Giant left atrium in a patient with Marfan syndrome.

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A 41-year-old man with Marfan syndrome, complicated by retinal detachment, heart failure with mildly reduced ejection fraction, and atrial fibrillation for one year, presented to the cardiology clinic due to worsening dyspnea. He had no significant family history. On physical examination, vital signs were normal. He had an ectomorphic, dolichocephalic physique with no jugular venous distension. Heart auscultation revealed irregular heart sounds and a mesosystolic murmur at the mitral focus. The Walker-Murdoch wrist sign was positive. Laboratory analysis showed no abnormalities. The electrocardiogram indicated atrial fibrillation and left ventricular hypertrophy. Coronary angiography revealed slow coronary flow. The transesophageal echocardiogram showed left atrial dilation with an anteroposterior diameter of 81.8 mm, volume of 793 ml, and volume index of 425 ml/m^2. Additionally, there was posterior leaflet prolapse of the mitral valve resulting in severe regurgitation classified as Carpentier type IIA. MRI measurements showed the left atrium measured 113 x 83 mm, with an area of 122.2 cm^2.

Due to a decline in functional status attributed to severe mitral regurgitation, the case was reviewed by the Heart Team, who approved mitral valve replacement surgery. Currently, the patient is classified as NYHA functional class II, with the heart rate controlled by a beta-blocker. The incidence of a giant left atrium is 0.3% to 0.6%, predominantly secondary to
rheumatic mitral disease in up to 92% of cases, with non-rheumatic etiologies being rare (1).

A retrospective study associated Marfan syndrome with left atrial dilation, suggesting primary atrial involvement in this syndrome (2).

References


Figure legends:

Figure 1. Giant left atrium in a patient with Marfan Syndrome. Panel A: Walker Murdoch wrist sign; Panel B: the funduscopic examination showing myopic choroidosis with chorioretinal atrophy; Panel C: chest X-Ray showing cardiomegaly (cardiothoracic ratio 0.81); Panel D: transesophageal echocardiogram (left side) showing giant left atrium comparing left ventricle, with posterior valve prolapse of the mitral valve (purple arrow), with color Doppler showing severe regurgitation (right side). Panel E: cardiac magnetic resonance imaging showing giant left atrium and left ventricle in a coronal axis.

MVP: Mitral Valve Prolapse; LA: Left Atrium; LV: Left Ventricle.
Supplementary Material.

Video Legends

Video 1. Transesophageal echocardiogram in a two-chamber view with color Doppler, showing the severe mitral regurgitation jet.

Videos 2 and 3. Cardiac magnetic resonance imaging in cine mode, demonstrating the giant left atrium and left ventricle in sagittal and transverse views, respectively.

Figure 1
313x208 mm (x DPI)