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CASE REPORT
Congenital heart disease

When the left atrium becomes a monster: a case report

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Background Congenital left atrium (LA) aneurysms are extremely rare entities in clinical practice and most frequently involve the atrial appendage and rarely arise from the body of LA, We report a case of giant LA aneurysm compressing heart and presenting in a very late stage.

Case summary A 31-year-old male, who was diagnosed to have dextrocardia, rheumatic heart disease, and atrial fibrillation and was kept on medical treatment long time ago, presented with congestive heart failure symptoms and cardiogenic shock. Emergency transthoracic echocardiography was done revealing situs solitus with aneurysmally dilated LA pushing heart to the right side (dextro-posed heart), moderate mitral regurgitation, and severe pulmonary hypertension, however, pulmonary artery anatomy could not be properly visualized so computed tomography (CT) was preformed confirming diagnosis and revealing compressed pulmonary arterial tree by the dilated LA, unfortunately patient died before proceeding to surgical intervention.

Discussion Congenital left atrial aneurysms are extremely rare anomaly and may be associated with significant morbidity. And, therefore, should be remembered as a potential anatomic cause of atrial arrhythmias or embolic phenomena, or both. The diagnosis may be easily established through non-invasive complementary techniques, such as echocardiography, CT, and cardiac magnetic resonance imaging. Symptomatic patients, those with large aneurysm or compelling indications for surgery should undergo surgical resection.

Keywords Case report • Left atrium aneurysm • Congestive heart failure • Dextro-position • Rheumatic heart disease • Mitral regurgitation • Atrial fibrillation

Learning points

- Congenital aneurysms of the left atrium are extremely rare anomalies.
- Echocardiography, computed tomography, and cardiac magnetic resonance imaging are crucial in the diagnoses and management.
- Surgical resection is the mainstay of treatment.

Introduction

Aneurysms of the left atrium (LA) are a rare anomaly,[1] that could be either congenital or acquired, congenital aneurysms are present as an isolated pathology whereas acquired cases are secondary to mitral valve disease, left ventricular (LV) dysfunction, or conditions that lead to elevated left atrial pressure.[2]

Congenital LA aneurysms are extremely rare entities in clinical practice and most frequently involve the atrial appendage or rarely arise from the body of LA.

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Timeline

Since childhood	Diagnosed to have rheumatic heart disease and kept on long-term penicillin
At the age of 25 years	Diagnosed to have atrial fibrillation and kept on anticoagulation
At age of 30 years (1 year before presentation)	Exertional dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, and bilateral lower limb oedema
At age of 30 years and 6 months (6 months before presentation)	Progressive weight loss and dysphagia
At presentation	Congestive heart failure symptoms and cardiogenic shock

Case presentation

A 31-year-old male presented with congestive heart failure (CHF) symptoms and cardiogenic shock, his condition started since childhood when he was diagnosed having rheumatic heart disease (RHD) and was kept on long-term penicillin. At the age of 25 years, he developed a recurrent attacks of palpitation, diagnosed at that time to have atrial fibrillation (AF) and was kept on anticoagulation. One year before presentation to our hospital, he started complaining of exertional dyspnoea (New York Heart Association Class II–III), orthopnoea, paroxysmal nocturnal dyspnoea, and bilateral lower limb oedema. Transthoracic echocardiography (TTE) was done at that time stating that he had dextrocardia and rheumatic appearance of the mitral valve leaflets with moderate mitral regurgitation (MR), normal mitral valve opening with no mitral stenosis and peak/mean diastolic gradient of 4/2 mmHg, respectively, dilated LA and severe pulmonary hypertension (PHT). Six months before presentation, the patient started to develop progressive weight loss and dysphagia. On presentation, he was complaining of dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, bilateral lower limb oedema up to knee, severe weight loss, and dysphagia. The patient has no past medical history of relevance. He was on warfarin 5 mg daily with International normalized ratio adjusted at 2.5 and long-term penicillin.

