Case Report

A Rare Combination of Giant Right Coronary Artery Aneurysm

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Abstract

Context: Giant coronary artery aneurysm (CAA) in adults is a rare clinical entity with an estimated incidence of 0.02%. CAA is commonly found in the right coronary artery with significant number of cases associated with fistula formation. **Case Report:** We describe a rare case of an 87 year-old man with large CAA with fistulous drainage into the right ventricle (RV) along with RV free wall vegetation as a cause of chronic weakness and lethargy. **Conclusion:** Giant CAA with fistulous drainage to the RV could present in the form of infective endocarditis. Early detection and surgical treatment would provide a significant benefit to these patients.

Keywords: Coronary artery fistula, Coronary artery aneurysm, Giant coronary artery aneurysm, Infective endocarditis, Right ventricular fistula

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Introduction

Coronary artery aneurysm (CAA) is an uncommon disease with a predilection of the right coronary artery. The prognosis depends on size of the aneurysm with the highest morbidity and mortality risk when the internal diameter is >8 mm. The most common cause is atherosclerosis followed by Kawasaki's disease and congenital aneurysms. We describe a rare case of giant CAA with right ventricle (RV) fistulous drainage, which was also found to have RV free wall vegetation and responded to intravenous antibiotics.

Case Presentation

An 87-year-old Caucasian male with past medical history of pulmonary embolism, purulent pericardial effusion, and post-pericardial window presented to the emergency department complaining of weakness, and lethargy for 1 month. On presentation temperature was 101 F, blood pressure 116/65 mmHg, pulse 110 beats/min, and respiration 24 breaths/min. Examination revealed bilateral (B/L) diminished breath sounds, B/L basal capitations, normal heart sounds, and no murmur or gallop.

Laboratory data showed white blood cell (WBC) count 18.8 cells/μL with left shift, brain natriuretic peptide (BNP) 1,040 ng/L, and troponin-I 2.26 μg/L. Electrocardiogram (EKG) did not show significant ST-T wave changes. Chest X-ray showed severe cardiomegaly [Figure 1]. Computed tomography (CT) of the chest revealed an unusual soft tissue mass anterior to the RV [Figure 2]. Transthoracic

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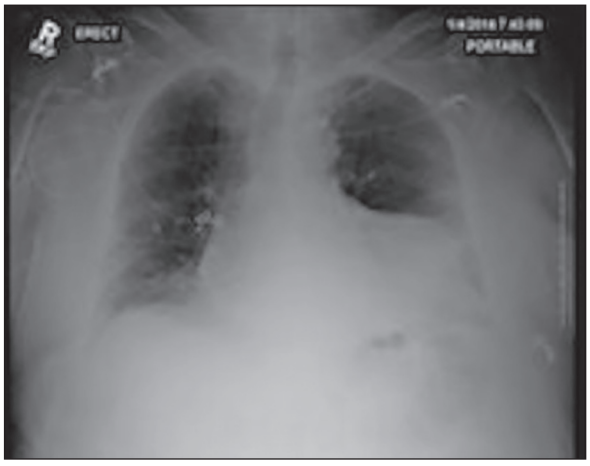

Figure 1: X-Ray chest consistent with severe cardiomegaly

echocardiogram revealed ejection fraction of 45%, moderate RV enlargement, moderate RV hypokinesis, and a large echo dense mass anterior to the RV compressing the RV free wall. Magnetic resonance imaging (MRI) of the chest showed a T2-signal intensity lobulated mass in the anterior epicardial space. Transesophageal echocardiogram showed a large (4 cm) complex structure exterior to the RV, consistent with right CAA. Color flow signal was noted near the RV outflow track consistent with fistulous drainage from the CAA. At the point of fistulous drainage there was large (18 × 12 mm) vegetation attached to the RV free wall [Figure 3]. The patient underwent left heart catheterization that showed a large CAA with possible fistulous drainage into the RV and triple vessel disease [Figure 4]. Surgical treatment was advised for exclusion of the CAA, closure of the fistula, excision of the vegetation, and coronary artery bypass grafting; but the patient refused surgical intervention. Subsequently, blood cultures revealed methicillin sensitive *Staphylococcus aureus*. Patient's overall condition improved with medical management and he was discharged to the skilled nursing facility with intravenous antibiotics and standard anti-ischemic medications.

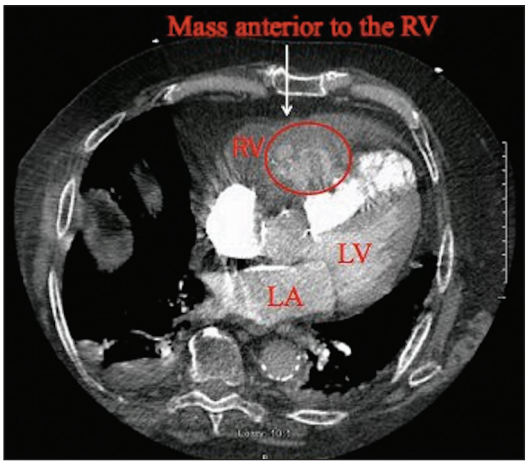

Figure 2: Computed tomography of the chest showing a soft tissue mass anterior to the RV. RV = Right ventricle, LV = Left ventricle, LA = Left atrium

