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Original Research Article

Surgical for Coronary Artery Fistulas: Review of Eight Cases from Single Institution

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Abstract: Coronary artery fistula (CAF) is a rare congenital anomaly that can be complicated by endocarditis, myocardial infarction, or coronary aneurysms. Our study summarized the clinical characteristics and surgical outcome of eight patients with CAF at department of cardiothoracic surgery, Guilin Medical University Hospital. From 2009 to 2016, eight patients (age, 8 months to 54 years were diagnosed with CAF by echocardiography, Multidetector CT or surgery. Four were female and four were male. Seven patients with isolated CAF were asymptomatic. One patient with associated anomaly (coronary artery aneurysm, aortic valve prolapse with severe aortic regurgitation) had dyspnea on exertion and palpitation. Four fistulas originated from the right and the other four originated from the left coronary artery. Drainage was to the right ventricle (four), right atrium (three) and left ventricle (one). All patients had surgical ligation. In the symptomatic patient, in addition to excision of aneurysm, aortic valve replacement was performed. There was no operative or late death. Follow-up evaluation (range, 2-7 years; mean, 4.1 years) showed no evidence of recurrent or residual CAF. Surgical management of CAF is a safe and effective treatment resulting in 100% survival and closure rate.
Keywords: Coronary artery fistula, Surgery, Coronary vessels.

INTRODUCTION

The incidence of isolated coronary artery fistula (CAF) is reported as 1:50,000 live births [1, 2]. The clinical presentation of CAF includes a continuous heart murmur, fatigue, dyspnea, arrhythmias, angina and even sudden death. CAFs that have not been closed or detected in childhood have been reported to become symptomatic in adulthood because of chronic volume load and coronary artery ischemia [3].The symptomatic CAFs have been associated with substantial morbidity and mortality at all ages [4]. Although existing guidlines recommend elective closure of a symptomatic CAF, but treatment for asymptomatic patients is controversial [5]. This study was performed to determine the clinical characteristics and surgical outcome of patients with CAFs.

PATIENTS AND METHODS

Patients

The chart databases of the department of cardiothoracic surgery, Guilin Medical University Hospital, were reviewed for all patients with the diagnosis of CAF noted on echocardiography or Multidetector CT (MDCT) from 2009 to 2016. Patients with CAFs or sinusoids associated with important congenital cardiac malformation, such as pulmonary atresia with intact ventricular septum, or mitral stenosis and aortic atresia were excluded. Eight patients met the inclusion criteria. Patients' charts were reviewed for demographic, symptoms, clinical findings, indication for echocardiography, associated diagnosis and radiologic findings at presentation. When performed, surgical or cardiac catheterization reports were reviewed. Surgical repair was approached via a median sternotomy and cardiopulmonary bypass. At follow up, evidence of myocardial ischemia, congestive heart failure, and arrythmias was sought clinically and with electrocardiography.

Table-1: Clinical, and paraclinical findings and outcome data of 10 patients with CAF

Patient No.	Gender	Age (years)	Reason for echo	Cardiomegaly on chest X-Ray	Associated cardiac anomaly	Fistula origin	Exit site	Follow-up (years)
1	Male	3	physical examinations	NO	-	right coronary artery	right atrium	3
2	Female	10	physical examinations	NO	-	left circumflex	right ventricle	3
3	Male	41	physical examinations	YES	-	right coronary artery	right atrium	2
4	Male	54	Dyspnea and palpitation after exertion for 2 years	YES	coronary artery aneurysm+aortic valve prolapse	right coronary artery	left ventricle	7
5	Female	0.8	physical examinations	YES	-	left circumflex	right ventricle	4
6	Male	2	physical examinations	YES	patent foramen ovale	right coronary artery	right atrium	2
7	Female	19	physical examinations	YES	-	left coronary artery	right ventricle	6
8	Female	6	physical examinations	YES	-	left circumflex	right ventricle	6

RESULTS

Eight patients had an incidental finding of a CAF detected accidentally by echocardiography or cardiac CT. The clinical demographic, clinical, echocardiographic, and outcome data are summarized in Table 1. The mean age at diagnosis was 17 years (range, 8 months to 54 years). The primary indication for echocardiography at presentation was murmur in 8 patients. Dyspnea on exertion and palpitation in addition to murmur were noted in one old patient. This patient, in addition to CAF, had right coronary aneurysm and aortic valve prolapse. A systolic murmur was audible in 8 patients. No patient had symptoms suggestive of angina or congestive heart failure.

Associated cardiac abnormality was present in 2 patients (Table 1). Cardiomegaly was present radiographically in 6 patients. The origin of the CAF was clearly defined by color Doppler, MDCT, angiography or intracardiac surgery in 8 patients. The origin of the fistula was from the right coronary artery system in 4 patients, from the left coronary artery in 4 patients. The fistula drained into the right ventricle in 4 patients, right atrium in 3 patients, and left ventricle in 1 patient. Cardiac catheterization was performed in 1 patient. Seven cases had surgical ligation of CAF by midsternotomy approach and cardiopulmonary bypas except that one case needed aortic valve replacement and coronary artery aneurysm resection. All patients had follow- up from 2 to 7 years (mean, 4.1 years). None had evidence of recurrent or residual fistula.

