

Coronary pulmonary artery fistula: A case series with review of the literature

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Abstract

Objective: To describe the clinical, angiographic profile and management strategies of patients of coronary pulmonary arterial fistulas presenting to a tertiary care center in a developing country.

Methods: All patients with coronary pulmonary artery fistula (CPAF) diagnosed using coronary angiogram in last two years i.e. 2011-2013 in a tertiary care center in South India were included in the study. Ten adult patients were treated for coronary pulmonary artery fistulas.

Results: Mean age was 56 ± 7.7 years (range 45-80 years) with no gender preponderance. Chest pain was the predominant symptom in 60 % of patients followed by giddiness and syncope. Only 20 % patients were found to have continuous or systolic murmur on auscultation. Majority of the fistulas were found to be originating from the left anterior descending artery (LAD), most commonly from proximal segment (n = 5). Majority (n = 9) responded to conservative management while one patient required surgical intervention.

Conclusion: Coronary pulmonary arterial fistulas are rare coronary anomaly which often goes unnoticed. CPAF was most frequently seen in middle age with male preponderance arising from proximal LAD. Patients present with diverse clinical presentations and subtle clinical findings. Majority of them being functionally insignificant, need only conservative measures.

Keywords: Coronary pulmonary artery fistula; Coronary angiogram; surgical closure.

1. Introduction

Coronary arteriovenous fistulas (CAVFs) are present in 0.002% of the general population [1-2]. Clinical presentations are variable depending on the type of fistula, shunt volume, site of the shunt, and presence of other cardiac conditions. Bilateral coronary fistulas between coronary arteries and the pulmonary artery are very rare. Over 90% of these fistulas drain into the systemic venous side of the circulation. Drainage of the fistula into the pulmonary trunk has been reported in 17% of cases.[3] We present our experience in seven adult patients with CPAFs, clinical profile, diagnostic evaluation and management strategies.

This report describes one of the largest hitherto published experiences in a tertiary care center in a developing country with most reports reported in recent times are mainly as anecdotal single case reports or as small series.

2. Methods

All patients with coronary pulmonary artery fistula (CPAF) diagnosed using coronary angiogram in last two years i.e. 2011-2013 in a tertiary care center in South India were included in the study. The medical records of all

patients were reviewed including clinical presentation, chest x-ray, electrocardiogram, treadmill, echocardiography and coronary angiograms were analyzed. Median follow up was 6 months. The data were entered in MS Excel. Descriptive statistics, i.e., means, standard deviations, frequencies, and percentages, were used to describe the study variables.

3. Results

Over a period of 2 years, 10 patients were found to have coronary pulmonary arterial fistula on coronary angiogram. Mean age of patients was 56 ± 7.7 yrs. In our study out of 10 patients, 5(50 %) were males and 5 (50 %) were females. 4 (40%) had hypertension, and 3 (30%) patients had diabetes. Chest pain was the predominant symptom in 60% of patients followed by easy fatigability, giddiness and syncope. Only 2 out of 10(20 %) patients were found to have continuous or systolic murmur on auscultation. 3 patients (30%) had ST-T changes on electrocardiogram and 4 (40%) were found to have evidence of provokable ischemia on non-invasive stress test. 8 out of 10 fistulas were found to be originating from the left anterior descending artery (LAD) with majority being from proximal segment (n = 5) and least

from distal segment (n = 1) (Table 1). Only one patient had fistula arising from the right coronary artery and one from left circumflex artery. All fistulas drained into the main pulmonary artery. Majority i.e. 7(70%) had normal coronary anatomy. 1 (10%) had associated coronary artery disease and

2 (20%) had myocardial bridging of left anterior descending artery (LAD). Majority i.e. 9 out of 10 (90%) responded to conservative management while one patient required surgical intervention.

Table 1: Origin, drainage site of the CPAFs and the treatment performed

Patient	Age	Sex	Origin	Drainage	Management
1	45	M	proximal LAD	PA	medical
2	80	M	mid LAD	PA	medical
3	47	M	proximal LAD	PA	medical
4	50	F	proximal LAD	PA	medical
5	51	M	distal LAD	PA	medical
6	47	M	proximal LAD	PA	medical
7	59	F	Left circumflex	PA	medical
8	62	F	RCA	PA	medical
9	63	F	mid LAD	PA	medical
10	56	F	proximal LAD	PA	surgical closure

M, male; F, female; LAD, left anterior descending coronary artery; RCA, right coronary artery; PA, pulmonary artery; SC, surgical closure



Figure 01: Coronary artery fistula

4. Discussion

Coronary artery fistulae (CAFs) are rare congenital malformations of coronary circulation. It is defined as an abnormal communication between an epicardial coronary artery and a cardiac chamber, any major vessel or with other vascular structure like coronary sinus or mediastinal vessels.^[1-3] This anomalous connection was first described in 1865 by Krause [4], but ever since has been reported more frequently with advancement in our imaging modalities and angiographic procedures. It has been classified as a major coronary anomaly under Ogden’s classification. [5] 0.002% of our general population is estimated to have this anomaly. Most of the fistulas are small and found incidentally during coronary angiography.

