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Short communication

Bilateral multiple coronary artery fistulae with angina pectoris and syncope

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ABSTRACT

Coronary artery fistulae (CAF) are rare cardiac anomalies. They frequently arise from the right coronary artery (RCA) with fistulae originating from the left anterior descending artery (LAD) or from multiple arteries being less common. They do not usually cause symptoms and are incidentally diagnosed on routine cardiac imaging. We report a 70 years old male patient presenting with chest pain and syncope during physical activity. Diagnostic coronary angiography revealed bilateral multiple CAF originating from both the LAD and RCA. As high blood flow output was recognized in these large vascular anomalies contributing to 'steal phenomenon' surgical intervention was planned. This manuscript aimed to present the case and review the current literature for the management and treatment of these coronary anomalies.

Introduction

A 70 year old male patient was admitted to our hospital as having typical chest pain and a syncope episode during climbing up the stairs of his apartment. He had experienced typical chest pain for about last one month. Syncope occurred while the patient was climbing up the stairs of his apartment. Prodromal symptoms were chest pain, pallor and dizziness. His relatives saw him recovering immediately after fainting. As he continued to have chest pain and dizziness an ambulance was called and brought to our hospital. He had not experienced any palpitations, involuntary contraction of the body and no urine and stool incontinence. He had been treated for hypertension and hyperlipidemia for about ten years. Ramipril 5 mg/day and Atorvastatin 20 mg/day were his daily medication. At admission he looked pale, his blood pressure was 110/80 mmHg and electrocardiography showed a sinus rhythm with no ischemic signs and a heart rate of 117/min (Fig. 1).

Respiratory examination was unremarkable. During heart auscultation a continuous murmur of 3/6 grade was heard all over the cardiac area. No carotid murmur was heard on auscultation. Carotid and vertebral artery doppler ultrasound examinations revealed no stenosis or plaque. His neurological examination and cranial computed tomography imaging were in normal limits. Neurology consultation stated no further neurologic tests for syncope. Abdominal examination was normal. No anemia or infection was found in his blood and urine tests contributing to sinus tachycardia. According to typical angina symptoms and 2013 European Society of Cardiology 'Guidelines in Myocardial Revascularization' a diagnostic coronary angiography was performed. After a single intracoronary injection of 25 ml of radio opaque substance (Iopamidol), the distal portion of LAD was not visualized (Fig. 2). Contemporarily all of the large and multiple vascular anomalies originating from the proximal portion of LAD were dyed. This showed a high blood flow output through these vascular anomalies. During opaque injection into RCA multiple, thin lumened and tortuous vascular anomalies originating from the proximal portion of RCA were visualized (Fig. 3). Minimal plaque on RCA and LAD was detected. In order to find the drainage of these vascular anomalies a cardiac catheterization was performed. A step-up saturation was revealed in the right atrium and superior vena cava and Qp/Qs were calculated as 1.7:1. Their high blood flow output showed a 'steal phenomenon' and was considered as the cause of angina and syncope. Echocardiography showed a normal ejection fraction and a mild enlargement of the right ventricle with normal systolic pulmonary artery pressure and no valvular dysfunction. A 64 slice multidetector computed tomography (MDCT) showed multiple coronary artery fistulas originating both from LAD and RCA. Their erratic morphology and chaotic irregularity made their drainage location hard to be determined (Figs. 4–5). As a result we suggested the patient to have a surgical repair of these vascular anomalies causing ischemia and syncope. Although detailed explanation, patient refused surgical procedure and then an outpatient follow-up was decided. He was discharged with aspirine 100 mg/day, atorvastatin 20 mg/day, ramipril 5 mg/day, metoprolol 25 mg/day and was advised for prophylactic antibiotics for prevention of endocarditis.

