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Case Report

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A Rare Case of Left Circumflex Coronary Artery Fistula to Coronary Sinus

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Abstract

Arteriovenous coronary malformations are rare in the adult patient population with challenging diagnosis and management. This is a complex case of a left circumflex artery to coronary sinus fistula in a patient with history of bioprosthetic aortic valve replacement presenting with new onset of shortness of breath.

Keywords

Fistula, Coronary Sinus, Bio Prosthetic Valve, Shunting

Abbreviations

AF - Atrial Fibrillation; AR - Aortic Regurgitation; BP - Brain Natriuretic Peptide; CAF - Coronary Arteriovenous Fistula; CCTA - Cardiac Computed Tomography with Arteriography; CS - Coronary Sinus; EKG - Electrocardiogram; LCx - Left Circumflex Artery; LV - Left Ventricle; PA - Pulmonary Artery; RA - Right Atrium; RV - Right Ventricle; RCA - Right Coronary Artery; TTE - Transthoracic Echocardiography

Introduction

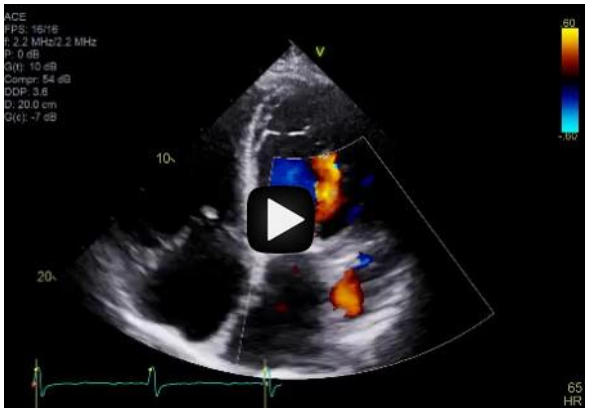
A 67-year-old man with a history of atrial fibrillation and hypertension presents with subacute dyspnea on exertion and palpitations that started 6 weeks prior to presentation.

Patient underwent aortic valve replacement with a 29 Carpenter-Edwards bioprosthetic valve and mitral valve repair 8 years prior due to endocarditis. Vital signs were normal and his physical exam was notable for a 2/6 diastolic murmur on the left sternal border. EKG showed sinus rhythm with regular rate, and laboratory data was notable for normal serial high sensitivity troponins and an elevated brain natriuretic peptide of 578pg/ml. Transthoracic echocardiography (TTE) showed an ejection fraction of 55%, left ventricular (LV) end diastolic dimension of 6.9cm, right ventricular (RV) and atrial (RA) enlargement, left atrial (LA) volume index of 68ml/m2, normal appearing bioprosthetic valve with moderate anteriorly directed eccentric aortic regurgitation jet, pressure half-time of 400msec and Vmax of 2m/s. Notably, a dilated coronary sinus (CS) and circumflex (LCx) artery were observed on TTE with turbulent flows (**Video-1**, **Video-2** and **Video-3**).

Medical History

Patient reported a history of coronary arteriovenous malformation diagnosed at the time of his aortic valve replacement and mitral valve repair secondary to endocarditis. Upon further review of the records, a left heart catheterization reported an aneurysmal dilatation of the LCx and possible fistula to the CS.

Further work up or interventions were not performed at the time. Patient was subsequently lost to follow up.

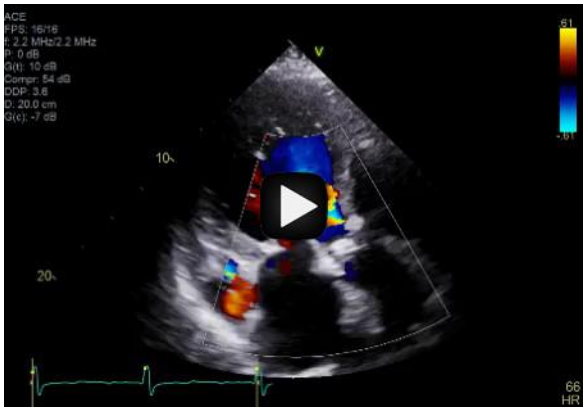


To watch the complete Video-1 online, click on the image
Video-1:

Transthoracic Echocardiogram, Apical 5 chamber view with Color Doppler. Enlarged coronary sinus with turbulent flow

To watch the complete Video-2 online, click on the image
Video-2:

Transthoracic Echocardiography, Parasternal Long Axis View, Increased Left Ventricular end end diastolic dimension of 6.9cm, Right ventricular, right atrial and left atrial enlargement (left atrial volume index of 68ml/m)



To watch the complete Video-3 online, click on the image
Video-3:

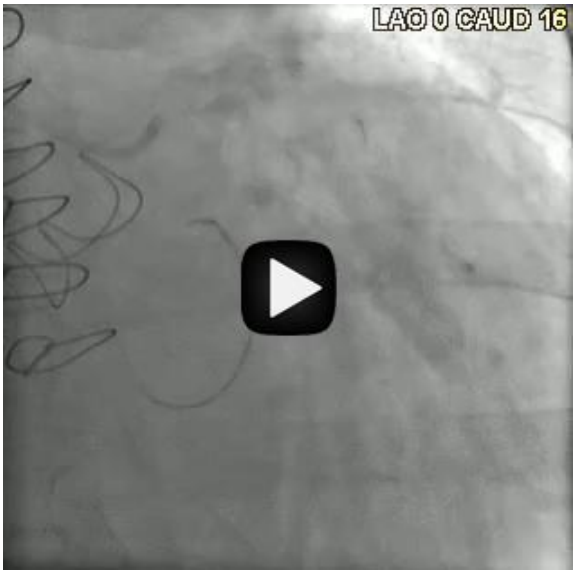
Transthoracic Echocardiography, Apical 3 Chamber View, with Color Doppler, Normal appearing bioprosthetic valve with moderate anteriorly directed eccentric aortic regurgitation jet, left atrial and coronary sinus enlargement with turbulent flow in the coronary sinus

Differential Diagnosis

The differential diagnosis included worsening aortic bioprosthetic valve regurgitation vs high output heart failure due to coronary arteriovenous fistula (CAF).

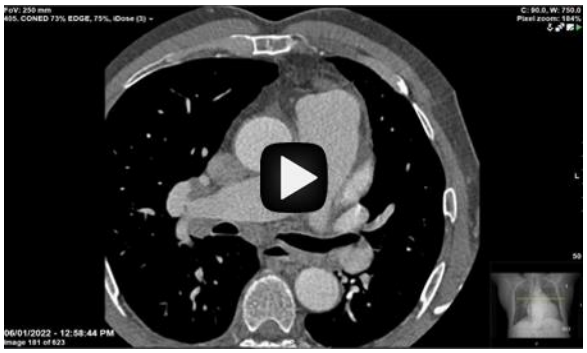
Investigations

A right heart catheterization was performed. Pressures were: RA 7mmHg, RV 41/7 mmHg, pulmonary artery (PA) 39/13 mmHg, mean 23mmHg, wedge 16 mmHg. There was oxygen saturation step up in the PA compared to superior vena cava (SVC) of 19% (53% to 72%); the site of step up was the RA and the Qp:Qs ratio was 2:1, which was consistent with a significant left to right (L-R) shunt. Left heart catheterization and coronary arteriography showed severely dilated and tortuous left main coronary artery and LCx, measuring up to 15.6mm with immediate opacification of coronary sinus confirming an extracardiac shunt (**Fig-1** and **Video-4**). Cardiac computed tomography with arteriography (CCTA) showed a 2cm LCx aneurysmal dilation with fistula to the CS (**Fig-2**, **Fig-3** and **Video-5**). Patient underwent surgical closure of the fistula, excision of the aneurysmal portion of the circumflex artery, aortovenous grafting to a single branch of the circumflex artery as well as bioprosthetic valve replacement.



