Giant Circumflex Artery Aneurysm With a Coronary Sinus Fistula

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Giant coronary artery aneurysms are extremely rare entities, and the authors are aware of only 13 case reports of such aneurysms of the circumflex artery. This case report is provided to add evidence of the natural history and pathophysiology and describe the successful surgical treatment of this condition.

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Giant coronary artery aneurysms (GCAAs) are rare entities; only 22 cases of aneurysms considered to have a diameter greater than 2 cm in diameter were reported between 1996 and 2009 [1]. GCAAs may manifest with cardiac symptoms of angina and myocardial infarction, symptoms of superior vena cava syndrome, or features of mediastinal mass and cardiac compression [1, 2]. Most coronary artery aneurysms affect the right coronary artery, and a few affect the circumflex (Cx) artery. Thirteen case reports of Cx GCAAs have been identified [1]. In this report we describe a case of a patient with Cx GCAAs who presented with previously unreported pericardial effusions, as well as the pathophysiology and anatomy of this lesion and subsequent successful surgical treatment.

A 56-year-old man presented a 7-month history of exertional dyspnea. A chest roentgenogram demonstrated an enlarged cardiac shadow with a cardiothoracic ratio of 0.84 (Fig 1). Examination by echocardiography identified a large pericardial effusion, which when drained was found to have a volume of 3 L. The effusion was confirmed to be a transudate, and pathologic investigations revealed no evidence of renal and liver dysfunction as a cause and no evidence of inflammatory, infective, or malignant causes. Investigation by computed tomography (CT) scan also revealed no evidence of malignant disease or infection but showed a large, aneurysmal Cx artery. Investigations into the patient's history of frequent travel to African countries revealed no evidence of infestations such as with *Strongyloides* and *Filaria* or infections such as tuberculosis or Lyme disease.

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The pericardial effusion rapidly recurred after drainage. The patient was therefore treated by surgical creation of a pericardial window and placement of a long-term drainage catheter in the pleural space. Tissue obtained during the procedure revealed no abnormalities. Studies were performed to investigate a potential causal link of the Cx GCAA with the effusion. Direct coronary angiography failed to opacify of the left coronary system because blood flow was extremely high, suggesting the presence of a high-flow arteriovenous malformation. A gated cardiac CT scan with three-dimensional (3D) reconstruction did demonstrate the detailed anatomic features (Fig 2). The left main and left anterior descending (LAD) arteries were of normal caliber, but 1.9 cm distal to its origin the Cx artery became aneurysmal, measuring 4.75 cm. It then continued on a tortuous pathway and terminated adjacent to and communicated with the coronary sinus, which was dilated to 1.92 cm. A 3D reconstruction scan also revealed that the Cx artery was commutating with the coronary sinus (Fig 3).

An initial path of conservative management failed after approximately 8 months as symptoms typical of angina developed that prompted further investigation by CT scan. The scan showed that the giant aneurysm had grown to 5.5 cm in diameter and was compressing the LAD artery. Rapid enlargement of the GCAA that raised the risk of rupture and caused further cardiac ischemia indicated the need for interventional treatment.

Percutaneous occlusion of the GCAA–coronary sinus fistula by cardiac catheterization of the fistula was attempted to reduce blood flow but failed. It was not possible to cannulate the coronary sinus.

Operative intervention was undertaken through a median sternotomy. Cardiopulmonary bypass was instituted using standard aortic cannulation and bicaval cannulation, moderate systemic hypothermia, and cold blood cardioplegia. Through a right atriotomy an

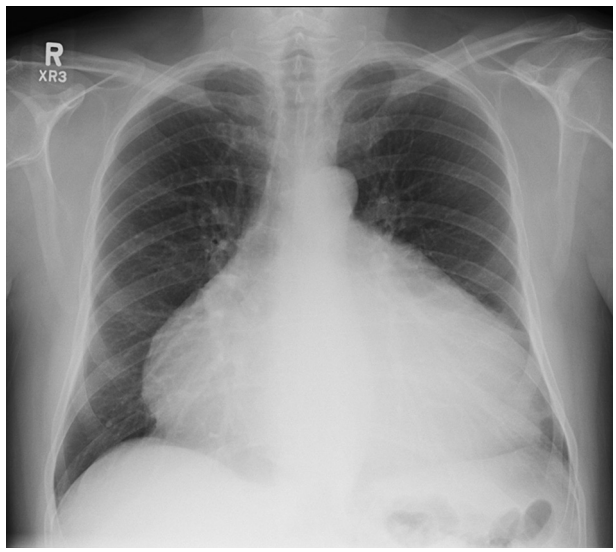


Fig 1. Chest roentgenogram showing a grossly enlarged cardiothoracic ratio of 0.84 suggesting the presence of a large pericardial effusion.

