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Case Report

Giant coronary artery fistula: A case report☆,☆☆

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ABSTRACT

Coronary artery fistulas (CAFs) are abnormal connections of coronary arteries where venous circuits bypass the normal capillaries in the myocardium. CAFs are rare, and most patients are asymptomatic. However, CAFs are the most common coronary artery anomalies affecting coronary hemodynamics. While most CAFs are asymptomatic in young patients, symptoms and complications become more frequent with increasing age. CAFs are characterized by variable clinical manifestations based on their size, origin, and drainage site. We describe a 35-year-old woman presenting with the shortness of breath after walking. Despite attempting medical treatment, the patient continued to experience dyspnea, fatigue, fainting the and chest pain episodes. After admission, cardiac imaging was immediately performed and recorded symptomatic CAFs. Percutaneous transcatheter closure treatment was indicated. The patient was discharged with clinical recovery. The treatment of symptomatic CAFs often requires the clear cardiac imaging and endovascular approach to achieve the best clinical results.

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Introduction

Coronary artery fistulas (CAFs) and coronary arteriovenous fistulas are congenital or acquired abnormal connections of coronary arteries with cardiac chambers or any systemic or pulmonary circulation segment without an intervening capillary network [1]. CAFs are uncommon coronary artery abnormalities, accounting for 0.3% of congenital heart diseases [1]. CAFs are classified by their drainage site, such as cardiac chambers, the coronary sinus (CS) and its tributary veins, or great vessels. They rarely drain into the CS and may frequently be associated with aneurysmal dilatation [2]. CAF prevalence based on computed tomographic angiography (CTA) is as high as 0.9%, higher than the previously reported prevalence of 0.002%-0.3% based on invasive angiography [3]. While CAFs have historically been evaluated by conventional invasive angiography, computed tomographic angiography (CTA) of the heart has emerged as a noninvasive method for quick diagnosis. Furthermore, 3-dimensional volume-rendered CTA facilitates accurately assessing the CAF's complex anatomy, including its origin, drainage site, and fistulous tract numbers and sizes. In adulthood, CAFs generally present after the second decade, with symptoms ranging from mild dyspnea to ischemic events or heart failure related to the left to right shunt [4]. The myocardial ischemia, heart failure, arrhythmia, and infective endocarditis are the most common complications of CAFs in adults [4]. If CAFs cause symptoms, vascular ligation or percutaneous closure is often recommended. Fistula closure is recommended in cases with large dimensions, a pulmonary-systemic shunt ratio of >1.5, or ischemic events. Since coronary aneurysm thrombosis may occur as a postoperative complication, anticoagulation and antiplatelet therapies are recommended [5].

Case presentation

A 35-year-old woman was referred to Can Tho S.I.S. General Hospital (Can Tho city, Vietnam) with shortness of breath. She suffered from dyspnea, fatigue, chest pain episodes, fainting, and shortness of breath after walking. Her medical history showed that she experienced acute heart failure despite no family history of cardiac diseases after her second pregnancy 2 years ago. Her physical examination showed jugular venous pressure, continuous murmur grade 3/6 in the second intercostal space left sternal margin, a heart rate of 100 beats/min, blood pressure of 110/70 mm Hg, and peripheral oxygen saturation of 98%. An electrocardiogram (ECG) showed a tachycardia of approximately 110 beats/min with a type 1 atrioventricular (AV) block (Fig. 1).

Chest radiography showed an enlarged heart shadow and a cardiothoracic ratio >0.5 (Fig. 2). Transthoracic echocardiography had abnormal parameters: an ejection fraction (EF) of ~50% (based on the modified Simpson's method), a dilated left ventricle (58 mm), a mitral valve regurgitation level of 2/4 (based on annulus dilatation), a tricuspid valve regurgitation level of 2/4, and a pulmonary arterial pressure of 50 mm Hg (Fig. 3). We found a severe dilation of the proximal right

