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Multiple coronary- Artery fistulas with chronic occlusion of right coronary artery acquired or congenital? : Two cases and literature review

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Abstract
Coronary artery fistulas are rare abnormalities of artery termination. Most of the coronary fistulas are congenital, acquired forms may rarely due to atherosclerosis. The diagnosis of these anomalies increased with the routine use of coronary angiography and multi detector computed tomography in the investigation of chest pain.
We report two rare cases with multiple coronary artery fistula developed in the right coronary artery which is severely occluded, diagnosed when performing a coronary angiography for acute coronary syndrome.
The aim of reporting these cases is to emphasize the involvement of severe atherosclerosis in development of acquired multiple coronary artery fistulas.

Keywords: coronary artery fistulas, atherosclerosis, coronary angiography, acquired, angiogenesis

Introduction

Coronary artery fistulae (CAFs) are defined as an abnormal direct communication between one or more coronary arteries with a cardiac chamber, great vessel, or other vascular structure bypassing the capillary network [1]. First description for this anomaly was in 1865 by the anatomist W. Krause. While the majority of CAFs are congenital, Acquired forms are extremely rare and usually iatrogenic, posttraumatic, or caused by atherosclerosis [4]. In view of the increased use of endovascular procedures, acquired fistulas are reported increasingly [21]. We report two cases of multiple CAFs with chronic occlusion of right coronary and we discuss whether they are congenital or acquired.
Our case report was written according to CARE guidelines [23].

Case 1

A 45 year-old man, with a history of hyperlipidemia, presented to the emergency room, after one hour and thirteen minutes of an anterior chest pain which typically radiated to the left arm, without other associated complaints. The patient reported a typical angina class II of the Canadian Cardiovascular Society (CCS) grading of angina pectoris during the past five years.His vital signs were stable and physical examination was normal. An immediate electrocardiogram registered sinus rhythm at 75 with ST elevation in anterior leads. Coronary angiography was indicated but was not feasible immediately. Therefore, the patient underwent successful thrombolysis after eliminating the contraindications. Echocardiogram showed wall motion abnormalities in the left anterior descending artery (LAD) area (akinesia in mild segment of septal wall and hypokinesia in the apex and anterior wall), moderate left ventricular dysfunction (ejection fraction estimated at 40%). The troponin level was clearly positive with a high LDL level, the rest of the laboratory assessment was unremarkable. Coronary angiography was performed before 24 hours. It revealed a severe lesion in mild segment of LAD with TIMI III flow, a total occlusion of RCA, a severe lesion in the ostium of the left circumflex coronary artery (LCX) and three fistulous communications. The first fistula arose from the first segment of the right coronary artery with drainage located on the mediastinal vessel (figure1), the second communicated both, conus branch and RCA with the right atrium, while the third one was connecting conus artery with the great cardiac vein and coronary sinus (figure2). The conus branch was selectively cannulated and took over the flow after the second occluded segment of the RCA (WERNER class III). The patient underwent surgery for coronary artery bypass grafting (CABG) left internal mammary artery to the left anterior descending, right internal mammary to the right coronary artery and saphenous vein graft to the obtuse marginal artery and closure of coronaro cameral fisula. The outcome was unfortunately unfavorable and the patient died postoperatively from refractory cardiogenic shock.

Case 2

A 57-year-old male, presented to our emergency care with typical chest pain. He reported a history of smoking and hyperlipidemia that was well-balanced on statin. His physical examination was normal: blood pressure was 130/70 mm Hg and heart rate at 73 beats/min. No signs of heart failure and no added sounds or murmurs were heard on cardiac auscultation. The electrocardiogram (ECG) showed sinus rhythm at 73 b/m and ST segment

depression in 8 leads coupled with ST segment elevation in aVR. Troponin I at admission was 3.15 ng/ml (normal values <0.028 ng/ml), the remaining laboratory tests were normal. Echocardiography showed a preserved systolic function (EF at 60%) without wall motion abnormalities. Moderate enlargement of the left atrium (area at 22cm) was noted. The percutaneous coronary angiography, performed with right radial artery access, revealed a chronic occlusion of the right coronary artery in its second segment with presence of three fistulous communications, the first one originating from RCA and draining into the right atrium (coronaro cameral fistula) (figure 3) and two small arteriovenous fistula connecting the right coronary artery with the coronary sinus and the sinus branch of RCA with pulmonary trunk (figure4). The RCA is receiving collaterals from the left anterior descending artery (LAD), which had a high grade stenoses in the middle segment. The left circumflex coronary artery was normal. The patient had a successful CABG. Ligature of fistulas has not been envisaged due to their small diameter.

