JACC: CASE REPORTS VOL. 3, NO. 1, 2021

EDITORIAL COMMENT

Straight Through the Heart

A Rare Cause of Coronary Artery Fistulae*

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Coronary artery fistulae (CAF) are rare clinical findings and challenging to diagnose. CAF are the development of an irregular connection of the coronary artery to any of the 4 chambers of the heart or the connection of a coronary artery to 1 of the great vessels (1). Although CAF are uncommon, accounting for an estimated 0.2% to 0.4% of all cardiac abnormalities, they are often a congenital defect or associated with another congenital heart defect. Pediatric patients are often asymptomatic but can become symptomatic as they grow (2). However, CAF can also be acquired later in life due to infection, cardiac surgery, biopsies, pacemaker implantation, and trauma (2). After orthotopic cardiac transplantation, the incidence increases to approximately 8%, which is likely related to repeated endomyocardial biopsies usually taken from the right ventricular portion of the interventricular septum (3,4). Although CAF can occur in any of the 3 major coronary arteries, most commonly these fistulae are related to the right coronary artery or the left anterior descending artery (right coronary artery in approximately 55% of cases, left coronary artery in 35%, and both in 5%). More than 90% of coronary artery fistulae drain into the low-pressure venous system resulting in a left-to-right shunt (5,6). As imaging modalities have recently improved, along with interventional and surgical advancements, more patients are being identified with this pathology, which can often be overlooked due to its rarity.

In this issue of *JACC: Case Reports*, Rodriguez et al. (7) describe a case in which a patient developed multiple coronary fistulae after receiving multiple penetrating chest wounds. This patient presented with signs and symptoms of heart failure, having previously received a diagnosis of heart failure with preserved ejection fraction and atrial fibrillation (AF). Frequent rehospitalizations due to acute decompensation with fluid overload raised a red flag that a more in-depth clinical workup was needed. Initially an electrocardiogram was performed, showing AF, as previously reported in the patients' medical history, and standard heart failure biomarkers (high-sensitivity troponin I and N-terminal pro-B-type natriuretic peptide concentrations) were evaluated. The first indication of the fistulae was found using transthoracic echocardiography with color Doppler imaging, which showed turbulent flow from the left main coronary trunk. Further investigation by coronary angiography and computed tomography coronary angiography revealed that the patient had a dilated left main trunk, a fistula between the left main trunk and the left atrium, and another fistula between the circumflex and left atrium. Several imaging modalities have been proven to be useful for the management of CAF. Echocardiography can be used safely without ionizing radiation to assess the anatomy of CAF and hemodynamic changes. Coronary angiography used to be the gold standard for assessing CAF due to its high spatial and temporal resolution. However, it is an invasive procedure which can involve procedure-related complications and is often limited in delineation of complex abnormal anatomical connections, with a reported correct diagnosis rate of 35% to 50% (8). Additional information about the exact anatomy, including the origin, course, and drainage site of CAF, are often needed for planning treatment strategies. Cardiac

*Editorials published in *JACC: Case Reports* reflect the views of the authors and do not necessarily represent the views of *JACC: Case Reports* or the American College of Cardiology.

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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.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.12.001

magnetic resonance angiography and computed tomography coronary angiography are useful imaging modalities for collecting this crucial information. Although cardiac magnetic resonance angiography does not involve the use of ionizing radiation or iodinated contrast material, computed tomography angiography requires shorter acquisition times and yields higher temporal and spatial resolution. Furthermore, multiplanar reconstruction with 3-dimensional (3D) volume-rendered imaging yields excellent anatomic information (9). Single-photon emission computerized tomography (SPECT), which is a noninvasive nuclear test, can be used to perform myocardial perfusion imaging (10). SPECT was initially considered in this case to assess the flow distribution of the left ventricle and tissue viability, but because the patient had a history of AF and rapid ventricular flutter, it was not used.

The recurrent signs and symptoms of heart failure were most likely explained by a coronary steal mechanism. Another case report described similar symptoms, which were related to diastolic volume overload and coronary steal phenomenon after heart transplantation and massive fistulation from all major coronary arteries into to the left ventricle (4). In the current case report, the patient underwent surgery and had successful ligation and revascularization of the coronary defects. This patient's pathology was hypothesized to have developed after a coronary laceration secondary to thoracoabdominal stab wounds and was unique in the presentation affecting the circumflex and the left atrium. Surgical options for patients with CAF vary based on clinical presentation. Recent advancements in minimally invasive surgery have made this an option, especially for treating pediatric patients (11). Transcatheter closure is also an option for patients, especially those who are older or have other comorbidities that could negatively impact their surgical outcome (11,12). In very rare cases, spontaneous closure of the CAF has been reported (1% to 2% of cases) (13).

CONCLUSIONS

Rodriguez et al. (7) provided a thorough report of how they built an imaging plan specific to this patient and highlighted how the clinical course of action was designed to exclusively fit this patient. The presentation of CAF was not the obvious cause of cardiac dysfunction in this patient, but careful dissection of the clinical history and thorough investigation allowed for successful diagnosis and treatment. This is an example of personalized medicine at the most basic definition for a rare pathology with a unique presentation or abnormality in each patient.

AUTHOR DISCLOSURES

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

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KEY WORDS chronic heart failure, coronary circulation, coronary vessel anomaly, diastolic heart failure