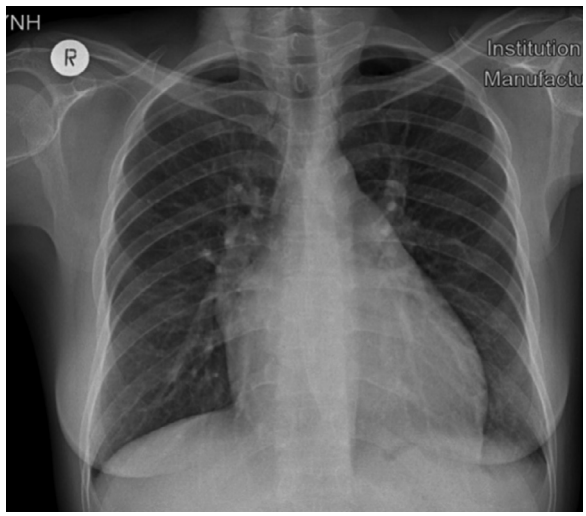

Fig. 2 – Chest radiography. An enlarged heart shadow and a cardiothoracic ratio of > 0.5 are present.

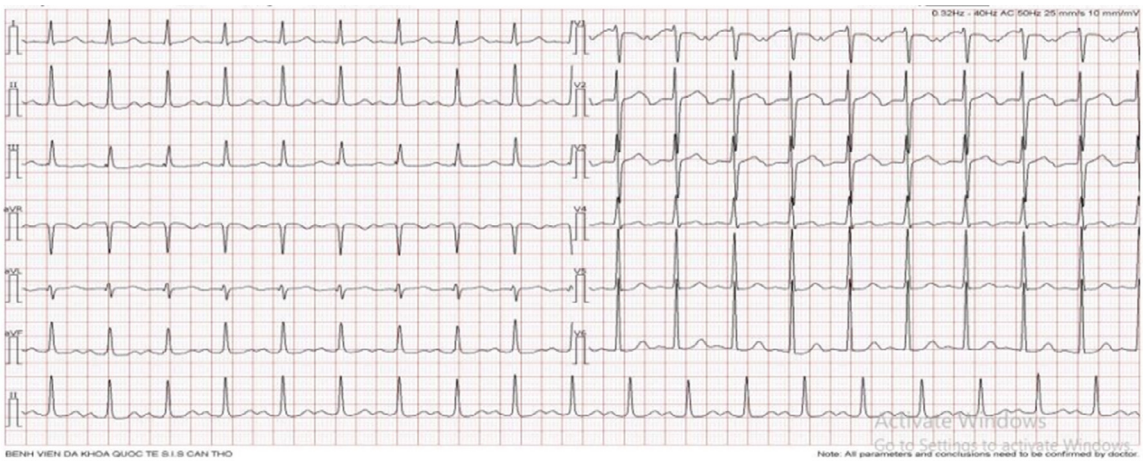

Fig. 1 – Preoperative ECG. The heart rate was ~110 beats/min with a type 1 AV block.

