Cor medusae trinity: Unraveling congenital coronary artery fistula complications through multimodality imaging

Bruno Bragança[1], Rafaela G Lopes[1], Inês Oliveira[1], Isabel Cruz[1], Glória Abreu[1], Adriana Pereira[1], Jorge Casanova[2], João Azevedo[1], Aurora Andrade[1]

[1]Serviço de Cardiologia, Unidade Local de Saúde Tâmega e Sousa (ULSTS), Penafiel, Portugal
[2]Serviço de Cirurgia Cardio-Torácica — Unidade Local de Saúde São João, Porto, Portugal

Correspondence to:
Bruno Bragança, MD, MSc, PhD, Serviço de Cardiologia, Unidade Local de Saúde Tâmega e Sousa (ULSTS), Avenida do Hospital Padre Américo 210, 4564-007 Penafiel, Portugal,
phone: +35 12 557 14 000,
e-mail: bbraganca14@gmail.com

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We report a case of a 62-year-old woman presenting with acute resting dyspnea, palpitations, and chest discomfort. She also reported progressive dyspnea and unintentional weight loss over the last 3 months. Her history included overweight, dyslipidemia, and a positive ischemic stress test demonstrating a small reversible defect in the left ventricle (LV), and dilation of atria and the LV on transthoracic echocardiography (TTE) (Figure 1A–C). Examination revealed a diastolic heart murmur and bi-basal crackles on lung auscultation. B-type natriuretic peptide was 1512 pg/ml, and high-sensitivity troponin I was 543 pg/ml. Electrocardiogram showed atrial fibrillation; chest computed tomography angiography (CTA) ruled out pulmonary embolism or aortic syndrome. TTE revealed severe aortic regurgitation, moderate mitral regurgitation, LV dilation (ejection fraction [EF] 53%), right chamber dilation, and an abnormal flux in a dilated coronary sinus (CS) (Figure 1D–E; Supplementary material, *Video S1–S4*). Transesophageal echocardiogram identified a vegetation (5 × 4 mm) on the right coronary cusp, causing 2 eccentric regurgita-

