estimated incidence of 0.002%, and 0.12% of all coronary anomalies [1, 5]. It is usually an isolated anomaly, only 25-30% of cases are associated with other structural cardiac lesions [6–9] including aortopulmonary window, truncus...
arteriosus, anomalous subclavian artery, tetralogy of Fallot, constrictive pericarditis, bicuspid aortic valve, and mitral regurgitation [4].

Pathophysiology of the disease depends on the direction of blood flow in coronary artery and pulmonary vascular resistance. It is well tolerated at the neonatal period because of high pulmonary vascular resistance which helps forward perfusion of abnormal coronary artery from pulmonary artery [6]. When the pulmonary vascular resistance falls, three possible scenarios ensue: 1) Insufficient collateralization between the right and left coronary arteries results in ischemia and death; 2) Adequate collateralization produces a steal phenomenon due to relative differences in the diastolic pressures between the pulmonary and systemic arterial beds, the fully oxygenated blood from abnormal coronary artery is stolen by pulmonary trunk results in chronic myocardial ischemia, or 3) Massive collateralization may maintain adequate myocardial perfusion even in the presence of the steal phenomenon [7].

Patients with ARCAPA are usually asymptomatic, when symptomatic; the clinical presentations are non-uniform, with congestive heart failure 30%, dyspnea 17%, angina 17%, fatigue 13%, myocardial infarction 9%, and sudden cardiac death 17%. The mechanism of sudden death is not completely understood, but may be related to the ischemia in myocardial territory supplied by RCA,
because of insufficient collaterals from left anterior descending and circumflex artery [5].

In our case, multiple collateralizations from left coronary artery have been developed that’s why the patient was asymptomatic.

Most of the patients demonstrate nonspecific ECG changes, they do not often exhibit typical features of
ischemia [8], it may be normal or shows left ventricular hypertrophy or deep Q-waves in inferior leads [1].

The diagnosis is mainly incidental, with advances in diagnostic technologies, diagnosis of ARCAPA during infancy and childhood has been increased [5]. Echocardiography is an essential diagnostic tool. CT angiography, MR angiography and coronary angiography are the gold standards for diagnosis, providing excellent visualization of coronary artery anomaly [6].

Our case diagnosed incidentally which he was admitted to hospital for chest infection, during physical examination we detect systolic murmur, after performing transthoracic echocardiography we suspect of having abnormal origin of right coronary artery, and we could confirm it by cardiac catheterization.

A corrective operation is recommended for all ARCAPA cases even if they are asymptomatic, because of risk of abnormal life events [8]. The correction can be done from neonatal period to adulthood. The surgery consists of RCA translocation and re-implantation to the aorta [3]. This technique provides double ostium coronary system and has the potential benefit of normalizing coronary flow reverse and lowering risk of sudden cardiac death [8].

At 3 year of age, our patient operated on successfully for re-implantation of RCA to the aortic cusp.

When anatomical considerations don’t allow for re-implantation technique, simple ligation of RCA at pulmonary artery or ligation of RCA with saphenous vein bypass grafting are alternative surgical techniques [1].

CONCLUSION

ARCAPA is a rare congenital anomaly which may be asymptomatic or presents in a varying range of symptoms. Diagnosis is usually made by echocardiography, CT angiography, MR angiography and coronary angiography. The treatment of choice is re-implantation of RCA to the aortic cusp with good overall outcomes.

REFERENCES


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Author Contributions

Aso Faeq Salih – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Chnar Mohammed Saeed – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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