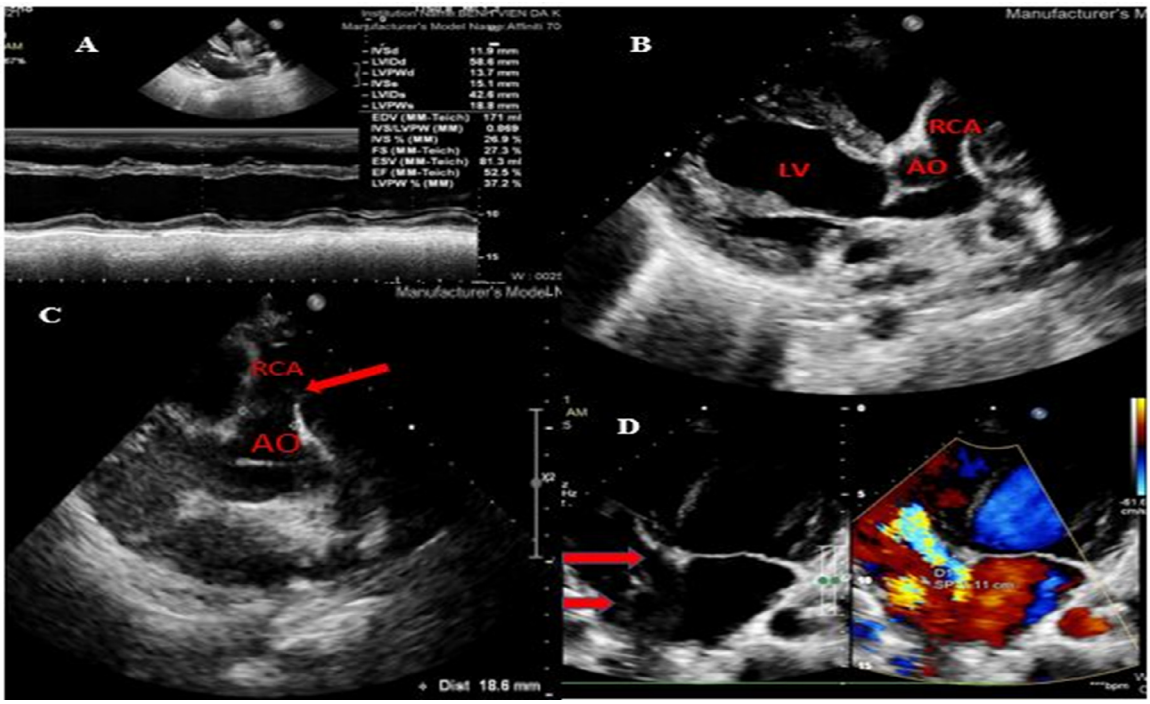

Fig. 3 – Preoperative transthoracic echocardiography. (A) Measured left ventricular parameters. (B–C) Severe dilation of the proximal RCA (red arrow). (D) An aneurysm of the middle RCA and a fistula to the right atrium with 2 branches (red arrows). Key: AO, aorta; LV, left ventricle; RCA, right coronary artery.

coronary artery (RCA; 18.6 mm), an aneurysm of the middle RCA (30 mm), and a fistula to the right atrium with 2 branches; the left-main coronary artery was normal with a diameter of ~3 mm.

The CTA confirmed the diagnosis, showing a giant, tortuous aneurysm originating from the proximal RCA and draining into the right atrium, an enlarged proximal (18 mm) and middle (40 mm) RCA, and the functional coronary artery originating near the proximal RCA (Fig. 4).

We performed percutaneous transcatheter closure. We placed the 6F sheath in the right femoral artery and right femoral vein. The heparin dose was 70 IU/kg. A 5F pigtail catheter was used for right coronary angiography at the root of the right artery. We determined coronary artery anatomy, fistula size, and functional RCA location. The 0.035″ guidewire was inserted from the aorta through the RCA into the right atrium. Then, a snare was used to catch the guidewire and pull it into the inferior vena cava and out into the right femoral vein to create a pathway from the aorta through the vena cava. A SteerEase Introducer 7F was inserted from the right femoral artery into the root of the aorta and slipped into the proximal and middle RCA. The first instrument (Amplatzer Vascular Plug II [16 mm]) was released at the fistula's end, and the second instrument (LifeTech Konar M VSD Occluder [14/12 mm]) was released in the middle of the RCA after the functional coronary artery. Angiography showed decreased shunt through the fistula. We proceeded to remove the tool. The summary report is shown in Fig. 5.

The dilated RCA is indicated by a yellow arrow, and the LifeTech Konar-MF VSD Occluder (14/12 mm) device is indicated by a red arrow.

The patient's postoperative course was uneventful. The medicine prescription included aspirin (160 mg/d), losartan (25 mg/d), and spiromide (20/50 mg/d). After 1 week, the patient felt dizzy and lightheaded, and her blood pressure was 90/60 mm Hg. The ECG showed junctional bradycardia (Fig. 6), but her high-sensitivity cardiac troponin I was normal. Continuous treatment included theophylline (100 mg; ½ pill twice daily) and oral anticoagulation therapies (acenocoumarol [4 mg]; ¼ pill per day) to maintain an international normalized ratio of 2-3.