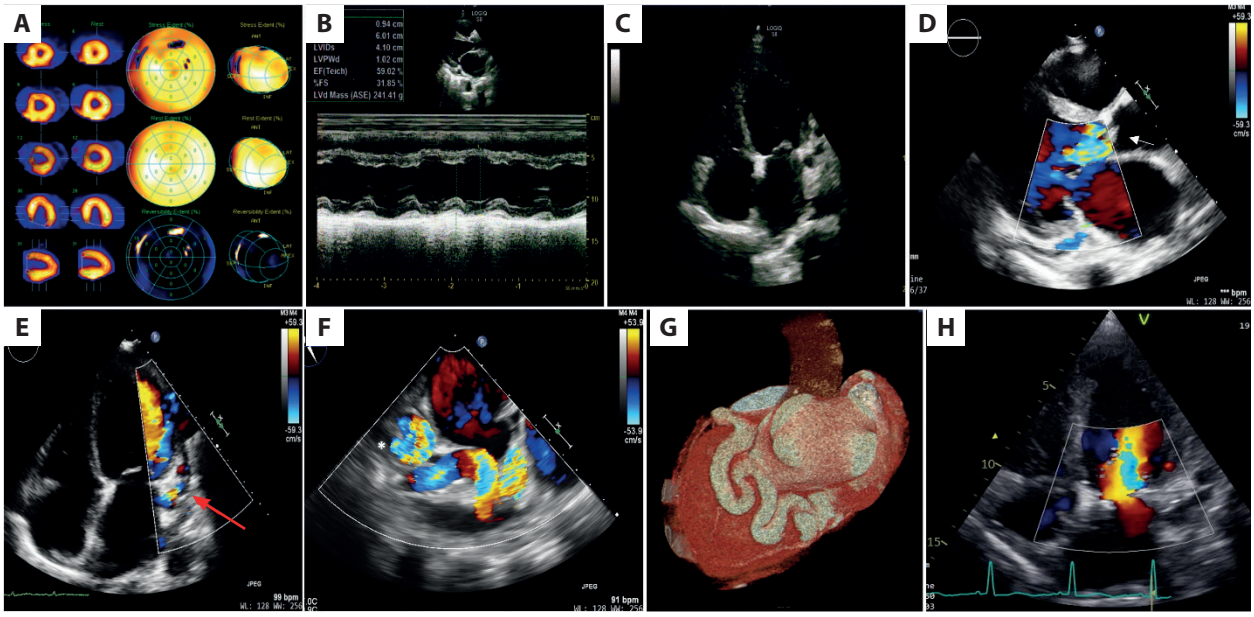

Figure 1. Concomitant complications of a giant congenital coronary artery fistula (CCAF) of the left circumflex coronary artery (LCx). **A.** Myocardial scintigraphy with mild reversible perfusion defect involving the anterior wall of the left ventricle (LV), consistent with a steal phenomenon in the left anterior descending artery, performed prior to infective endocarditis. **B** and **C.** Transthoracic echocardiography (TTE) demonstrating dilation of the atria and LV prior to infective endocarditis. **D.** TTE showing aortic vegetation (white arrow) associated with severe eccentric aortic regurgitation, and the abnormal flux in a dilated coronary sinus (CS; red arrow). **E.** TTE revealing LV dilatation with mild systolic dysfunction, right cardiac chamber enlargement, and an abnormal flow in the posterolateral LV wall. **F.** Transesophageal echocardiography showing a turbulent and tortuous inflow into CS originating from an aneurysmal LCx fistula (asterisk). **G.** Coronary computed tomography angiography showing CCAF of the LCx draining into the CS (14 × 17 mm) and in the right atrium. **H.** Four-month control TTE after surgical repair and medical therapy for HF demonstrating reverse remodeling of the LV and absence of abnormal flux in the posterolateral LV wall

tion jets (right coronary cusp perforation and noncoronary cusp prolapse). An aneurysmal left coronary artery with a turbulent flow to the CS was also noted (Figure 1F; Supplementary material, *Video S5*). Blood cultures identified penicillin-sensitive *Streptococcus mutans*. Coronary computed tomography angiography confirmed a congenital coronary artery fistula (CCAF; 14 × 17 mm) from an aneurysmal left circumflex (LCx) coronary artery draining into the CS and right atrium (Figure 1G). After discussion among the Heart Team, bioprosthetic aortic valve replacement, mitral and tricuspid annuloplasty, and LCx ligation with saphenous vein graft bypass were performed. Postoperative LVEF was 40%, prompting up-titration of medical therapy for heart failure with reduced ejection fraction. At 4 months, LV function improved (Figure 1H; Supplementary material, *Video S6*), and at one year, the patient remained stable with no signs of heart failure.

CCAFs are abnormal connections between coronary arteries and surrounding epithelialized structures due to anomalous epicardial and myocardial development, forming low-resistance pathways that may interfere with coronary hemodynamics [1]. With an estimated prevalence of 0.1% [2], CCAF-related complications such as myocardial ischemia, infective endocarditis (IE), and ventricular dysfunction are exceptionally rare [1], but they more often seen in larger CCAF draining into CS or right cardiac chambers [2]. In this case, LV remodeling was likely driven by volume overload secondary to CCAF and left-to-right shunt [3]. The coexistence of CCAF and IE is increasingly reported, but whether CCAF predisposes to IE or acts as a coincidental finding remains uncertain. *Streptococcus* species was the most frequent isolated agent, with either right- or left-sided valves affected regardless of CCAF anatomy [4]. Closure of CCAFs can be performed *via* percutaneous or surgical techniques. Surgery was chosen due to required intervention in the aortic valve. Surgical closure would still be preferred in isolated CCAF intervention because catheter-based closure of fistulas originating from distal coronary arteries with diameter exceeding 10 mm has a heightened risk of myocardial infarction due to thrombosis caused by stagnant flow in the aneurysmatic bed [1].

This case uniquely illustrates 3 simultaneous CCAF-related complications: myocardial ischemia, ventricular dysfunction, and IE — and it emphasizes the key role of multimodality imaging in diagnosing and managing such rare anomalies.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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