To watch the complete Video-4 online, click on the image
Video-4:

Coronary Angiography, Left Anterior Oblique and Caudal View, Prior Sternotomy Wires with Bioprosthetic Aortic Valve and Tricuspid Annuloplasty Ring, Left Main Coronary artery engagement and contrast injection showing a dilated and tortuous Left Circumflex Artery with delayed opacification of the coronary sinus



To watch the complete Video-5 online, click on the image
Video-5:

Coronary Angiography, Left Anterior Oblique and Caudal View, Prior Sternotomy Wires with Bioprosthetic Aortic Valve and Tricuspid Annuloplasty Ring, Left Main Coronary artery engagement and contrast injection showing a dilated and tortuous Left Circumflex Artery with delayed opacification of the coronary sinus

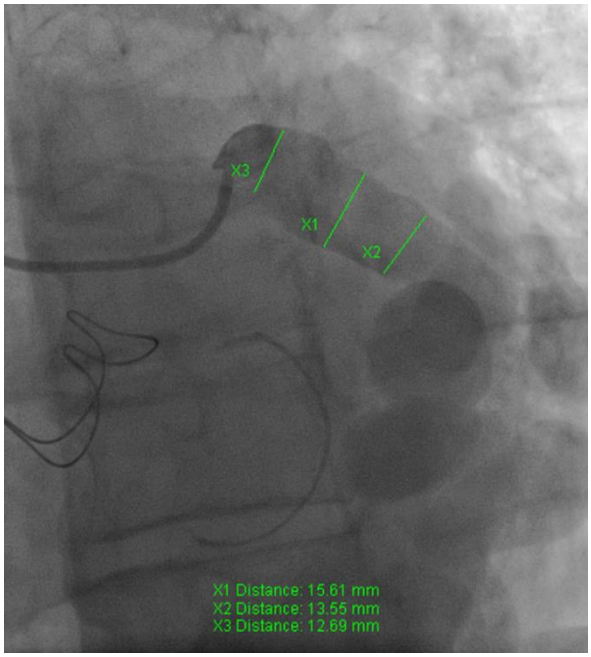


Fig-1: Left Heart Catheterization

LAO 22 CRAN 3 view, radial approach, 6 tip catheter, dilated left main (x1) and left circumflex arteries

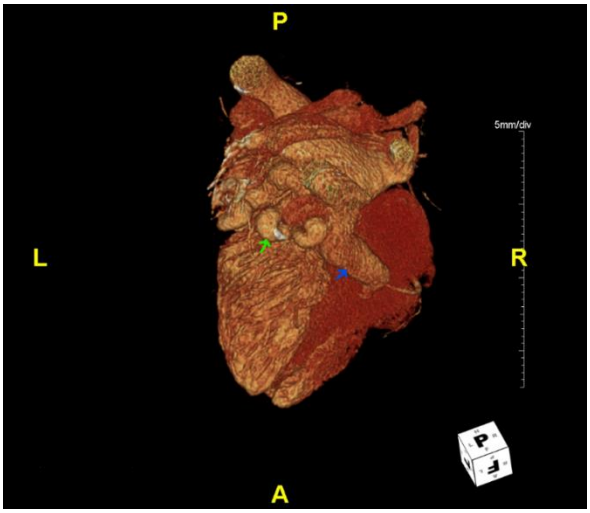


Fig-2: Cardiac CTA 3D Model Reconstraction

Left inferoposterior view showing dilated coronar sinus as well as dilated and tortuous left circumflex and great cardiac/oblique left atrial veins.

