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Circumflex Coronary Artery Fistulae Draining into Right Atrium: A Case Report

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Abstract

Coronary artery fistula (CAF) is an anomaly making a coronary artery communicate with a heart cavity or a great vessel, thus bypassing the myocardial capillary bed. CAF is frequently diagnosed as an incidental finding. Herein, we present the case of a 4-year-old boy. He was referred for a pediatric cardiology assessment due to a continuous murmur at the middle sternal border. Echocardiogram showed dilated left coronary artery and an abnormal diastolic flow in the right atrium. The right-sided chambers were slightly enlarged, but ventricular contractility was normal. CAF was suspected. Angiography and CT confirmed the diagnosis of coronary fistula from the circumflex coronary artery to the right atrium. Successful transcatheter closure with an Amplatzer Piccolo Occluder was performed with complete occlusion.

Keywords

Coronary Artery Fistula, Circumflex Coronary Artery, Congenital Heart Disease

1. Introduction

Coronary artery fistula (CAF) is an anomaly making a coronary artery communicate with a heart cavity or a great vessel, thus bypassing the myocardial capillary bed [1] [2]. This malformation is rare and affects 0.002% of the population. It represents 0.4% of congenital heart disease [3]. In most cases, it is a congenital lesion and is asymptomatic in early life. Fistulas can be diagnosed by echocardiography with the visualization of a dilated coronary artery or by Doppler evaluation of the fistula itself. Imaging tests such as coronary computed tomography angiography and cardiac magnetic resonance imaging can play a significant role in the diagnosis and definition of the therapeutic strategy for this pathology. However, coronary angiography is the method of choice for diagnosis [4]. Spontane-

ous fistula closure is very rare, although cases have been described [5]. Small asymptomatic fistulas require no treatment and are followed up for complications. But in the cases of moderate and large CAFs complications such as coronary steal syndrome, myocardial infarction, heart failure, or tamponade can manifest following the abnormal communication that the fistula creates between the coronary arteries and cardiac chambers or major vessels and the subsequent shunt. The management is complicated and recommendations are based on cases of very small retrospective series. Treatment can be conservative, surgical, or more recently through transcatheter closure, depending on local experience and the morphology of the fistula. We present the rare case of a pediatric patient with a coronary artery fistula from the circumflex coronary artery to the right atrium. Transcatheter closure with an Amplatzer Piccolo Occluder was performed with complete occlusion. There were no complications early after closure and at 2 years follow-up.

2. Observation

A 4-year-old boy was referred to our center for further evaluation of a continuous parasternal murmur. He was asymptomatic during daily physical activities. The murmur was incidentally diagnosed during physical examination. On physical examination, a grade-2 continuous murmur was heard, most loudly at the left sternal border. An electrocardiogram showed no evidence of ischemia, and chest radiographs were normal. Transthoracic echocardiography (TTE) revealed a turbulent systolic-diastolic flow in the right atrium. The origin of the left main coronary artery and circumflex coronary artery was dilated (**Figure 1**). There was mild right atrial and ventricular dilation, and the patient's systolic pulmonary artery pressure was normal.

CT angiography demonstrated the fistula originated from circumflex coronary artery and ran toward the right atrium (**Figure 2**).

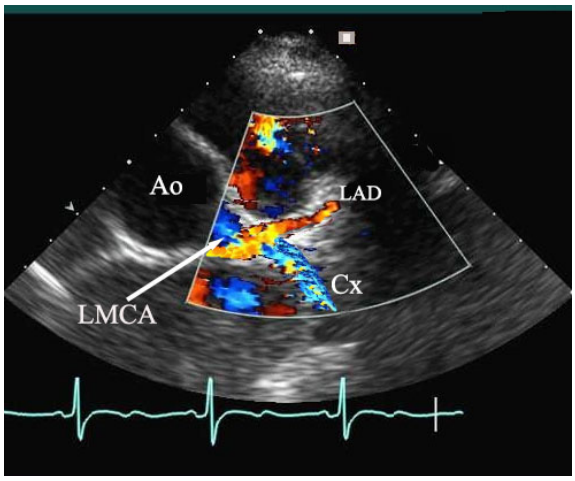

Figure 1. 2D transthoracic echocardiography, short axis view. LMCA: dilated left main coronary artery; Cx: dilated circumflex coronary artery; LAD: normal left anterior descending coronary artery; Ao: aorta.

