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Giant left atrium in patient with Barlow syndrome.

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An 80-year-old man was admitted to our hospital with weakness and resting dyspnea. His medical history included dilated cardiomyopathy, mitral regurgitation (MR), and persistent atrial fibrillation diagnosed 10 years ago. Physical examination revealed signs of pulmonary congestion, massive bilateral peripheral edema, pulse wave irregularity, and systolic 3/6 murmur at apex radiating to the parasternal region. Marked cardiomegaly and right-sided pleural effusion was seen on chest radiography. (Fig. 1 A). The transthoracic echocardiography (TTE) showed a giant left atrium (LA), with a diameter in the parasternal long axis view (LAX) of 13cm, and severe MR. Both leaflets of the mitral valve were thickened and prolapsed into the LA (Fig 1 B). The medial papillary muscle could not be visualized. Suspecting a mitral valve apparatus anomaly, we performed cardiac magnetic resonance (CMR) (Simens Symphony 1,5T) and transesophageal echocardiography 3D (Hitachi Lisendo 880).

CMR revealed monstrous LA (195x105x155mm), with a volume of 3100 ml. (Fig. 1C). The left ventricle was also enlarged (ejection fraction, 55%), and MR was severe. The TEE 3D examination found the mitral annulus was dilated to 55 mm. The lateral papillary muscle exhibited numerous chordae tendineae to both leaflets, and the medial papillary muscle had two heads with one chorda tendinea to the ventricular septum, one to the lateral papillary muscle, and three to the anterior leaflet. Both leaflets were thickened and prolapsing. The anterior leaflet was perforated in the A1 segment (Fig 1D).

The patient’s case was discussed during a multidisciplinary consultation. Mitral valve repair or replacement is considered the gold standard treatment for patients with severe MR[1,2] He refused surgery. Treatment consisted of standard pharmacotherapy of heart failure included ramipril, spironolactone, furosemide, carvedilol and warfarin, thoracocentesis, and daily weight control. After four months of follow-up, the patient is in heart failure class NYHA II.
Barlow’s syndrome is characterized by bilateral prolapse or billowing leaflets, chordae elongation, and annular dilatation. In some cases the lack of coaptation of leaflets leads to MR and subsequent LA enlargement. However, its exact etiology is still unknown [3]. LA larger than 8 cm in the LAX is considered to be giant.

We present this case with monstrous LA, as its volume from CMR was estimated at 3100 ml. In the available literature we found only one case report of similar LA volume described at the beginning of the 20th century [4]. Chordae tendineae abnormalities are rarely described [5]. In our case two chordae tendineae of medial papillary muscle were incorrectly attached, but paradoxically it could improve mitral valve function.

In most cases TTE remains the first line investigation for Barlow’s syndrome; however, in some cases it has limitations. Echocardiography TEE 3D plays a central role for evaluation of these patients. CMR offers an alternative modality to study the morphology and function of the mitral apparatus. In the presented case, clinically-relevant details have been evaluated using echocardiography TEE, TEE 3D, and CMR. A multimodality imaging approach provides precise diagnosis of our patient with giant left atrium.
References:
Figure 1: Giant left atrium.