Giant Left Atrium in a Triple Rheumatic Heart Disease

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Introduction
Several definitions and dimensions have been used to determine a giant left atrium (GLA). The range of dimensions for GLA reported in previous studies was from 6 to beyond 10 cm of antero-posterior diameter in the parasternal long axis view [1]. However, other investigators defined the GLA as a cardiothoracic ratio on chest X-ray (CXR) of > 0.7 combined with a left atrial anterior-posterior diameter of > 8 cm on trans-thoracic echocardiography (TTE) [2]. The GLA is a rare condition, commonly associated with rheumatic mitral valve disease, and very rarely with non rheumatic heart disease (nRHD). The triple valvular heart disease with involved mitral, aortic and tricuspid valves is quite uncommon. A 47 year female patient with a past medical history of rheumatic heart disease (RHD) and known severe mitral stenosis presented with severe breathlessness (NYHA class IV). She had undergone mitral valve commissurotomy and tricuspid valve annuloplasty 12 years previously. Transthoracic echocardiography revealed a giant left atrium, moderate to severe mitral valve restenosis, severe mitral regurgitation, moderate aortic regurgitation and severe tricuspid regurgitation, associated with severe secondary pulmonary hypertension and a markedly dilated right heart chambers. The patient was considered inoperable by the heart team, because of advanced pulmonary hypertension predicting a very high risk for open heart surgery. The final treatment decision was a difficult and complex issue.

Case presentation
A 47 year female patient with a past medical history of RHD with known severe mitral stenosis was admitted to our Department complaining of severe breathlessness. She underwent mitral valve commissurotomy and tricuspid valve annuloplasty 12 years ago. Five years ago she suffered an ischaemic stroke, but recovered without remaining neurological deficit. On admission she presented with signs and symptoms of heart failure, qualified as functional NYHA class IV, an irregularly irregular pulse of about 110 beats/minute, blood pressure of 110/70 mmHg and body temperature of 37°C. On physical examination patient was tachyarhythmic, tachypnoeic, severely orthopnoeic, with notably elevated jugular venous pressure and with bilateral distention of jugular veins (Fig.1). Palpitations, fatigue, shortness of breath, recurrent paroxysmal nocturnal dyspnea were reported, too. Heart examination revealed pansystolic murmurs in the whole precordium, with loudest intensity at the apex and palpable systolic thrill. There were no complaints of voice hoarseness, dysphagia or any other gastrointestinal symptoms. In the postero-anterior view the chest X-ray (Fig.2), showed significant enlargement of the entire heart silhouette with cardiothoracic ratio of 0.8 and hilo-pulmonary congestion. The costo-diaphragmatic recesses were free from fluid. Electrocardiogram (ECG) showed atrial fibrillation (Fig.3), with well controlled ventricular response (heart rate of 90 beats/minute),
LV hypertrophy and signs of digitalization with downsloping ST depression and characteristic “sagging” appearance, and short QT interval.

Transthoracic echocardiography revealed an impressive enlargement of left atrium, whereas in the apical 4-chamber and parasternal long axis views reaching a diameter of 10.5x10 cm, and LA area was 85.6cm² (Fig.4). On 2-dimensional and M-mode echocardiography, the mitral valve leaflets were qualified as thickened, with severely limited motility. A moderate to severe mitral valve restenosis, with a mitral valve hemodynamic area of 1.3 cm² and planimetric area of 1.38 cm², combined with severe regurgitation was registered on color-Doppler examination. The aortic cusps also were thickened and moderately calcified and were associated with moderate aortic regurgitation (Fig.4). A severe tricuspid regurgitation and severe secondary pulmonary hypertension assessed by color-Doppler, with a systolic pulmonary pressure up to 120 mmHg assessed by tricuspid pressure drop (Fig. 4D), was also registered. The tricuspid valve dysfunction was associated with thickened and fibrotic leaflets. Left ventricular end-diastolic diameter (LVEDD= 4.8 cm), kinetics and LV ejection fraction (LV EF ~ 55%) were normal. In contrast, right heart chambers were markedly dilated; the transversal diameter of the right ventricle and right atrium were 4.9 cm and 6.0 cm, respectively.

The patient was considered inoperable by the heart team, because of advanced pulmonary hypertension, predicting a very high risk for open heart surgery. Consequently she was discharged with a diagnosis of valvular cardiomyopathy and heart failure with preserved ejection fraction. We discharged the patient under the conventional medical therapy for heart failure (furosemide, ACE inhibitors, beta blockers, digoxin, spironolactone and warfarin), adding the sacubitril/valsartan therapy, which was recently added in ESC guidelines for heart failure treatment [5].

Discussion

We report this case as a triple rheumatic valve disease with associated GLA, considering that it represents an entity of rarity. GLA, which is a deformed structure of the LA, mainly was reported as a result of mitral valve regurgitation as a core mechanism of its enlargement. In contrast, our patient underwent a surgical correction 12 years ago for the stenotic deformities of mitral valve. She presented with moderate mitral restenosis associated with severe mitral and tricuspid regurgitations, while aortic valve reveals a significant level of anatomical and functional deterioration. However, despite these findings, our main objective was the further treatment of this very complex patient. In fact, there are few studies, particularly observational, that tried to show the benefit of the different treatments of valvular heart disease (VHD) [6, 7]. In addition, the prevalence of VHD is higher in the elderly nowadays, and it has a high prevalence of comorbidities and increased risk
associated with intervention [8]. Another important aspect of contemporary VHD is the growing proportion of previously-operated patients [6]. Therefore the final treatment decision is quite difficult and complex in patients with VHD. The heart team considered this patient as inoperable, due to very advanced pulmonary hypertension. We discharged the patient under the medical therapy, according to the newest ESC guidelines for heart failure treatment [5]. Despite the fact that the therapy with sacubitril/valsartan is indicated only in patients with heart failure and reduced left ventricular ejection fraction in recent guidelines, we introduced this therapy in our patient as we believe that it could be beneficial for our end-stage patient. Moreover, the ongoing trials are testing this drug also in patients with heart failure and preserved ejection fraction [9], expecting improvement in survival in these patients.

Conclusion
We present a patient with triple rheumatic heart disease, giant left atrium and severe pulmonary hypertension. Final treatment decision came from the heart team, as a difficult and complex issue. She was considered inoperable, and was discharged with conventional medical therapy.

Abbreviations
GLA - Giant left atrium;
RHD - rheumatic heart disease;
LA - left atrium;
LV - left ventricle;
VHD - Valvular heart disease.

Declarations of interest
The authors declare no conflicts of interest.

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