After 2 weeks, the patient had decreased dizziness, an ECG showed a sinus rhythm and heart rate of ~80 beats/min (Fig. 7), and echocardiography showed an intradevice shunt.

After 1 year of follow-up, echocardiography showed a normal left ventricular function EF (55.6%) and decreased left ventricular dilation (54.5 mm; Fig. 8). The CTA showed the successful closure of the fistula without any relevant changes to the coronary arteries (Fig. 9).

Discussion

We presented a case with severe proximal RCA dilation (18.6 mm), an aneurysm in the middle RCA (30 mm), and a fistula to the right atrium with 2 branches. The RCA is the most common CAF origin site, accounting for 50%-55% of all cases (Sakakibara CAF classification) [6].

Coronary-cameral fistulae involve the entry of a coronary artery into any of the cardiac chambers. They are the most common CAF type detected by coronary angiography [7]. Coronary-cameral fistulae commonly originate from the

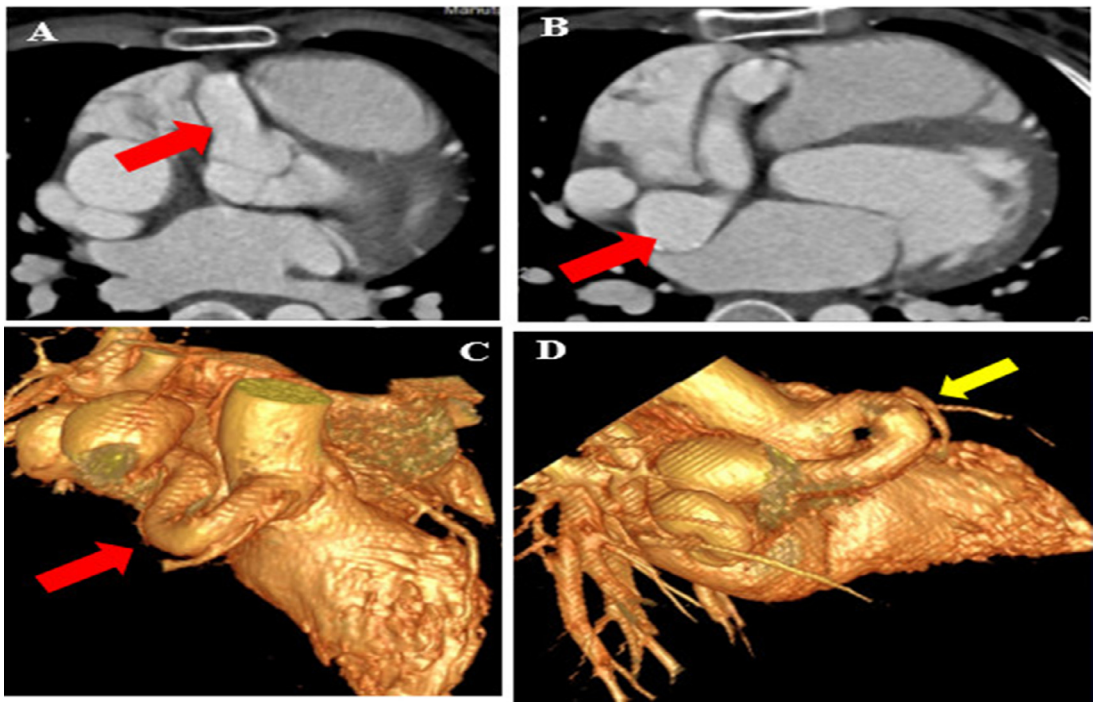

Fig. 4 – Preoperative CTA, (A–C) A fistula from the RCA to the right atrium (red arrow). (D) A functional RCA supplied blood to the right ventricle (yellow arrow).

