

Transcatheter treatment of an unusual coronary arteriovenous malformation with a fistulous sac in the interventricular septum

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ABSTRACT

Coronary artery fistula (CAF) is a rare congenital anomaly with a reported incidence of 0.2%–0.6%. There is a wide variation in the clinical presentation depending on the size and the site of the fistula. Transcatheter closure is currently the treatment of choice in these patients. We report a case of CAF with an unusually large fistulous sac within the interventricular septum. The fistula had connections with all the three major coronary arteries, namely, left anterior descending (LAD), left circumflex, and right coronary arteries but did not have any exit resulting in to and fro movement of blood within the sac and the feeding vessels. The patient was managed successfully by transcatheter coil embolization.

Keywords: Coil closure, coronary aneurysm, device closure, percutaneous closure

INTRODUCTION

Coronary artery fistula (CAF) is a rare congenital anomaly in which one of the major coronary arteries opens directly into one of the cardiac chambers or venous structures around heart like coronary sinus. Although, most of the patients remain asymptomatic, a few of them present with heart failure, angina, myocardial infarction, infective endocarditis and very rarely with the rupture of the fistulous sac. Transcatheter closure remains the mainstay of treatment. Surgical closure is considered only in those where the transcatheter intervention is not feasible or has failed or has resulted in some complication. Despite a successful closure, most of the patients need to be under long term medical surveillance in order to prevent complications related to dilated feeder arteries and fistulous sac. We present a rare case of CAF which had multiple feeders with no distal communication requiring a different strategy for transcatheter closure.

CASE REPORT

A 24-year-old female presented with complaints of chest pain and dyspnea on exertion (New York Heart Association II/IV) of 3 months duration. On examination, her vitals were stable with an oxygen saturation of 98% in room air. Auscultation revealed a 3/6 systolic murmur over the lower part of the precordium. Electrocardiogram (ECG) showed T wave inversion in leads II, III, aVF, and V3-V6 [Figure 1]. Two-dimensional (2D)-echo showed a large cystic mass within the interventricular septum measuring 3.2 cm × 2.4 cm [Figure 2] along with the dilation of the coronary arteries. Cardiac magnetic resonance imaging revealed similar findings of an interventricular sac and connections to all the three coronaries [Figure 3]. Coronary angiography was performed, which showed a large fistulous sac at the distal end of the left anterior descending (LAD) artery [Figure 4a]. It also received

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feeding vessels from the obtuse marginal branch of the left circumflex (LCx) and posterior descending branch of the right coronary artery (RCA) [Figure 4b]. The sac did not have any exit for drainage, producing a to and fro flow in the feeding vessels.

Considering the symptomatic status of the patient, and given the size of the fistulous sac, micro coil occlusion of the fistulous sac along with the feeders was planned. The fistula sac was entered through the LAD feeder using a 0.014" (BMW) guidewire. To our surprise, the wire was easily passed into the other 2 feeders arising from the LCX and the RCA. Thus, there was no need to cannulate them separately through the LCX and the RCA, respectively. It was decided to occlude the smallest feeder arising from the LCX first. A microcatheter was passed over the wire stationed into the OM branch of the LCX through the fistulous sac, and the feeder was closed using a Concerto Helix eV3 2 mm × 4 cm coil. Thereafter, the RCA feeder was similarly entered through the sac and occluded with Concerto Helix eV3 3 mm × 8 cm and 4 mm × 8 cm coils [Figure 5a]. The sac was then partially filled using two 20 mm × 50 cm Helix eV3 coils. The last part of the second of the two coils was deployed in the feeder from the LAD, thus occluding it [Figure 5b].

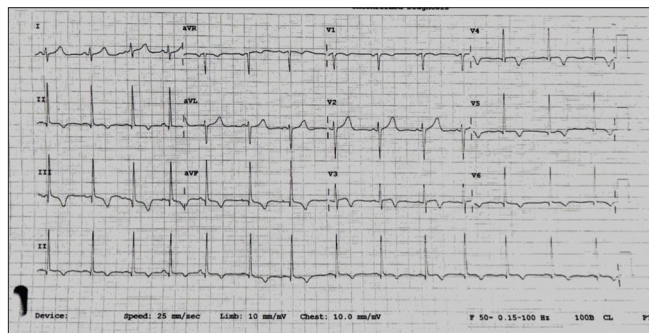


Figure 1: Electrocardiogram showing T inversion in II, III, aVF, and V3-V6

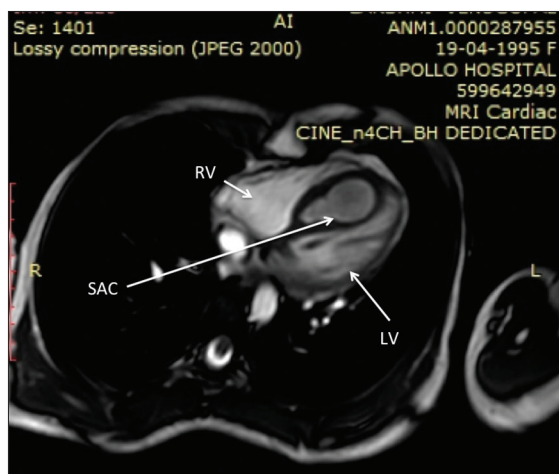


Figure 3: Magnetic resonance imaging showing the fistulous sac within the interventricular septum

