International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Anomalous Origin of Right Coronary Artery Originating from the Pulmonary Trunk (ARCAPA): A Rare Incidental Finding in a Patient with Chest Pain at a Tertiary Care Hospital in Rajasthan

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Abstract: ***Anomalous origin of the right coronary artery originating from the pulmonary trunk (ARCAPA) is a rare congenital coronary anomaly. Mostly the patients are symptoms free and this anomaly is an incidental finding during evaluation for other problems. We present a case of a 53years male with no known comorbidities, presenting to the emergency department with intermittent chest pain and heaviness for three days. The initial electrocardiogram (ECG) demonstrated sinus tachycardia with q waves in inferior leads. Echocardiography was suggestive of fair ejection fraction. He underwent coronary angiography that was not associated with any stenosis. Contrast reflux was seen from the right coronary artery (RCA) ostium to the pulmonary artery along with retrograde filling of the RCA. There was no any significant obstruction of the RCA when it was traced via left to right collaterals. Right heart catheterization and pulmonary angiography were performed which confirmed the origin of the RCA from the pulmonary trunk. The patient was kept on medical management for the time being and was advised for surgery. Later he was discharged home with slight improvement in his symptoms. Origin of the RCA from the pulmonary artery (ARCAPA) is a rare congenital malformation. Most of the patients with ARCAPA remain asymptomatic. In this case report, the chest discomfort was probably unrelated to the coronary malformation. However, sudden cardiac death may have been linked to ARCAPA (if associated with Myocardial Infarction) and therefore a corrective surgical procedure is recommended even for the patients who are symptoms free. Surgical techniques which are available include: (a) Simple ligation of the RCA (b) Ligation of the RCA with saphenous vein bypass grafting and re - implantation of the RCA into the aorta (this method is considered to be better for the restoration of blood supply in myocardium). However, its long - term benefits have not been conclusively demonstrated.***

Keywords: ARCAPA, coronary anomaly, chest pain, pulmonary trunk, surgical techniques

1. Introduction

Anomalous origin of the right coronary artery originating from the pulmonary trunk (ARCAPA) is a rare congenital coronary anomaly (an estimated prevalence of 0.002%) [1].

Mostly the patients are asymptomatic & usually, these anomalies are detected on incidental basis. A review done by Modi, et al in 2010, twelve cases were diagnosed in infants ≤ 1 year of age, 44 cases were diagnosed in children ≤18 years of age, 17 cases were in adults >60 years of age, and in 8 cases, the age was not recorded [2].

Patients who are associated with cardiac defects are diagnosed early in life compared to patients with isolated ARCAPA. Those patients who are without any associated cardiac defects, may present with a murmur in heart on auscultation, congestive cardiac failure symptoms, and sudden cardiac death or may remain asymptomatic. Detection of ARCAPA is usually incidental on evaluation (like, angiography of coronaries for evaluation of the cause of chest pain [2].

We describe the case of a middle - aged male who presented with intermittent chest pain and heaviness, was diagnosed with ARCAPA. Informed consent statement was obtained for this.

2. Case Presentation

A 53year male, of Indian origin, with no known comorbidities, presented with intermittent chest pain and heaviness for three days. It was gradual in progression and associated with diaphoresis. Family history was not significant (no history of sudden cardiac death and premature coronary heart disease). He was a farmer by occupation and was non - smoker and non - alcoholic. No history of use any illicit drugs.

On examination, he was not comfortable due to intermittent chest discomfort. Blood pressure was 140/70 mmHg and the heart rate 92 beats per minute. There was no elevation in jugular venous pressure or carotid bruit in the neck.

Cardiac examination was not significant except for tachycardia and breath sounds were equal bilaterally. No pedal edema. A complete blood count and basal metabolic panel were within normal range. HisECG demonstrated sinus tachycardia with q waves in inferior leads.

Echocardiogram revealed the normal ejection fraction (approximately in the range of 50% - 55%). There were no regional wall motion abnormalities.

On coronary angiography (CAG), there was no any stenosis of the left main/left anterior descending artery. The right coronary artery (RCA) ostium was not seen and there appeared to be left to right collaterals with retrograde filling of the RCA. The opening of the RCA was seen in the superior cardiac structures most likely in the pulmonary artery.

There was no any significant obstruction of the RCA when viewed via left to right collaterals. Right heart catheterization and pulmonary angiography were performed to confirm the origin of RCA.

