Case report - Congenital
Tortuous right coronary artery to coronary sinus fistula

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Abstract

We are reporting the successful surgical treatment of a 23-year-old female with a giant right coronary artery to coronary sinus fistula. This woman had complaints of chest pain and dyspnea on exertion for few months. Transthoracic echocardiography (TTE) showed a large tortuous right coronary artery and a dilated coronary sinus. Preoperative multi-detector computed tomography (MDCT) coronary angiography and cardiac catheterization confirmed the diagnosis of a right coronary artery to coronary sinus fistula. The patient underwent surgical closure of the fistula and division of the communication between the right coronary artery and the coronary sinus with the use of cardiopulmonary bypass. The patient was discharged home on postoperative day 5 and at one-year follow-up is symptom-free.

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1. Introduction

Coronary artery fistula (CAF) is the most common coronary arterial malformation but is generally a rare cardiac anomaly. Congenital fistulous connection of the coronary artery into a cardiac chamber or major vessel often causes a marked dilation of the donor coronary artery leading to aneurysm formation.

Conventional angiography is the golden standard of diagnosing this abnormality. The recent development of electrocardiographically gated multi-detector computed tomography (MDCT) coronary angiography has allowed accurate and non-invasive depiction of coronary artery diseases and malformation.

The most frequent symptoms and fistula-related complications are dyspnea on exertion, palpitations, congestive heart failure, myocardial infarction, infective endocarditis and death [1]. Surgical closure of the CAF is indicated to prevent progressive congestive heart failure, endocarditis, coronary aneurysm formation with rupture or embolization, and when left to right shunt (Qp/Qs) is >1:1.5 in asymptomatic patients.

We describe the case of a female who underwent successful surgical treatment of a severely enlarged tortuous right coronary artery (RCA) with a fistula draining into the coronary sinus (CS).

2. Case report

A 23-year-old female patient was admitted to our hospital with chest pain and dyspnea on exertion for the past 12 months. Her vital signs were unremarkable. Transthoracic echocardiography (TTE) showed dilation of the proximal and middle portion of the RCA as well as an abnormal vessel-like structure around the CS. The left ventricular ejection fraction (LVEF) was 60% with no valvular abnormalities. Cardiac catheterization showed a dilated tortuous RCA with communication to the CS (Fig. 1a). It was unclear if there were one or more points of communication. Further imaging with MDCT coronary angiography, showed a dilated RCA connecting to a giant CS (Fig. 1b,c).

In this case, indications for surgery included a symptomatic patient, large CAF characterized by the hemodynamically significant shunt, significant aneurysmal formation from RCA and a giant CS.

The surgical approach to the heart was achieved via a bilateral submammary skin incision and median sternotomy. The intraoperative findings were a dilated tortuous RCA with communication to the CS. Therefore, the heart was arrested and the distal RCA was dissected and opened longitudinally at its bifurcation and the communication points were identified from inside (Fig. 2a,b). This was double ligated and divided with 4/0 prolene proximal to the RCA and distally to the CS. Cross-clamp time and cardiopulmonary bypass time were 60 min and 70 min, respectively. The postoperative course was uneventful and the patient was discharged home in an excellent condition on postoperative day 5. At one-year follow-up she is asymptomatic, and the
0.2% in the adult population referred for cardiac catheterization [3]. CAF is classified as an abnormality in the termination of a coronary artery. It can communicate with any cardiac chamber, the RCA being the most frequent (55% of the cases) [4]. Fistulous drainage to the left heart occurs in 11% and to the CS only in a minority of cases (approx. 7%) [1]. Associated congenital cardiovascular abnormalities have been reported in the literature in 5–30% of cases [5]. CAF is defined as a direct precapillary connection between a branch of a coronary artery and the lumen of a cardiac chamber, the CS or superior vena cava, or a pulmonary artery or pulmonary vein close to the heart [6].

Clinical symptoms and age at manifestation of a congenital CAF depend on the underlying anatomy and on the size of the fistulous connection to the left or right side of the heart. Most patients with CAF are asymptomatic, however, some studies have emphasized that the incidence of symptoms and complications increases with age, particularly after the age of 20 [1, 7]. The longstanding left to right shunt can produce significant volume overload, with progressive dilatation of both tricuspid and mitral valve annuli, with consequent severe mitral valve regurgitation (MR) and tricuspid valve regurgitation (TR). Rarely sudden death has been attributed to these anomalies especially in the young.

**Fig. 1. Imaging findings:** (a) coronary angiography (tortuous right coronary artery). (b,c) Image of the multi-detector computed tomography coronary angiography.

**Fig. 2. Intraoperative view:** (a) tortuous right coronary artery (black arrow). (b) Open right coronary artery to coronary sinus fistula.

repeat echo showed a patent and normal size CS, with LVEF 60%. The aesthetic result was very gratifying.

**3. Discussion**

CAF was first described in 1865 by Krause [2]. It is a rare congenital anomaly with a reported incidence of 0.1% to
[8]. Rupture of coronary artery aneurysms in the pericardium with tamponade is a very rare but life-threatening complication of CAF. Early surgical correction is indicated because of the high prevalence of late symptoms and complications, especially when the shunt is significant (Qp/Qs) ratio ≥ 1.5.

A markedly enlarged coronary artery can usually be detected with TTE and transesophageal echocardiography (TOE). Magnetic resonance imaging (MRI) and CT cardiac coronary angiography are useful, non-invasive, and accurate imaging techniques for the detection of major coronary artery anomalies. Conventional coronary angiography has been the diagnostic modality to detect CAFs, but MDCT is now being widely applied for diagnosing cardiovascular anomalies and the number of incidentally detected CAFs on MDCT has been increasing.

