An Asymptomatic Case of a Right Coronary Artery Aneurysm With Right Atrial Fistula

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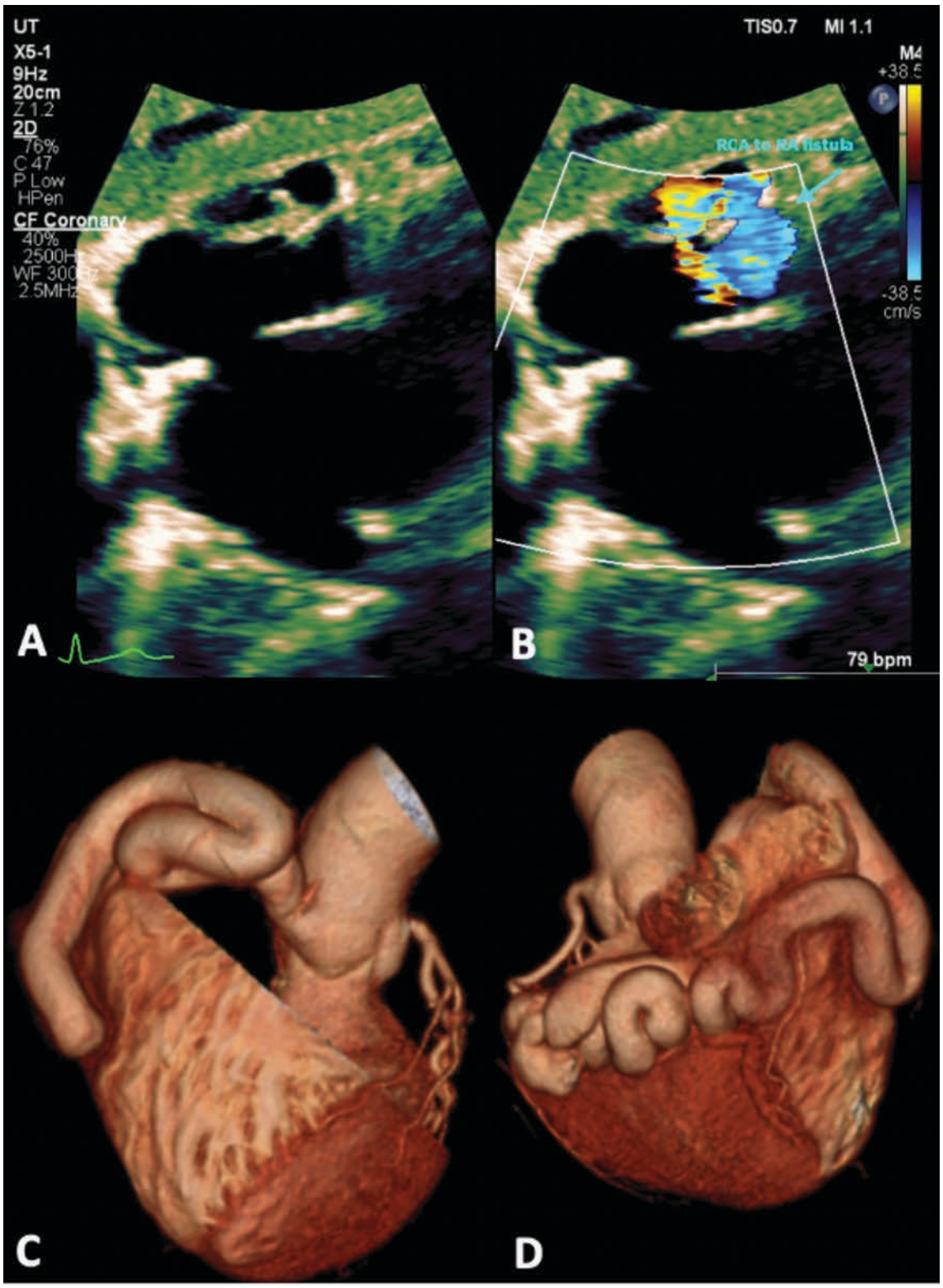

Figure 1. **(A) Subcostal view of right coronary artery (RCA) to right atrial (RA) fistula. (B) Doppler overview of subcostal view revealing fistula flow pattern. (C) 3D image showing significate dilatation of the RCA off the right aortic root. (D) 3D image showing significant RCA and coronary sinus dilatation in the transverse pericardial sinus.**

Coronary artery aneurysms are defined as a localized luminal dilation measuring at least 1.3 to 2 times the diameter of an adjacent, normal vessel segment.[1] If the coronary artery aneurysm is large enough, it can be classified as a giant coronary artery aneurysm. Different definitions of giant coronary artery aneurysms have been proposed, as aneurysms exceeding a diameter of 20, 40, or 50 mm; a definite diagnostic diameter for giant coronary artery aneurysm has, however, not been established.[2-6]

The most common etiology associated with coronary artery aneurysms in the adult population is atherosclerosis.[1,2] Some researchers have found coronary artery aneurym linked to cardiac risk factors such as hypertension, lipid abnormalities, smoking history, and diabetes, and other studies have found systemic hypertension as the largest risk factor.[4] Other etiologies include autoimmune (systemic lupus erythematosus, Kawasaki disease), infectious (syphilis), illict drug use (cocaine), connective tissue diseases (Marfan syndrome, Ehlers-Danlos syndrome), congenital birth defects, trauma, and iatrogenic.[3] These etiologies are also geographically variable. In western countries, atherosclerosis is the most common cause of coronary artery aneurysm at 50%, with congenital (17%) and infectious causes (10%) being second and third, respectively.[1] The leading hypothesis for these three risk factors is an underlying destruction or inflammation process that occurs at the level of atrial media. This destruction or inflammation leads to thinning of the atrial wall and increased wall stress, believed to promote the development of coronary artery aneurysms.[5]

Case Presentation

A 69-year-old Caucasian female presented to her primary care physician for a routine wellness visit. There were no complaints at this visit. A 3/6 holosystolic murmur best auscultated in the 2nd intercostal space at the right sternal border was appreciated. The rest of her exam was unremarkable including signs of cyanosis. She denied any chest pain, shortness of breath, paroxysmal nocturnal dyspnea, and neurological symptoms. This prompted a follow-up electrocardiogram and stress test. The exercise stress test revealed a low risk for significant cardiovascular event and no suspicious reversible defects were seen. An estimated ejection fraction of 63% was noted.