On pulmonary angiography the origin of RCA from the pulmonary trunk was evident. The visualized portion of pulmonary artery appeared patent.

The patient was discharged home on medical management with an advice for surgical correction. There wasslight improvement in his symptoms also.

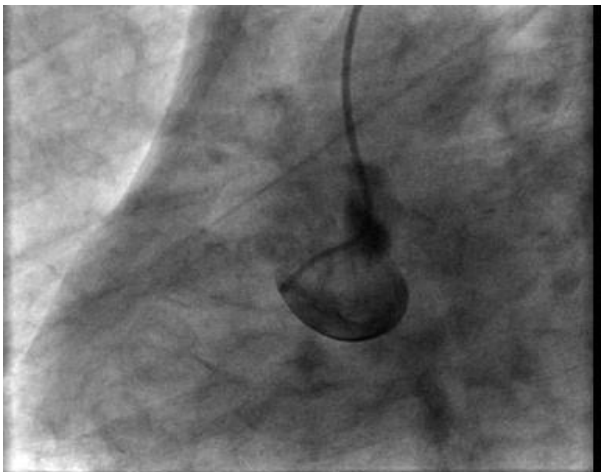

Figure 1: (No ostium of RCA seen)

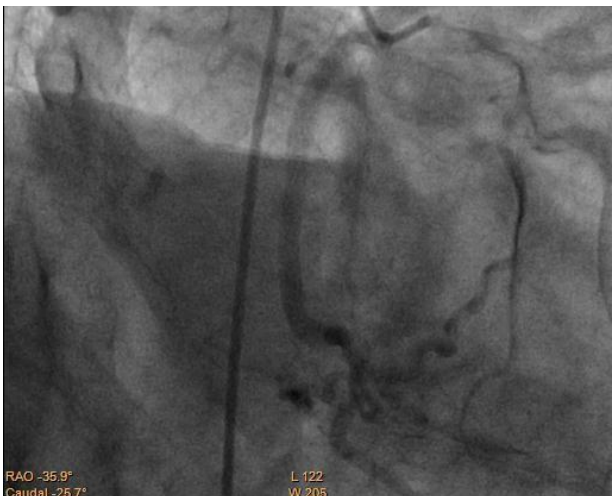

Figure 2: (Angiogram of the LAD showing retrograde filling of RCA, via collateral vessels. Reflux of contrast seen from RCA ostium to the pulmonary artery)

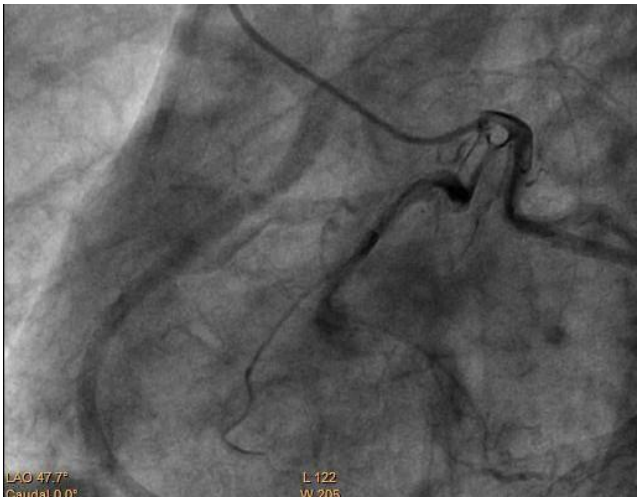

Figure 3: (Showing Retrograde filling of RCA in a different view, reflux of contrast seen from RCA ostium to the pulmonary artery)

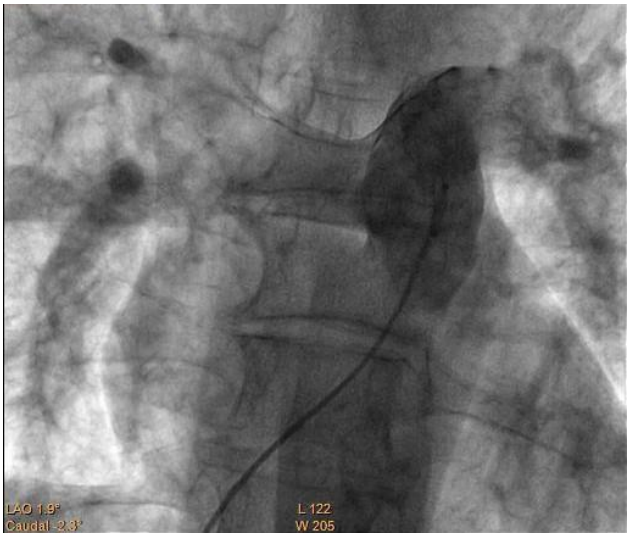

Figure 4: (Contrast in Pulmonary artery)

3. Discussion

Congenital coronary artery anomalies are rare, with an incidence of 0.3%–0.9% increasing up to 36% in patients with congenital heart disease [3].