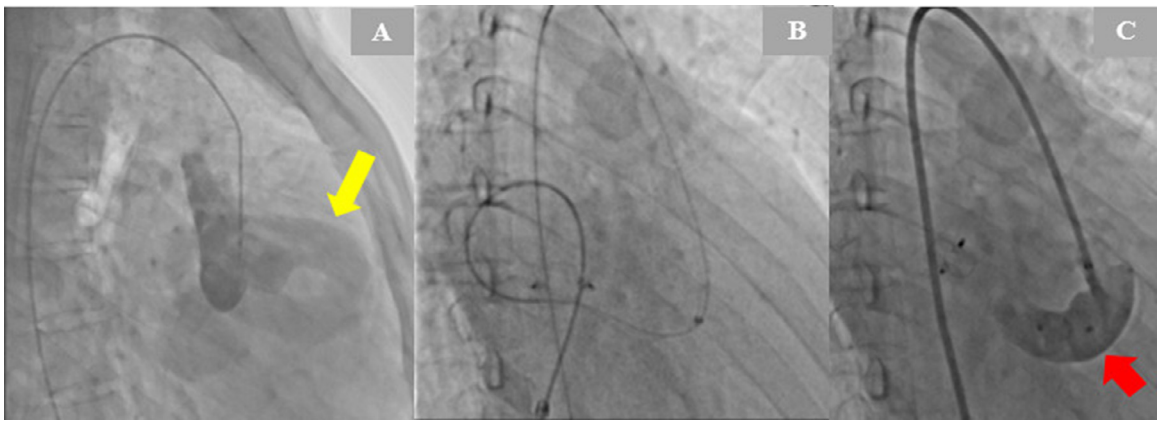

Fig. 5 – Percutaneous transcatheter closure report. (A) The sheath was placed in the right femoral artery and vein using a pigtail catheter for right coronary angiography at the root of the right artery. (B) The guidewire was inserted from the aorta through the RCA into the right atrium. The snare was used to catch the guidewire and pull it into the inferior vena cava and out into the right femoral vein to create a pathway from the aorta through the vena cava. (C) A SteerEase Introducer 7F was inserted from the right femoral artery into the root of the aorta and slipped into the proximal and middle RCA. The instruments (Amplatzer Vascular plug II [16 mm] and LifeTech Konar-MF VSD Occluder [14/12 mm]) were released in the middle of the RCA after the functional coronary artery.

RCA (55%), the left anterior descending artery (35%), or both. The most commonly involved chamber is the right ventricle (41%), followed by the right atrium (26%), and the left atrium and ventricle (3%-5%) [8]. Constant high blood flow through coronary arteries may lead to aneurysmal dilatation of arterioluminal coronary-cameral fistulae. A CAF into the right heart chamber (ie, left-to-right shunt) leads to increased right heart volume and pulmonary hypertension [9]. Most coronary-cameral fistulas are asymptomatic and detected incidentally. However, for large hemodynamically significant fistulae, surgical or percutaneous procedures are often required [2].

The management strategy for patients with CAFs depends on fistula size and anatomy, symptoms, patient age, and associated cardiovascular abnormalities [10]. While the spontaneous closure of a fistula is rare, it occurs in 1%-2% of cases. Therefore, regular follow-up is suggested [10]. Small asymptomatic CAFs are often treated with antiplatelet therapy and antibiotics and monitored for complications without inter-

