

Isolated Rheumatic Mitral Regurgitation with Giant Left Atrium in Sinus Rhythm – A Case Report

MD. FAISAL IBN KABIR, MD. RASULAMIN, MOHAMMAD RAYHAN MASUM MANDOL, MD. ARIFUL ISLAM JOARDER, SM EARE MAHABUB, MD. FAKHRUL ISLAM KHALED, MUSTASHIRUL HAQUE, PRASHANT BAJRACHARYA, KHURSHED AHMED, MD. HARISUL HOQUE

Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka

Address of Correspondence: Dr Md Faisal Ibne Kabir, Assistant Professor, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka.

Abstract

A 30-year-old woman, patient of severe MR with pulmonary hypertension presented with symptoms. On evaluation, giant left atrium was found on echocardiography (65 mm). Most strikingly, the patient was still in sinus rhythm. She was managed medically and after stabilization of symptoms, discharged with advice for mitral valve surgery and regular follow-up.

Keywords: Isolated Rheumatic Mitral Regurgitation, Giant Left Atrium, No atrial fibrillation

Introduction:

Isolated mitral regurgitation is a relatively uncommon presentation of chronic rheumatic heart disease¹. Chronic nature of rheumatic mitral regurgitation causes gradual dilatation of the left atrium with little increase in pressure and therefore relatively fewer symptoms. However, occasionally the dilatation of LA is so severe, it is termed giant LA. Giant left atrium is a rare condition, with a reported incidence of 0.3%, and occurs following mainly rheumatic mitral valve disease². Atrial fibrillation is almost always present³ predisposing to thromboembolic complications. A subset of patients may develop severe dysphagia from esophageal compression or hoarseness from laryngeal nerve impingement⁴.

Here we are presenting a case of isolated mitral regurgitation with giant left atrium in sinus rhythm.

Case Report|:

A 30-year-old woman normotensive, non-diabetic presented with the complain of SOB and palpitation for 2 ½ months and leg swelling for 10 days. 2 ½ months back, after 10 days of delivery of her 3rd child, she developed SOB which was initially present on exertion (NYHA-II) and relieved by rest, but since last 10 days, it was present on minimal exertion or rest (NYHA III-IV) with orthopnea but no PND. She also had dry cough but no hemoptysis or diurnal and seasonal variation. She also had palpitation which occurred with SOB during exertion and was relieved by rest. It was regular and not associated with dizziness or syncope. Since last 10 days, she also developed swelling of legs which was gradually increasing, associated with decreased urine output but no puffiness of face or abdominal swelling. She didn't have history of chest pain,



Fig.-1: ECG of patient showing LVH with left atrial enlargement.

fever, dysphagia or hoarseness of voice. She had no history suggestive of rheumatic fever in childhood or significant past illness. She is a housewife and mother of 3 children, all normal vaginal delivery and in good health.

On physical examination, the patient was ill-looking, dyspneic, lying on propped up position. She had anemia and bilateral pitting pedal edema. Her pulse was 104 beats/min, regular, normal volume, BP 120/80 mm Hg, JVP raised (6 cm above sternal angle) and RR-22/min. Precordial examination revealed apex beat shifted to left 5th ICS, 1 cm lateral to MCL, thrusting in nature. Parasternal heave and palpable P2 present. S1 – soft, P2 – loud in pulmonary area, pansystolic murmur in mitral area with radiation to left axilla, grade 3/6 and another pansystolic murmur in tricuspid area. Bilateral basal crepts present in the lungs. Other systemic examination revealed no abnormalities.



