Original article

Dual origin multiple plexus-like coronary to pulmonary artery fistulas – Consideration of their etiology and therapeutic strategy

Hisashi Sugiyama (MD, PhD)*, Tetsuko Ishii (MD), Toshio Nakanishi (MD, PhD, FJCC), In-Sam Park (MD, PhD)

Pediatric Cardiology, Heart Institute, Tokyo Women’s Medical University, Tokyo, Japan

A R T I C L E   I N F O

Article history:
Received 7 April 2016
Received in revised form 29 June 2016
Accepted 19 July 2016
Available online 21 August 2016

Keywords:
Coronary artery fistula
Vascular plug
Detachable coil
Plexus-like formation

A B S T R A C T

Background: Multiple plexus-like coronary to pulmonary fistulas (CAPFs) originating from bilateral coronary arteries are a rare type of coronary artery fistula (CAF). Their etiology and therapeutic strategy are discussed.

Methods and results: Three patients were diagnosed with dual origin plexus-like CAFs drained to the pulmonary artery trunk. Their ages ranged from 40 to 78 years. Enhanced computed tomography could clearly demonstrate three-dimensional anatomy of CAPFs. Four catheter interventions were performed in all 3 patients. Antegrade approach was applied in 2 procedures and retrograde approach in 2 procedures. A detachable coil was used in 3 procedures and a combination of a vascular plug and detachable coils for 1 procedure. The effective occlusion was achieved in all patients without complications. Follow-up myocardial scintigraphy showed no perfusion defect in all patients.

Conclusions: CAPFs in our cases, which developed in their adulthood, anatomically correspond with the vasa vasorum of the proximal of the great arteries. The dual origin multiple plexus-like CAPFs might develop from the vasa vasorum of the proximal portion of the great arteries with age. The antegrade approach would be effective because even dual origin multiple plexus-like CAFs converged and drained to a single major exit of the pulmonary trunk. Catheter occlusion could be feasible and safe for dual origin multiple plexus-like CAPFs.

© 2016 Japanese College of Cardiology. Published by Elsevier Ltd. All rights reserved.

Introduction

A coronary artery fistula (CAF) is a rare coronary abnormality. Incidental detection of a CAF in asymptomatic patients has been reported as 0.1–0.2% on coronary angiography [1,2]. The prevalence of a CAF on coronary computed tomography (CT) was 0.3–0.9%, which is higher than that based on coronary angiography [3,4]. Fifty percent of CAFs originated from the right coronary artery (RCA), 42% from the left coronary artery (LCA), and 5% from both coronary arteries. Drainage was opened into the right ventricle in 41%, into the right atrium in 26%, into the pulmonary artery in 17%, into the coronary sinus in 7%, into the left atrium in 5%, into the left ventricle in 3%, and into the superior vena cava in 1% [5]. Meanwhile in the coronary CT study, the most common type of CAFs was multiple plexus-like coronary to pulmonary fistula (CAF). The discrepancy in those may depend on the difference of the imaging modality. Previous case reports revealed that a dual origin multiple plexus-like CAF had distinctive features [6–9].

We review three recent patients with dual origin multiple plexus-like CAPFs and discuss their etiology and therapeutic strategy.

Materials and methods

Three patients were diagnosed with dual origin multiple plexus-like CAFs drained to the pulmonary artery trunk. The patients’ characteristics are shown in Table 1. Their ages ranged from 40 to 78 years. Multiple plexus-like fistulas originated from both LCA and RCA, and drained to the pulmonary artery trunk in all patients. Before the catheter intervention, 3DCT and coronary angiography were evaluated. The indications for closure were based on their clinical symptoms, features of fistula, and hemodynamics. In the present series, all had dyspnea on exertion and aneurysmal formation which put the patients at risk of an unexpected rupture. Case 1 had left ventricular volume overload

* Corresponding author at: Pediatric Cardiology, Heart Institute, Tokyo Women’s Medical University, 8-1 Kawada, Shinjuku, Tokyo 162-8666, Japan.
Fax: +81 3 3356 0441
E-mail address: psugiyama@hj.twmu.ac.jp (H. Sugiyama).

http://dx.doi.org/10.1016/j.jjcc.2016.07.016
0914-5087/© 2016 Japanese College of Cardiology. Published by Elsevier Ltd. All rights reserved.
with 1.4 of pulmonary systemic flow ratio by Fick principle and had ST depression in chest leads on her electrocardiogram. Therapeutic strategy was discussed in cardiac team with cardiac surgeons. All three patients underwent catheter intervention for the CAF. Case 1 underwent 2 sessions of the procedure. Details of the catheter procedures and the outcomes were reviewed.

Written informed consent was obtained from all patients or their guardians prior to the procedure. Tokyo Women’s Medical University institutional review board approved the present retrospective review.