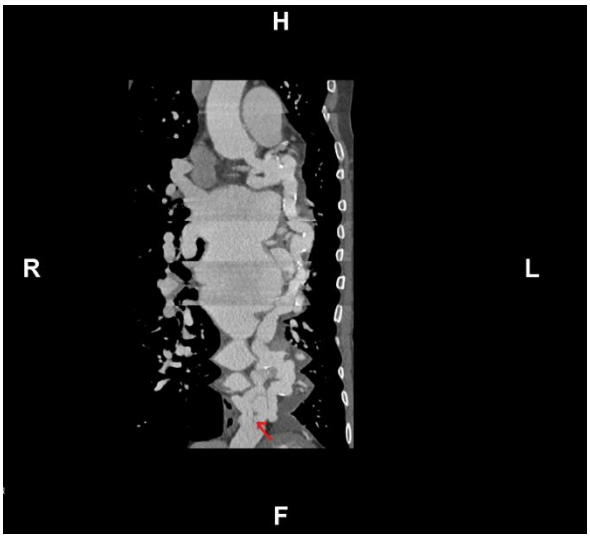


Fig-3: Cardiac CTA

Left Circumflex artery reconstruction. Dilated and tortuous with demonstration of fistula at the level of coronary sinus

Discussion

We present a case of a CAF between the left circumflex coronary artery and the CS. It is a rare entity with the overall prevalence of CAF in the general population reported to 0.9% [1]. Most of the patients are asymptomatic with a small proportion presenting with endocarditis, arrhythmias, heart failure symptoms and chest pain [2]. In our case the presence of moderate AR confounded the presentation urging further work up with a right heart catheterization. CAFs can be classified according to their origin and drainage site to coronary-cameral fistulas (connections between coronary arteries

and any of the heart chambers) or coronary arteriovenous malformations (connections that occur between coronary arteries and parts of the systemic/pulmonary vessels) [3]. They can originate from each of the main coronary arteries; right coronary artery (RCA) 50-55%, LAD 35-40%, and LCx 5-20% of reported cases [4]. The most common drainage sites are the pulmonary trunk (89%), right ventricle (41%), right atrium (26%), coronary sinus (7%) and the left ventricle/atrium (5%/3%) [4]. CAFs can be congenital, traumatic or iatrogenic [3].

In our case, the documentation of CAF prior to his first operation classifies it as congenital rather than iatrogenic. The gold standard for the diagnosis of CAFs is coronary angiography. Nonetheless, extreme aneurysmal dilation of the coronary vessel does not allow for proper opacification and identification of the fistula. Coronary computed tomography with angiography offers the advantage of more accurate anatomical and functional assessments. These patients should be evaluated by experts to determine the role of medical, percutaneous or surgical closure [5]. Fistula size, presence of significant shunt or symptoms are some of the closure indications [6,7]. Percutaneous closure is relatively safe, however, in older patients with tortuous and distal CAFs, there is a higher risk of post-operative thrombosis resulting in procedural myocardial infraction [6]. Medical therapy with aspirin and/or anticoagulation has not been vigorously studied, thus their use should be chosen on a case-by-case basis [6]. While surgery was initial closure strategy for CAF patients, it was later found to have high rates of fistula recurrence [3]. In the present case, surgical closure was chosen because of a degenerating bio-prosthetic valve, presence of a significant CAF and surgical expertise at our institution.

Follow up:

The patient had an uneventful postoperative course, his symptoms improved and was discharged to cardiac rehab. His follow up echocardiogram at 5 months post-operatively showed significant improvement of his LV dilation.

Conclusion

CAFs are rare with various and insidious presentations and their diagnosis requires a high level of suspicion in the presence of other ongoing processes. Coronary CT angiography is the best imaging modality for diagnosis and characterization. Referral to a center with high level of expertise in percutaneous coronary embolization or surgical closure of these lesions is highly recommended.

Learning Objectives

1. CCTA is a powerful non-invasive diagnostic tool for coronary fistulas that is extremely helpful to the surgical team.
2. Closure of a coronary fistula is recommended in cases of large dimensions/aneurism, relevant left–right shunt, or ischemic events.

Informed Consent

Patient gave full permission for the publication and other use of audio-visual material and textual material in all editions of the above-named product.

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None

Conflict of Interest

The authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

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