Multiple right coronary artery fistulas to coronary sinus: A case report and literature review

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Abstract

BACKGROUND: Coronary arteriovenous fistula is a rare congenital or acquired abnormal connection between a coronary artery and any of the great vessels or any of the heart chambers. Most of them are diagnosed during routine coronary angiography.

CASE REPORT: This case report illustrates a successful surgical ligating of multiple right coronary artery and circumflex artery fistulas to coronary sinus.

CONCLUSION: According to our experience and literature review, it can be concluded that to prevent potential complications in various cases of coronary arteriovenous fistula, early surgical management, just after their condition has been diagnosed, is the best choice.

Keywords: Coronary Arteriovenous Fistula, Right Coronary Artery, Dilated Coronary Sinus

Introduction

Coronary arteriovenous fistula (AVF) is an uncommon malformation.1,2 Most fistulas are single communications, but multiple fistulas have been identified too. It has variable presentations, such as asymptomatic, acute onset or chronic progression of symptoms.3 Most of the times patients do not have any significant signs and symptoms and are diagnosed during routine coronary angiography. The potential complications of coronary fistula include congestive heart failure, thrombosis, myocardial infarction and even sudden death.4 Spontaneous closure is rare.1

Symptomatic patients or those with severe shunts may be treated with surgical closure, although percutaneous closure with coil embolization may also be tried.3 This case report illustrates a successful surgical ligation of multiple right coronary artery and circumflex artery fistulas to coronary sinus.

Case Report

Our patient was a 42 year old man. He has been found to have a machinery murmur at the left lower sternal border during routine checkup by his family physician. The patient had experienced chest discomfort and dyspnea on exertion which was precipitated over the last two years. Laboratory tests were normal.

A mild right-heart border expansion was seen on chest x-ray. Electrocardiography was normal. Computed tomography angiographies showed dilated left circumflex and right coronary arteries and tortuosity in a whole course that was fistulized to the posterior and inferior aspects of coronary sinus respectively. Right coronary artery (dominant artery) was fistulized to inferior aspect of coronary sinus via posterior left ventricular (PLV) branch and coronary sinus, mid cardiac and greater cardiac veins were dilated. Left ventricular hypertrophy was noted and mild pericardial effusion was present. Left heart catheterization and selective coronary angiography showed large AVF from circumflex to coronary sinus and huge right coronary artery. The ejection fraction was 50%. Surgery was performed under cardiopulmonary bypass with separate bicaval cannulation. On direct observation, a large, dilated, tortuous right coronary artery on the surface of the heart was seen (Figures 1-3) which was draining to a dilated coronary sinus. Long time presence of multiple right coronary artery and circumflex artery fistulas to coronary sinus has led to severe dilatation of the coronary sinus (Figures 4 and 5).
After the right coronary artery was opened, coronary sinus was unroofed and had a severely stenotic ostium. Two fistulas communications were ligated. The postoperative course was uneventful. Right after the surgery, the continuous murmur at the left sternal border could not be heard anymore.

In follow-up visits 1 week and 1 month after the surgery, the patient's condition improved gradually so that in the second visit, he felt good and did not experience dyspnea on exertion anymore.

**Discussion**

Coronary arteriovenous fistulas were first described by Krause in 1865.5,6 These are rare congenital or acquired abnormal connections between a coronary artery and any of the great vessels or heart chambers that can bypass the myocardial circulation.7,8 Most of coronary AVFs are diagnosed at routine coronary angiography.4 The incidence of AVF in patients undergoing diagnostic cardiac catheterization has been reported to be 0.1%. Although both left and right coronary arteries are common origins, the right coronary artery involvement is more often (50%-55%).6 Actually the right coronary artery is the most common origin and right ventricle is the most common distal connection site. Drainage to the coronary sinus has been found in seven percent of surgical cases. Multiple coronary artery fistulas have been found in five percent of patients.7

The course of this malformation is usually benign but significant complications can occur.7 Clinical symptoms include angina, fatigue and dyspnea which are thought to be caused by a phenomenon known as coronary steal, where blood is drawn away from the distal coronary vasculature...
by the fistulous connection.\(^8\) Size of the fistula, degree of associated shunt and complications determine disease presentation. For example, the associated shunt can cause high output heart failure. Besides, arrhythmia, embolization, rupture, myocardial infarction, infective endocarditis, and sudden death are the other potential complications.\(^4,9\) Because of these potential complications, timely intervention is indicated.\(^8\) Direct ligation of the fistulous communication at the point of entry to the cardiac chamber is usually recommended. There is a 3.6% risk of postoperative myocardial infarction.\(^4\) In this patient, a rare kind of multiple coronary AVFs was reported with an unroofed severely dilated coronary sinus that made it difficult to find the site of coronary sinus orifice.

El Watidy et al. reported a similar case of coronary AVFs in a patient which also developed mitral and tricuspid regurgitation due to longstanding left to right shunt with a 7-year interval between the diagnosis and the corrective surgery.\(^4\) This time in our patient was around one year and there was no significant volume overload or mitral and tricuspid valves regurgitation. According to our experience and literature review, it can be concluded that to prevent potential complications in various cases of coronary AVFs, early surgical management, just after the condition has been diagnosed is the best choice.\(^4,7\)

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**Conflict of Interests**

Authors have no conflict of interests.

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