Fig.-2: Chest X-ray PA view showing borderline cardiomegaly with features of left atrial enlargement (bulging of LAA and double right heart border)

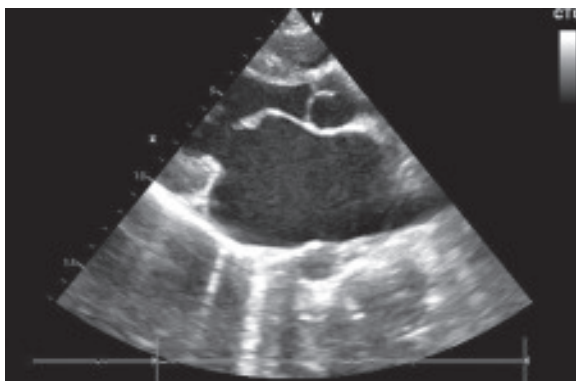


Fig.-3: PLAX view showing enlarged LA, thickened and calcified mitral leaflets

On investigations, chest X-ray reveals borderline cardiomegaly with bulging of LAA and double right heart border. ECG is suggestive of LVH with left atrial enlargement. Echocardiography shows severe mitral regurgitation with thickened mitral leaflets and hugely dilated left atrium (65 mm). Her LV was dilated (LVIDD=58mm) but LV systolic function was normal (LVEF=58%). She also had moderate tricuspid regurgitation and moderate pulmonary hypertension (PASP – 64 mm Hg). Her ASO titre was raised (401 IU/ml). Other routine investigations were within normal limits.

The patient was diagnosed as a case of chronic rheumatic heart disease with severe mitral regurgitation, moderate tricuspid regurgitation, moderate pulmonary hypertension with congestive cardiac failure and managed medically with diuretics, ACEIs, rate-limiting B-blockers, antiplatelets and oral penicillin. She improved in medical management and discharged to home with advice for mitral valve surgery and regular follow-up.

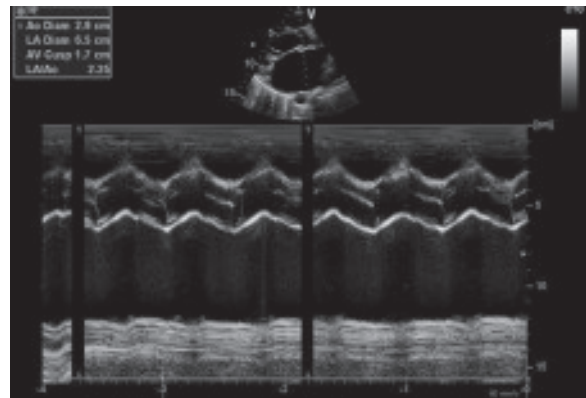


Fig.-4: M-mode at the level of aortic valve showing dilated LA (65 mm)

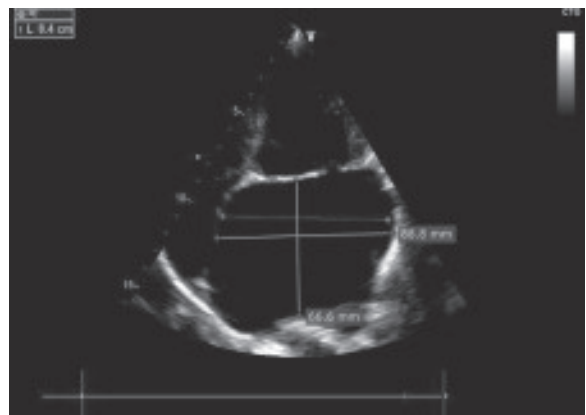


Fig.-5: Apical 4-C view showing dilated LA (66 mm X 88 mm)