The patient had a past medical history of osteopenia and gastroesophageal reflux disease that was well controlled with omeprazole. She did not drink alcohol or smoke. She did not use illicit drugs and had no family history of cardiac disease.

The initial differential diagnosis of a holosystolic murmur in an otherwise healthy middle-aged female commonly includes valvular heart disease, atrial septal defect, ventricular septal defect, and functional murmur, all causes further identified with a follow-up echocardiogram.

A right coronary artery aneurysm with right atrial fistula is an exceedingly rare condition and can present with a wide range of symptoms.

The echocardiogram results revealed an intact right coronary cusp and a normal ejection fraction, estimated to be 60%-65%. Mild tricuspid regurgitation was noted. Most notably, a right coronary aneurysm was appreciated, which communicated with the right atrium through a fistula (Figure 1A-D). A dilated coronary sinus was also noted.

Computed tomography angiography (CTA) of the coronary arteries revealed a stable/moderate tortious right coronary to right atrial fistula with a maximum transverse dimension of 15 mm distal to the ostia. A right heart catheter revealed a pulmonary to systemic flow ratio (Qp:Qs) of 1.4, signifying moderate shunting between the right coronary artery and the right atrium.

The patient continued regular office visits and echocardiograms to monitor for heart failure symptomatology, which would develop from progression of increased fistula flow. She was also prescribed aspirin for her presumed atherosclerosis that had led to the coronary artery aneurysm. Thus far, the patient has had no further adverse effects from her underlying right coronary artery aneurysm with the associated right atrial fistula.

Discussion

While the etiologies of coronary artery aneurysms are atherosclerosis, autoimmune, infectious, congenital birth defects, and trauma, our patient seemed to fall in the most common subgroup, atherosclerosis; however, inflammation was not fully ruled out in this case. Coronary artery aneurysms have more recently begun to be reported as a result from percutaneous interventions, including stenting, atherectomy, and coronary artery angioplasty.[5] Predisposing factors are believed to be use of oversized stents or balloons, certain techniques, and laser angioplasty.[5] The reported occurrence rate of an aneurysm is 10% post direct atherectomy, 3.9% post-percutaneous coronary angioplasty, and 3.5%-5% post stent placement.[6]

A wide array of imaging techniques have been used to view coronary artery aneurysms. Noninvasive options include echocardiography, CT, and magnetic resonance imaging.[7] If the aneurysm is large enough, it has been visualized on chest radiography.[8] In terms of defining size, shape, type, and location, coronary angiography continues to be the gold standard.[7]

Treatment remains controversial. Surgical correction is currently the generally accepted treatment for giant coronary artery aneurysms. These surgical corrections include isolated coronary artery bypass grafting, aneurysm plication, aneurysm ligation with distal bypass grafting, and saphenous vein patch repair of the aneurysms.[8] These surgical approaches require median sternotomy, cardiopulmonary bypass, and myocardial revascularization. Therefore, certain patients will require other options that include percutaneous intervention and medical management. Dependent variables for surgical corrections include the size of the aneurysm and accompanying fistula if present, risk of rupture during surgery, and concomitant coronary artery disease.[1]

Some papers support medical management with the use of antiplatelet or antithromobotic treatment for all large aneurysms, in the absence of surgical correction.[1] There has been little study in terms of success of medical management and decreased risk of thrombotic events. Despite the lack of evidence, warfarin has continued to be the drug of choice for anticoagulation in patients that are unlikely surgical candidates.

Percutaneous options continue to be the newest development for treatment without the risks that are involved with the surgical approach of sternotomy and cardiopulmonary bypass. Percutaneous options include stent placement with and without coil embolization. There have been cases of polytetrafluoroethylene (PTFE)-covered stents that have been successful.[9] The less invasive percutaneous approach is hypothesized to be equipped for dealing with aneurysms greater than 5mm and less than 10 mm.[9] Further development on techniques and stent technology may lead to increased future success of this approach.

The prognosis remains controversial. The clinical sequelae include thrombus formation and embolization of said thrombi, fistula formation (as is represented in our case), and rupture.[7] The most common sequela is thrombus formation due to the laminar blood flow that evolves from the dilatation and leads to increased coagulation within the aneurysm.[1] It has been reported that 25% of giant coronary artery aneurysms have an associated fistula.[3] Rupture of the aneurysm continues to be rare and unpredictable. The 5-year survival rate with a coronary artery aneurysm was calculated to be 71%.[10] There has been no association in 5-year survival in patients with a coronary aneurysm and its size.[10] There has also been no reported difference in 5-year survival of patients with an aneurysm either with or without obstructive coronary artery disease.[10]

Conclusion

An RCA aneurysm with RA fistula is an exceedingly rare condition and can present with a wide range of symptoms. Management is typically surgical and is influenced by a multitude of factors. Treatment is generally surgical, but in asymptomatic patients, treatment remains unclear. ■

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