On general examination, signs of shock were present with low arterial blood pressure (70/40 mmHg), tachycardia with heart rate of 120 b.p.m. on AF. In addition dyspnoea, orthopnoea, cachexia, congested neck veins with absent A wave and prominent V wave, enlarged tender liver, and bilateral lower limb oedema up to knee were very evident reflecting CHF. His cardiovascular examination revealed apex to be at the third right intercostal space outside midclavicular line with Grade III/VI pan systolic murmur of MR best heard over the apex.

Informed consent was obtained from the patient. He was admitted to coronary care unit, intravenous inotropes were started (noradrenaline 0.1 μg/kg/min—dobutamine 20 μg/kg/min), diuretic (frusemide 40 mg bid), and warfarin 5 mg were continued.

Emergency TTE was performed revealing situs solitus with normal connexions (atrioventricular and ventriculoatrial concordance), dextro-posed heart, hugely dilated LA pushing heart to the right side (*Figure 1*), mild diffuse thickening of both mitral valve leaflets with no restriction of mobility of any of them. There was eccentric mitral insufficiency of moderate intensity which could be either primary due to mitral valve disease or secondary to dilation of the mitral ring and lack of cooptation of both leaflets, which was properly distorted by the aneurysm of the LA resulting in moderate eccentric MR (*Figure 2*). Doppler assessment of the mitral flow revealed peak/mean diastolic gradient of 4/2 mmHg, respectively (not coinciding with atrial dilatation) and MR with vena contracta of 4 mm. Other quantitative measurements for assessment of severity of MR was not performed by the time of examination due to emergency state of the patient and whether those parameters can be validated in such enormous LA enlargement was questioned. The subvalvular mitral apparatus was normal with intact cords and papillary muscles. There was evidence of heavy spontaneous echocardiographic contrast with no evidence of thrombi in LA appendage, normal tricuspid valve morphology with severe tricuspid regurgitation (*Figure 3*) and PHT with estimated right ventricular systolic pressure (RVSP) of 62 mmHg, normal aortic valve morphology, and normal LV and RV size with good biventricular systolic function. However, pulmonary arterial tree could not be properly visualized.

Multi-slice computed tomography (CT) was performed to assess pulmonary arterial tree, assess situs, and relation of LA to surrounding structures. Multi-slice CT confirmed being situs solitus with dextro-version, hugely dilated LA measuring 130 mm × 150 mm (*Figure 4*), occupying left chest cavity, extending from diaphragm downward to neck upward, pushing heart completely to the right side. The LA was compressing main pulmonary artery and extremely compressing the right pulmonary artery (RPA) towards aorta causing significant RPA stenosis (*Figure 5*), explaining the elevated RVSP. Few

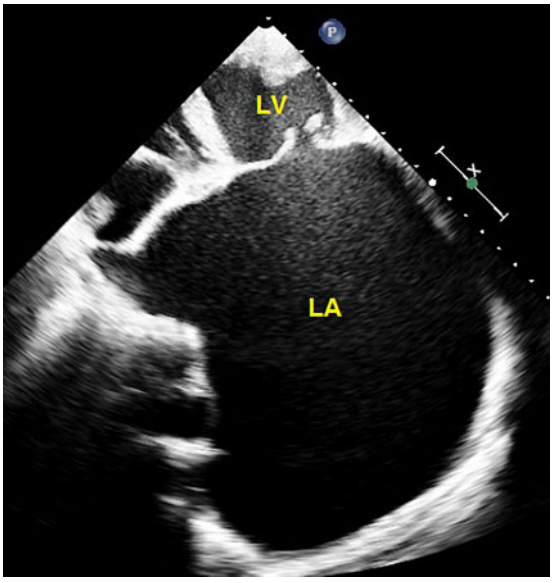

Figure 1 Transthoracic echocardiography apical four-chamber view showing aneurysmally dilated left atrium. LA, left atrium; LV, left ventricular.

