An Audaciously Aneurysmal Atrium

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INTRODUCTION

Giant left atrium (GLA) is enlargement of the left atrium beyond sizes seen in routine clinical practice. GLA is defined by anterior-posterior measurement at end-systole on transthoracic echocardiography with a minimum diameter of 6.5 to 10 cm, depending on the authorship of the various case reports and series.[1-6] GLA is the sequela of pressure and volume overload of the left atrium, which has been described as a complication of mitral stenosis, mitral insufficiency, and mitral prolapse.[7,8] However, the vast majority of cases result from rheumatic heart disease (RHD) with mitral involvement.[3,9] In this case report, we describe a case of RHD-associated GLA that resulted in the largest left atrial dimensions described to date.

CASE PRESENTATION

A 69-year-old Asian man with RHD presented to our institution reporting 2 weeks of progressive dyspnea. Results of physical examination were remarkable for elevated jugular venous pressure, bilateral rales, and a 3/6 holosystolic murmur at the apex followed by a faint diastolic rumble. His bedside cardiac monitor and electrocardiogram revealed that he was in atrial fibrillation, with a heart rate of 61 beats/min. Chest radiography depicted a cardiothoracic ratio of 0.77 (Figure 1), and computed tomographic imaging revealed that the patient's left atrium measured 21 cm in transverse diameter and 17 cm in superoinferior diameter (Figures 2 and 3). Computed tomography also disclosed severe bullous emphysematous changes of the lungs complicated by pneumothorax in addition to multiple pulmonary emboli (Figures 4–6, Videos 1 and 2). The patient's clinical condition improved after noninvasive ventilation for 18 hours, antibiotics for suspected community-acquired pneumonia, chest tube insertion, and anticoagulation for his pulmonary embolism. His transthoracic echocardiographic findings included a mean transmitral gradient of 10 mm Hg, moderate mitral regurgitation, and, remarkably, a left atrium with an anteroposterior diameter of 9.2 cm (Figure 7, Video 3). Using computed tomographic dimensions, we calculated the volume of the left atrium using the standard formula for calculating volume of ellipsoid objects, $V_{\text{ellipsoid}} = (4/3)\pi(r_1 \times r_2 \times r_3)$; we found that the left atrial volume was approximately 1,600 cm^3 (Figures 2 and 3). The left atrium was measured to be 10.6 cm in anteroposterior diameter on cardiac magnetic resonance imaging, and sludge was evident on both magnetic resonance imaging and transthoracic echocardiography (Figures 8 and 9, Videos 4–8).

DISCUSSION

GLA, as described by Hewett in 1849, is defined as an aneurysmal dilation of the left atrium and contraction of the left atrioventricular opening in the context of mitral valve disease.[10] Using the echocardiographic criterion of an anteroposterior diameter $\geq$ 8 cm, the occurrence of GLA among patients with RHD was 0.6%.[3] Complications include atrial fibrillation, spontaneous echocardiographic contrast, sludge, or thrombus leading to thromboembolism. In addition, there may be compression of adjacent structures, including an interatrial septal bulge leading to partial inferior vena cava obstruction, compression of main bronchus causing right middle lobe collapse, or the esophagus leading to dysphagia,[11] for which a patient may undergo left atrial volume reduction surgery.[3] Our patient had severe mitral stenosis due to RHD leading to GLA with the presence of sludge. Fortunately, imaging did not exhibit any compressive complications. Nevertheless, the massive enlargement of the left atrium may restrict lung capacity as it competes for intrathoracic space. Because of our patient's severe comorbidities, he was deemed at very high risk for surgical intervention. The large amount of sludge and mitral regurgitation precluded percutaneous mitral valve balloon valvuloplasty. He will be managed with continued rate control of his atrial fibrillation in addition to anticoagulation with a vitamin K antagonist. This case presentation illustrates the tremendous remodeling capability of the left atrium and the complexity of managing RHD with GLA.

CONCLUSIONS

GLA is a rare condition most commonly due to rheumatic mitral disease, and this largest described left atrium to date demonstrates the remarkable extent of remodeling possible due to pressure and volume overload. Patients with GLA often require chronic anticoagulation because of left atrial thrombus or atrial fibrillation. Valve repair remains the mainstay of surgical intervention for patients with GLA. In patients with compressive symptoms, consideration should be given for resection of redundant atrial tissue.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2017.10.005.

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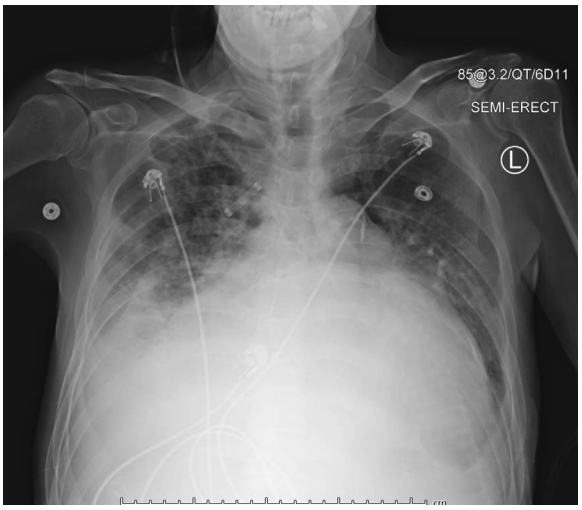

Figure 1 AP chest radiograph. Anteroposterior chest x-ray revealing alveolar and interstitial opacities in the lung fields, left-sided pneumothorax, and enlarged cardiac silhouette with a cardiothoracic ratio of 0.77.