Multiple classifications have been proposed on the basis of anatomy, angiographic, and hemodynamic findings [3].

According to Greenberg, et al., major anomalies leading to abnormal myocardial perfusion are an origin of coronary artery from opposite or non - coronary sinus, anomalous origin from the pulmonary artery, coronary artery fistula and myocardial bridging [4].

Amongst anomalous origin from the pulmonary artery, four variations of this condition have been described:

1) ALCAPA: An origin of the left coronary artery from the pulmonary artery.
2) ARCAPA: An origin of the right coronary artery from the pulmonary artery.
3) An origin of an accessory coronary artery from the pulmonary artery.

4) An origin of the entire coronary circulation from the pulmonary artery [3].

ALCAPA is more common than ARCAPA and is fatal in infancy [5]. The incidence of ARCAPA is estimated to be 0.002% of the population and represents 0.12% of coronary anomalies [1].

When the abnormality was recorded the age of the patients ranged from 1st day to 90 years. As most of the patients remain asymptomatic, the exact incidence of this coronary malformation is expected to be higher.

In contrast, the Bland–White–Garland syndrome (BWGS), patients often show mitral insufficiency and signs of anterolateral myocardial infarction early in childhood [6].

When symptomatic, the patients presenting with an ARCAPA is not consistent, and may include dyspnea (17%), fatigue (13%), congestive heart failure (30%), angina (17%), myocardial infarction (9%), and even sudden cardiac arrest (17%) [6]. The severity and the time of onset of the symptoms depend on the type of anomaly, the direction of blood flow in the anomalous vessel and the presence of collaterals. Normally, there is a retrograde flow in the anomalous artery due to the difference in pressure between systemic and pulmonary circulation which may lead to coronary steal phenomena.

Any increase in demand of oxygenmay lead to exhaustion of the physiologic reserve which may result in ischemia or infarction leading to sudden cardiac arrest [5].

ECG in ARCAPA, may be normal or it may show left ventricular hypertrophy or deep Q - waves in the inferior leads. Other diagnostic modalities include cardiac CT, CAG and cardiacMRI which provide visualization of anomalies of coronary arteries and also provide detailed anatomic information of the origin, course, and relationship of the anomalous coronary artery [5].

Adverse outcomes include increased risk of myocardial infarction and sudden cardiac death which are described in ARCAPA patients, regardless of symptoms, surgical correction is recommended whenever this anomaly is diagnosed [7].

Surgical corrections which are available include simple ligation of the RCA, ligation of the RCA with saphenous vein bypass grafting, and re - implantation of the RCA into the aorta [8]. The aim is to eliminate the left - to - right shunt and establish dual coronary circulation (to prevent the risk of myocardial ischemia from coronary steal). The location of the ostium of the RCA in the pulmonary artery will affect the technique used for surgical repair [3]. Transfer of the anomalous vessel to the aorta is the treatment of choice (since it provides establishment of dual coronary circulation that allows for normalization of coronary flow reserve and for greater protection against secondary coronary bypass graft changes because of age and atherosclerosis) [9].

When anatomical considerations preclude re - implantation of the RCA into the aorta, ligation of the abnormal pulmonary origin of the coronary artery or arterial bypass grafting should be considered as alternative therapeutic options [4]. However, surgical and pathological reports describe the anomalous RCA as being thin - walled, dilated and vein like structured; therefore, it is thought that the RCA does not tend to normalize in diameter after surgical correction of ARCAPA [8]. Also, symptoms and myocardial ischemia persisted in patients after reimplantation along with dilated coronary arteries with persistent slow runoff into the periphery [8].

4. Conclusions

This case described the incidental findings of a rare coronary anomaly in a patient who presented with chest pain and heaviness. Hissymptoms was probably unrelated to the coronary malformation. Sudden cardiac death has been linked to the anomalous origin of the right coronary artery originating from the pulmonary trunk (ARCAPA) and therefore a corrective operation is recommended regardless of symptom status.

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