Unusual presentation of a huge right coronary artery fistula; Technical issues with device closure

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Received 6 June 2013; accepted 11 July 2013
Available online 7 August 2013

Abstract  Coronary artery fistula (CAF) is considered an embryologic persistence of primitive intra-trabecular spaces which allow the developing coronary artery to communicate with the other cardiac chambers or vascular structures. It is observed in 0.05–0.25% of coronary angiographic studies, most of which drain into a right heart chamber or into the pulmonary artery, while a congenital right coronary artery (RCA) into a left heart chamber is less frequent. In this study, we describe an unusual case treated by closure device in the right coronary artery fistula to the left ventricle, and associated literature is reviewed. A 40-year-old female presented with chronic cough, otherwise asymptomatic. Echocardiogram revealed unusual flow into the left ventricle. The decision was to close this fistula with device through the RCA into the left ventricle. The management of this unusually large fistula is described with focus on technical issues with device closure.

1. Introduction

Coronary artery fistula (CAF) is considered an embryologic persistence of primitive intra-trabecular spaces which allow the developing coronary artery to communicate with the other cardiac chambers or vascular structures. It is observed in 0.05–0.25% of coronary angiographic studies, most of which drain into a right heart chamber or into the pulmonary artery, while a congenital right coronary artery (RCA) into a left heart chamber is less frequent. In this study, we describe an unusual case treated by closure device in the right coronary artery fistula to the left ventricle, and associated literature is reviewed.

2. Case report

A 40-year-old female presented with chronic cough, otherwise asymptomatic. Transthoracic echocardiogram revealed a mild to moderately dilated aortic root, mild aortic regurgitation, mild to moderate left ventricular dilation with unusual flow in the left ventricle. Trans-esophageal echocardiogram suggested a coronary fistula with flow entering the left ventricle just...
below the posterior leaflet of the mitral valve. A 64 multi-slice CT scan was performed that confirmed the diagnosis and delineated the anatomy of the fistula. The right coronary artery itself was massively dilated measuring up to 25 mm in diameter, it took a very tortuous course then communicated with the left ventricle just below the atrio-ventricular ring (Fig. 1).

3. Cardiac catheterization

The procedure was performed under general anesthesia. Access was obtained via the right and left femoral arteries and the right femoral vein. TEE was performed prior to commencing the catheterization to access the relationship between the entrance of the fistula into the LV and the mitral valve. The fistula entered into the posterior medial aspect of the LV near the inter-ventricular septum, just superior to the posterior-medial papillary muscle, but relatively distant from the posterior leaflet of the mitral valve. There was a mild aortic regurgitation and a trivial mitral regurgitation. The LV appeared mildly dilated with good ventricular systolic function. The RCA/fistula was injected in multiple projections to better delineate the anatomy of the fistula and to determine the take off of major tributaries and branches particularly the posterior descending coronary artery. A 7F balloon angiographic catheter (Arrow international, Reading, PA) was used so as not to traumatize the fistula during catheter manipulation and during the injections. The fistula formed a large C shape as it took off the RCA and measured 25 mm in it is widest dimension, it then tapered to a point around 10 mm, then opened into a larger mushroom shaped area measuring 19 mm and then narrowed again to about 14 mm as it entered the LV (Fig. 2). Fig. 3 reveals the posterior descending coronary artery well away from the final mushroom part of the fistula. After precise delineation of the anatomy it was elected to close the fistula as far distally as possible which would be in this mushroom shaped terminal part. As the largest part of this area measured 19 mm, we elected to use a 22 mm ASD device, knowing that the left sided disk of the device is 36 mm. The fistula was made from the aortic end using a 6F JR 3.5 100 cm catheter (Cook Inc., Bloomington, IN) and a Wooley Wire (Covidien, Plymouth, MN). The Wooley wire made it through the fistula into the LV,

Figure 1 3D reconstruction showing the dilated right coronary artery arising from the right coronary cusp then running a surpigenous course until it enters the left ventricle just inferior to the atrio-ventricular ring. Note the origin of the posterior descending artery (PDA).

Figure 2 (a) Angiogram in the lateral projection reveals the hugely dilated right coronary artery entering the LV. (b) Pictorial representation of the RCA and fistula; note the two areas of narrowing the mushroom shape of it is terminal end.

Figure 3 Angiogram revealing the dilated right coronary artery entering the LV and delineating the take off of the posterior descending coronary artery and its distance from the mushroom shaped part of the fistula.
but the JR was too short to reach the LV. The Wooley wire was removed and a .035 Teflon exchange wire (COOK Medical Inc.) was advanced into the LV. A Snare catheter (B Braun Interventional systems, Bethlehem, PA) was introduced into the ventricle via the left femoral artery, however it did not have the correct angle needed to snare the wire, so the snare was placed in a JR catheter that was used to snare the wire, this facilitated the torque of the snare. The wire was exteriorized out of the left femoral artery sheath and a wire loop was created (Fig. 4). A 10F 90 cm Flexor sheath (Cook Medical Inc.) was introduced over the wire from the LV side. The sheath barely reached the fistula and was ~7 mm short from being where we wanted to be. It was elected to leave the wire in place so as not to loose access while introducing the device next to it (buddy wire technique) (Fig. 5a). The left sided disk of the device opened in the mushroom shape of the fistula, but

Figure 4  (a) Cine in the lateral projection reveals the exchange wire entering the fistula from the aortic end and protruding into the LV. A snare is seen entering the LV via the aorta. (b) Cine in the lateral projection reveals the exchange wire snared and being pulled out of the aorta.

