JACC: CASE REPORTS VOL. 3, NO. 5, 2021

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

CASE REPORT: CLINICAL CASE

A Continuous Murmur as the Only Clinical Sign for Complex Coronary Artery Fistulas Diagnosis

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ABSTRACT

An asymptomatic patient presented at our hospital exhibiting a Brugada electrocardiography pattern with coronary artery fistulas. Coronary artery fistula is a congenital or acquired rare abnormal condition with increased symptoms and complications over time. In the absence of the therapeutic consensus, we discuss the association and management for this condition. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:740–4) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A healthy 28-year-old man was referred to our hospital for a cardiac murmur at the left sternal border and a Brugada-like electrocardiography (ECG) pattern after consulting a general practitioner for fatigue and remittent fever. Given the situation of his influenza-positive colleagues and early-onset of fever, we performed a rapid influenza test and fever workup without items for autoimmune diseases. In addition to the negative influenza test result, the workup showed normal results of chest x-ray imaging and uranalysis with very low-grade inflammation (**Table 1**). He was diagnosed as having influenza afterward. The continuous murmur and ECG abnormality remained significant after recovery from influenza. Transthoracic echocardiography (TTE) demonstrated normal left ventricular function and no evidence of infectious endocarditis (IE).

At admission, the patient was asymptomatic, and his blood pressure was 103/73 mm Hg, heart rate was 52 beats/min, and body temperature was 36.6°C. The laboratory blood test results showed no evidence of systemic inflammation and myocardial ischemia (**Table 1**). The continuous murmur of Levine II/VI was still observed (**Figure 1,** Supplemental Figure 1). ECG showed sinus rhythm at 56 beats/min and the Brugada-like ECG pattern (**Figure 2**).

LEARNING OBJECTIVES

- To re-realize the importance of physical examination, including auscultation.
- To consider the treatment strategy for coronary artery fistulas.

MEDICAL HISTORY

There was no medical history, including any chest wall trauma and invasive cardiac procedures.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received May 27, 2020; revised manuscript received February 8, 2021, accepted March 12, 2021.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2021.03.007

TABLE 1 Blood Test Results

	At First Visit	At Admission	Normal Range
White blood cells, per μl	6,400	4,500	3,300–8,600
Hemoglobin, g/dl	15.8	13.5	13.7–16.8
Platelets, $\times 10^4/\mu l$	22.9	23.8	15.8–34.8
Potassium, mmol/L	3.7	4.0	3.6–4.8
C-reactive protein, mg/dl	0.58	0.02	≤0.14
Troponin I, pg/ml	<10.0	<10.0	<30.0
N-terminal pro-brain natriuretic peptide, pg/ml	N/A	52	<400

DIFFERENTIAL DIAGNOSIS

Continuous murmurs are commonly observed in patients with patent ductus arteriosus, ruptured aneurysm of the sinus of Valsalva, and aortic regurgitation with ventricular septal defect. Brugada syndrome (BrS), Brugada phenocopy (BrP) and its underlying conditions are also included to the diagnosis list in view of the Brugada-like ECG pattern (1).

INVESTIGATIONS

Subsequent cardiac computed tomography angiography (CCTA) demonstrated complex fistulas, consistent with the auscultation point of continuous murmur. One of them arose from the proximal left anterior descending artery (LAD) and drained into the left bronchial artery (BA): coronary artery-to-bronchial artery fistula (CBF), accompanied by its tortuous and aneurysmal (5.6-mm-diameter) changes (**Figures 3A and 3B**). Another fistulous communication took off from the left main trunk (LMT) and merged into the CBF (**Figure 3C**). Additionally, the early arterial phase CCTA images identified contrast material in the pulmonary trunk (PT), suggesting a communication with the CBF (**Figure 3D,** Supplemental Figure 2). TTE confirmed an insignificant left-to-right shunt (Q_p/Q_s ratio: 1.0) and no signs of ventricular overload.

