Coronary Cameral Fistula and its Complications: A Case Report

Abstract

Coronary cameral fistulas (CCFs) are rare arteriovenous malformations that may be congenital or acquired. The presentation of CCFs varies from asymptomatic in early age to symptomatic and start of complications upon aging. Although percutaneous closure with embolization can also be done, surgical closure of CCFs is a gold standard of treatment. We present the case of a 20-year-old patient with a fistula connecting right coronary artery and the right atrium, along with aortic valve endocarditis and congestive cardiac failure.

Keywords: Congestive heart failure, coronary cameral fistula, endocarditis, right atrium, right coronary artery

Introduction

Coronary cameral fistulas (CCFs) are rare cardiac malformations, seen only in 0.002% of the population. Congenital fistulas are more common than acquired.[1,2] Coronary arteriovenous malformations connect one or more coronary arteries and a cardiac chamber or greatt vessel.[3] Hemodynamically significant fistulae are seen in 19% of patients who are less than 20 years of age and 63% in older age group.[4] We present the case of a 20-year-old male with coronary cameral fistula from RCA to right atrium (RA) with infective endocarditis as well.

Case Report

A 20-year-old male presented with fever since last 8 months. The fever was mild to moderate in severity, intermittent, and associated with headache. He had dyspnea on exertion which was insidious upon onset, progressive in nature, and coursed from NYHA grade II to III in last 8 months, for which he had multiple hospital admissions.

On examination, the general condition was moderate with a pulse rate of 106/min, regular, low in volume; blood pressure of 90/40 mmHg (right upper limb) upon dopamine infusion @ 6 mcg/kg/min. Cardiovascular examination revealed continuous murmur over precordium and diastolic murmur over aortic area. Rest of the physical examination was normal.

The patient was anemic and the blood culture was positive for fungal growth; however, other blood investigations were normal. ECG was suggestive of normal sinus rhythm with left ventricular hypertrophy (LVH).

Transthoracic echocardiography (TTE) revealed dilated left atrium and ventricle. Aortic valve had vegetations along with aortic root abscess and severe regurgitation. Patient also had moderate mitral regurgitation. There was a coronary cameral fistula from right coronary artery to right atrium. Left ventricle ejection fraction was estimated to be 45%.

Cardiac CT findings confirmed a coronary cameral fistula of the dilated sinonodal artery (20.9 × 17 mm) arising from RCA and draining into right atrium (RA) and cardiomegaly with dilatation of all four chambers. There were signs of pulmonary hypertension with a heart failure pattern. Multiple focal consolidations suggestive of secondary infection were seen in lung.

With the above-mentioned symptoms, positive blood culture, and imaging, the patient was diagnosed with coronary cameral fistula with infective endocarditis. He was started on antifungal agent amphotericin-B for three weeks. After the course of antibiotic and upon negative blood culture report, the patient was posted for aortic valve replacement (AVR) and coronary-cameral fistula (CCF) repair.

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The patient was preoxygenated followed by induction of anesthesia with Inj fentanyl 150 mcg and Inj Etomidate 6 mg titrated to loss of consciousness. Inj Rocuronium 50 mg was given as a muscle relaxant and endotracheal intubation was done. Patient was maintained with O2/Air/Sevoflurane.

Surgery inspection revealed dilated RA and RCA having a tortuous course over the right atrioventricular groove. Dilated sinoatrial nodal branch was also seen [Figure 1]. Intraoperative transesophageal echocardiographic (TEE) examination revealed that all four cardiac chambers were dilated. A dilated RCA was noted with 5.2 mm diameter at origin, with the tortuous course and fistula terminating into RA [Figure 2a-c]. Additionally, vegetations were noted on the aortic valve [Video 1] and the spectral Doppler suggested severe aortic regurgitation [Figure 3].

Surgical steps include aorto-bicaval cannulation, ostial as well as retrograde cold cardioplegia, and hypothermia (28–30 degree celsius) on CPB to achieve good myocardial protection. Fistula opening was closed by suturing the distal end towards RA. The aortic valve was replaced with a prosthetic valve. Post CPB, TEE showed normal functioning prosthetic aortic valve without any valvular and paravalvular leaks. The closed distal end of fistula was seen as echo-density in Figure 4. Proximal course was confirmed same as pre-operative. Total aortic cross clamp time was 90 min and the CPB time was 120 min. No significant postoperative complications were experienced.

**Discussion**

CAFs are usually of congenital origin. These may be acquired from trauma, infection, or iatrogenic injury.[1,5-7] Coronary fistulas with the cardiac chambers (Cameral fistulas) are rare congenital vascular anomalies reported in approximately 0.08% to 0.3% of unselected patients undergoing diagnostic coronary angiography.[8,9]

CAFs draining into the right heart structures are more common than left heart structures.

In our case, CAF originated from RCA draining into RA. Most CAFs would be small and asymptomatic due to the uncompromised myocardial blood flow. But small CAFs tend to grow large with age. If untreated, these fistulas can lead to clinical symptoms.

Diastolic runoff can occur in large size fistulae which may lead to coronary steal. There is a left-to-right shunt if the fistula drains to the systemic venous side of the circulation. Distal myocardial stealing of the site of connection is the pathophysiological mechanism responsible for the symptoms of coronary fistula.[10,11]

CAFs can give rise to increased left ventricular end-diastolic pressure, left ventricular hypertrophy, congestive heart failure, atrial fibrillation, ventricular tachyarrhythmias, chronic myocardial ischemia, and myocardial infarction. Valvular regurgitation secondary to papillary muscle dysfunction has been described in children and adults with CCFs. Hemopericardium can result as a result of the rupture of an associated aneurysm which is a life-threatening complication and hence it is important to control the arterial pressure during laryngoscopy. Pulmonary hypertension is also reported in some cases in which CCFs were subjected to increased pulmonary flow.[8] Our patient presented with infective endocarditis, pulmonary hypertension, and severe aortic regurgitation leading to congestive heart failure.
There is no consensus on the treatment of choice of symptomatic fistulae due to its rarity. Surgical repair, catheter closure, and medical management can be tried.\cite{12}

The anesthetic management of these patients involves prevention of coronary seal and perioperative myocardial ischemia, which can occur with an increased left to right shunt. Symptomatic patient and those presenting with complications need special attention and anesthesia administration to maintain hemodynamics. Intraoperative ECG monitoring is useful in detecting perioperative ischemia along with TEE that can detect intraoperative fresh regional wall motion abnormalities and guide surgical correction as well. The concern during cardiopulmonary bypass is to maintain adequate perfusion to the myocardium beyond the fistula.\cite{5}

**Conclusion**

CAFs are rare entity. These can be managed with standard anesthesia techniques, after ensuring the availability of advance monitoring techniques in the form of TEE and invasive pressure monitoring along with adequate ionotropic support. Medical management of aggravating factors such as sepsis or endocarditis should be dealt prior to a major surgery for optimal results. Elective ventilation is advisable as such patients have pulmonary hypertension and will be benefited by controlled ventilation in postoperative period as well.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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