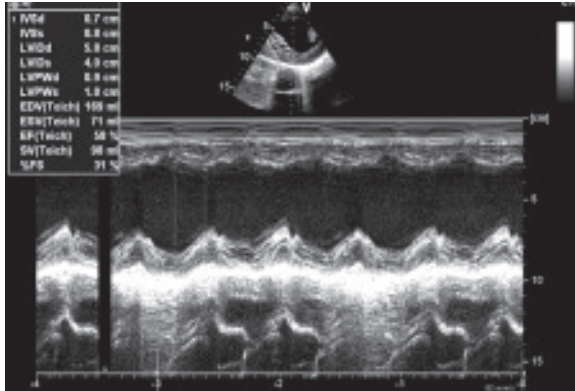


Fig.-6: LV study shows dilated LV and good LV systolic fcn

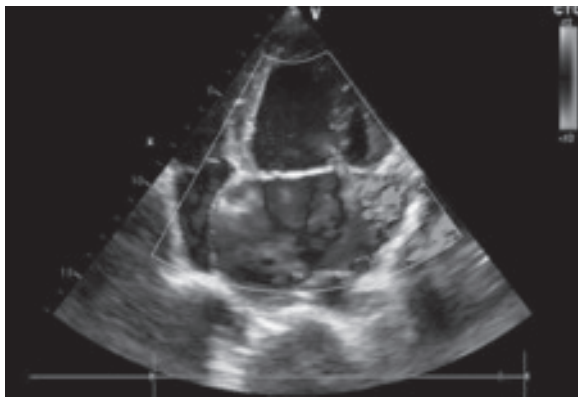


Fig.-7: Color flow mapping in A4C view showing severe MR

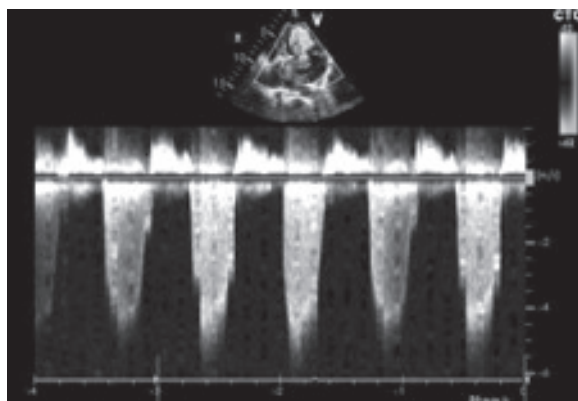


Fig.-8: CW Doppler study showing severe MR

Discussion:

Chronic rheumatic heart disease presents as isolated MS in 25% and combined MS and MR in 40%¹. So isolated MR in this patient is an uncommon presentation. The thickening of the mitral leaflets with mild involvement of the aortic cusps supports the rheumatic origin of MR. Other possible causes of MR in this patient like mitral

valve prolapse, post-partum cardiomyopathy, infective endocarditis, connective tissue disorder, congenital defects, degenerative and coronary artery disease have been ruled out by history, examination and investigations narrowing down the diagnosis to rheumatic heart disease.

Giant left atrium has been ascribed to left atrial enlargement >6 cm in diameter⁵. Left atrial dilatation occurs due to various causes such as mitral valve disease, left ventricular systolic as well as diastolic dysfunction and others. In general it is assumed that the more severe and chronic the cardiac condition, the larger is the left atrium⁶. However, giant LA is found mainly in rheumatic mitral valve disease. The hypothesis behind this is that rheumatic pancarditis damages the entire heart, including the left and right atria. The damaged left atrium then dilates more easily when mitral regurgitation fills the atrium with a large volume of blood leading to development of giant LA³. In this patient the left atrium anteroposterior diameter in PLAX view using M-mode is 65 mm which meets the criteria of giant left atrium. The measurements are more prominent in apical 4-chamber view (66 mm X 88 mm). The cause of rheumatic MR as the cause of this giant left atrium is apparent from the echocardiography. Also the chest X-ray shows cardiomegaly with features of left atrial enlargement (bulging of LAA and double right heart border) and ECG shows LVH with left atrial enlargement. The patient had only moderate pulmonary hypertension (PASP – 64 mmHg) which could be due to the large and expandable left atrium blunting the rapid rise of pulmonary venous pressure³.

Despite the giant left atrium, the patient hasn't developed atrial fibrillation which is pretty unique as almost all cases of giant left atrium have AF³. There is also no sign of thrombus or mechanical complication like dysphagia and hoarseness of voice. The patient had presented with congestive cardiac failure which had been precipitated by the increase in both venous return and vascular resistance seen in postpartum period in MR patients⁷. She was managed medically and responded well to the management. The patient being a case of symptomatic severe MR has class I indication for mitral valve surgery, preferably mitral valve repair⁸. The patient was discharged to home with advice for surgery and regular follow-up.

Conclusion:

Isolated rheumatic mitral regurgitation with giant left atrium patient in sinus rhythm is very rarely found in clinical practice. For these patients, along with the medical management, early intervention (mitral valve surgery)

could be a better option so as to prevent the onset of atrial fibrillation and its consequences.

Conflict of interest: None

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