Discussion

CAFs are rare congenital malformations that can be defined as direct vascular connections from a coronary artery to a cardiac chamber or

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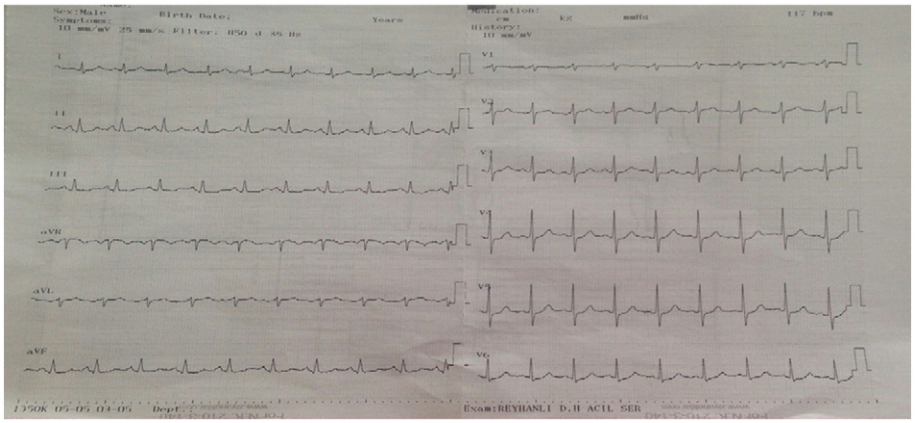

Fig. 1. Electrocardiography: Sinus rhythm with a heart rate of 117/min.

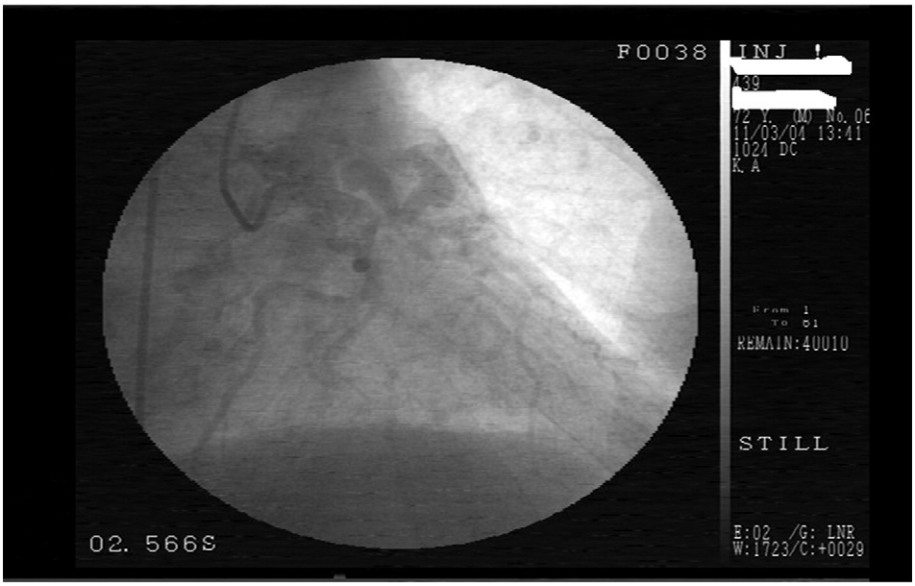

Fig. 2. Left anterior oblique view of coronary angiography. Mid and distal portion of LAD is not visualized due to high blood flow output of coronary artery fistula originating from the proximal portion of LAD.

