A Rare case of Coronary A-V Fistula Draining into SVC

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Abstract:
A Congenital coronary arteriovenous fistula is a direct communication between a coronary artery and the lumen of any of the cardiac chambers, the coronary sinus, the pulmonary artery, or the superior vena cava. The number, origin, and course of the coronary arteries are otherwise normal. Coronary arteriovenous fistula is a rare congenital anomaly that is seen in 0.1 to 0.2 of coronary angiograms. They are classified into 3 categories abnormalities of origin, distribution, and termination. Coronary artery fistulae are considered to be termination abnormalities. They are uncommon and are seen in only 0.1 to 0.2 of coronary angiograms. In the majority of reported cases, coronary fistulae were found to originate from the right coronary artery (52 of cases) and to drain into the right ventricle (40 of cases). Fistulae originated from the circumflex artery in 18 of cases and drained into the pulmonary artery in 17 of cases. In the majority of cases (over 90 of patients), the coronary fistula results from an abnormal coronary artery system with aberrant termination (coronary artery fistula or coronary-cameral fistula) rather than arteriovenous fistula. In more than 90 of reported cases, the fistula terminates in the right side of the heart (approximately 40 in the RV, 30 in the RA, and 20 in the PA). It rarely terminates in the left side of the heart, but when it does, the majority enter the left atrium (LA). We present a rare case of coronary A-V fistula drains into SVC. Our patient is 42 year old not a known heart disease presented with class II dyspnea and palpitation. Clinical examination revealed continuous murmur over right sternal border. Echocardiogram showed turbulence present in LCx origin and its course. Coronary angiogram and CT angiogram showed hugely dilated tortuous left circumflex artery draining into posterior aspect of superior vena cava.
INTRODUCTION:

A coronary arterial fistula is a connection between one or more of the coronary arteries and a cardiac chamber or great vessel. This is a rare defect and usually occurs in isolation. Its exact incidence is unknown. The majority of these fistulas are congenital in origin although they may occasionally be detected after cardiac surgery. They do not usually cause symptoms or complications in the first two decades, especially when small. After this age, the frequency of both symptoms and complications increases.

CT angiogram showing Coronary A-V fistula arises from Left Circumflex Artery and draining into Superior Vena Cava
CT angiogram of Coronary A-V fistula arises from Left Circumflex Artery and draining into Superior Vena Cava

**CASE REPORT:**

42 year old female presented to our hospital with NYHA class II dyspnea for one year and palpitation on and off for six months duration. No history of chest pain or syncope. She is not on treatment for any heart ailments previously. She is not a known diabetic. She had given birth to two children with uneventful antenatal and natal history. Clinical examination of cardiovascular system showed apical impulse felt in left 6th intercostal space hyperdynamic in nature. There was Grade 3 continuous murmur present in 3 & 4 Right intercostal space close to sternum. All other system examinations were normal. We proceeded with investigations. Standard 12 lead electrocardiogram showed rhythm of atrial fibrillation (Fig 1). Color Doppler Echocardiogram revealed dilated left circumflex artery with high volume flow and turbulence at its origin and along its length (Fig 2). Doppler showed continuous flow signal (Fig 3). Conventional coronary angiogram was performed to identify the course and drainage site. Power injection of dye in aortic root showed Left circumflex artery is hugely dilated and tortuous appears draining into right atrium and superior vena cava junction (Fig 4). LAD and RCA were normal in course and size (Fig 5).