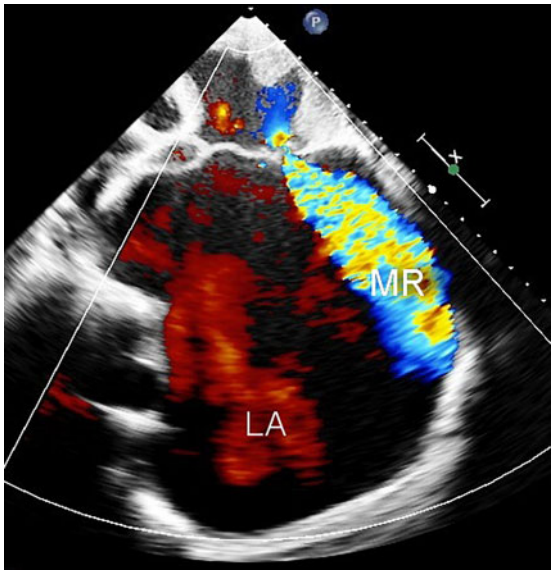

Figure 2 Transthoracic echocardiography apical four-chamber view showing moderate mitral regurgitation. LA, left atrium; MR, mitral regurgitation.

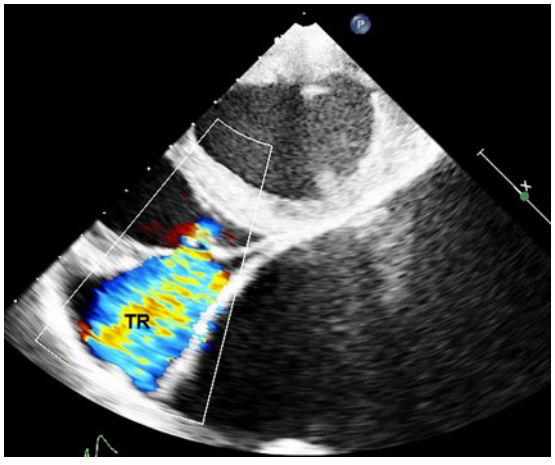

Figure 3 Transthoracic echocardiography apical four-chamber view showing severe tricuspid regurgitation. TR, tricuspid regurgitation.

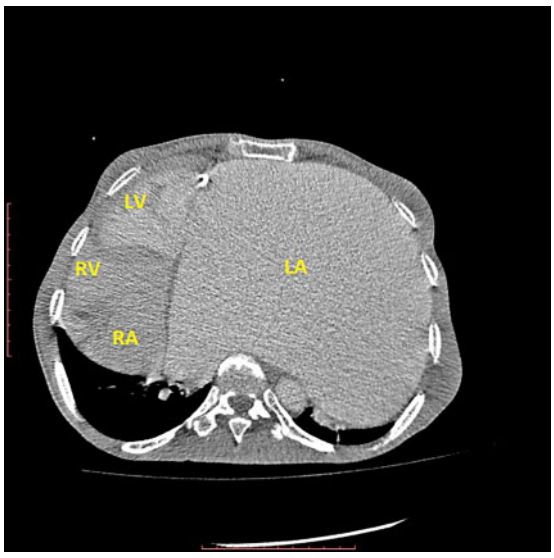

Figure 4 Multi-slice computed tomography showing aneurysmally dilated left atrium. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

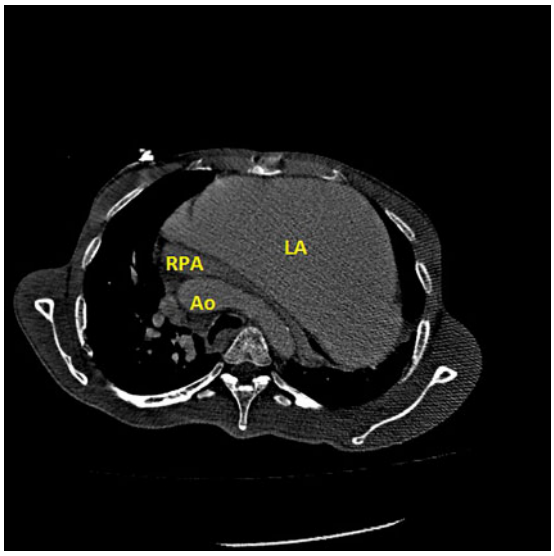

Figure 5 Multi-slice computed tomography showing left atrium compressing right pulmonary artery. Ao, aorta; LA, left atrium; RPA, right pulmonary artery.

hours after presentation, the patient developed Brady-asystole cardiac arrest. Cardiopulmonary resuscitation was performed for 45 min with no response.

Discussion

Isolated aneurysm of the atrium is a rare congenital malformation seen in clinical practice and occurs in either the left or right atrium including bilaterally. It was first described in 1938 by Semans and Taussig.[1] There are about 50 cases reported in the literature[3] with the majority of cases affecting LA and of congenital origin without evidence of predisposing factor, inflammatory, or degenerative processes. Acquired cases are usually associated with inflammatory process. The cause has been postulated to be dysplasia of the pectinate muscle and related bundles of muscles of LA. The aneurysm most commonly involves the LA appendage rather than the whole left

atrial body, similar to our case. These aneurysms can be seen with intact pericardium[4] or with a pericardial defect[5] causing aneurysmal protrusion resembling dog's ears.[6] The definite rate of growth of LA has not been identified yet.

In Egypt, RHD is endemic and, because of fear of the condition, there are problems with over-diagnosis of RHD. The patient stated that he was diagnosed since childhood to have RHD, although at the time of presentation, there was no document confirming that previous diagnosis. However, given the increased thickness of mitral valve leaflets and having RHD endemic in Egypt, rheumatic mitral valve disease could be contributing to the LA dilatation.

The age of presentation is variable ranging from few months till sixth or seventh decade with the most common being second or third decade. The enlarging LA aneurysm does not only compress adjacent cardiac structures but can also cause serious symptoms mainly arrhythmias or thromboembolic manifestations. The patients can also present with heart failure symptoms related to compression of the pulmonary veins, chest pain related to compression of the left coronary artery,[7] or cardiac tamponade due to limitation of diastolic expansion of left ventricle. To the best of our knowledge, this case is the first in literature presenting with this giant LA and in a very late stage being in cardiogenic shock and CHF symptoms. This is due to blood pooling in the aneurysmally dilated LA, with compression of the main PA and RPA causing severe PHT, stagnation of blood flow in the LA causing cardiogenic shock and mostly compression of the pulmonary veins resulting in CHF and dysphagia which may be caused by CHF or compression of the oesophagus.

The basis of diagnosis is imaging modalities mainly TTE and transoesophageal echocardiography.[8] Cardiac magnetic resonance imaging (CMRI) and CT can play an important role in ruling out differential diagnosis, detect relation of the aneurysm to coronary arteries, pulmonary veins, and other cardiac structures.

Transthoracic echocardiography is the most feasible and commonly used for rapid diagnosis especially in the setting of emergency. In the context of marked LA dilation, can we depend on the standard echocardiographic parameters for assessment of severity of MR including vena contracta width, effective regurgitant orifice area, regurgitant volume, regurgitant fraction, and pulmonary vein flow? I think this should be further studied.

Role of medical treatment is only limited to anticoagulation for prevention of thromboembolism and anti-arrhythmic for management of arrhythmias. Because of the serious morbidity and mortality related to the aneurysm, the treatment of choice is surgical resection.[9]

Conclusion

Congenital left atrial aneurysms are extremely rare anomaly and may be associated with significant morbidity. Therefore, it should be remembered as a potential anatomic cause of atrial arrhythmias or embolic phenomena, or both. The diagnosis may be easily established through non-invasive complementary techniques, such as echocardiography, CT, and CMRI. Symptomatic patients, those with large aneurysm or compelling indications for surgery should undergo surgical resection.

Lead author biography

Khaled A. Shams, graduated from the Faculty of Medicine, Ain Shams University in 2005. He was a trainee resident in the Cardiology Department, Ain Shams University Hospitals from 2007 to 2010. He obtained a Master of Science of Cardiology in 2010 and obtained medical doctorate of Cardiology in 2010. He was a visiting fellow in Congenital and Structural Heart Disease Unit, Cardiology Department, Ain Shams University Hospitals, and become lecturer of Cardiology at Cardiology Department, Helwan University in 2018.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: no conflict of interest .

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