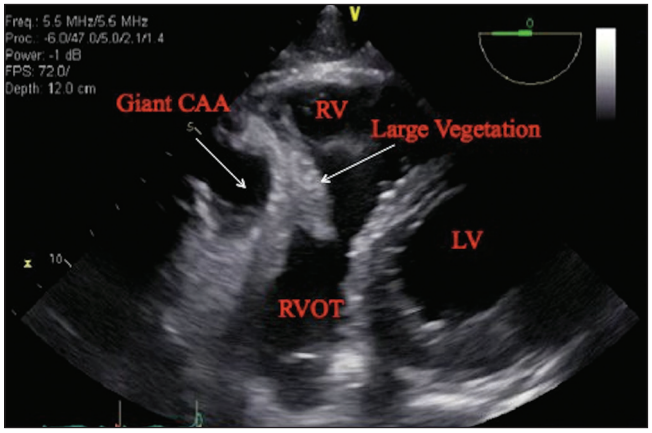

Figure 3: Transesophageal echocardiogram: A giant CAA is noted exterior to the RV (arrow). Large vegetation attached to the RV free wall is also seen (arrow). CAA = Coronary artery aneurysm, RVOT = Right ventricular outflow track

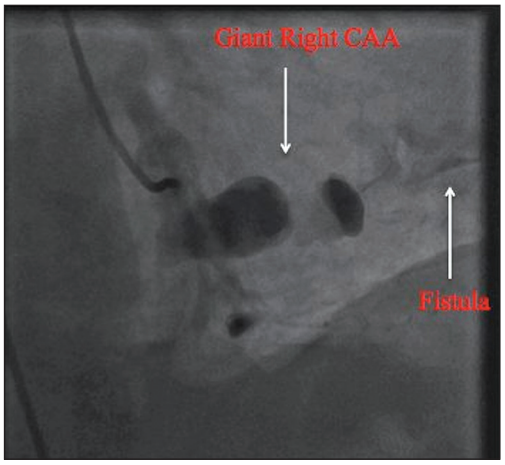

Figure 4: Left heart catheterization showing a giant right CAA along with fistula formation (arrows)

Discussion

CAA is defined as a coronary artery dilation that exceeds the diameter of normal adjacent segments of coronary artery or 1.5 times greater the diameter of the largest coronary artery.[1] Estimated incidence of CAA is 0.3-5% among patients undergoing coronary artery angiography.[1] Although exact definition of the "giant" CAA is still lacking, it generally refers to a dilatation that is greater than four times of the diameter of reference vessel, or has a diameter exceeding 8 mm.[2] In fact, diameter varies from 50 to 150 mm in the reported cases of giant CAAs in adults.[2]

In adults giant CAA's are extremely rare entities with an estimated incidence of 0.02%in the atherosclerotic cases.[2] In infants and children, Kawasaki's disease is the most common cause of giant CAA.[3] The combination of giant CAA, fistulous drainage to the RV, and further complicated with infective endocarditis as described in the index case is extremely rare.

Non-atherosclerotic causes of giant CAA include connective tissue disorders, vasculitis, infections, drug abuse, and trauma.[4] Right coronary artery is reported to be the most common location of CAA's.[5] A 15-20% of the cases of CAA's are also associated with fistula formation.[5]

Although CAA generally remains asymptomatic, it may present with unstable angina, myocardial infarction, congestive heart failure, and even sudden cardiac death.[6] Aneurysmal size is considered to be the most important predictor of myocardial infarction.[6] Giant CAA's may present as a superior vena cava syndrome,

intracardiac mass or extracardiac mass with recurrent hemoptysis due to local compression of the bronchial tree.[7] In patients with familial hypercholesterolemia, giant CAA has also been associated with spontaneous coronary artery dissection.[8]

Cardiac catheterization remains the gold standard diagnostic tool for detection of CAA: Provides information about the shape, size, location, and coexisting coronary artery disease or coronary artery fistula; and is also helpful for planning the subsequent management.[9] Noninvasive modalities include echocardiography, coronary computed tomographic angiography; which is particularly helpful in detection of coexisting fistula and cardiac MRI.[10]

Management of the CAA is variable according to the etiology, size, symptoms, and associated coronary artery disease or coronary artery anomalies. In literature, medical treatment, stent implantation, and surgical treatment have been described.[11] Generally small, asymptomatic aneurysms without associated coronary artery disease or anomalies are managed with conservative treatment, which consists of anticoagulant and antiplatelet drugs to avoid thromboembolic events.[11] Giant CAA's, especially with symptoms are usually managed with surgical exclusion using a resection or ligation.[12] Closure of the fistula is also mandatory if giant CAA's are combined with fistula.[13] Percutaneous intervention may be an alternative to the surgical treatment in patients with high-predicted perioperative risk.[14] Although the natural history and prognosis remain obscure, the overall 5-year survival of giant CAA's is almost 71%.[9] In conclusion, giant CAA combined with fistulous drainage to the RV may provide a focus for the inflammation, which can rarely present in the form of infective endocarditis. Ideally these patients would benefit from surgical resection and closure of the fistula.

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