Figure 2. CT angiography. Cx: dilated circumflex coronary artery; RA: right atrium.

Diagnostic cardiac catheterization and coronary angiography revealed a mild 1.4:1 left-to-right shunt, normal pulmonary artery pressure, normal epicardial coronary arteries, and a coronary fistula that arose from circumflex coronary artery and drained into the right atrium. The diameter of the narrowest part of the fistula, as measured by coronary angiography, was 1.5 - 2 mm (**Figure 3**).

The patient was readmitted to the cardiac catheterization laboratory two month later. The intention was to use the Amplatzer Piccolo Occluder ("ABBOTT") to close the fistula. We decided to perform the occlusion via femoral vein using arterio-venous wire loop. Vascular access was obtained through the right femoral artery (sheath 4F) and right femoral vein (sheath 5F). Heparin and antibiotic were administered. We passed the selective 4F diagnostic catheter into the left coronary artery. A guide wire 0.014 × 180 cm ("TERUMO" Runthrough Ns Floppy Guide Wire) was inserted into the dilated circumflex artery and then to the right atrium through the orifice. The wire was snared using an "Amplatz Goose Neck Snare" ("Medtronic") in the inferior vena cava and exteriorized in the right femoral vein for the creation of an arteriovenous wire loop (**Figure 4**).

The 4F (Amplatzer TorqVue LP Delivery Systems "ABBOTT") delivery system was inserted through the femoral vein into the CAF via the atrial end. The Amplatzer Piccolo Occluder was chosen to be approximately 2 mm greater in diameter than the diameter of the fistula and 9-PDAP-05-02-L Amplatzer Piccolo Occluder was loaded and delivered into the fistula (**Figure 5**).

Complete closure of the coronary artery fistula was observed by coronarography (**Figure 6**).

During 2-year follow-up he remained without any complaints or pathological symptoms. In control angio-CT, complete closure of the CAF was confirmed (**Figure 7**).

3. Discussion

A coronary artery fistula (CAF) is a rare congenital cardiac anomaly in which there is a connection between the coronary artery and a cardiac chamber or a great vessel, bypassing the myocardial capillary bed [1] [2]. The first description

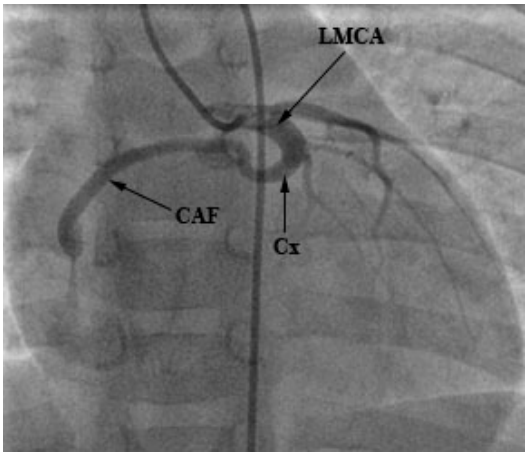

Figure 3. Coronarography of left coronary artery. LMCA: dilated left main coronary artery; CAF: coronary artery fistula; Cx: dilated circumflex coronary artery.

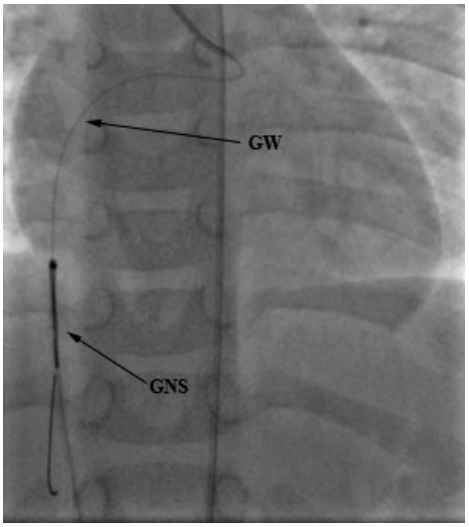

Figure 4. A guide wire was inserted into the circumflex artery and then to the right atrium through the orifice to the inferior vena cava. GW: "TERUMO" Runthrough Ns Floppy Guide Wire; GNS: "Amplatz Goose Neck Snare".