The outcome was favorable and the follow-up was without particularties. The patient had no more ischemic angina.

Discussion

Coronary artery fistulas (CAFs) are a rare coronary anomalies, with a prevalence estimated at 0.002% in the general population and approximately 0.05–0.25% among patients undergoing coronary angiography [2]. The anomaly accounts for only 0.4% of congenital heart defects and 20% of people with congenital CAFs have other concomitant cardiac anomalies (aortic and pulmonary atresia, patent ductus arteriosus and tetralogy of Fallot) [3]. Coronary-cameral fistula (CCF) is a fistula that arise from a coronary artery and terminate into a chamber heart (left or right), due to a fail of obliteration of the intramyocardial trabecular sinusoids system in embryological life [5]. CCF can be divided into two types, arterioluminal and arteriosinusoidal. The arterioluminal types have fistula connections directly into the cardiac chamber. Arteriosinusoidal types have an indirect connection to the cardiac chambers via a cardiac sinusoidal network [6]. CAFs to other vessels form is when one of the branches that should disappear actually persists and connects to the branch from the aortic sinus [7]. The major sites of origin are the right coronary artery (55%), the left coronary artery system (35%), and both coronary arteries (5%). Drainage occurs most frequently to the right ventricle, in about 40%, followed by the right atrium, coronary sinus, and pulmonary trunk. Less frequently they may drain into the superior vena cava, left atrium or left ventricle [8]. Clinical presentation depends on factors such as the age of patient, amount of shunting, development of cardiac ischemia, and resistance of recipient vessel or chamber [3]. Symptoms are present in 19–63% of patients, with the majority occurringxx after 18 years of age.The most common symptoms reported are dyspnea and chest pain [9]. Angina pectoris was often present in the absence of underlying coronary artery disease, aortic stenosis or hypertrophic cardiomyopathy and the ischemia is related to coronary steal phenomenon in which the fistula produces a shunt of blood flow away from the coronary artery runoff. This can lead to a demand-supply mismatch in the dependent myocardium, particularly during exercise [9, 10, 11]. Heart failure and pulmonary hypertension may be developed due to volume overload, when the fistula terminates in the right or the left side of the heart [7]. Rarely, pericardial effusion and sudden death can be a presenting feature [14]. In asymptomatic patients, the most common clinical presentation of CAF is a soft, continuous murmur that tends to be crescendo- decrescendo in both systole and diastole but louder in diastole. When fistulas terminate in a left-sided chamber the aortic run-off mimics aortic regurgitation murmur [9]. Coronary angiography remains the gold standard for the diagnosis, new methods, including multidetector computed tomog- raphy (MDCT) or multislice computed tomography (MSCT), magnetic resonance imaging (MRI), transesophageal echocardiography, trans-thoracic echocardiography and Doppler echocardiography, have also been beneficial [12]. The natural history of CAF is variable, with long periods of stability in some and sudden onset or gradual progression of symptoms in others. Spontaneous closure of CAF is uncommon but has been reported. Complications include bacterial endocarditis, thrombosis, distal embolization, aneurysm formation, dissection, rupture, premature atherosclerosis, pulmonary hypertension, atrial fibrillation, ventricular tachyarrhythmias, myocardial ischemia, or infarction [3, 13]. The main indications for closure of CAFs are clinical symptoms, especially of heart failure and myocardial ischemia and in asymptomatic patients with high-flow shunting, to prevent occurrence of symptoms or complications, especially in pediatric population [15]. The treatment options include transcatheter closure, surgical ligation and pharmacological treatment. Indications for surgical management are distal fistulas, large, high-flow fistulas, multiple complex communications, tortuous arteries, prominent aneurysms, a wide drainage site, associated clinically significant cardiac lesions, the need for simultaneous bypass, and the presence of large vascular branches that can be accidentally embolized, when the fistulas are diffuse; Intervention becomes more difficult or impossible and symptoms can be relieved by medical therapy [13]. Catheter closure can be performed with a proximally located fistula or a single drainage site and for those with a high perioperative risk and safe accessibility to the coronary artery supplying the fistula, a variety of materials have been used, including Gi- anturco coils, covered stainless-steel coils, detachable balloons, coaxial embolization with platinum microcoils, double-umbrella devices, the Gianturco Grifka vascular occlusion device and Amplatzer duct occluder [7, 14, 16]. Pharmacological treatment include antiplatelet therapy to prevent thrombosis, Warfarin in severe coronary artery dilatation (10 mm) and Beta-blockers or calcium channel blockers in angina induced by CAFs [13, 9, 3]. Luo *et al*. recommended Prophylaxis for bacterial endocarditis is recommended in all CAF patients and in patients after complete fistula occlusion for at least 1 year [3]. Hyper lipidemia, systemic hypertension,