Spontaneous closure of the fistula secondary to spontaneous thrombosis has been reported, although it is very uncommon (1–2% of cases) [9]. Current treatment options for CAF include surgical ligation alone (either with or without cardiopulmonary bypass), surgical ligation accompanied by coronary artery bypass surgery, and transcatheter closure. Techniques for transcatheter closure of CAF include; the use of various types of occlusion coils, vascular plugs, umbrella devices, covered stents and histoacryl resin. When surgery is contemplated it is of paramount importance to know the exact angiographic visualization of the anatomy of a CAF and the regular coronary vessels branching off proximally and distally of the fistula. In asymptomatic patients with high-flow shunting, closure is indicated to prevent occurrence of symptoms or complications, especially in the pediatric population. In most cases a median sternotomy and cardiopulmonary bypass are necessary to open the cardiac chamber in order to find and ligate the points of entry and to avoid postoperative hazards like myocardial ischemia and arrhythmias. Long-term follow-up is essential due to the possibility of postoperative recanalization, persistent dilatation of the coronary artery and ostium, thrombus formation, calcification, arrhythmias, and myocardial infarction. Surgery for CAF is a safe and effective procedure with a very low morbidity and mortality rate (0–4%) [6].

References


eComment: Surgical treatment of coronary arteriovenous fistulas

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In their well written paper, Mitropoulos and associates [1] report on the successful surgical management of a coronary arteriovenous fistula (CAF). We would like to share our experience related to two patients and to add a brief comment.

The first patient had a combination of double CAF associated with mitral and tricuspid regurgitation, PFO and chronic AF [2]. The patient successfully underwent suture ligation of CAF, combined mitral and tricuspid valve repair plus closure of PFO and modified Maze procedure with radiofrequency [2].

The second patient had multiple coronary-pulmonary fistulas with associated aortic valve regurgitation and ASD and successfully underwent AVR, repair of the ASD with autologous pericardial patch and obliteration of CAF and feeding vessels with surgical clips [3]. We found the additional manoeuvre of opening the pulmonary artery to be a helpful technique: this enabled direct identification of the entry points of all the fistulas into the pulmonary artery, and their direct closure with a patch of autologous pericardium sealed with Bioglue to avoid further recurrence of malformation [3].

CAVF is an uncommon entity and a major coronary anomaly according to Ogden’s classification, being present in 0.002% of the general population and representing 0.4% of all cardiac malformations [3, 4]. CAF of congenital origin can arise due to persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow in the early embryologic period [3].

Nomenclature is based on a descriptive analysis of the vessel origin and the chamber of termination. According to the angiographic classification by Sakakibara et al., there are types A and B: type A – proximal type, proximal coronary segment distal to the origin of the fistula, distal end normal; type B – distal type, coronary artery dilated over entire length, terminating as a fistula into the right side of the heart (end-artery type), proximal to fistula, regular branching of coronary [3, 4]. The surgical implications of this classification are: type A fistula may be treated by epicardial ligation of the fistula, maintaining normal branch flow or, alternatively, ligation of the coronary artery proximal and distal to the origin of fistula may be necessary. Type B fistula requires ligation by intracaval purse-string sutures at the site of termination with CPB [4].

Coronary steal phenomenon is the primary patho-physiological problem seen in CAF without outflow obstruction. There is general agreement that symptomatic patients should be treated. The surgical obliteration of the fistula is the cornerstone of surgical treatment, first described by Björk in 1947, and remains until now the most effective treatment [3].

Anatomy of CAVF and associated anomalies/pathologies determines which technique should be chosen. CABG may be performed if the coronary circulation has been compromised during the procedure and the entire procedure can be done on or off-CPB [3, 5]. CAF, combined mitral and tricuspid valve repair, or if the size of CAF is not too large.

In addition, intraoperative TOE greatly contributes to the facilitation of surgical treatment by identifying the origin, the draining points for the fistula, the quantification of valve regurgitation and finally the efficacy of CAF obliteration [5].

References


fistula to the coronary sinus is the direct run-off of cardioplegic solution to the right atrium; consequently the myocardium is not protected correctly. It is advisable to administer antegrade cardioplegia with digital compression of the fistula at the level of the coronary sinus [2, 3]. Retrograde cardioplegia represents a valuable alternative in the setting of a coronary artery fistula to the coronary sinus. Our technique would have been to administer antegrade cardioplegia with external compression of the coronary sinus then, after heart arrest, to perform a 2 cm longitudinal incision on the external aspect of the coronary sinus immediately prior to its distal extremity. This opening, which is obligatory to identify the entry point of the fistula, also allows us to introduce a retrograde cardioplegia cannula.

References


We read with great interest the article by Mitropoulos and colleagues [1] concerning the successful surgical treatment of a giant right coronary artery fistula to coronary sinus. Fistulas between coronary arteries and coronary sinus are unusual. Volume overload due to the fistulous connection may cause a marked dilation of the coronary artery, which ultimately leads to aneurysm formation.

However, we would like to comment on an important point regarding the surgical management in the abovementioned case report. The authors did not describe the technique used for myocardial protection. One of the main drawbacks of antegrade cardioplegia in the presence of a large fistula to the coronary sinus is the direct run-off of cardioplegic solution to the right atrium; consequently the myocardium is not protected correctly. It is advisable to administer antegrade cardioplegia with digital compression of the fistula at the level of the coronary sinus [2, 3]. Retrograde cardioplegia represents a valuable alternative in the setting of a coronary artery fistula to the coronary sinus. Our technique would have been to administer antegrade cardioplegia with external compression of the coronary sinus then, after heart arrest, to perform a 2 cm longitudinal incision on the external aspect of the coronary sinus immediately prior to its distal extremity. This opening, which is obligatory to identify the entry point of the fistula, also allows us to introduce a retrograde cardioplegia cannula.