Fistulae from distal part of Right Coronary Artery to Right pulmonary Artery: Case report and review of literature

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ABSTRACT:

A congenital coronary arteriovenous fistula (A-V-Fistulae) is a direct communication between a coronary artery and the lumen of any one of the four cardiac chambers, the coronary sinus or its tributary veins, or the SVC, pulmonary artery or pulmonary veins close to the heart.

We are presenting a case of fistulae from distal part of RCA to right pulmonary artery. This form of fistula is very rare.

Abreviations: RCA: Right coronary artery.

RPA: Right pulmonary artery

LCX: Left circumflex artery.

Congenital coronary arteriovenous fistula is a direct communication between a coronary artery and the lumen of any one of the four cardiac chambers, the coronary sinus or its tributary veins, or the SVC, pulmonary artery or pulmonary veins close to the heart.

More than 90% of fistulae open into right heart chambers or their connecting vessels. About 20% connects to pulmonary artery. In surgically treated cases the fistulous opening when single is seldom larger than 2 to 5 mm, although uncommonly it may itself be aneurismal. In anomalous connection of left coronary artery to pulmonary trunk, the whole of the left main coronary artery or only the LAD or circumflex branch connects anomalously to the proximal pulmonary trunk or very rarely to the proximal right pulmonary artery. Branching pattern of the anomalously connecting left coronary artery remains normal. The right coronary arises normally from the aorta and has a normal branching pattern.collatertals from the RCA feed the left coronary artery in which flow is reversed, so that the left coronary artery drains into the pulmonary artery. Very rarely both coronary arteries connect to the pulmonary artery by a single trunk.

CASE REPORT:

A 57 y/o man admitted with dyspnea and chest pain since 3 years ago that was aggravated by activities. He had palpitation from that time and he was in FC III.There was not any positive family history and he was well since that time .he used drugs as nitrocontine,simvastatin and aldacton,lasix and digoxin.In physical exam : BP=140/80 PR=80 RR=12 T=37.0C

JVP was distended. Lung was clear. There was a III/VI systolic murmur at LBS.

Abdomen and extremities were normal, and there was not organomegally, or peripherals edema, and peripheral arteries were normal. Chest x-ray showed cardiomegally and C/T ratio was about 60%.

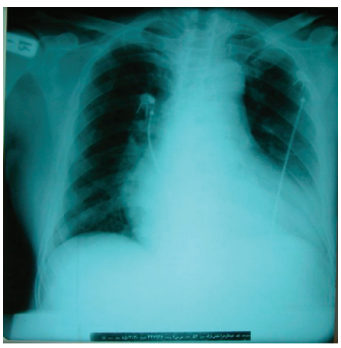

Figure 1: *Chest X-ray*

ECG findings were: NSR, RVH and clockwise rotation of heart.

TEE findings were:

1/sever LV dilatation. 2/sever diastolic LV dysfunction. 3/moderate MR. 4/sever AI. 5/sever pulmonary hypertension. 6/RCA fistula probably to RCA. In angiography there was significant LAD lesion, RCA was dilated and there was absence of LCX, and a fistula from distal RCA to RPA was seen. There was sever AI with

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bicuspid aortic valve.
The above finding was also seen by CT- Angiography