diabetes and tobacco use are associated with an increased risk of complications with CAFs. Therefore, control of these factors is important [7].

Most of CAFs are congenital, acquired fistulae had been reported in cases of chest trauma and as a complication of percutaneous coronary intervention, permanent ventricular pacing, endomyocardial biopsy, myocardial infarction and cardiac surgery [17].

Coronary atherosclerosis was postulated as a cause for arterioatrial shunting by Searcy *et al* in two patients who had drainage from the right coronary artery to the right atrium [18]. In the presence of severe narrowing of the coronary artery, myocardial hypoxia could stimulate collateral vessel development by angiogenesis process, as an important alternative mechanism adapted by the hypoxic tissue. These collaterals channels can "lost their way" or direction and terminated in a cardiac chamber directly by thebesian veins or another vessel. It is suggested also, that arteriovenous channels dilate and permit enough shunting of blood to be visualized during coronary arteriography, in severe right coronary artery stenosis [17, 18, 19]. SALAH. A *et al* suggested that there is no difference between congenital and acquired CCFs in terms of vessel involvement, the acquired CCFs showed no specific or unique symptomatology and the majority of patients were be treated medically [19].

CAFs in our cases had a small diameter and they were all developed over of the RCA occlusion which had a normal proximal size. However, in congenital fistulas, the right coronary artery proximal to the fistulous communication were usually tortuous and aneurysmal [20]. Also in our patients there were multiple CAFs: two coronary arteriovenous fistulas, arising from RCA and terminating in pulmonary trunk and coronary sinus and one Coronary cameral fistula communicating proximal RCA with right atrium via a sinusoidal network, while rarely reported in congenital fistulas [19]. Because of the age of the patients, small size of the fistulas, the rarity congenital multiple fistulas, the lack of dilatation of the proximal right coronary and findings of literature review we believe that these fistulas are due to severe atherosclerosis and the most likely mechanism is the development of aberrant collaterals that led to fistula formation.

In the best of our knowledge, these are the first two cases in the literature of acquired multiple coronary artery fistulas secondary to severe atherosclerosis.

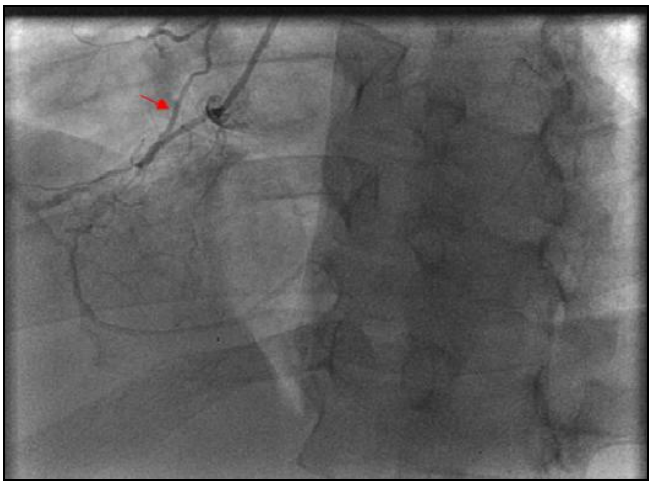

Fig 1: Coronary angiography showing a fistula arising from the right coronary artery with probable drainage located at the mediastinal vessels (red arrow)