DISSCUSION

CAF is a very rare anomaly in congenial heart diseases. It is one of the most common among the coronary artery anomalies. It was firstly described by Krause in 1865 [1, 6]. The first surgical treatment was also performed by Bjork and Crafoord in 1947 [7]. CAFs are associated with other congenital heart disease in 20% to 45% [8]. The left, right, or both coronary arteries may be involved. CAFs predominantly drain into the low-pressure structures of the heart (92%): into the right ventricle in 41%, the right atrium in 26%, the coronary sinus in 7%, the pulmonary artery in 17%, and the superior vena cava in 1% of cases. Drainage into both ventricles is uncommon [9]. The communications can occur in isolation or in association with other cardiac anomalies.

Isolated CAFs are usually asymptomatic [10]. The prenatal diagnosis of isolated CAF has been reported in few cases in the literature [11-13]. Some CAFs may disappear spontaneously during their childhood [6]. Some CAFs can present with angina or myocardial infarction due to the coronary steal phenomenon. CAFs are also one of the reasons of the non-atherosclerotic acute coronary syndrome in the adult 15]. A large left-to-right shunt can lead to right ventricular enlargement, congestive heart failure, pulmonary hypertension, associated aneurysm, even rupture of the fistula.

CAFs can be diagnosed as non-invasive by transthoracic echocardiography. Echocardiography was able to show the abnormal dilated coronary arteries and the drainage site of the fistula in our patient. But the reliability of echocardiography in CAFs diagnosis is limited. In some cases, the complex structure of neighboring the fistula, course of the fistula, and the site of drainage may not be accurately detected due to two-dimensional monitoring. In spite of its invasive nature, conventional coronary angiography (CAG) is a standard diagnostic method for CAFs. MDCT is also useful in non-invasive evaluation of the coronary anomalies. MDCT is a more accurate imaging method that can successfully provide details regarding the CAF's morphological features and relation with adjacent heart structures in two or three dimensions [16] MDCT is also a non-invasive tool with its capability of whole acquisition in single breath hold [17,18]. MDCT can not only provide accurate assessment of the size and location of CAF and aneurysm, but also show the course and drainage site of the fistula. It is also helpful in preoperative planning by showing relationships between the complex anatomy of the coronary artery fistula and great vessels [19]. MDCT has many advantages over echocardiography and CAG, because of its ability to demonstrate the fistula separate from surrounding cardiovascular structures along with any aneurysm or obstruction in its course [20]. We recommend that it is necessary to use several cardiovascular imaging modalities when investigating some complicated cases, including echocardiography, MDCT and invasive coronary angiography.

The surgery for CAFs, remains the most effective treatment until now [8]. Surgical correction is strongly recommended to prevent the development of congestive heart failure, angina, myocardial infarction, and pulmonary hypertension as well as rupture of coronary aneurysm. Treatment of adult asymptomatic patients with non-significant shunting to prevent fistula-related complications is still a matter of debate. Surgical intervention for these cases should be determined by considering the operative risk and the patient's wish.

In recent years, improved interventional devices have made catheter closure applicable to 80–90% of patients in some institutions with extensive experience [21]. Various devices have been used, including various types of occlusion coils [22]. Selection of the occlusion device should be based on the anatomic features of the fistula and the anatomical relationship between the coronary branch and the occlusion site of the CAFs [23]. The device complications include acute thrombosis and periprocedural myocardial infarction.

The therapeutic strategy, including the timing of surgical treatment, is not well defined, especially in asymptomatic patients [24]. Regarding surgical intervention, whether to leave or exclude the CAF, in addition to ligation of the fistula, remains an issue, considering the risk of later rupture when leaving the CAF and sacrifice of the native coronary circulation when excluding the aneurysm, which could be fatal from a myocardial infarction. When the fistula is hard to localize epicardially, cardiopulmonary bypass and endocardial approaches are necessary. Endocardial closure is achieved by suture closure or patch placement. When there is a single epicardial fistula on the lateral surface of the heart, closure may be accomplished via thoracotomy. The simplest method of closure is by epicardial interruption using ligatures or clips. Large tortuous fistulae may have fragile walls, especially in case of aneurysmal change. In such cases plication or excision may be required. Coronary artery bypass grafting is required when the native coronary artery is extensively malformed and likely to thrombose after closure. Hybrid approaches to locate drainage sites by placing a coronary wire through the fistula before surgery has been described [25]. Precise preoperative evaluation of the CAF morphology, and the possible presence of an aneurysm, is necessary when considering surgery indication and options. MDCT and coronary angiogram can have an additive value in preoperative evaluation of this cardiac anomaly [26].

Complications after surgical closure for CAFs are higher than after transcatheter closure because surgery are usually not amenable to transcatheter closure or are associated with comorbid cardiac conditions requiring surgery. Some complications include thrombosis or incomplete closure. If the fistula seen on echocardiography before the procedure, CAG may be used to confirm closure post procedure. We do not routinely perform CAG. It is important to exploration the mitral and aortic valves during fistula closure. When the ventricles are dilated, late valvular regurgitation occurs in a significant proportion of individuals and may require additional surgery after the CAF procedure.

CONCLUSIONS

Despite the limitations of small sample size and incomplete anatomic and functional evaluations of coronary arteries during follow-up, our study illustrates successful surgery for CAFs. In these patients, surgical for CAFs is a safe and an effective treatment modality. In conclusion, MDCT is a valuable non-invasive technique for diagnosing, monitoring and follow-up of the patients with CAFs.

CONFLICT OF INTERESTS

None declared.

AUTHOR'S CONTRIBUTION

Haiyong Wang and Xianzhu Liang wrote the paper. Tianci Qian, Fang Lei, Fugui Ruan, Jiangbin Sun, Jianfei Song and Zhenzong Du supervised the

composition of the paper. All authors read and approved the final paper.

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