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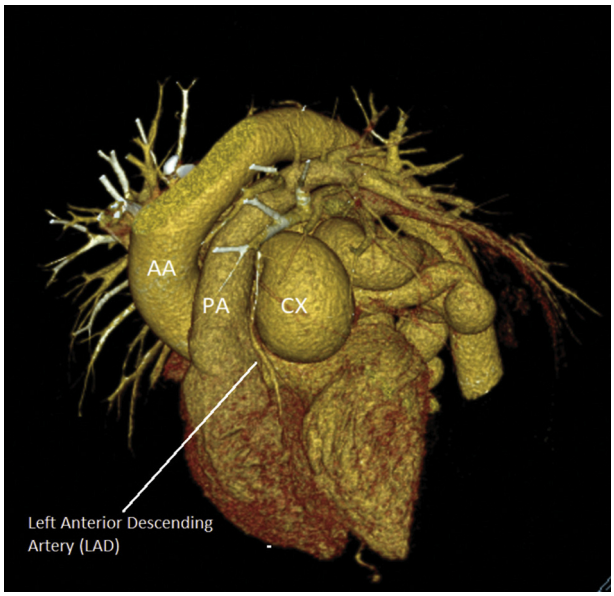


Fig 2. Gated cardiac computed tomography three-dimensional reconstruction of the heart identifying the circumflex artery and its torturous course. The left anterior descending artery is seen arising from between the circumflex artery (CX) and the pulmonary artery (PA). (AA = ascending aorta.)

unsuccessful attempt to cannulate the coronary sinus was made; progress of instruments beyond the ostium was prohibited.

An attempt was made to identify and fashion coronary artery bypass grafts using saphenous vein to two large obtuse marginal (OM) arteries evident on the preoperative CT angiogram. OM1 was readily identified and grafted, but OM2 was identifiable only after a longitudinal arteriotomy was made in the aneurysmal Cx artery to identify this vessel from within the lumen. Two further lateral vessels were identified and grafted, thus providing comprehensive revascularization of the left ventricle. The entrance and exit of the GCAA just lateral to the left main stem were occluded from within the lumen by a layered closure with a continuous suture reinforced by interrupted sutures buttressed with Teflon (Chemours, Wilmington, DE) pledgets. Weaning from cardiopulmonary bypass required support with an intraaortic balloon pump and infusion of inotropes.

Postoperative progress was rapid, and hospital discharge achieved on day 8. Follow-up examination 6 months postoperatively identified full resolution of symptoms and return to normal activities. A CT angiogram demonstrated patent bypass grafts and an absence of the Cx GCAA. An echocardiogram demonstrated normal ventricular function and associated reduction of right ventricular size when compared with preoperative studies.

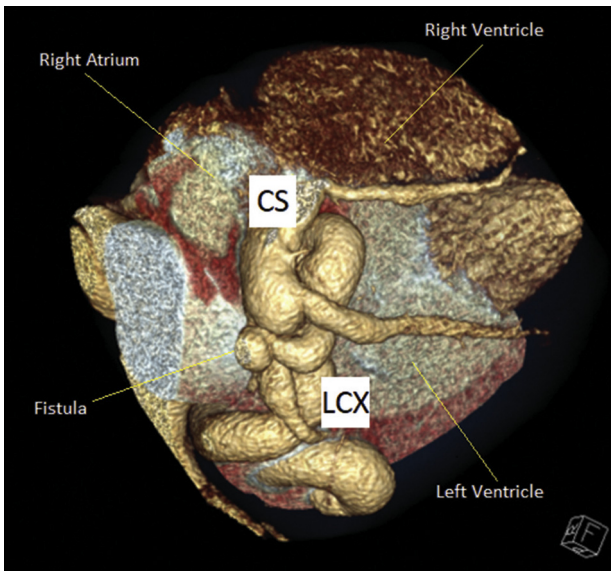


Fig 3. Gated cardiac computed tomography three-dimensional reconstruction showing the inferior surface of the heart and highlighting the tortuous giant left circumflex artery (LCX) aneurysm connecting with a dilated coronary sinus (CS).

Comment

Coronary artery aneurysms are identified in less than 5% of coronary angiographic series [3]. GCAAs were reported in only 22 cases between 1996 and 2009, and the defining characteristic was an aneurysmal diameter greater than 2 cm [1]. Although there is no consensus definition of GCAA, the American Heart Association has defined GCAA as an aneurysm greater than 8 mm in diameter [4].

GCAAs most commonly affect the right coronary artery (89%); GCAAs of the LAD are rare, and only 13 cases of Cx GCAAs have been reported [5]. The primary cause of GCAA is atherosclerotic disease [1]; however, 10 of the 13 cases of Cx GCAAs were considered congenital because a fistulous communication of this GCAA with the coronary sinus was identified. Presentations of GCAA, perhaps related to high and abnormal flow through an arteriovenous fistula, include exercise-related dyspnea, palpitations, fatigue, angina, myocardial infarction, and hemopericardium. Other symptoms related to the mass effect include superior vena cava syndrome and symptoms of mediastinal mass and cardiac compression [1, 2]. We propose that our patient had recurrent pericardial effusions as a result of high flow causing transudation through the thinned Cx arterial wall. Angina was caused by compression of the LAD artery by the Cx aneurysm and possibly by a coronary artery steal phenomenon [6].

Why had the patient presented relatively late in early life when a coronary artery–coronary sinus fistula with a large arteriovenous shunt could cause early high-output cardiac failure? We suggest that the severity of arteriovenous shunting was moderated by a membrane at the outflow of the coronary sinus, a membrane that also caused failure of retrograde cannulation of the coronary sinus.

Successful operative treatment achieved comprehensive revascularization by using CT coronary angiography

to delineate the number and position of target vessels. A second vital strategic element was closure of the GCAA.

In conclusion, we presented the case of a 56-year-old man who presented with signs and symptoms, consisting of recurrent pericardial effusion and angina, of a congenital GCAA of the Cx artery. We reported the successful investigation and surgical management of the case.

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