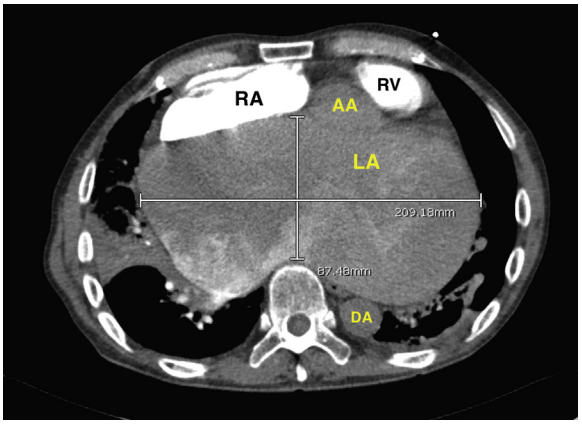

Figure 2 Transverse computed tomographic image of the chest. Computed tomography of the chest depicting the left atrial dimensions including a transverse diameter of 21 cm and an anteroposterior diameter of 8.7 cm. *AA*, Ascending aorta; *DA*, descending aorta; *LA*, left atrium; *RA*, right atrium; *RV*, right ventricle.

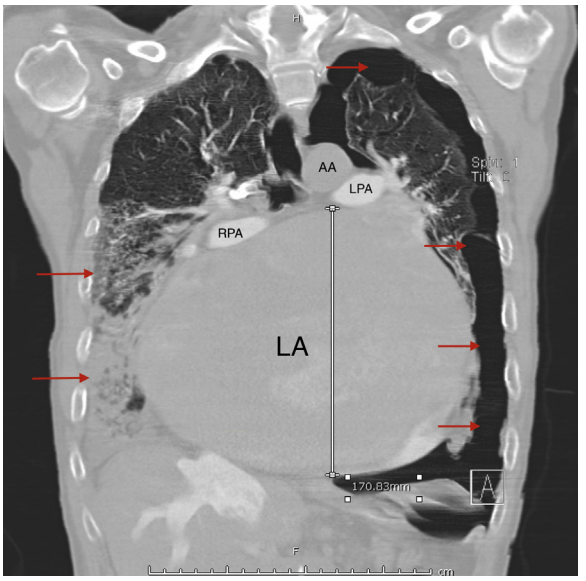

Figure 3 Coronal computed tomographic image of the chest. Computed tomography of the chest showing superoinferior measurement of the left atrium measured at 17 cm and left-sided pneumothorax (*red arrows* on the right of the image), and right-sided airspace and interstitial opacities of the lungs (*lower left and upper left arrows*, respectively). *AA*, Aortic arch; *LA*, left atrium; *LPA*, left pulmonary artery; *RPA*, right pulmonary artery.

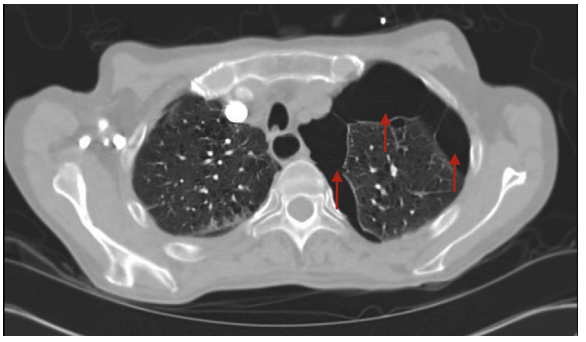

Figure 4 Transverse computed tomographic image of the chest in the lung window. Computed tomography of the chest in the transverse plane revealing multiple pleural blebs (*right and left arrows*), and a large anterior pneumothorax (*center arrow*).

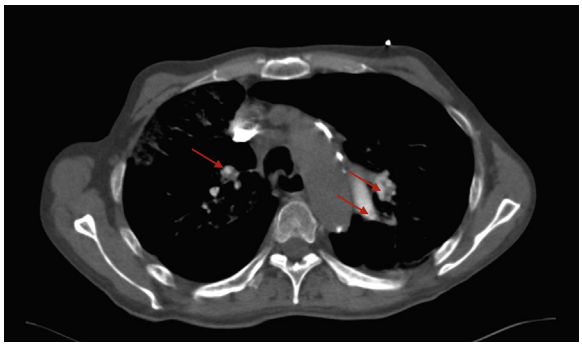

Figure 5 Transverse computed tomographic pulmonary angiogram. Computed tomography pulmonary angiographic image showing filling defects in bilateral pulmonary artery branches representing pulmonary emboli (*red arrows*).