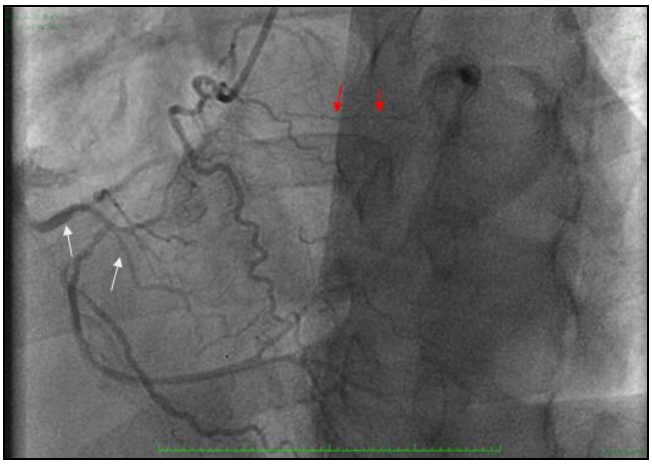

Fig 2: Coronary angiography showing two coronaro-fistula, the first (white arrow) communicating both, conus branch and RCA with the right atrium (coronaro-cameral fisrula) and the second arising from the conus artery with probable drainage located at the great cardiac vein and coronary sinus (red arrow)

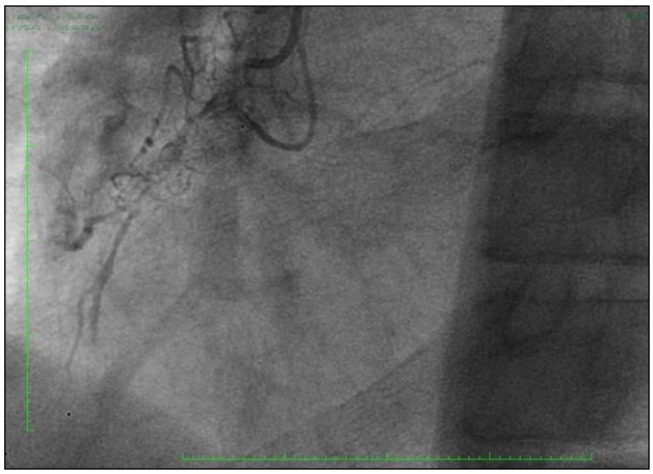

Fig 3: coronary angiograme (LAO) showed an occluded RCA and a coronaro cameral fistula between the RCA and the right atrium (white arraow)

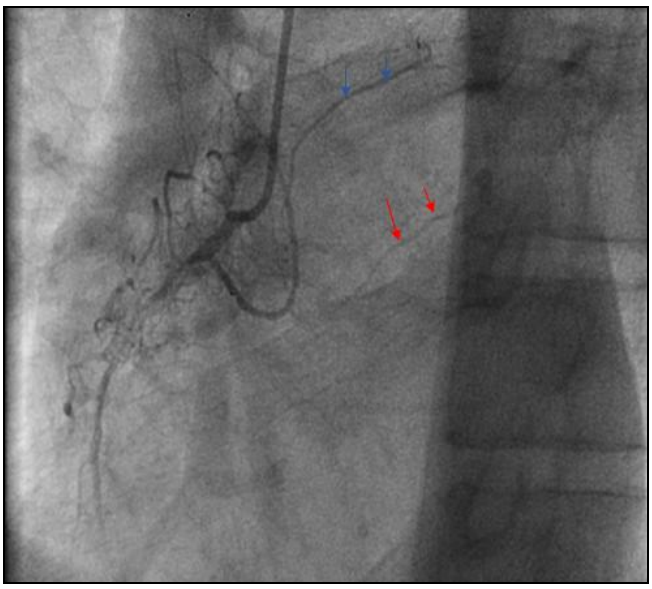

Fig 4: coronary angiograme (LAO) showing to two small arteriovenous fistula connecting the right coronary artery with the coronary sinus (red arrow) and the sinus branch of RCA with pulmonary trunk (blue arrow).

Conclusion

Severe atherosclerosis may occur development of coronary artery fistulas. Hypothesis of angiogenesis is most likely the mechanism. In the best of our knowledge this is the first cases of multiple CAFs due to severe atherosclerosis. Further studies are needed to investigate the precipitating factors and incidence of acquired CAFs to atherosclerosis.

Conflict of interest

None

Sources of funding

None

Ethical approval

N/a

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Mohamed Benasser: Study concept, Data collection, Data analysis, Writing the paper.
Mohamed Cherti: Supervision and data validation

Research registration
N/a

Guarantor
Mohamed Benasser

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