Case Report

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Anatomy Section

Congenital Bilateral Coronary Arteriovenous Fistula- A Case Report

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ABSTRACT

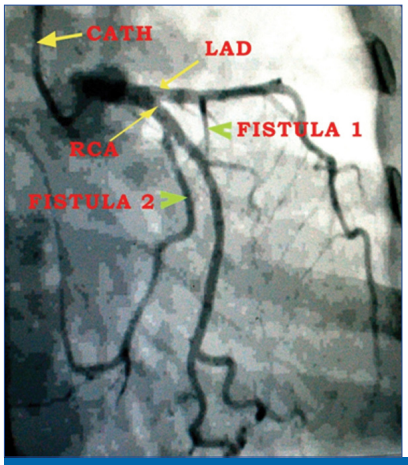
Coronary Artery Fistula (CAF) is an abnormal communication between the coronary artery and one of the chambers of the heart or any vessel close to the heart. We hereby, describe the possible anomalous connection between coronary arteries and the right atrium and coronary sinus of the heart in a 52 years old male which has been found through coronary angiogram. It is a very rare anomaly as the fistulous communication drains both into the right atrial chamber and also into the coronary sinus and as a result Coronary Arteriovenous Fistula (CAVF) was established. The origin of fistula is usually unilateral involving only one coronary artery either right or left. It is a very rare anomaly as the fistula is bilateral involving both the right and left coronary systems. The anatomy and morphological features of the fistula is very important in deciding the treatment method of CAF. It has been studied that physiology of CAF mimics various heart lesions. In this case where this fistulae that drains into right atrium and coronary sinus (systemic vein) mimics the physiology of an atrial septal defect.

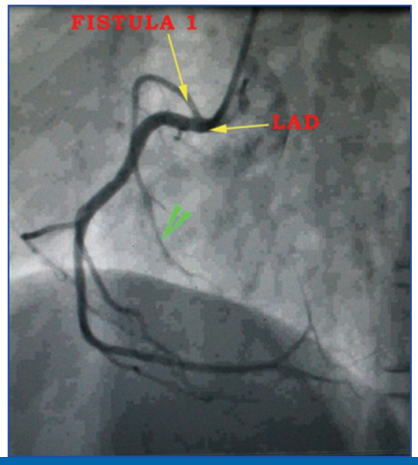
Keywords: Congenital cardiac anomaly, Congestive cardiac failure, Coronary cameral fistula

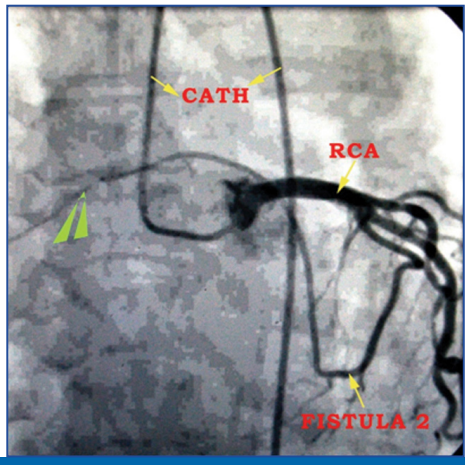
CASE REPORT

A 52-year-old male has been admitted to the hospital with complaints of exertional dyspnoea, fatigue and palpitation for two weeks. It was not associated with sweating, giddiness, nausea, vomiting or pedal oedema. Patient was hypertensive and on medication for past eight years. No significant family, personal history or other relevant medical history was reported by the patients. On examination, he was conscious and oriented. No pallor, icterus, cyanosis, clubbing and lymphadenopathy was found. Vital signs were normal.

Routine CVS examination was done. On inspection and palpation no abnormality was found. On percussion, borders of heart felt within normal limits. On auscultation no murmurs were heard.







[Table/Fig-1]: Angiogram showing the presence of bilateral CAVF. **[Table/Fig-2]:** Angiogram showing the closer view of fistula 1. FISTULA 1: Communication between Left Anterior Descending artery (LAD) and right atrial chamber (green arrow head) indicates the fistula opening into the right atrial chamber. **[Table/Fig-3]:** Angiogram showing the closer view of fisutla 2. FISTULA 2: Communication between Right Coronary artery (RCA) and Coronary Sinus CATH – Catheter. (Green arrow head indicates the opening of Fistula 2 to coronay sinus).

The patient was subjected to cardiac investigation. ECG was normal but chest X-ray showed mild cardiomegaly. Coronary angiogram revealed that both right and left coronary arteries arose from the aorta. There was a fistulous branch of left anterior descending artery which communicates directly with the right atrial chamber [Table/Fig-1-3].

There is a fistulous branch of right coronary artery which directly ends in the coronary sinus [Table/Fig-2].

The caliber of these fistulae was found to be smaller than the normal caliber of the coronary arteries, indicating that the fistula is less significant.

From all the observations, the final diagnosis was made to be congenital arteriovenous fistula. The patient is then sent to cardiac centre for further management which is likely to be surgical intervention.

DISCUSSION

CAVF is a very rare anomaly in the age group above 20 years with the prevalence of 0.002% in general population and is more frequently visualised in 0.25% of patients undergoing catheterisation and its symptoms seem to increase with age (1-5). CAVF was first described by Krause in 1865 and the first surgical treatment was done by Bjork and Crafoord in 1947 in a patient with pre operative diagnosis of patent ductus arteriosus [1].