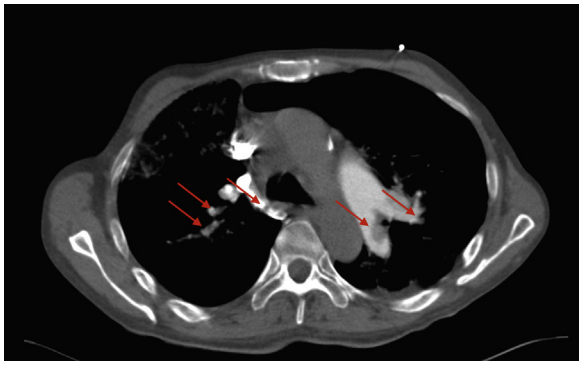

Figure 6 Transverse computed tomographic pulmonary angiogram. Computed tomography pulmonary angiographic image showing filling defects in bilateral pulmonary artery branches representing pulmonary emboli (*red arrows*).

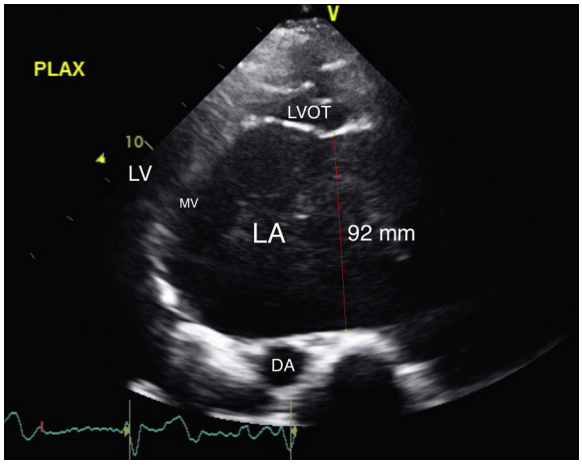

Figure 7 Transthoracic echocardiogram in parasternal long-axis view. Transthoracic echocardiogram image in parasternal long-axis view depicting a left atrial diameter of 9.2 cm measured in the anteroposterior dimension. *DA*, Descending aorta; *LA*, left atrium; *LV*, left ventricle; *LVOT*, left ventricular outflow tract; *MV*, mitral valve.

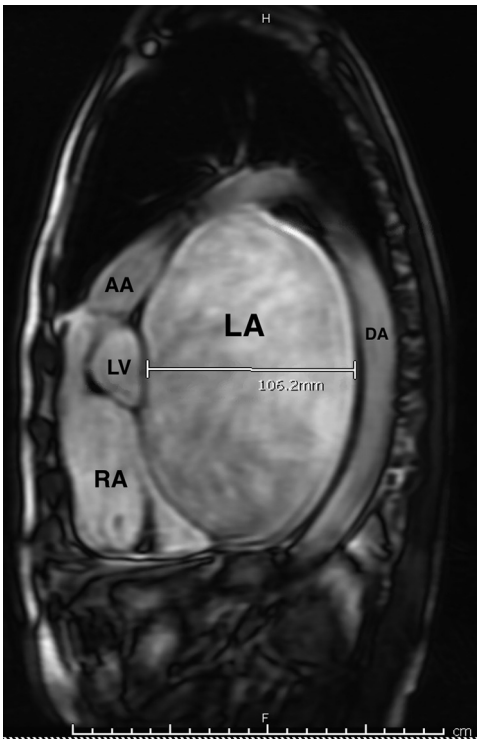

Figure 8 Sagittal image of cardiac magnetic resonance images with depicting anatomy. Sagittal slice of cardiac magnetic resonance imaging revealing the giant left atrium with an anteroposterior diameter of 10.6 cm. *AA*, Ascending aorta; *DA*, descending aorta; *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium.

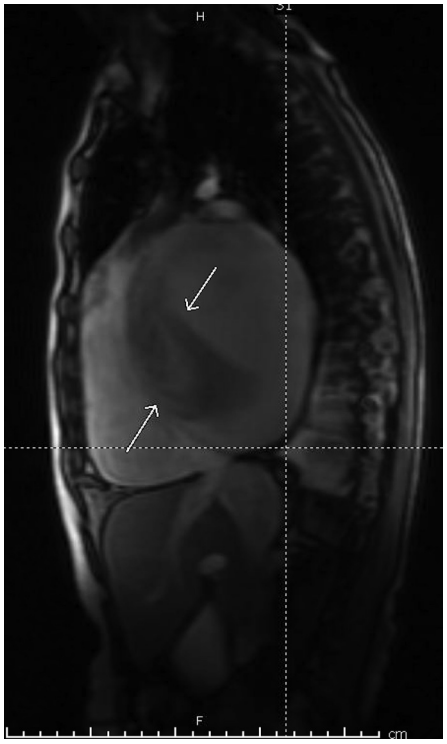

Figure 9 Cardiac magnetic resonance images of left atrial thrombus burden. Cardiac magnetic resonance imaging in the sagittal plane revealing the large burden of left atrial thrombus and microthrombi (*white arrows*).

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