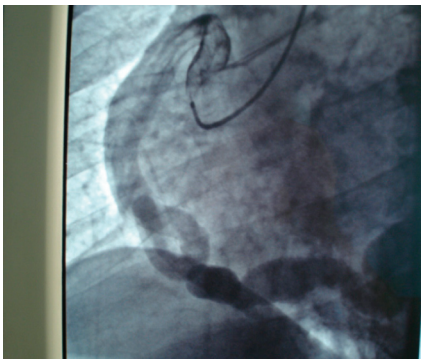

Figure2: *Angiography of RCA*

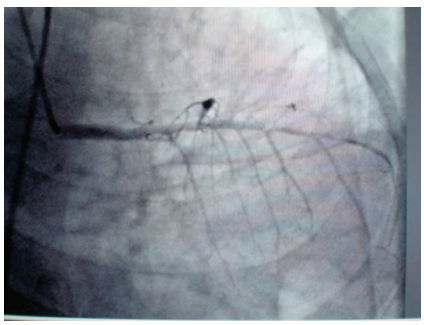

Figure 3: *Angiography of Left main coronary artery.*

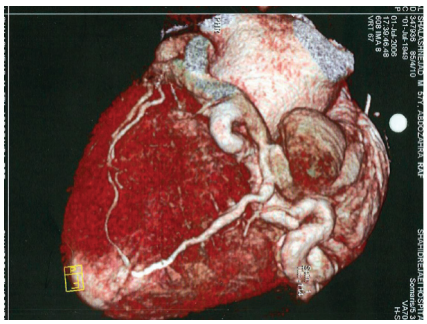

Figure4: *CT-Angiography, PDA and the fistulae*

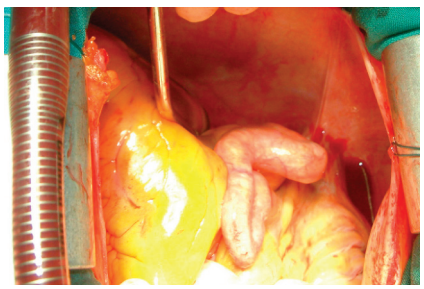

Figure5: *Dilated RCA*

DIAGNOSIS:

We operated this patient with diagnosis of CAD with LAD lesion, RCA to RPA fistula, sever AI and mild to moderate MR with sever dilatation of LV Accompanied with sever LV diastolic dysfunction.
Under CPB and moderate hypothermia and cardiac arrest we performed a LIMA to LAD and SVG to Diagonal. anastomosis, then AVR because of sever AI and bicuspid aorta ,with NO:21 metallic valve was done and the fistula was explored via RPA and was closed from the inside of RPA .then it was ligated and sewed from RCA. CPB was discontinued with mild dose of dopamine. Pump time was 160 minutes and clamp time was 123 minutes. Patient was extubated at first day in ICU and discharged from ICU in day 3.In post-op visit 3 months later he was in function class 1 without any symptoms

Discussion:

Coronary artery fistulas are rare abnormalities with an estimated frequency of 0.27-0.4% of all congenital cardiac lesions. The causative factors are unknown, but most are thought to originate as congenital anomalies or, less commonly, as a result of injury during coronary intervention or surgical procedures [2]. They may develop when enlargement of the capillary network occurs during the embryogenesis of coronary circulation or when the main coronary arteries remain attached to the pulmonary trunk at the time of their separation [3].
Most coronary artery fistulas are asymptomatic. Symptoms are more likely to develop in older patients or in those with larger fistulas[2].Patients can,however,present with myocardial ischemia,angina,congestive heart failure, bacterial endocarditis,cardiac arrhythmia,or fistula rupture with or without associated chest pain and tamponade and in the other hand it steals blood from coronary circulation. There is general agreement that symptomatic patients should be treated. Patients with coronary artery fistulae and another complex cardiac lesion for which an operation would be indicated and which should undergo surgical closure of fistula. Our patient was the first case who had fistula from distal part of right coronary artery into the right pulmonary artery .he had sever AI that can explain his heart failure symptoms and we can explain his chest pain because of his coronary artery lesions, but a large fistula in the other hand can explain some of his symptoms

and fistula closure reduced the flow and the shunt from his heart and could reduce the work of heart at all and recovered the 02 saturation in this patient.

Conclusion:

Fistulae from distal part of RCA to RPL are a rare condition and closure of this type of fistulae can eliminate symptoms of patient. We think that because the LCX was absent in this patient the origin of fistulae in this patient was LCX artery that anomalously originated from RCA(the most popular anomaly of coronary arteries) during embryogenesis of coronary circulation or when the coronary arteries remain attached to the pulmonary trunk and did not separate from pulmonary trunk.

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