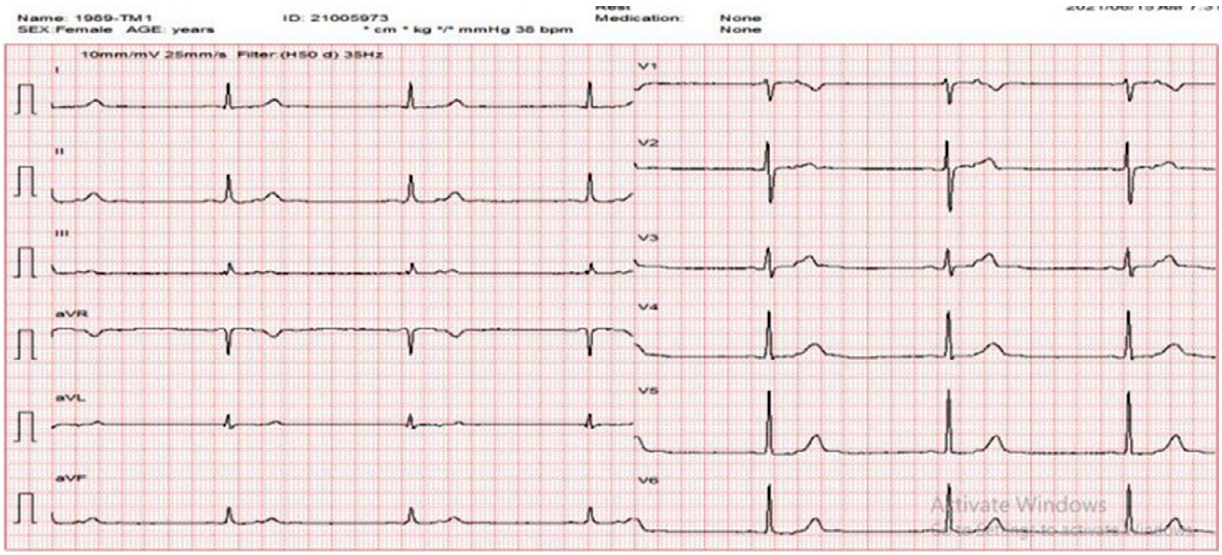

Fig. 6 – ECG after 1 week. Junctional bradycardia is apparent.

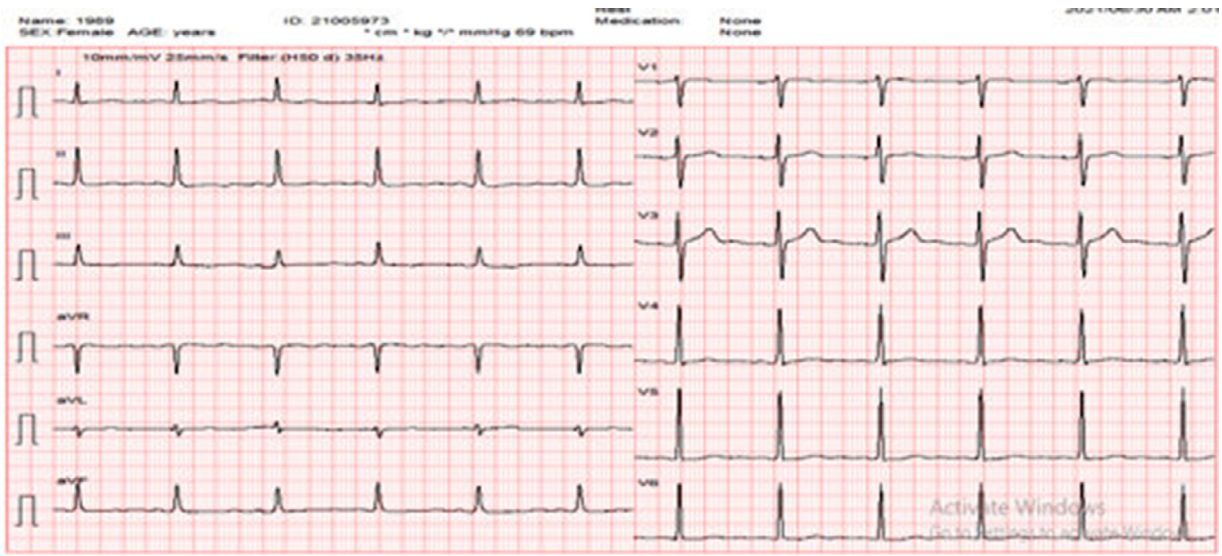

Fig. 7 – ECG after 2 weeks. A sinus rhythm and heart rate of ~80 beats/min are seen.

vention [11]. The American College of Cardiology and American Heart Association guidelines highly recommend interventional management for large CAFs regardless of symptoms and small-to-moderate fistulas with symptoms such as myocardial ischemia, arrhythmia, ventricular dysfunction, and endarteritis [12]. Treatment options include surgical ligation and percutaneous transcatheter closure [12].

