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Coronary Cameral Fistulae- An Unusual Cause of Acute Coronary Syndrome

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1. INTRODUCTION

Coronary-cameral fistulae are abnormal connections between a coronary artery and one of the four chambers of the heart. Sixteen per cent occur as multiple fistulae but only 5% involve both coronaries. We report a rare case of multiple coronary-cameral fistulae, involving both coronary arteries and both ventricles, that presented as an acute coronary syndrome and associated with concentric left ventricular hypertrophy. A review of the underlying pathophysiology and management of this rare condition ensues.

2. CASE REPORT

A 61-year-old gentleman presented with a one-week history of shortness of breath and central chest pain on exertion. His past medical history included chronic obstructive airway disease (COAD) and epilepsy. Examination revealed no cardiovascular abnormalities. The presenting electrocardiogram showed new deep T-wave inversions in leads V1-V6 and leads II, III, and AVF. Chest X-ray showed hyper-inflated lung fields. Cardiac markers revealed two mildly elevated Troponin I eight hours apart (0.49ng/ml, 0.59ng/ml). The patient was admitted to a monitor bed as the main diagnosis at this point was acute coronary syndrome. No arrhythmias were recorded on monitor. A decision was taken to proceed to coronary angiography. This was carried out via a right radial approach. All coronary arteries were free from atherosclerosis but as contrast was injected into the left coronary artery, there was diffuse shunting into the left ventricular cavity (Figure 1). This was most evident distally. The same phenomenon occurred when contrast was injected into the right coronary artery. Contrast was shunted mostly into the left ventricle; however a small proportion was also seen draining into the right ventricle. Left ventriculography showed normal systolic function but left ventricular hypertrophy was evident. A transthoracic echocardiogram was therefore carried out which confirmed concentric left ventricular hypertrophy. The presence of micro-fistulae was detected on echocardiography using colour flow Doppler (Figure 2). The patient was started on ACE inhibitors and remained pain free during

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admission. He was seen at outpatients department 3 months after being discharged from hospital. He remained asymptomatic.

3. DISCUSSION

Coronary artery fistulae (CAF) are abnormal communications between a coronary artery and any of the four chambers of the heart (coronary-cameral) or any of the great vessels (Manesh and Davis, 2008), bypassing the myocardial capillary bed. They were first described in 1865 by Krause and are thought to result from persistence of the embryological myocardial microcirculation.

CAF are a rare but recognized abnormality and are present in 0.002% of the population. They represent 0.2-0.4% of all cardiac malformations. The majority are congenital in origin however some may be acquired and are a result of trauma, infection or iatrogenic injury such as cardiac surgery and transcutaneous catheter techniques used for myocardial biopsy.

Fistulae originating from the right coronary artery are the commonest (50-60%). 25%-42% originate from the left anterior descending artery and18.3% from the left circumflex artery. A single fistula is most common (74%-90%). Multiple fistulae occur in 10.7%-16%. Both coronaries are involved in 5% of cases. Over 90% of fistulae drain into the right side of the heart (Chang et al., 2013; Lee et al., 2012). Our case is remarkable in view that the patient had multiple fistulae arising from both right and left coronary arteries and draining into both the left and right ventricles, but mostly into the left ventricle.

The majority of CAF are asymptomatic. In the rest, the most common symptoms are exertional dyspnoea, angina and fatigue. Congestive heart failure is common with a fistula to the coronary sinus. Some patients may be referred in view of a continuous murmur at the left lower sternal border. If the fistula connects to the left ventricle, an early diastolic murmur may be heard.

Complications from CAF include: excessive load to chambers resulting in congestive heart failure; coronary steal syndrome producing myocardial ischaemia due to diversion of blood; valvular regurgitation secondary to papillary muscle dysfunction; rarely infective endocarditis; thrombosis within the fistula and spontaneous rupture of aneursymal fistula causing haemopericardium have also been reported. In the case reported above, the main symptom was chest pain; however, the patient was symptom-free prior to the hospital admission. The chest pain could be explained by coronary steal syndrome caused by the fistuale.

Definitive diagnosis of CAF is made by coronary angiography. Transthoracic echocardiography (TTE) reveals coronary artery dilatation and chamber enlargement. In our case, TTE also showed concentric left ventricular hypertrophy. An association between congestive cardiac failure and hypertrophic cardiomyopathy has been reported in the literature. The exact mechanism of this association remains to be identified. The hypertrophy could be the result of chronic left ventricular volume overload through the coronary artery-ventricular shunt (Dresios et al., 2010).

Small asymptomatic fistulae do not require treatment. A small number of cases may undergo spontaneous closure. Indications for treatment include: large or increasing left-to-right shunt; left ventricular volume overload and/or dysfunction; myocardial ischaemia; congestive heart failure; and prevention of endocarditis. Current treatment options include surgical ligation or transcather closure (Oylumlu et al., 2014).