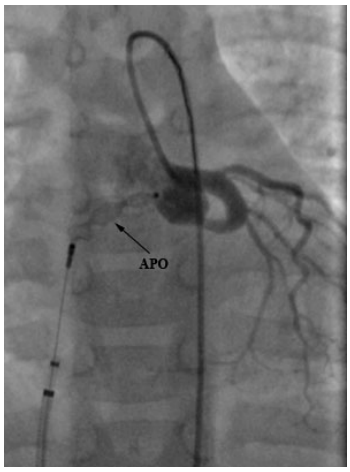

Figure 5. The Amplatzer Piccolo Occluder (APO) delivered into the fistula.

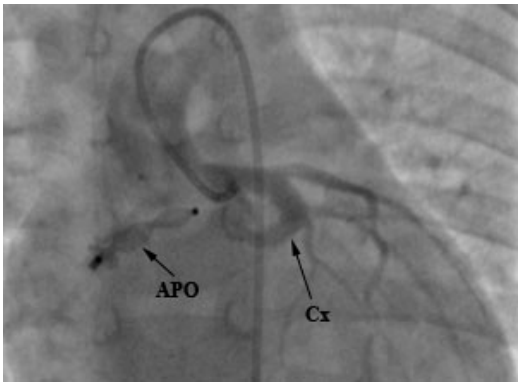

Figure 6. APO: Amplatzer Piccolo Occluder; Cx: circumflex coronary artery.

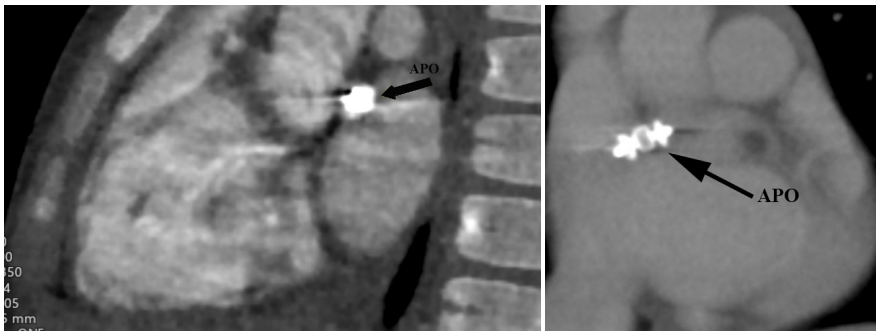

Figure 7. CT angiography shows the occluder in appropriate position. APO: Amplatzer Piccolo Occluder.

was by Krause in 1865 and the first surgical treatment was reported by Bjork and Crafoord in 1947 [3]. This malformation is rare and affects 0.002% of the population. It represents 0.4% of congenital heart disease [6]. They are often asymptomatic, so their diagnosis is often incidental. Coronary fistulas may be congenital or acquired. Most of the fistulas are congenital, and their embryological origin appears to be due to the persistence of sinusoidal connections between the lumens of the primitive tubular heart. The acquired forms may be divided into iatrogenic (during percutaneous coronary intervention, cardiac surgery, myocardial biopsy, and septal myectomy), traumatic, or related to a disease (such as myocardial infarction, Kawasaki disease) [7] [8] [9] [10]. The most common site of origination of congenital CAFs is the RCA with an incidence of 33%, followed by the left coronary artery (LCA) (34.9%), left anterior descending artery (6.3%), circumflex artery (4.8%), and finally, both RCA and LCA (1.6%) [11]. CAFs do not only arise from different coronaries but also terminate in different cardiac chambers and nearby vessels. The right ventricle (RV) is the most prevalent drainage site for CAFs, making up 34.9% of cases. The RA (27%) and pulmonary artery (PA) (27%) are the second most common sites of drainage and are followed by the left ventricle (LV) (6.3%), coronary sinus (CS) (3.2%), and finally LA (1.6%) [11].