No arrhythmias were observed during hospitalization, and a drug challenge test with pilsicainide (1 mg/kg) did not induce the type 1 Brugada ECG pattern.

MANAGEMENT

We performed coronary angiography (CAG) and cardiac single-photon emission computed tomography for further evaluation. No coronary atherosclerotic lesions and hypoplastic left circumflex coronary artery were found on CAG. CAG confirmed a clear communication between the fistulas from the proximal LAD and LMT (**Figure 4**). Exercise myocardial perfusion scintigraphy showed a reverse redistribution in the LAD territory (**Figure 5**). Given the lack of cardiac symptoms and myocardial ischemia in addition to the medium-sized aneurysm (2), this case was treated with a conservative therapy approach after further discussions with cardiac surgeons.

DISCUSSION

Coronary artery fistulas (CAFs) are congenital or acquired abnormal communicative tracts from coronary arteries to cardiac chambers or other vascular structures. The incidence was reported to be 0.002% in the

ABBREVIATIONS AND ACRONYMS

BA = bronchial artery
BrP = Brugada phenocopy
BrS = Brugada syndrome
CAF = coronary artery fistula
CAG = coronary angiography
CBF = coronary artery-to-bronchial artery fistula
CCTA = cardiac computed tomography angiography
ECG = electrocardiography
IE = infectious endocarditis
LAD = left anterior descending artery
LMT = left main trunk
PT = pulmonary trunk
TTE = transthoracic echocardiography

