Giant Left Atrium: Adaptive or Maladaptive?

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An 88-year old woman with a history of rheumatic heart disease (RHD), permanent atrial fibrillation and hypertension presented to our institute with increasing dyspnoea. A chest X-ray showed severe cardiomegaly (Image 1A). A transthoracic echocardiogram showed a rheumatic mitral valve (MV) with mild stenosis and severe MV regurgitation and a pulmonary artery systolic pressure 62 mmHg. The left atrium was massively dilated resulting in right atrial and ventricular compression (Image 1B). An ECG-gated cardiac CT confirmed a massive left atrium with an area of 119cm² and volume of 996 mls (673 mls/cm²) that occupied two thirds of the chest cavity and resulted in compression of the lung parenchyma and the right heart, traction on the pulmonary veins and displacement of the coronary veins (Images 1C-1F, Video 1). Given the technical challenges associated with any approach to the MV, the patient was managed with medical therapy.

Left atrial evaluation is a crucial component of cardiac function. It enables not only left ventricular filling but also left ventricular pressure regulation in left sided heart valve disease and cardiomyopathies [1,2]. Massive left atrial dilatation is strongly associated with RHD, but may also infrequently occur in patients with MV prolapse and restrictive cardiomyopathies. Although the precise mechanism is unknown, it has been postulated that in RHD, a pancarditis effecting the left atrium occurs [3]. This results in a reduction of its elastic properties and renders it more susceptible to pressure increases with MV stenosis or regurgitation. An alternative theory is that dilation of the left atrium represents a protective mechanism designed to shield the pulmonary vasculature against this pressure change [4]. This may explain why patients often remain asymptomatic until symptoms related to the compression of adjacent structures become apparent [5]. In this event, the treatment of choice remains mitral valve and left atrial reduction surgery.

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References:

Legends

Image 1 Legend

Panel A shows the severe dilatation of the cardiac silhouette on the chest X-ray. Panel B shows the severe dilatation of the LA on the transthoracic echocardiogram 4-chamber view. Panels C and D show the severe dilatation of the LA, the small left ventricular cavity size and compression of the RA and RV on ECG-gated cardiac CT. Panel E shows the calcified MV in the short axis view on ECG-gated cardiac CT in keeping with rheumatic mitral valve disease. Panel F shows the compression of the RVOT on ECG-gated cardiac CT.

ECG, electrocardiogram; CT, computed tomography; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; MPA, main pulmonary artery; MV, mitral valve; RVOT, right ventricular outflow tract.

Video 1 Legend

ECG-gated cardiac CT showing the severe dilatation of the LA and the compression of the right chambers in the 4-chambers view.

ECG, electrocardiogram; CT, computed tomography; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.