The clinical manifestations of CAFs depend on the diameter of the fistula and the pressure gradient between the systemic blood pressure affecting the coronary

artery and the blood pressure in the draining chamber. In some cases, CAFs are relatively small, and patients are asymptomatic. But the clinical manifestations increase with age. CAFs can result in the coronary artery steal phenomenon and deviate blood flow through the coronary arteries into the terminating chambers. Other complications may include thrombosis, embolism, cardiac failure, arrhythmias, aneurysmal dilatation and rupture, endocarditis, endarteritis or arrhythmias [12] [13].

Natural history is variable: some close spontaneously, while others persist. It may happen that the coronary artery which originates fistula gradually dilated up to frank aneurysm, while the fistula may be complicated by ulceration of the intima, degeneration of the media, atherosclerotic plaques, calcification, mural thrombus, and rarely rupture [3]. The clinical manifestations increase with age. The most frequent symptoms are dyspnea on exertion, angina, fatigue, palpitations, and paroxysmal nocturnal dyspnea. Echocardiography is an important primary non-invasive tool for identifying the anomalous origin of CAF. In general, echocardiography can show the location and type of the CAF, including the course and drainage site of coronary artery, while it didn't delineate the exact anatomy of the fistula [14] [15] [16]. The gold standard for the detection of coronary fistulas remains coronary angiography. MRI and CT may provide additional diagnostic elements thanks to 3D reconstructions. Transthoracic and transesophageal echocardiography is useful especially in the evaluation of the hemodynamic effects of the fistula on cardiac chambers [9] [10] [17].

There are many ways to classify CAFs. From a clinical standpoint, CAFs can be divided based on the presence of myocardial ischemia, into anomalies without ischemia, anomalies with episodic ischemia, and anomalies with obligatory ischemia. Despite this important functional assessment, physicians often categorize CAFs based on anatomic characteristics [7] [18]. The coronary arteriovenous fistulas are divided into five types according to the chamber or vessel into which they drain: Type I (draining into the right atrium), Type II (draining into the right ventricle), Type III (draining into the pulmonary artery), Type IV (draining into the left atrium), and Type V (draining into the left ventricle) [19].

The closure of the fistula is recommended when it is symptomatic, while the treatment in asymptomatic patients remains controversial. Large coronary fistulas should be closed by transcatheter or surgical treatment, regardless of symptoms, while small to moderate size fistulas should be treated only if they cause symptoms [16] [17]. The surgical approach is ligation of epicardial fistula. Transcatheter closure may be performed with various types of devices (stents, umbrellas, balloons, coils, etc.) but requires favorable anatomy, *i.e.* not tortuous artery with single fistula and accessibility of the distal portion to closure device [15] [16] [20]. The advantages of the transcatheter approach include less morbidity, lower cost, shorter recovery time, and avoidance of thoracotomy and cardiopulmonary bypass. In comparison with many devices that were previously used for the occlusion of coronary fistulae, the Amplatzer Piccolo occluder affords several advantages, including ease of delivery, a wide range of device sizes,

and the opportunity to reposition the device safely during and after initial deployment. In our case, the shape of CAF was favorable for transcatheter closure because of distal (right atrial end) stenosis of the one. Our choice was a device that was at least 50% larger than the narrowest segment of the fistula, in order to prevent the risk of a residual shunt. The successful results in our patient's case suggest that it is feasible and safe to apply a retrograde approach when placing an Amplatzer Piccolo occluder for transcatheter occlusion of coronary artery fistulae.

4. Conclusion

This case demonstrates a coronary cameral fistula, which was successfully treated by transcatheter closure with an Amplatzer Piccolo Occluder. The diagnosis of CCF can be challenging requiring multiple imaging modalities. This case highlights the utility of noninvasive echocardiography and color Doppler, which can be used in the initial detection of this defect [9] [10]. A cardiac CT angiogram is another modality for diagnosis. However, coronary angiography is still the gold standard of diagnosis.

Conflicts of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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