FIGURE 1 A Phonocardiogram at the Left Sternal Border

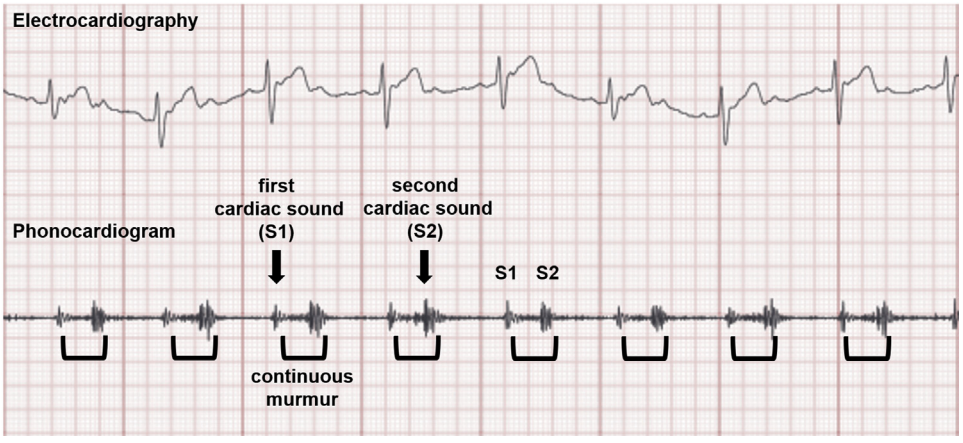
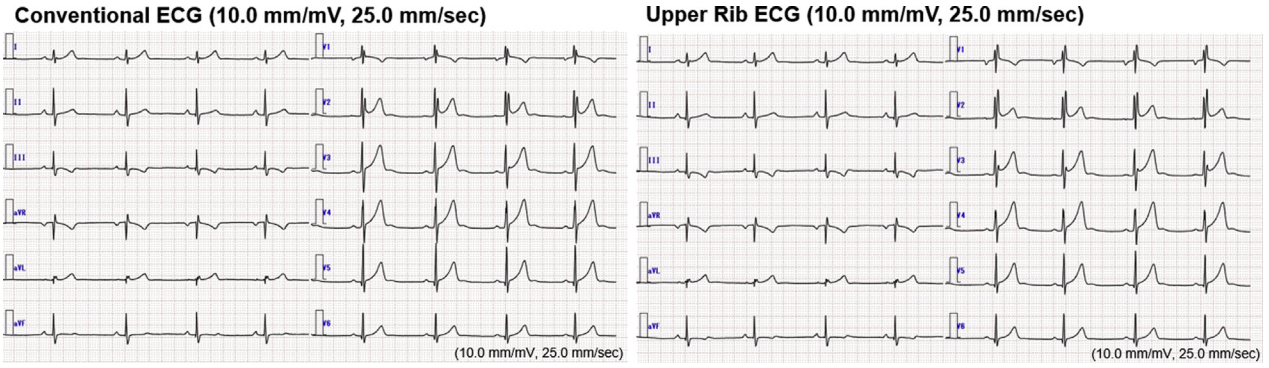
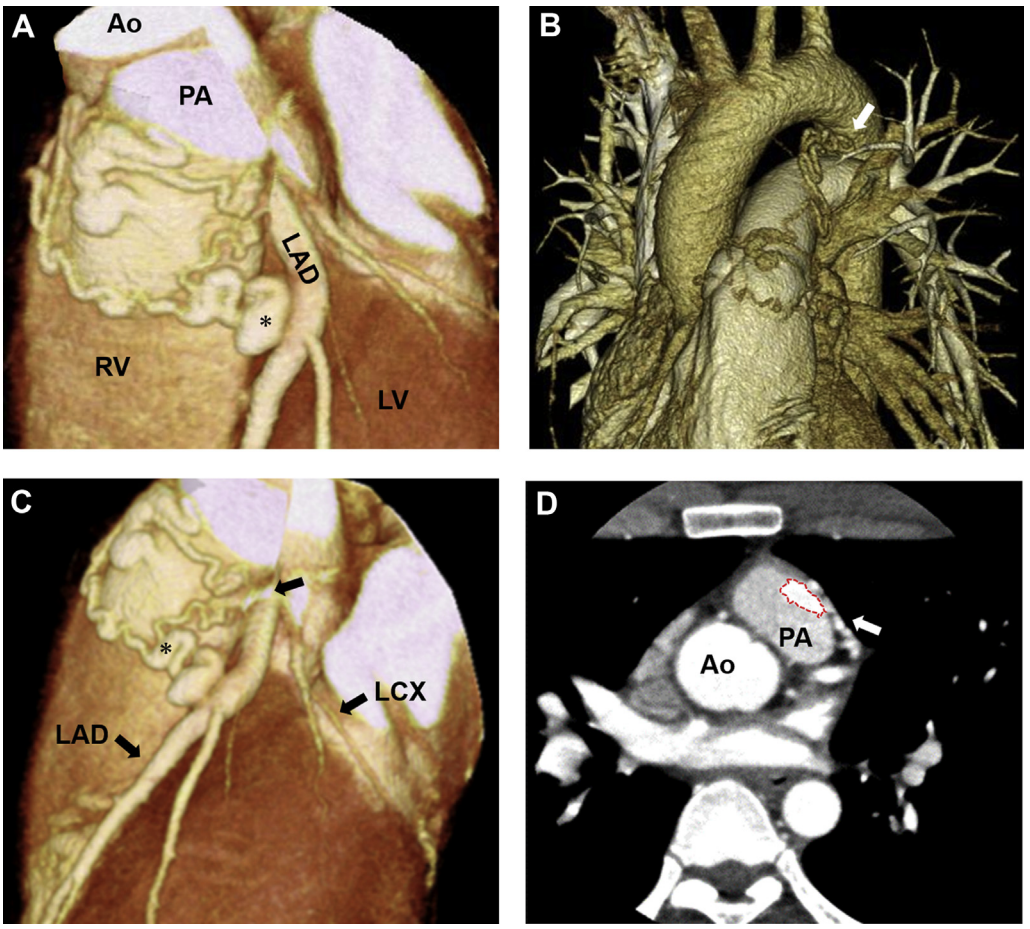