CAF are usually congenital or sometimes acquired following trauma, infection, iatrogenic or other injury. They are frequently associated with other congenital cardiac defects like atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, ventricular septal defect, and pulmonary atresia.[6-9] Edwards classification of primary and secondary fistulas define primary fistula as the main

pathological lesion and secondary fistula occurs as a consequence of other cardiac malformations such as aortic or pulmonary atresia. The secondary fistula are rare and we describe all cases of primary CAF in our case series.[10] However, in our series, we didn’t find any associated congenital cardiac defect.

Coronary pulmonary artery fistula (CPAF) constitute 15-30% of all CAFs. Multiple fistulas occur in 10.7-16% of all CAFs. Both coronary arteries are involved in about 5% of cases [1-3]. However none of the case in our series had both coronary artery involvement. Levin et al in their review of 363 cases concluded that coronary fistulas have slight right side predominance compared to left. Bilateral fistulas are unique in their tendency to terminate in the pulmonary artery and 17% of the unilateral fistulas drained into the pulmonary artery.[6]

In contrast with the majority of the literature, most CPAFs in our study group originated from the left coronary artery draining into the main pulmonary artery. This might be because our small population study group is only consisted of relatively older age individuals as shown previously by

Urrutia et al that drainage into the main pulmonary artery are a relatively more common with advancing age.[9]

Symptoms and sign of CAF are variable and may depend on size and site of drainage. It may be asymptomatic in many patients and may be detected incidentally. Some asymptomatic patients may come to clinical attention due to continuous or systolic murmur along the upper left parasternal border in cases of CPAF. A continuous murmur if present is highly suggestive. Differential diagnosis includes persistent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect and systemic arteriovenous fistula.

Chest pain, angina, dyspnea, fatigue, myocardial infarction (MI) may be the presenting symptom in patients with CPAF. 3-7% of patients may have angina or MI due to coronary steal phenomenon. In our series we have 60% patients who presented with angina. Fistula draining into right side of heart typically produces symptoms of pulmonary congestion while those draining to left can cause left ventricular overloading. Congestive heart failure (CHF) and endocarditis have been reported. Symptoms and risk of these lethal complications increase with age. In rare cases, CPAF may present with spontaneous rupture leading to hemopericardium and cardiac tamponade. [11]

Electrocardiogram and chest xray does not yield much information to aid in diagnosis and may show non specific changes. Angiography remains the gold standard for diagnosing CPAF which can delineate the origin, course, termination of vessels as well as associated anomalies; however use of non-invasive procedures is increasingly becoming popular. Transthoracic echocardiogram, transesophageal echocardiogram as well as MRI may be useful aid in diagnosing and follow-up of CPAF. Multidetector computed tomography is an emerging non-invasive tool which provides good anatomical correlation with coronary angiogram.[12]

Spontaneous closure of CAF is very rare but has been reported sporadically. The management in most cases is guided by the hemodynamic consequences of the fistula and symptoms. Surgery is indicated in symptomatic patients with CHF or MI and in asymptomatic patients with significant shunts. Management of asymptomatic patient with insignificant shunts is a matter of debate; however most favor a conservative approach in trivial shunts without any symptoms. [13-16]

Concomitant cardiac diseases are important in the planning the management approach for CPAF. In patients requiring surgical closure of the concomitant cardiac disease, the closure of the CAVFs should be performed to reduce postoperative early and late complications.

Surgery with ligation of vessels traditionally remains the method of choice for closure of CAFs and remains safe and effective with good reported success. Detachable coil and

balloons and many newer devices are in line for therapeutic management of CAF.

Transcatheter closures of CAF ever since its first use in 1980s have been increasingly used as a therapeutic modality for closure of CAF. In cases with favorable anatomy (e.g. non tortuous vessel, easily accessible distal part and the fistula should be unique with distal narrowing to avoid embolism to the drainage site), it has become preferred modality. [17,18] Results from surgery and transcatheter closure studies have found both approaches to have similar efficacy, morbidity and mortality. [19]

5. Conclusion

Coronary pulmonary arterial fistulas although an uncommon congenital anomaly often goes unnoticed, untreated and under reported. CPAF was most frequently seen in middle age with male preponderance. Patients present with diverse clinical presentations, subtle clinical findings and low prevalence of coexisting cardiac diseases in our series. Majority of them being functionally insignificant, need only conservative measures. Coronary angiogram remains the gold standard diagnostic tool. Surgery is recommended for significant symptomatic fistulas.

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