If fistulae are bilateral, it is recommended to manage as for single fistula. Intervention is difficult or impossible when fistulae are diffuse. Many times, despite ischaemia, intervention is not considered. Symptoms are usually relieved by medical therapy such as beta-blockers and calcium channel blockers (Arsian et al, 2009). In the above mentioned case, since the fistulae were diffuse, neither surgery nor trancatheter occlusion was a feasible treatment option. B-blockers were contra-indicated in this patient in view of brittle COAD. He was started on ACE inhibitor. He remained asymtomatic and so no further management was necessary.

4. CONCLUSION

Coronary-cameral fistulae are very rare causes of acute coronary syndrome. Because of the rarity of the condition, there is lack of evidence regarding the underlying pathophysiology. Left ventricular hypertrophy in association with congestive heart failure was noted in our case; thus a suggested mechanisms is left ventricular volume overload. There is also insufficient data with regards the best management strategy, especially for bilateral small microfistulae in view that available literature is only in the form of case-reports. Medical treatment is only suggested if patients are symptomatic; undoubtedly, further research is needed to unravel the best therapeutic modalities for CAF.

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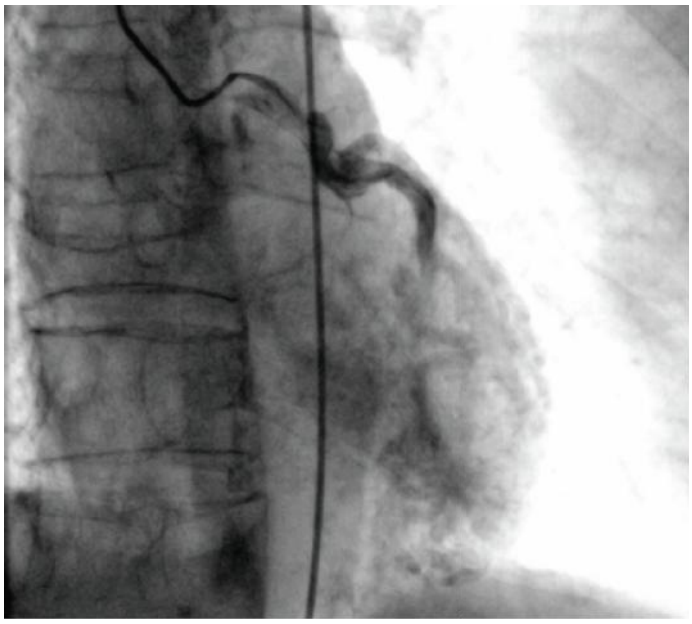

Figure 1

Shunting of contrast from the LCA into LV cavity through micro-fistulae

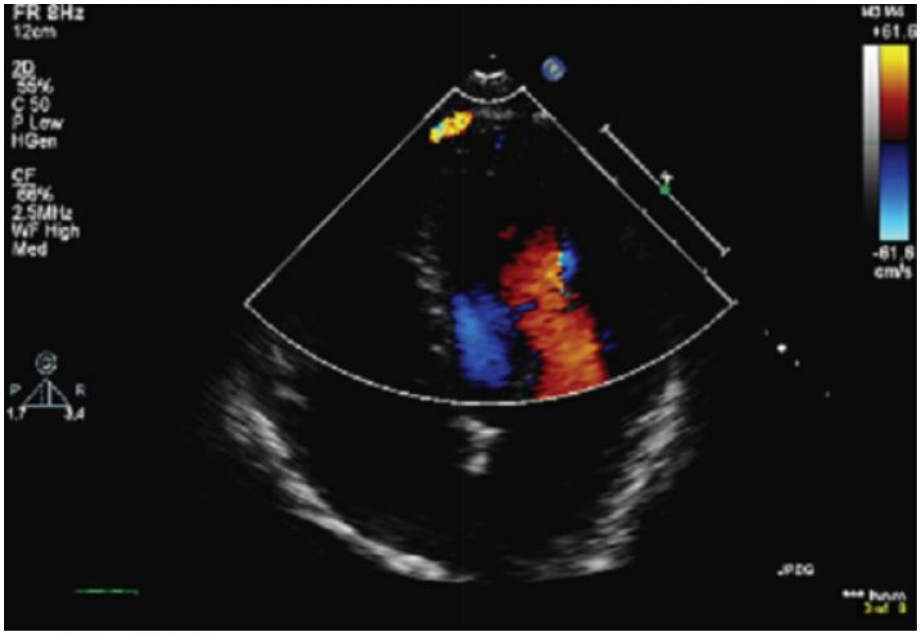

Figure 2

Colour flow doppler on trans-thoracic echocardiography demonstrating micro-fistulae

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