FIGURE 2 Brugada-Like Electrocardiogram Pattern



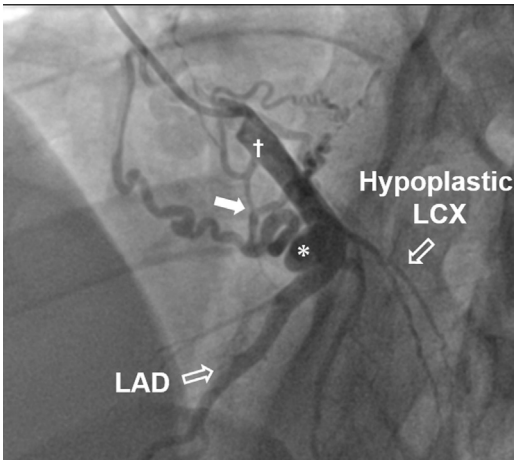
The upper rib electrocardiogram shows the type 2 Brugada-like electrocardiogram (ECG) pattern.

FIGURE 3 Coronary Artery Fistulas



(A) Complex fistulas with the form of tortuous and aneurysmal **(asterisk)** changes. **(B)** Drainage site of the CBF. **(C)** Another communication from the LMT **(arrow)** with the CBF **(asterisk)**. **(D)** Contrast material **(dashed round region)** in the PT from the CBF **(arrow)**. Ao = aorta; CBF = coronary artery-to-bronchial artery fistula; LAD = left anterior descending artery; LCX = left circumflex coronary artery; LMT = left main trunk; LV = left ventricle; PA = pulmonary artery; PT = pulmonary trunk; RV = right ventricle.

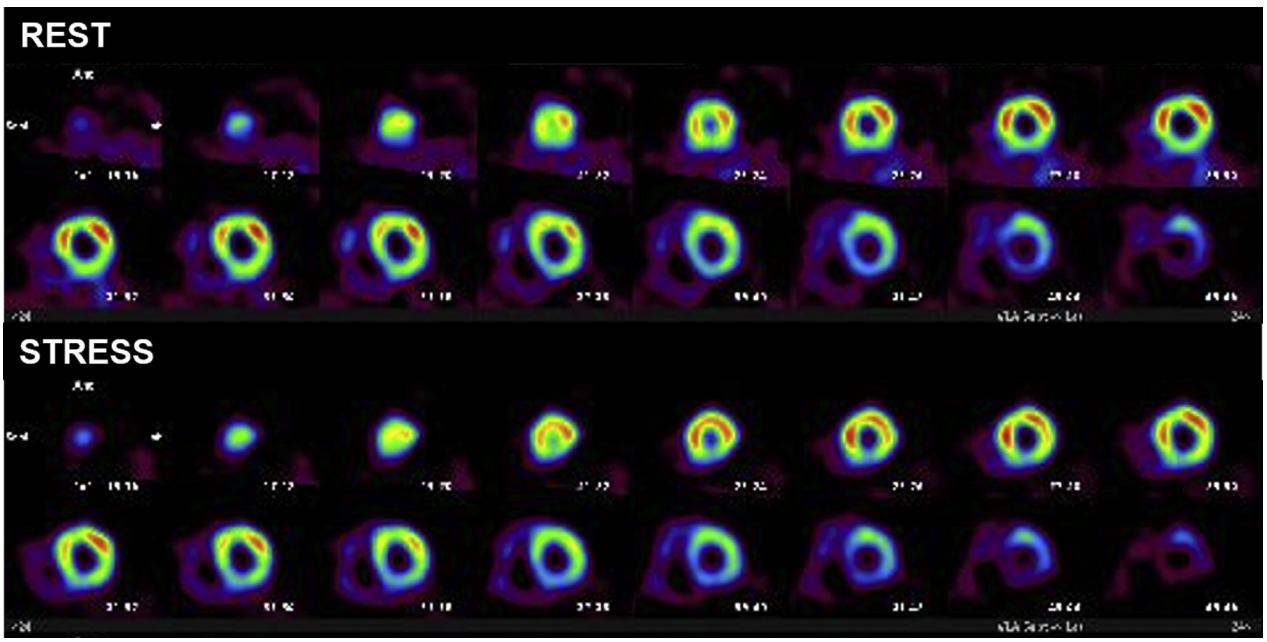
FIGURE 4 A Clear Communication Between the 2 Fistulas