Results

The chief complaint was dyspnea on exertion and chest discomfort in all patients. Pulmonary systemic flow ratio by Fick principle was 1.4 in case 1 and 1.1 in cases 2 and 3. Left ventricular function was normal in cases 1 and 3, but ejection fraction was reduced to 0.51 in case 2 on echocardiography. Atherosclerosis was suspected in case 3 with high blood pressure, high low-density lipoprotein cholesterol level, the higher value of pulse wave velocity, and 65% stenosis of plaque on the carotid arteries on vascular echo assessment. No one had family history of hemangioma and CAF. And also no one had findings of systemic hemangioma.

Three-dimensional CT study

Three-dimensional CT showed plexus-like vessels developed around the anterior ascending aorta, right ventricular outflow, and pulmonary artery trunk in all cases (case 1: Fig. 1, case 2: Fig. 2, and case 3: Fig. 3). The fistula formed a sacculated aneurysm just before the drainage to the pulmonary artery trunk in cases 1 and 3, and a large dilatation just before the exit orifice in case 2 (case 1: Fig. 4, case 2: Fig. 5, case 3: Fig. 6). In case 3, size of the aneurysm developed from 22 mm × 24 mm 5 years previously to 24 mm × 27 mm at 3 years previously, to 25 mm × 30 mm at present.

Catheter intervention

Angiographic findings and catheter intervention are summarized in Table 2. In case 1, LCA angiography showed multiple tortuous fistulas from the left anterior branch (LAD) and fistulas formed plexus vessels with aneurysm (Fig. 4a). RCA angiography showed a CAF, which went into the plexus vessels (Fig. 4b). Initially, two small fistulas from LAD were occluded with detachable coils (Orbit Galaxy, DePuy Synthes, West Chester, PA, ...
A vascular plug-I (12 mm, St Jude Medical, Plymouth, MN, USA) was implanted for 10 mm × 12 mm diameter of the aneurysm (Fig. 4c). In addition, detachable coils were implanted for small CAFs originating from the RCA.

In case 2, LCA angiography showed multiple tortuous fistulas from LAD and RCA angiography showed a small CAF, which connected to the plexus fistula from LAD (Fig. 5a). Considering the experience of case 1, an antegrade approach was adapted. Under guidance of the selective coronary angiography and 3DCT, a multifunction catheter was advanced into a large dilated fistula. Four pieces of detachable coils (Target XL, Stryker Co. Ltd., Kalamazoo, MI, USA) were implanted into 13.7 mm × 5.8 mm size of dilated fistula (Fig. 5b).

In case 3, LCA angiography showed multiple tortuous fistulas with several aneurysms from LAD and RCA angiography showed a small CAF, which connected to the plexus fistula from LAD (Fig. 6a). An antegrade approach could not be applied because the largest saccular aneurysm adjusted to the pulmonary artery was already occluded with a thrombus and another exit of a CAF could not be clearly detected on the angiography. Five pieces of detachable coil (Target XL, Stryker, Co. Ltd.) were implanted into the aneurysmal fistula by the retrograde approach (Fig. 6b). The small fistula from RCA remained but the shunt was considered to be negligible. Warfarin was administered to keep 1.5–2.5 of prothrombin time-international normalized ratio for at least 6 months after the procedure and 100 mg of aspirin was also administered as antiplatelet.

To prevent thromboembolism, activated clotting time was routinely measured at 30-min intervals and was kept more

Fig. 3. 3DCT showed multiple thread-like coronary fistula, which originated from both RCA and LAD (black arrow), and formed aneurysms and one huge aneurysm of those (white arrow) was calcified and almost occluded with a thrombus in case 3.

Fig. 4. (a) LCA angiography showed multiple twisting fistulas (white arrow), which formed plexus vessels with an aneurysm (black arrow) from left anterior branch in case 1. (b) RCA angiography showed coronary artery fistula (black arrow), which went into plexus formation (white arrow) in case 1. (c) Vascular plug-I (black arrow) was implanted for 10 mm × 12 mm diameter of the aneurysm by the guiding catheter (white arrow) through pulmonary artery in case 1.

Fig. 5. (a) LCA angiography showed multiple entries and twisting fistulas from the left anterior branch (white arrow) in case 2. (b) Four pieces of detachable coil (black arrow) were implanted into the aneurysm (14 mm × 6 mm) by the guiding catheter (white arrow) through pulmonary artery in case 2.
than 250 s by heparin sodium injection during the procedure. Furthermore, coronary guiding catheter was gently manipulated to be parallel to axis of the coronary artery for prevention from coronary dissection. Neither ST change on electrocardiogram nor elevation of myocardial enzyme occurred with the procedure in any of the patients. In case 1, ST change improved after catheter intervention. Symptoms were relieved in all patients. After the procedure, follow-up myocardial scintigraphy by 201 TlCl or 99mTc-MIBI with excise or adenosine loading showed no significant findings in all patients.