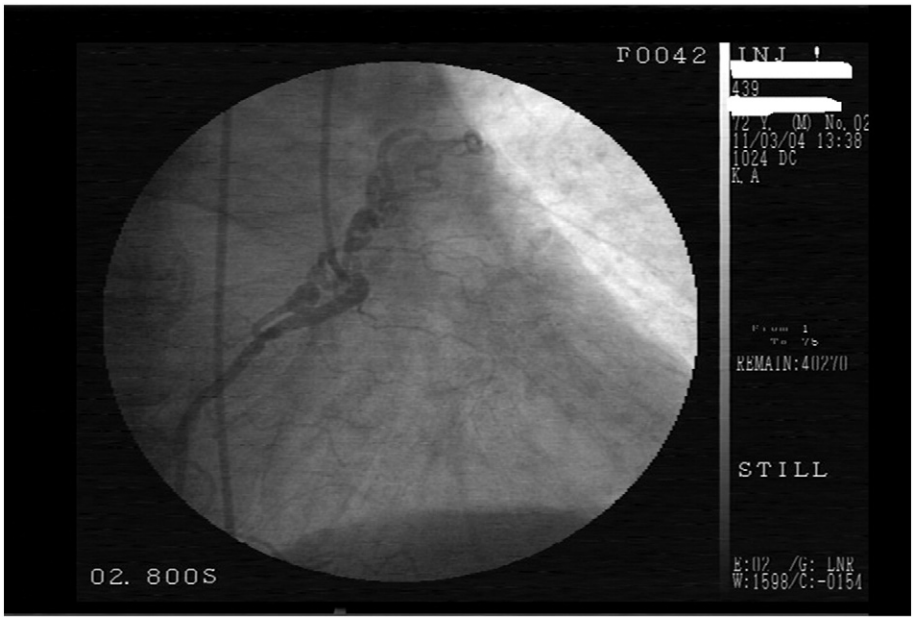

Fig. 3. Right anterior oblique view of coronary angiography. Multiple CAF originating from the proximal portion of RCA. Its irregularity and tortuosity are noticed.

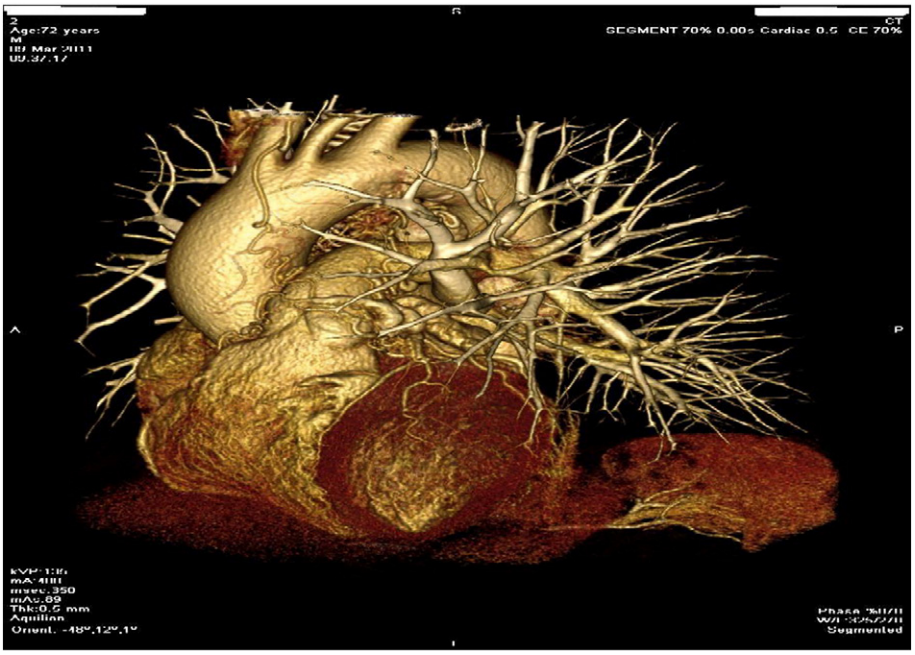

Fig. 4. 64 slices MDCT showing relevant arteriovenous fistula like structures originating from LAD. Their tortuosity and irregularity make uncertain determination of the drainage point.

major central blood vessel without an intervening capillary bed.[1] The first CAF was described by Krause in 1865.[2] The majority arises from RCA in 50%, from LAD in 42% and both in 5%.[3] Drainage may occur into the right ventricle (41%), right atrium (26%), pulmonary artery (17%), coronary sinus (7%), left atrium (5%), and left ventricle (3%) and rarely into the superior vena cava (1%).[3] According to cardiac catheterization we noticed a rare drainage point and especially for CAF originating from LAD. Its rareness being an important finding as Hassan Zamani et al. showed in their 18 year period retrospective study that the most common drainage of LAD originating CAF was the main pulmonary artery in 35%.[5] This abnormality can be diagnosed at any age mainly as an incidental finding on coronary angiography, echocardiography or MDCT. Several diagnostic studies revealed an incidence of 0.1%.[4] Most of them are single communications but rarely multiple fistulas have been reported, as in our case.[5]