Postcoiling angiogram of the LMCA and RCA showed a significant reduction of the flow into the sac. There was a slow flow in the RCA, but the flow in the LAD and LCx was brisk. The slow flow was managed using IV heparin for the next 48 h along with long term oral dual antiplatelet agents. Post-procedure echo showed coils within the sac [Figure 6a and b] without any flow into the sac on color Doppler. Ventricular function was normal, and there was no evidence of regional wall motion abnormality. There were no fresh ECG changes during or after the procedure.

At 1 month follow-up the patient was totally symptom-free. ECG remained unchanged with no evidence of any fresh changes. A 2D echo showed a reduction in the size of the fistulous sac, which was filled with coils. There was no blood flow seen in the sac on color flow mapping. There were no regional wall motion abnormalities, and the ventricular function remained normal.

Coronary angiogram was repeated after 6 months, which showed complete occlusion of flow to the fistulous sac. The coronary arteries showed reversed remodeling with normalization of their size, shape, and caliber with normal flow [Figure 7].

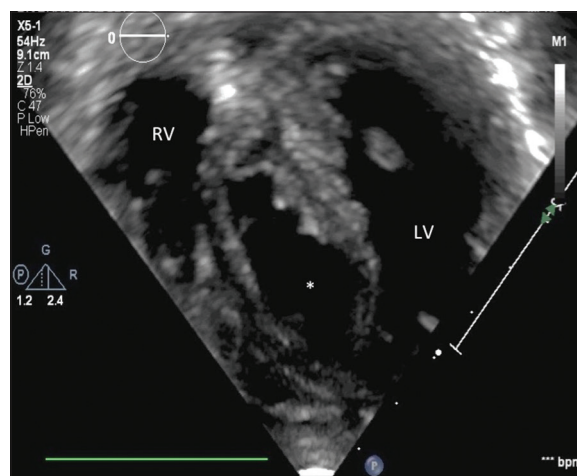


Figure 2: Two-dimensional echo four-chamber view showing the right ventricle and left ventricle with a fistulous sac (*) burrowing the interventricular septum

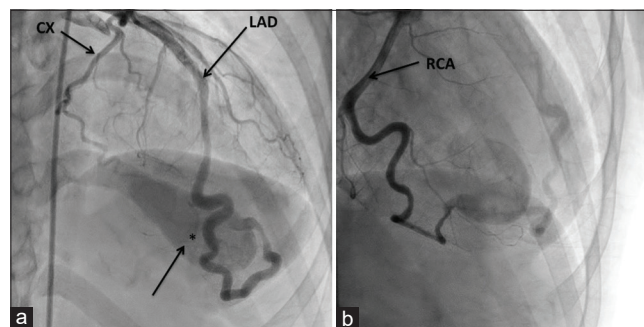


Figure 4: Coronary angiogram showing feeders into the sac (*) from the left anterior descending and circumflex (a) and from right coronary artery (b)

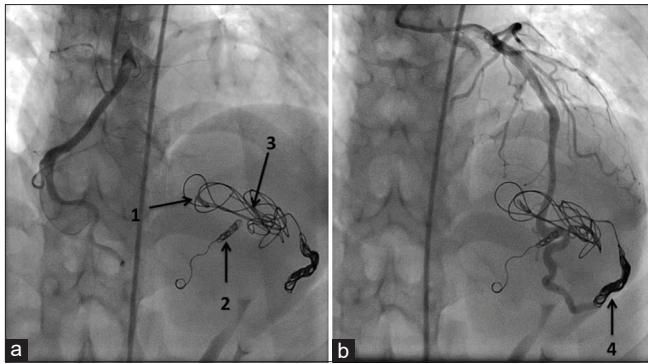


Figure 5: Concerto Helix eV3 (2 mm × 4 cm) coil occluding feeder from the left circumflex (1), (3 mm × 8 cm and 4 mm × 8 cm) coils occluding right coronary artery feeder (2), 20 mm × 50 cm coil within the fistulous sac and (3) in panel A and 20 mm × 50 cm coil occluding left anterior descending feeder (4) in panel B