Percutaneous transcatheter closure is a noninvasive replacement for treating CAFs. It is indicated in patients whose anatomical fistulae are favorable for the procedure, including a single narrow drainage site, a proximal origin, an absence of multiple fistulas or large branch vessels, and an absence of concomitant cardiac disorders [6]. It is preferred for older patients who are at risk for perioperative complications. Various devices are used in percutaneous transcatheter procedures, including coils, detachable balloons, umbrella intravascular devices, vascular plugs, covered stents, and duct occluders [6].

There are 2 interventional methods. The first is obstructing the proximal fistula using a retrograde arterial approach. The second is obstructing the distal CAF using an antegrade venous approach [12].

In our case, percutaneous closure was performed proximally, near the origin or distal of the drainage site, for proximal CAFs. The postoperative course was uneventful, and no procedures were required. There are currently no post-treatment follow-up guidelines for adult patients with congenital CAFs, and ECG changes and arrhythmia are common during and after transcatheter closure [10]. We prescribed some medicines to prevent thrombus formation and heart failure. After 1 week, our patient felt dizzy and lightheaded. Her blood pressure had decreased, and an ECG showed arrhythmia (junctional bradycardia), but there was no evidence of thrombus formation (troponin I-hs was negative). Therefore, we added theophylline to stimulate the heartbeat and

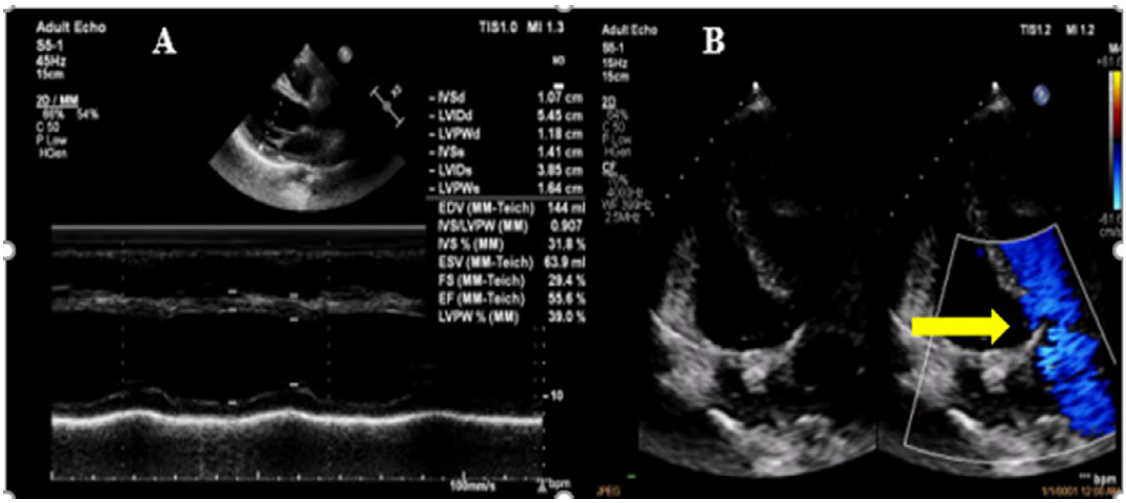

Fig. 8 – Postoperative transthoracic echocardiography. (A) Left ventricle parameters after closure. (B) Mitral valve regurgitation (yellow arrow).