Asterisk indicates aneurysmal change, **dagger** marks a fistula from left main trunk, and **arrows** show the merging point. Abbreviations as in Figure 3.

general population (3). A variety of CAFs are classified by the drainage sites. CBF is a subtype of CAFs with an incidence of 0.1% to 0.6% in patients undergoing CAG/CCTA, originating mainly from the left circumflex coronary artery, unlike the present case. CAFs occasionally have more than 1 origin and drainage sites with complex communications (4). In our case, 2 distinct origins, the LMT and proximal LAD, were identified, and the CBF communicated with the PT in addition to the left BA. CAFs dilate over time with increasing risk of shunt enlargement, myocardial ischemia, arrhythmia, thrombosis, rupture, and IE, although CAFs remain silent in most cases. The management strategies of conservative and invasive therapies have not reached consensus in the 2018 American Heart Association and American College of Cardiology guidelines (5). A prior guideline (6) recommended interventional management for large CAFs regardless of symptom presence or absence and for small to moderate-size CAFs with symptoms. Buccheri et al. (3) advocated that an invasive approach appears to be reasonable in the presence of significant left-to-right shunt, leading to ventricular overload, myocardial hypertrophy, and myocardial ischemia. Additionally, CAF-related aneurysms of ≥30 mm in diameter are considered to be at risk of rupture (7). Furthermore, the latest European Society of Cardiology guideline (8) published in 2020 referred to the indications for invasive approaches: the presence of symptoms, complications, and a significant shunt. These

FIGURE 5 Exercise Myocardial Perfusion Scintigraphy



Reverse redistribution in the left anterior descending territory and respiratory artifacts in the inferior wall were identified.

recommendations could support our conservative approach to an asymptomatic, medium-sized CAF without significant shunt and documented myocardial ischemia. Meanwhile, close monitoring should be mandatory even after the invasive therapy is performed due to post-procedural complications, such as residual leakage, recanalization, persistent dilatation or aneurysmal change, and thrombus formation (4).

BrS presents with a typical type 1 Brugada ECG pattern due to a congenital sodium-channel abnormality, resulting in increasing cardiac arrhythmia and sudden death. The negative provocative challenge test result served to differentiate this case from BrS, suggesting different pathophysiological mechanisms. Our case remained the saddleback type ECG after pyretolysis. BrPs are clinical entities that show the Brugada-like ECG patterns induced by reversible heterogeneous clinical conditions, including mechanical compression of the right ventricular outflow tract (1). The observed CAFs, which run across the epicardial surface of the anterior wall of the right ventricular outflow tract, may be associated with our Brugada-like ECG pattern. Additionally, a case report recently demonstrated a similar association of CAFs with a Brugada-like ECG pattern (9). The similar drainage sites with that of our case, the BA, are of considerable interest.

FOLLOW-UP

The patient was asymptomatic at the 6-month follow-up, and close monitoring is scheduled to detect any signs of the CBF progression. Maintenance of good oral health status and use of prophylactic antibiotics under specific situations should also be implemented to prevent IE.

CONCLUSIONS

In our case, a continuous murmur and a Brugada-like ECG pattern were the only clues to the further investigation, highlighting the importance of auscultation. It is important to take the underlying diseases of BrPs into consideration for any future further evaluation. Our conservative approach requires close monitoring to prevent the development of later complications. Given the limited data on CAFs, future studies are required to establish the appropriate management strategy.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS coronary vessel anomaly, electrocardiogram, murmur

APPENDIX For supplemental figures, please see the online version of this paper.