**DISCUSSION:**

The CAFs were first described by Josef Hyrtl in 1851 (Friedman et al., 2007). The incidence of the CAFs is changes according to genetic or ethnic racial factors or either to different geographical regions. The true incidence is difficult to evaluate because about most of the cases may be asymptomatic and clinically indetetable until an echocardiogram or catheterization is performed. The incidence of CAFs is 0.3–0.8% in patient’s undergone diagnostic cardiac catheterizations (Angelini, 1999; Cebi et al., 2008). Echocardiographic studies estimated the incidence of congenital CAFs in children at 0.06 to 0.2% (Sherwood et al., 1999; Hsieh et al., 2002). Sex predilection is controversial. In a study it is found out that males are more affected than females, with a ratio of 2.3 to 1 (Ata et al., 2009). Another study shows that there is no sex predilection for CAF (Chiu et al., 2008). A Congenital coronary arteriovenous fistula is a direct communication between a coronary artery and the lumen of any of the cardiac chambers, the coronary sinus,
Coronary arteriovenous fistula is a rare congenital anomaly that is seen in 0.1% to 0.2% of coronary angiograms. They are classified into 3 categories: abnormalities of origin, distribution, and termination. Coronary artery fistulæ are considered to be termination abnormalities. They are uncommon and are seen in only 0.1% to 0.2% of coronary angiograms. This is a rare abnormality and usually occurs in isolation [1]. Its exact incidence is unknown. The majority of the fistulas have a congenital origin, but may occasionally be detected after cardiac surgery, such as valve replacement, coronary artery bypass grafting and after repeated myocardial biopsies in cardiac transplantation. The feeding artery of the fistula may drain from a main coronary artery or one of its branches and is usually a dilated and tortuous artery terminating in one of the cardiac chambers or a vessel. The more proximal the feeding artery originates from the main coronary artery, the more dilated it tends to be. If the fistula drains to the right atrium with a proximally arising feeding artery, it tends to be considerably dilated but less tortuous. If there is a more distal origin of the feeding artery, and in particular when the fistulas originate from the left coronary artery and drain to the left ventricle, they may be very tortuous, presenting a challenge for catheter closure. However, in the less frequently encountered right coronary artery to coronary sinus drainage, the fistula vessel may be large and very tortuous. It is important to note that there may be multiple feeding arteries to a single coronary arterial fistula drainage point or there may be multiple drainage sites [2]. The fistulas originate from the right coronary artery in about 52% of cases, the left anterior descending coronary artery being the next most frequently involved in approximately 30% of cases and the circumflex coronary artery in about 18% of cases [3]. Over 90% of the fistulas from either coronary artery drain to the right side of the heart and the remainder drain to the left side of the heart [4]. In the right heart, drainage occurs most frequently to the right ventricle (in about 40%), followed by the right atrium, coronary sinus, and pulmonary trunk rarely into SVC. Multiple fistulas between the three major coronary arteries and the left ventricle have also been reported [5]. In adults, occasionally fistulas may be encountered which originate from both the coronary arteries and drain into the pulmonary trunk. These fistulas may cause angina and require closure. When the fistula drains to the right side of the heart, the volume load is increased to the right heart as well as to the pulmonary vascular bed, the left atrium and the left ventricle. When the fistula drains into the left atrium or the left ventricle, there is volume overloading of these chambers but no increase in the pulmonary blood flow. This results in different echocardiographic appearances of dilation of different cardiac chambers due to the shunts. The size of the shunt is determined by the size of the fistula and the pressure difference between the coronary artery and the chamber into which the fistula drains. Occasionally congestive cardiac failure occurs and very rarely, in adults, myocardial ischaemia may occur. Coronary arterial fistulas are usually asymptomatic in the first two decades, especially when they are haemodynamically small. Indeed, a small number may close spontaneously. After this, the frequency of both symptoms and complications increases [6].
Complications include 'steal' from the adjacent myocardium causing myocardial ischaemia, thrombosis and embolism, cardiac failure, atrial fibrillation, rupture, endocarditis/endarteritis and arrhythmias [7,8]. Thrombosis within the fistula is rare but may cause acute myocardial infarction, and atrial and ventricular arrhythmias [9]. Spontaneous rupture of the aneurysmal fistula causing haemopericardium has also been reported [10]. The largest shunts occur when the coronary artery connects to the right side of the heart rather than the left heart chambers. The treatment options for coronary arterial fistulas include surgery or catheter closure. Surgery involves internal closure of the fistula within the receiving chamber or vessel whenever feasible, but when the fistula is associated with a large aneurysm of the feeding artery, it may need to be ligated from within the aneurysm. Surgery is not risk free: it is associated with a low morbidity and mortality rate ranging from 0 to 6%. Catheter closure of the fistulas is now considered to be an effective and safe alternative to surgery. The aim of catheter closure is to occlude the fistula artery as distally and as close to its termination point as possible, so as to avoid any possibility of occluding branches to the normal myocardium. Whatever technique of catheter closure is used, the occlusion should be at a precise point. Different types of embolisation materials can be used such as detachable balloons, stainless steel coils or platinum microcoils [2,19-23]. The technique may be influenced by several factors such as the age of the patient, the morphology of the feeding arteries and the location of the fistula. Many types of equipment should be available in the catheterisation laboratory for closing the fistulas. These include a selection of non-tapered catheters, balloon catheters, Tracker or Ferret catheters, different types of floppy or superfloppy coronary guidewires of 0.014" calibre, different types and sizes of coils (conventional Gianturco coils and controlled-release coils), detachable balloons and a variety of occlusion devices. In some patients, it may be easier to enter the fistula easily from the right side of the heart. These may be suitable for occlusion with an Amplatzer occluder type of devices, such as a vascular plug, a duct occluder or an atrial or ventricular septal occluder [11,12,13,14]. Our case is middle aged women with features of significant left to right more than 2:1 with signs of failure and having large exit size of fistula measuring 18mm size drains into superior vena cava surgery is the treatment of choice.

REFERENCE:


