

Erlenmeyer-shaped Heart in a Patient with Giant Left Atrium due to Mixed Mitral Valve Pathology: A Neglected Case in a Rural Area in Indonesia

Aprilia Romdiyana*, R. Mohammad Budiarto, Evelyne Chandra

Aprilia Romdiyana*, R. Mohammad Budiarto, Evelyne Chandra

Department of Cardiology and Vascular Medicine, Airlangga University, Dr Soetomo General Hospital, Surabaya, INDONESIA.

Correspondence

Aprilia Romdiyana

Department of Cardiology and Vascular Medicine, Airlangga University, Dr Soetomo General Hospital, Surabaya, INDONESIA.

E-mail: apriliaromdiyana@gmail.com

History

- Submission Date: 02-08-2022;
- Review completed: 19-09-2022;
- Accepted Date: 14-10-2022.

DOI : 10.5530/pj.2022.14.175

Article Available online

<http://www.phcogj.com/v14/i6>

Copyright

© 2022 Phcogj.Com. This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International license.

ABSTRACT

Giant left atrium (GLA) is a rare condition typically found in patients with rheumatic mitral valve disease. On chest radiographs, this condition often mimics another cause of cardiomegaly. A 52-year-old man was admitted to the hospital with complaints of shortness of breath and orthopnea. Mid-diastolic and grade IV pansystolic murmurs could be heard at the apex radiating to the axilla on cardiac auscultation. The electrocardiogram showed a junctional tachycardia rhythm, while the chest radiograph displayed extreme cardiomegaly with Erlenmeyer-shaped heart. Massive pericardial effusion was suspected. Transthoracic echocardiography (TTE) was then performed, revealing GLA with a left atrial volume index (LAVi) of 1101 ml/m². Meanwhile, transoesophageal echocardiography (TEE) confirmed calcification of P2 and P3 extending to the border of the mitral leaflet up to one third of the chordae tendineae, with moderate mitral stenosis and eccentric mitral regurgitation jet due to prolapse of A2. The patient was treated with optimal medical therapy while awaiting cardiac surgery. GLA is a rare complication of mitral valve disease and an almost forgotten cause of cardiomegaly. Diagnostic imaging procedures with echocardiography are a less invasive and readily available modality to establish the diagnosis of cardiomegaly.

Keywords: Erlenmeyer-shaped heart, Giant left atrium, Mitral stenosis, Mitral regurgitation, Junctional tachycardia.

INTRODUCTION

The prevalence of Rheumatic Heart Disease (RHD) remains high to date, especially in developing countries. One of the most common complications of RHD is chronic mitral valve disease. In some patients, this leads to left atrial enlargement, which can grow into gigantic proportions.¹

Giant Left Atrium (GLA) is defined as a huge enlargement of the left atrium, with a diameter exceeding 65 mm. It is a rare condition with a reported incidence of 0.3%.² The rare incidence of GLA results in diagnostic errors as its presentation on chest radiographs often mimics another cause of cardiomegaly.

This case report reveals an enlargement of cardiac silhouette due to a giant left atrial chamber in a patient with a history of moderate mitral stenosis and severe mitral regurgitation. The echocardiography showed GLA filled with spontaneous echo contrast, but without obvious thrombi.

CASE REPORT

A 52-year-old man, with a history of RHD, was referred to the emergency department of a tertiary hospital in Indonesia with main complaints of worsening shortness of breath and orthopnea. The patient also complained of bilateral leg oedema, whereas any complaints of chest pain on exertion, hoarseness, or difficulty swallowing were denied. Meanwhile, there was no history of diabetes mellitus, hypertension, coronary heart disease, or stroke. Prior to admission, the patient was on a regimen of diuretics, digoxin, and warfarin.

Physical examination revealed blood pressure 116/66 mmHg, regular pulse 109 beats per minute, and a respiratory rate of 28 breaths per minute with jugular vein distention. On chest auscultation, a regular heart sound was heard, with mid-diastolic and grade IV pansystolic murmurs at the apex radiating to the axilla. There were inspiratory crackles (rales) at both lung bases, slight abdominal swelling, and pitting oedema in both legs. Furthermore, laboratory examinations revealed mildly elevated liver enzymes and potassium levels. Electrocardiogram examination showed a junctional tachycardia rhythm of 110 beats per minute, while chest radiograph showed extreme cardiomegaly, nearly complete opacification, with a cardiothoracic ratio of 90%, an Erlenmeyer-shaped heart, and suspected massive pericardial effusion (Figure 1).

Transthoracic echocardiographic (TTE) examination showed an extremely huge left atrium of 13.9 mm x 12.3 mm on the apical four-chamber view (Figure 2), with LAVi of 1101 ml/m² (Figure 3). The left atrium was filled with spontaneous echo contrast but no definite thrombi. Mitral valve was calcified with moderate MS (valvular area of 1.4 cm² and mean PG of 7.15 mmHg) and Wilkins Score of 1-2-2-2, combined with severe mitral regurgitation due to restrictive opening of the posterior mitral leaflet (PML). Left ventricular systolic function was preserved (EF 73%), whereas the right ventricular systolic function was reduced (TAPSE 1.4 cm).

Transoesophageal echocardiography (TEE) was performed to determine the morphology of mitral valve disease. This examination revealed thickening and calcification of P2 and P3, extending to the border

Cite this article: Romdiyana A, Budiarto RM, Chandra E. Erlenmeyer-shaped Heart in a Patient with Giant Left Atrium due to Mixed Mitral Valve Pathology: A Neglected Case in a Rural Area in Indonesia. *Pharmacogn J.* 2022;14(6): 829-832.



Figure 1: Erlenmeyer-shaped heart, and suspected massive pericardial effusion.

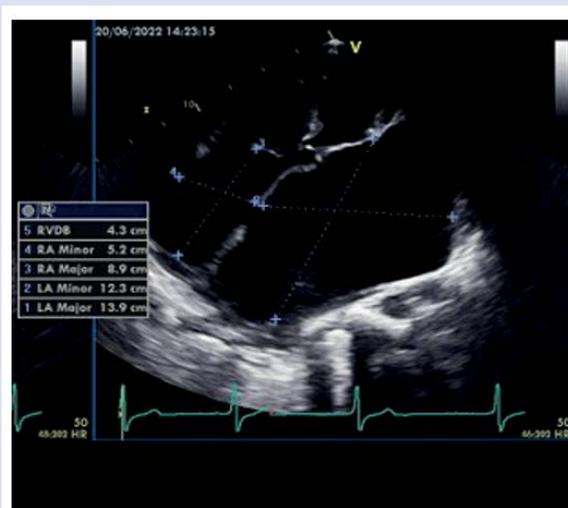


Figure 2: Transthoracic echocardiographic (TTE) examination showed an extremely huge left atrium of 13.9 mm x 12.3 mm on the apical four-chamber view.

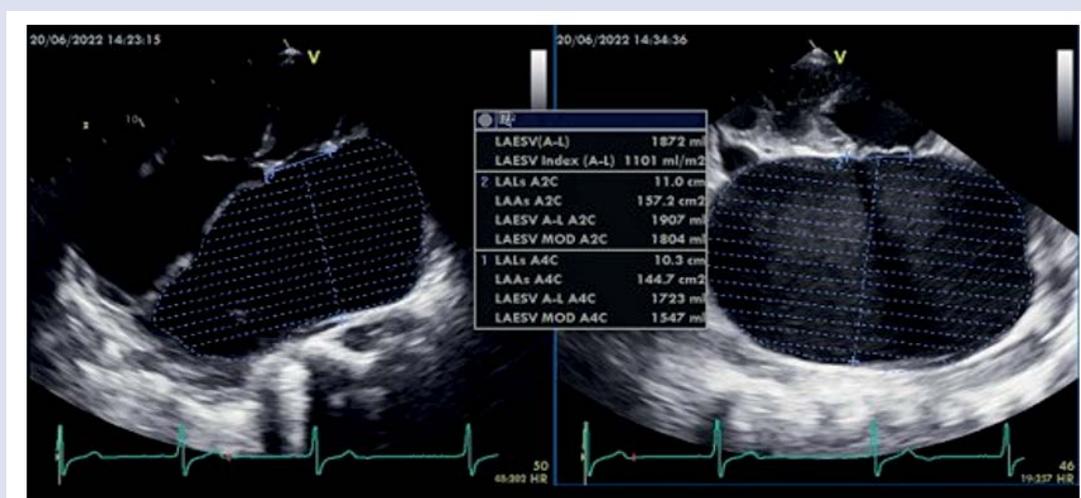


Figure 3: Transthoracic echocardiographic (TTE) examination showed an extremely huge left atrium of 13.9 mm x 12.3 mm on the apical four-chamber view with LAVi of 1101 ml/m²

of the mitral leaflet and reaching one third of the chordae tendineae, accompanied by prolapse of A2 with severe mitral regurgitation.

The patient was admitted to the hospital for congestive heart failure according to the clinical presentation and treated with intravenous diuretic agents to reduce congestion, as well as with anticoagulants to prevent thrombus formation. After aggressive diuresis, there was improvement in symptoms and hemodynamic. Coronary angiography with normal coronary arteries was then carried out to prepare the patient for surgical intervention.

DISCUSSION

The left atrium can be massively enlarged, known as GLA, which may result from a long-standing sequela of RHD with mitral valve-related complications, predominantly mitral regurgitation.³ However, patients with chronic mitral valve disease are not always associated with GLA; only 19% of them can develop such condition.⁴ Long term mitral valve disease causes left atrial enlargement as a compensatory mechanism for prolonged intracavitary pressure and volume overload. This has a reservoir effect as it reduces pulmonary congestion, thereby protecting the lungs from pulmonary hypertension and oedema.⁵

On chest radiographs, left atrial enlargement initially manifests as posterior chamber enlargement with subsequent progression to a rightward bulge, causing a double contour of the right heart border ('double contour sign' on posteroanterior chest radiography) as well as splaying or widening of the carina.⁶ As left atrial dilatation progresses, GLA should always be considered in patients with a history of rheumatic mitral disease who develop a lateral extension of the heart border, reflecting an enlarged left atrium towards the right lateral side of the chest wall.³ Such condition is frequently misdiagnosed as mass lesion or pleural or pericardial effusion. In this case report, the patient has an Erlenmeyer-shaped heart mimicking a massive pericardial effusion.

Echocardiographic assessment of left atrial size by M-mode or two-dimensional echocardiography in parasternal long-axis view is a measurement of its anteroposterior linear dimension. Although it has been shown to correlate with angiographic measurements and has been used extensively in clinical practice and research, this two-dimensional measurement is a less accurate and unreliable representation of the true left atrial size.⁷ Assessment of the left atrial by linear measurement assumes a constant relationship between all left atrial dimensions, which may not be correct, especially in the presence of asymmetric left atrial enlargement in the superior-inferior and medial-lateral axes. This underlies the recommendation of the current guidelines for determining left atrial volume in the assessment of left atrial geometry.⁸ Three-dimensional echocardiography provides a more accurate and reproducible estimate of left atrial size, compared to reference standards such as magnetic resonance imaging (MRI), biplane contrast ventriculography, and cine computed tomography (CT).⁹ In addition, the strength of the association between cardiovascular diseases and left atrial size is stronger for left atrial volume than for left atrial linear dimensions.¹⁰

Left atrial enlargement is associated with blood stasis and thrombus formation. Furthermore, huge enlargement of the left atrium is prone to develop various complications including compressive symptoms of dysphagia and hoarseness, thrombus formation, and thromboembolic events. The risk of thromboembolism increases with left atrial size regardless of anticoagulation.¹¹ In addition, left atrial enlargement or LA dilatation leads to thinning and fibrosis of myocardium and conducting fibers, thus resulting in electrical tissue inhomogeneity,

impaired electrical activation, and contractility. Every 5 mm increase in left atrial diameter escalates the development of atrial fibrillation by 39%.¹²

Surgical treatment of giant left atrium aims to correct mitral valve abnormalities, relieve compression symptoms, prevent thromboembolism, and reverse atrial fibrillation to normal sinus rhythm. Two procedures have been applied in the management of mitral valve surgery with a giant left atrium: (1) performing mitral valve surgery with left atrial volume reduction (debulking); and (2) having mitral valve replacement alone.²

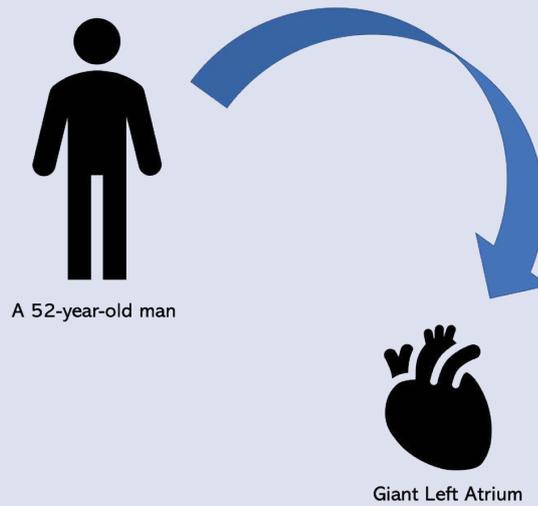
CONCLUSION

A correct diagnosis of GLA is at times not possible to reach with routine chest radiography alone and the condition may be misdiagnosed as another condition causing massive cardiomegaly. Therefore, it is extremely important to link the medical history of patients with the findings on chest radiography and to further evaluate the patients with more detailed imaging studies prior to performing any invasive procedures.

REFERENCES

1. EL Maghraby A, Hajar R. Giant left atrium: A review. *Heart Views*. 2012;13(2):46-52.
2. Apostolakis E, Shuhaiber JH. The surgical management of giant left atrium. *Eur J Cardiothorac Surg*. 2008;33(2):182-90.
3. Hurst JW. Memories of patients with a giant left atrium. *Circulation*. 2001;104(22):2630-2.
4. Di Eusanio G, Gregorini R, Mazzola A, Clementi G, Procaccini B, Cavarra F, *et al.* Giant left atrium and mitral valve replacement: risk factor analysis. *Eur J Cardiothorac Surg*. 1988;2(3):151-9.
5. Schwammenthal E, Vered Z, Agranat O, Kaplinsky E, Rabinowitz B, Feinberg MS. Impact of atrioventricular compliance on pulmonary artery pressure in mitral stenosis: An exercise echocardiographic study. *Circulation*. 2000;102(19):2378-85.
6. Shah BN, Rubens M. Giant left atrium: A forgotten cause of cardiomegaly. *Postgrad Med J*. 2012;88(1045):673-5.
7. Wade MR, Chandraratna PA, Reid CL, Lin SL, Rahimtoola SH. Accuracy of nondirected and directed M-mode echocardiography as an estimate of left atrial size. *Am J Cardiol*. 1987;60(14):1208-11.
8. Lang RM, Bierig M, Devereux RB, Flachskampf FA, Foster E, Pellikka PA, *et al.* Recommendations for chamber quantification: A report from the American Society of Echocardiography's Guidelines and Standards Committee and the Chamber Quantification Writing Group, developed in conjunction with the European Association of Echocardiography, a branch of the European Society of Cardiology. *J Am Soc Echocardiogr*. 2005;18(12):1440-63.
9. Rodevan O, Bjornerheim R, Ljosland M, Maehle J, Smith HJ, Ihlen H. Left atrial volumes assessed by three- and two-dimensional echocardiography compared to MRI estimates. *Int J Card Imaging*. 1999;15(5):397-410.
10. Tsang TSM, Abhayaratna WP, Barnes ME, Miyasaka Y, Gersh BJ, Bailey KR, *et al.* Prediction of cardiovascular outcomes with left atrial size: Is volume superior to area or diameter? *J Am Coll Cardiol*. 2006;47(5):1018-23.
11. Goldstein LB. Left atrial enlargement: A cause of stroke? *Can Med Assoc J*. 2011;183(10):1129-30.
12. Vaziri SM, Larson MG, Benjamin EJ, Levy D. Echocardiographic predictors of nonrheumatic atrial fibrillation. The Framingham Heart Study. *Circulation*. 1994;89(2):724-30.

GRAPHICAL ABSTRACT



ABOUT AUTHORS



Aprilia Romdiyana is a student in Department of Cardiology and Vascular Medicine, Airlangga University, Surabaya, Indonesia.

Cite this article: Romdiyana A, Budiarto RM, Chandra E. Erlenmeyer-shaped Heart in a Patient with Giant Left Atrium due to Mixed Mitral Valve Pathology: A Neglected Case in a Rural Area in Indonesia. *Pharmacogn J.* 2022;14(6): 829-832.