CAF's are the most common coronary artery abnormalities causing hemodynamic compromise. Although CAF are usually congenital some of them are acquired due to multiple factors such as Takayasu arteritis, trauma, coronary atherosclerosis, endomyocardial biopsy etc. Syncope is a really challenging sign regarding the differentiation of cardiac and neurologic diseases. Coronary steel phenomenon should be recognized as an important mechanism for myocardial ischemia as well as for a rare presentation of syncope in this case.[6] Clinical importance of CAF is due to the risk of several complications including heart failure, infective endocarditis, myocardial ischemia and dysrhythmias like atrial tachycardia, ventricular tachycardia, and bundle branch block, first degree atrioventricular, block prompting immediate and appropriate treatment. Patients with hemodynamic compromise, signs like angina, heart failure and high blood flow shunt (Qp: Qs > 1, 5:1), should be addressed to intervention treatments.[7] Percutaneous coronary closure

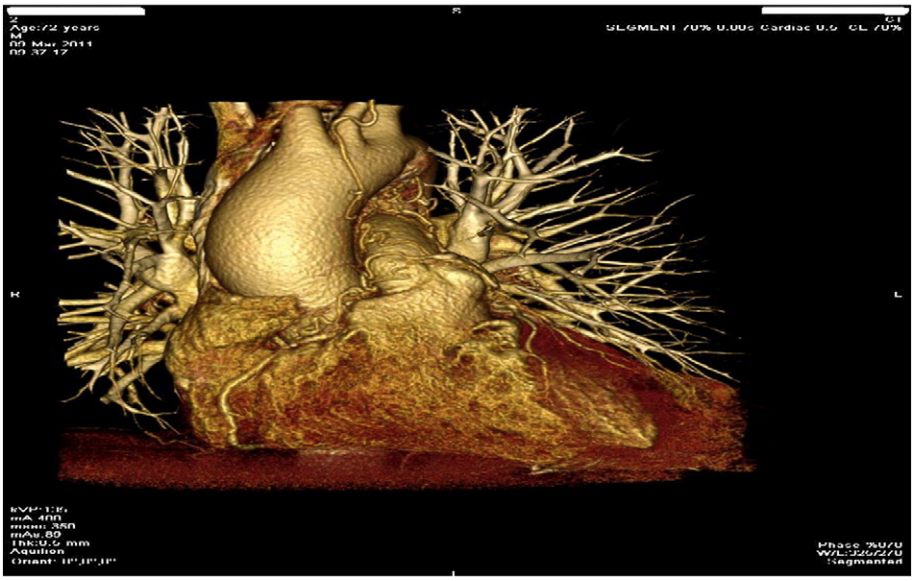

Fig. 5. 64 slice MDCT showing relevant arteriovenous fistula like structures originating from RCA. Their tortuosity and irregularity makes uncertain determination of the drainage point.

of CAF has been increasing in frequency as an effective and safe alternative to surgery, but only to anatomically appropriate and single CAF. It may be unsuitable in select cases of extreme vessel tortuosity, multiple drainage sites and coronary branches at the site of optimal device positioning.[8] Multiple and irregular CAF that are symptomatic, especially originating proximally from the coronary system, are usually corrected surgically by particularly exploring the origin and termination and appropriately ligating them.[9] Long term outlook of multiple fistulas is sketchy and long term outcomes of patient after fistula closure is woefully lacking. This lack of information warrants following patients with CAF whether closed or not indefinitely.

Conclusion

CAF's are the most common congenital malformations of the coronary artery system that can cause hemodynamic compromise. Close follow up of patients with CAF, closed or not should be carried out indefinitely regarding complications that can occur and lack of information of long term outcomes.

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