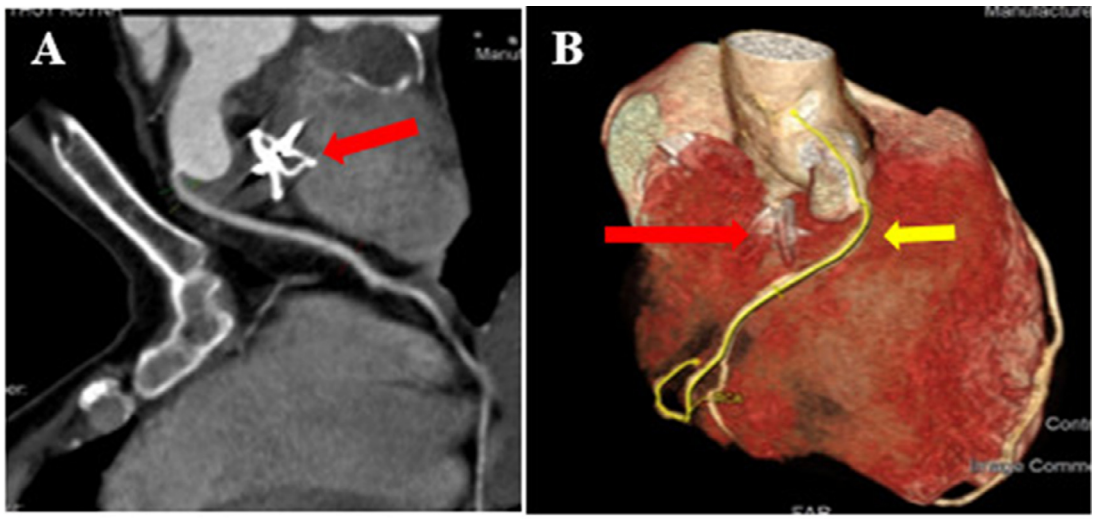

Fig. 9 – Postoperative CTA. (A) Perfect fistula closure by equipment (red arrow). (B) Functional RCA (yellow arrow).

oral anticoagulation therapies (acenocoumarol [4 mg]; ¼ pill per day) to prevent thrombosis. The patient improved, and her ECG became normal, with a sinus rhythm and heart rate of ~80 beats/min.

After 1 year of follow-up, echocardiography showed normal left ventricular function EF and decreased left ventricular dilation. CTA is often used since it is a noninvasive examination for follow-up. While the results vary, residual leakage from CAF recanalization is present in 10% of transcatheter or surgically managed cases [3]. Therefore, we performed a CTA that showed the successful closure of the fistula without any relevant changes to the coronary arteries. Comparing pre- and postprocedure images helps detect postprocedural complications such as recanalization, thrombus, and myocardial ischemia.

With this case report, we would like to emphasize the importance of a collaborative environment for selecting the most appropriate strategy to treat such complex patients. All guidelines identify changes in blood flow characteristics (rate, velocity, and turbulence) caused by fistula closure as a risk factor for intracoronary thrombosis in the aneurysmal segment. Therefore, early-, mid-, and long-term follow-up plans must be prioritized after coronary fistula closure with an aneurismal segment to detect complications such as coronary thrombosis and myocardial ischemia.

Conclusion

CAFs are rare and can have abnormal or variable clinical courses, from no symptoms to severe complications, including heart failure and myocardial infarction, depending on the extent of the shunt. CTA with 3D reconstruction can

accurately assess the CAF's complex anatomy: origin sites, drainage sites, and associated anomalies. Percutaneous transcatheter closure is indicated for patients whose anatomy is favorable for the procedure. However, since post-procedure complications can occur, patients must be closely monitored.

Statement of ethics

Ethical approval was not necessary for the preparation of this article.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Data availability statement

All data generated or analyzed during this study are included in this article and/or its online supplementary material files. Further enquiries can be directed to the corresponding author.

Authors' contribution

Pham Thi Thao Trang, Tran Chi Cuong, and Nguyen Minh Duc contributed to write original draft. Do Nguyen Tin, Nguyen Manh Cuong, and Tran Chi Cuong contributed to undergo percutaneous transcatheter closure procedure, collect and interpret the imaging. Pham Thi Thao Trang, Nguyen Manh Cuong, Nguyen Tran Tran, and Le Minh Thang made substantial contributions to collect patient data and clinical data analysis. All authors have read, revised, and approved the final published version of the manuscript. All authors were responsible for submission of our study for publication.

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