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Anomalous left anterior descending artery to coronary sinus fistula with associated localized ischemia: A clinical dilemma

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

EF **Frank D. Russo**
EF **Homayoun R. Ahmadian**
DEF **Ahmad M. Slim**

Cardiology Service, San Antonio Military Medical Center, San Antonio, TX, U.S.A.

Corresponding Author: Ahmad M. Slim, e-mail: ahmad.m.slim.mil@mail.mil

Patient: Female, 57
Final Diagnosis: Coronary sinus – venous fistula
Symptoms: Dyspnoea
Medication: —
Clinical Procedure: —
Specialty: Cardiology

Objective: Rare disease

Background: Coronary arterial fistula, or arterio-venous malformation (AVM), is a connection between the coronary tree and a cardiac chamber or great vessel, having bypassed the myocardial capillary bed. Known complications from coronary artery fistulas may include “steal” from the adjacent myocardium, resulting in myocardial ischemia.

Case Report: We report the case of a 57-year-old Hispanic woman with abnormal preoperative electrocardiogram (ECG) and symptoms of dyspnea on exertion, who underwent a stress echocardiography demonstrating inferior distribution hypokinesis at peak exercise. Coronary computed tomography angiography (CCTA) demonstrated a venous fistula connecting the coronary sinus (CS) with the distal portion of the left anterior descending artery (LAD), occupying the territory of a left posterior descending artery (L-PDA) and corresponding in distribution with the patient's stress-induced wall motion abnormalities.

Conclusions: Anomalous left anterior descending artery to coronary sinus fistula with associated ischemia is a rare clinical dilemma with limited experience of success with either surgical or medical options.

MeSH Keywords: Arteriovenous Fistula • Left Anterior Descending Coronary Artery Fistula • Computed Tomography • Coronary Sinus

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Background

Coronary arteriovenous fistula has an incidence of 0.1–0.2%, with 90% draining into right-sided structures (the right atrium, the right atrial-superior vena cava junction, or the right ventricle) [1]. Of the 10% of the fistulae draining into the left-sided cardiac structures, 80% drain into the left atrium [2]. Patients are usually asymptomatic, but sometimes can present with angina, congestive heart failure, pulmonary hypertension, endocarditis, spontaneous fistula rupture, or arrhythmia; these clinical presentations are largely dependent on the fistula size, point of origin, and location of anastomosis, resulting in a “steal” phenomenon [3]. Small asymptomatic fistulae require no intervention, but large symptomatic ones should be corrected by either an open surgical or catheter-based approach [4].

Case Report

This is the case of a 57-year-old Hispanic woman with abnormal preoperative electrocardiogram (ECG) results and symptoms of dyspnea on exertion, who underwent a stress echocardiography for cardiac risk stratification for intermediate-risk non-cardiac, orthopedic surgery. Stress echocardiography demonstrated inferior wall hypokinesia at peak exercise with normal augmentation of ejection fraction. The patient was opposed to immediate invasive coronary angiography (ICA), and coronary computed tomography angiography (CCTA) was performed to assess the extent of disease as her medical therapy was being optimized.

CCTA demonstrated a venous fistula connecting the coronary sinus (CS) with the distal portion of the left anterior descending artery (LAD), occupying the territory of a left posterior descending artery (L-PDA) and corresponding in distribution with the patient’s stress-induced wall motion abnormalities (Figures 1 and 2). Her symptoms improved with addition of

Figure 1. Axial and Coronal images with Sequential gating, cardiac phases utilized of 40–80% on CCTA (kVp=120 with care dose variable mAs and slice thickness of 0.7 mm with 0.4 overlap) demonstrating the difference in opacification based on the Hounsfield Units (HU). In image (A) the coronary sinus HU is 309.3±28.3, where the HU is 159.0±24.8 in the right atrium and 379.3±12.8 in the left ventricle as shown in images (B) and (C), respectively. Image (D) highlights the course of the opacified fistula on maximal intensity projection images.

Figure 2. 3D rendered image as well as minimal intensity projection images with Sequential gating, cardiac phases utilized of 40–80% on CCTA (kVp=120 with care dose variable mAs and slice thickness of 0.7 mm with 0.4 overlap) of the course of the fistula connecting the coronary sinus (CS) and the distal left anterior descent (LAD) artery.

a Thiazide diuretic for blood pressure control with continuation of beta-blocker and aspirin therapies (ultimately achieving a BP of 119/74 and HR of 60). She declined invasive angiography to assess for possible surgical intervention and to assess the extent of cardiac shunting unless her symptoms should worsen despite maximally tolerated medical therapy. Ultimately, the patient declined her elective surgical operation. Her symptoms have remained stable on medical therapy.

Discussion

A coronary arterial fistula, or arteriovenous malformation, is a connection between the coronary tree and a cardiac chamber or great vessel, having bypassed the myocardial capillary bed [5,6]. Known complications from coronary artery fistulas may include “steal” from the adjacent myocardium, resulting in myocardial ischemia [7].

CCTA scan was obtained utilizing a 128-slice dual head scanner (Somatom Definition Flash CT®, Siemens, Erlangen, Germany) prospective sequential protocol with 40–80% image acquisition window. The scan was analyzed utilizing Vital® Images reconstruction protocol by a cardiologist with level III SCCT experience. CCTA demonstrated the presence of the CS to LAD fistula with the same level of opacification as the left ventricle, as would be expected in the presence of a cardiac shunt from a high to low pressure system. Thus, during peak exercise, we hypothesize that the increase in myocardial oxygen demand exceeds the coronary flow reserve to the inferior septum, providing a plausible explanation for the hypokinetic segment at peak exercise supplied by the fistula territory; otherwise, it would have been supplied by the congenitally absent L-PDA.

Several treatment options have been described with variable outcomes from coil embolization, occluder device deployment, to surgical ligation [8], depending on the anatomy favoring

different interventions. Our patient was offered cardiac catheterization with possible utilization of percutaneous or surgical options depending on suitability of device deployment during the evaluation, but she declined invasive intervention at this time due to her limited life-style and the success of medical therapy in controlling her symptoms.

Conclusions

Anomalous left anterior descending artery to coronary sinus fistula with associated ischemia is a rare clinical dilemma, with limited success with either surgical or medical options.

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However, our patient has shown clinical success on limited follow-up with guideline-directed maximally tolerated medical therapy aimed at minimizing myocardial demand and thereby minimizing ischemia burden due to cardiac shunting through the fistula.

Statement

The opinions in this manuscript do not constitute endorsement by San Antonio Army Medical Center, the U.S. Army Medical Department, the U.S. Army Office of the Surgeon General, the Department of the Army, Department of Defense, or the U.S. Government of the information contained therein.