Figure 5  (a) Cine in the lateral projection reveals the long sheath with both the cable from the device and the buddy wire in place after the device is released. (b) Cine in the lateral projection showing the buddy wire as it is being pulled from the aortic end next to the device.

Figure 6  Angiogram in the lateral projection with and without contrast reveals the 22 mm Amplatzer device well seated in the fistula; note how the device is elongated to take the shape of the terminal part of the fistula.
the right sided disk opened mostly in the LV. This was felt to be inadequate, while performing the TEE the device slipped into the LV (still attached). The device was pulled back into the sheath and the sheath was advanced further into the fistula by applying more traction on the side of the wire coming out of the right femoral artery. That allowed the sheath to end in a better position well within the mushroom part. The same device was advanced into the mushroom part of the fistula, this time all the left and most of the right sided disk was placed inside the fistula with only a small protrusion into the LV. TEE was then performed to confirm that the device was not interfering with the mitral valve posterior leaflet or apparatus. An attempt was made at removing the buddy wire while the device was still attached, but this was not possible due to crowding of the sheath with the device cable next to the buddy wire. It was elected to release the device and remove the device cable prior to removing the wire. The wire was pulled out toward the aorta, thus pulling the device more into the fistula than into the LV (Fig. 5b). Angiogram confirmed the device to be in the fistula with small amount of flow though the device (Fig. 6). Patient was maintained on low dose aspirin. At 6 month follow up patient symptoms resolved, the shunt through the device was trivial and LV size was significantly decreased.

4. Discussion

Coronary artery fistulas have been recognized since 1965. The etiology of CAFs may be congenital, traumatic or iatrogenic, that is, after coronary intervention or valve replacement. Communication between a coronary artery and a cardiac chamber is less frequent. The most prevalent receiving chamber is the right ventricle (45%), followed by the right atrium (25%) and the pulmonary artery (20%). However CAF between the coronary artery and a left chamber is a rare condition. The clinical diagnosis of CAF is difficult because clinical presentation, laboratory and electrocardiographic manifestations are nonspecific. Most patients with CAF, as in the current patient, present with typical angina pectoris without CAD. Both CAF and cardiomegaly can produce myocardial ischemia and angina, and their association could aggravate the ischemia. The main mechanism of myocardial ischemia seems to be related to the coronary steal phenomenon.

CAF mostly remain asymptomatic. The treatment of CAF is essentially medical; conservative management with continued follow-up of these patients appears to be appropriate. Surgical correction is exceptional and may be considered in only severe forms with refractory to medical treatment. In the presence of symptoms of congestive heart failure, significant left-to-right shunt and arrhythmias, other major cardiac lesions, concomitant CAD, elective closure of coronary fistula are generally accepted. Clinical symptoms of coronary ischemia, such as exertional angina or dyspnea are the primary indication for the closure of a fistula. The first successful transcatheter coil embolization of a coronary artery fistula was performed in 1982. Our experience shows that the transcatheter occlusion of coronary fistula is a safe and effective procedure that is at least as successful as surgical treatment. Nevertheless, the technique may not be successful in every case. It is essential to select the optimal embolization method in relation to the size and location of the fistula, and to prepare a range of devices to cope with unexpected requirements and avoid multiple occlusion procedures.

4.1. Technical issues

Coronary fistula are commonly tortuous, however because of the late presentation, the size of the patient and the unusually large size of the fistula, it seemed like nothing we used was long enough. It would have been easier if we had longer catheters and sheaths available. Coronary fistulae should be closed as far distally as possible to spare as many branches of the coronary artery as possible. The use of the buddy wire has been reported in coronary artery intervention, however its use in the scenario of coronary fistula to our knowledge has not been reported . Using the buddy wire was a fall back when the device was inadequately placed the first time around and made reintroduction of the sheath straightforward. The buddy wire however made it cumbersome after the device was placed; as it could not be removed until the device was released, which could have dislodged the device. For that reason the wire was pulled out toward the aorta, rather than toward the LV to avoid embolization of the device. Thrombus formation in the fistula after closure has been reported, it is our impression that this is caused by roughening of the intima of the coronary artery by excessive manipulation, for that reason whenever possible one should avoid leaving wires exposed in the fistula and avoid introducing the long sheath into the fistula, hence the snare technique to get access. One should always understand the anatomy of the fistula completely before attempting to enter or close it. In our case we studied the fistula with a 64 multi-slice CT prior to the catheterization procedure, this helped clarify the anatomy on the outside, however the relationship of the fistula to vital structures for example the mitral valve apparatus in our case could have only been delineated with trans-esophageal echocardiography. To achieve the best results one should use all available modalities to reach a clear understanding of the anatomy before embarking on a long and tedious procedure like this.

5. Conclusion

Coronary fistula may have very unusual presentations; heightened awareness facilitates diagnosis of these unusual cases. Device closure of huge coronary fistula is feasible and safe only when all precautions including clear understanding of anatomy and carefully thought through approaches are used.

Conflict of interest

None.

References