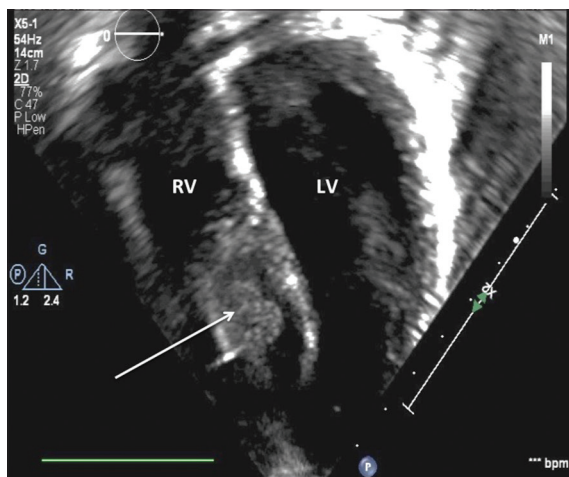


Figure 6: Two-dimensional echo four-chamber view showing the right ventricle and left ventricle with coils within the fistulous sac (*)

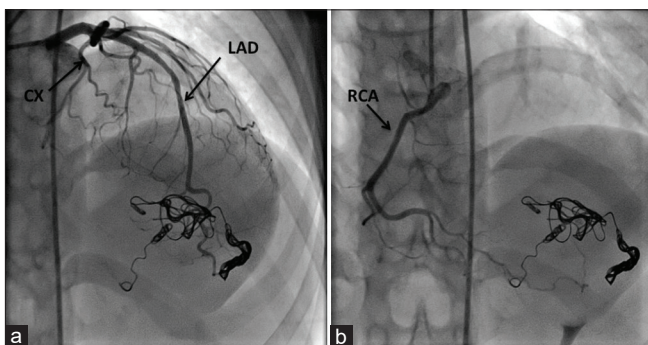


Figure 7: Coronary angiogram at 6 months of follow-up shows complete reverse remodeling of the (a) left anterior descending and circumflex arteries and (b) right coronary artery, with no flow into the fistulous sac

DISCUSSION

A coronary artery fistula (CAF) is a connection between one or more of the epicardial coronary arteries and a cardiac chamber or great artery or vein having bypassed

the myocardial capillary bed. Most coronary fistulas are congenital in origin, but they may occur after cardiac surgery or trauma.^[1] The fistulas originate from the RCA in about 52% of cases, the LAD artery being the next most frequently involved in approximately 30% of cases and the LCX artery in about 18%.^[1] 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.^[2] Our case was unique, with multiple feeders to the fistula, which had no exit resulting in huge, aneurysmal dilatation of the fistulous sac.

Clinical presentation of CAF depends upon the anatomy and the size of the fistulous connection, and thus, there is a tendency for symptoms to develop over time, with many of them remaining asymptomatic or presenting for the first time in adulthood as happened in our case. Complications include angina due to coronary steal from adjacent myocardium, thrombosis of the fistula due to blood stasis, heart failure due to volume overload, atrial fibrillation due to atrial enlargement, endocarditis, or endarteritis, rupture and arrhythmias.^[3] The 2008 American College of Cardiology/American Heart Association guidelines for the management of adults with congenital heart disease recommend closure of all large CAF regardless of symptomatology using transcatheter or surgical techniques and call for the closure of small to moderate fistulas only in the presence of symptoms (including ischemia, arrhythmia, and unexplained systolic or diastolic dysfunction).^[4] Our patient had features of angina probably as a result of a rapid increase in the size of the fistulous sac as well as due to the coronary steal as evident from the to and fro flow in the all 3 feeders. The ECG was also suggestive of ischemia in the inferior and anterior walls. Hence, it was decided to proceed with closure

Our case was unusual because it had no exit either in the right or the left-sided chambers. Thus it was mandatory to close the Coronary arteriovenous fistula (CAVF) retrogradely. Furthermore, the fistula was very distal and with the tortuous course and multiple feeders making it almost mandatory to use microcatheter and microcoils. Since we could enter all the feeders through the LAD via the fistulous sac, the procedure became all the more simple since the feeders from the RCA and the LCx could be occluded retrogradely through the sac. The retrograde entry into the RCA and LCX feeders would have been quite difficult even with a microcatheter given the distal and tortuous nature of these feeders. Since there was no exit to the fistula, the flow within all the 3 feeders was to and fro. This made the stability of the microcoils an important issue because the coils had the potential of embolizing retrogradely from one of the feeders through the sac into the other feeder. Hence, we used the detachable coils, which can be released in a controlled fashion only after confirming their stability.

These technical considerations helped in successful coil closure of this complex CAF with multiple feeders without any procedural complications and with the resolution of symptoms in the short term.

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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