CAF causes severe complications, it is mandatory to treat every CAVF whether it is symptomatic or asymptomatic [1]. The symptom in CAVF patient depends upon the size and also the site of drainage of the fistula. In the present case both the fistulae were draining into the venous structures of circulation (right atrium and coronary sinus) which correlates with Ata Y et al., who reported that mostly the fistula drain into venous structures of circulation which include right sided chambers, pulmonary artery, coronary sinus and superior vena cava rather than the left sided chambers [1].

CAF are classified based on the abnormalities of termination of coronary arteries [2]. Accordingly, CAF may be a coronary-cameral fistula or a CAVF depending on its termination into a chamber of the heart or into any segment of systemic and pulmonary circulation respectively [3]. In the present study we observed both the types of fistula in a single patient. Fistula 1 where there is a sizeable communication between the left anterior descending artery which bypasses the myocardial capillary bed and enters into the right atrium of the heart is case of a coronary-cameral fistula. Fistula 2 where there is a communication between the right coronary artery and the coronary sinus (systemic circulation) is a case of a CAVF. The simple presence of these fistulae though smaller in size may lead to severe complications like congestive cardiac failure due to volume overload [Table/Fig-2,3].

It can be inferred from the previous studies [1,4-6] that when the CAF is very small, there will be no symptoms and it remains undetectable. It will be diagnosed as an accidental finding while performing an echocardiography or coronary arteriography for an unrelated cause or during an autopsy. Usually, these small fistulas do not cause any complications and may sometimes resolve spontaneously. On the other hand larger fistula either symptomatic or asymptomatic should be treated in order to avoid fistula related complications which will increase with age because of the high risk of endocarditis, myocardial infarction and congestive cardiac failure. Larger fistula enlarges progressively with time and spontaneous closures are very rare.

Types of Coronary Artery Fistula

Types of CAF have been postulated based on their onset and angiographic study.

CAF may be congenital or acquired. In congenital fistulae, drainage is most often to a low pressure cardiac chamber; the Right Ventricle (RV), Right Atrium (RA), or the Pulmonary Arteries (PA) and less frequently to the superior vena cava, coronary sinus, and pulmonary veins [7].

In this present case, as there is no previous history of trauma or cardiac surgery, it may be considered as congenital CAF. The present case is a rare anomaly of CAF as it is an isolated finding with no associated heart disease. It is also noted that in Fistula 2 where it drains into coronary sinus is a very rare feature for congenital CAF.

The coronary artery branches become significantly enlarged proximal to the shunt site with increased flow of blood. Another classiification of CAF is of Sakakibara by angiography which categorises into two-Type A, proximal coronary segment dilated to the origin of the fistula with distal end normal and Type B, coronary dilated over entire length, terminating as a fistula in the right side of the heart (end-artery type) [7]. In this present case, as the entire length of coronary arteries are dilated it is considered as Type B CAF.

Anatomical and Physiological basis of Coronary Artery Fistula

The origin of a fistula may be unilateral or bilateral involving both left and right coronary artery systems. Fistulous opening into a chamber or the drainage is mostly single or rarely, double if both coronary artery systems are involved [2]. The present case is unique because it involves both the coronary artery systems (double fistula). The origin of Fistula 1 is from left anterior descending artery which is reported in 35% and termination is into the right atrial chamber which is found in 90% cases. The Fistula 2 showed a rare entity whose origin is from the right coronary artery (60%) and termination is in to the coronary sinus (3%) [8].The physiology of the clinical picture depends on the

resistance of the fistulous connection and on the site of fistula termination [7]. In this present case both the fistulae drains into the venous side of circulation, a left-to-right shunt occurs and it mimics the physiology of atrial septal defect.

CAF is an anomaly of coronary arteries which causes hemodynamic compromise. Usually, small fistulas have no role in it but large fistulae by means of coronary artery steal phenomenon causes ischemia of the segment of myocardium perfused by the coronary artery. The pathophysiologic mechanism of CAF is myocardial stealing or reduction in myocardial blood flow distal to the site of the CAF connection [2].

Embryological basis of Coronary Artery Fistula

In the primitive coronary circulation, the endothelial lined spaces or intertrabecular sinusoids between the embryonal heart muscle columns and of endothelial outgrowths towards the epicardial surfaces, freely communicate with the newly formed epicardial vessels and together form the new sinusoidal circulation [9]. Intracardiac coronary artery may terminate into the capillary plexus, into sinusoids of the myocardium or directly into a ventricle. As in the normal case, the outermost intertrabecular spaces shrink and fuse with the coronary arteries to form the capillary network and these intertrabecular vessels preserve their communications with the ventricle to form the thebesian veins of the adult heart. When the intertrabecular spaces connecting coronary arteries, veins and cardiac chambers do not close up, it give rise to persistent sinusoidal trabeculae, which may further develop into a CAF.

Another elucidation for CAF may be faulty development of the distal branches of the coronary artery ramified vascular network [2].

CONCLUSION

The etiology of congenital CAF remains obscure, although many theories have been postulated. As severe complications arise because of the CAF, it is mandatory that each CAF should be surgically corrected. In this present case, although CAF is isolated and there are no associated cardiac disorders, these fistulae should be surgically corrected as the CAF is bilateral and the patient presented with symptoms of volume overload which indicates that will lead to congestive cardiac failure sooner or later. This is a primary indication for surgical correction of CAF. The possible treatment modalities recommended may be invasive surgical closure or transcatheter closure of the fistulae.

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