**Discussion**

The etiology of CAFs is either congenital or acquired. Congenital fistulous connections between the coronary arteries to cardiac chamber appear to represent the persistence of embryonic intratrabecular spaces and sinusoids [10]. With regard to the etiology of congenital CAPFs, Vaidyanathan et al. proposed the Hackensellner involution-persistence hypothesis, which was originally for total anomalous origin of the coronary arteries from the pulmonary artery [9,11]. In our series, the hypothesis was not applicable because the appearance of fistulas in our series was obviously different. Furthermore, symptoms and heart murmur could not be noticed in their younger age. Those implied that etiology of the CAPF in our series was not congenital but acquired. CAFs can be acquired from trauma, inflammation, or can be iatrogenic. Clinically, they can occur as a result of intracardiac surgery, percutaneous myocardial biopsy, or as a complication of Kawasaki disease or Takayasu disease [12–16]. In the present cases, CAFs originated from both coronary arteries and connected to each other and the exit of their CAFs was located at the anterior to left lateral aspect of the pulmonary artery trunk. An aneurysmal or large dilated fistula developed just before the exit of the CAFs. Histologically, a saccular aneurysm was thought to be caused by medial necrosis, which was suspected to be inflammation, injury, or stenosis with atherosclerotic change [7,17]. Case 3 had evidence of atherosclerotic disorders but the other two patients had no evidence of atherosclerosis.

Anatomically, the vasa vasorum of the ascending aorta and pulmonary trunk are mainly supplied by both coronary arteries. Anterior aspects of the ascending aorta and pulmonary trunk are supplied by RCA, whereas anterior and left lateral aspects of the pulmonary trunk are supplied by LCA. These superficial vessels form a network in adventitia as well as intramural plexus [18,19]. Coronary 3DCT has the capability of demonstrating comprehensive complex anatomy of a CAF [3,4]. In our series, 3DCT clearly showed plexus vascular formation in front of the ascending aorta and pulmonary trunk. Previous studies revealed that CAPFs have different features from the other types of CAFs which drained to the cardiac chamber [6–9]. Similar to our cases, multiple CAFs tend to be supplied by both coronary arteries and terminate more often into the pulmonary artery [20]. Furthermore, some studies reported that a CAPF often has small shunt and tends to form an aneurysm [17,21,10]. Considering the analogy between the vasa vasorum of the anterior aorta and pulmonary trunk, and the appearance of CAPFs on 3DCT in our series, the dual origin multiple plexus-like CAFP could develop from vasa vasorum with age and eventually connect to the pulmonary artery.

The indications for closure are clinical symptoms, especially heart failure, myocardial ischemia, and large fistulas in asymptomatic patients, especially in pediatric age groups [3,12,20]. As in our series, dyspnea on exertion is the most common symptom [22]. Aneurysmal formation is also considered to be an indication for closure to prevent an unexpected rupture [7,17]. Meanwhile, treatment for asymptomatic small CAFs is still controversial [7,12,22–24]. With advances in the technology of imaging modalities, asymptomatic small CAFs were accidentally detected on coronary CT or echocardiography [3,4,12,21]. Long-term
follow-up studies in asymptomatic patients with small CAPFs revealed no complications and no cardiac events [12,21, 22,24]. Thus, an asymptomatic small size CAPF should be conservatively observed although the risk of infective endocarditis remains [23]. In our series, catheter intervention was based on the symptoms, an aneurysmal formation which was at risk for rupture, or hemodynamics.

Recently, catheter intervention for CAPFs became popular because a variety of devices such as conventional coils, detachable coils, and vascular plugs are available. The retrograde approach for multiple plexus-like CAPF was time-consuming and could result in inadequate occlusion. Meanwhile, the antegrade approach was effective because even multiple plexus-like fistulas converged and drained to a single major exit of the main pulmonary artery.

Conclusion

We have experienced three patients with dual origin multiple plexus-like CAPFs. Three-dimensional CT was remarkably useful to understand the comprehensive anatomy of a CAPF and could provide us with essential information for the catheter intervention. The dual origin multiple plexus-like CAPFs anatomically correspond with vasa vasorum of the proximal of the great arteries. Multiple plexus-like CAPFs might develop from vasa vasorum of the proximal portion of the great arteries with age. Catheter occlusion by antegrade approach is effective for a single major exit with aneurysm before the drainage site. Catheter occlusion could be feasible and safe for dual origin multiple plexus-like CAPFs.

Conflict of interest

The authors have no conflicts of interest to declare.

Acknowledgments

We thank all the catheter laboratory staff for their support.

References