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Case Report

Left atrial ball valve thrombus in restrictive cardiomyopathy and normal mitral valve: Loose cannon in heart

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ABSTRACT

Left atrial ball valve thrombus is an unusual condition, especially in patients with normal mitral valve. In the present case, we describe a 61-year-old female with restrictive cardiomyopathy who presented with a large left atrial ball valve thrombus, which subsequently embolized to right carotid artery and was treated with intravenous thrombolysis. This case provides useful insight into the genesis of such thrombi and highlights management dilemmas of a rare clinical problem.

1. Introduction

Ball valve thrombi almost always form in the setting of mitral valve disease.[1] There are only three previous reports of a ball valve thrombus in the left atrium in the presence of a normal mitral valve.[2–4] We describe one such case and discuss the insights this provides into the pathophysiology of ball valve thrombi.

2. Case report

A 61-year-old female presented with complaints of non-progressive breathlessness and palpitations on exertion of 20–25 years duration. These symptoms occurred on walking 2 km on plain ground or on climbing 8–10 steps. There was no history of palpitations at rest, giddiness, syncope, or chest pain. She gave history suggestive of three episodes of transient ischemic attacks. In each episode, she had sudden onset weakness of a lower limb lasting around 2 h. The first episode occurred 10 months earlier and the subsequent 2 episodes were at approximately 3 months interval each. She also had transient swelling of feet and body 3 weeks earlier that had subsided with treatment. She was not hypertensive or diabetic. She was a non-smoker, did not consume alcohol, and had no history of jaundice.

She was 6th among 7 siblings. Three of her siblings had heart problems, the details of which were not known. There was no family history of sudden death.

On examination, she was comfortable at rest. Heart rate was 110/min, irregularly irregular. Blood pressure was 120/70 mmHg supine and 100/60 mmHg standing. Jugular venous pressure was raised 10 cm above sternal angle. The apex beat was displaced outwards, being present in 5th intercostal space 1 cm

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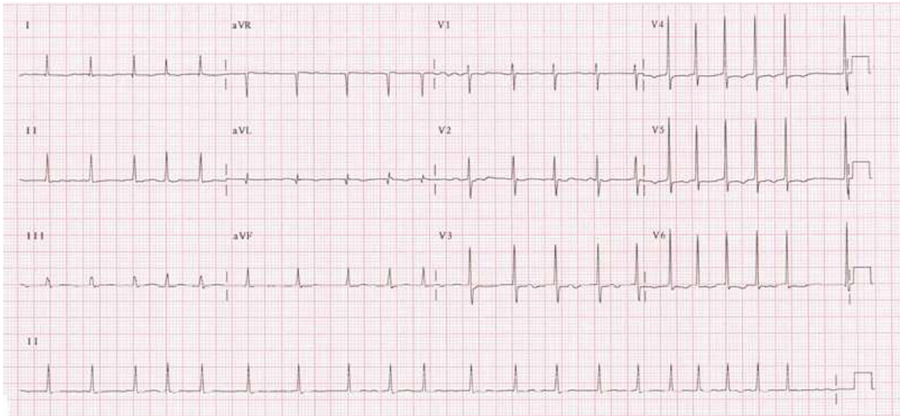

Fig. 1 – ECG showing atrial fibrillation with ventricular rate of 120/min and left ventricular hypertrophy.

outside the mid-clavicular line and there was a grade II left parasternal heave. Heart sounds were normal and there were no murmurs.

The ECG showed atrial fibrillation with a heart rate of 124/min and left ventricular (LV) hypertrophy (Fig. 1).

Echocardiographic examination showed biatrial enlargement (Fig. 2a). Left atrial dimension in the parasternal long axis view was 52 mm. There was mild LV hypertrophy with normal LV contractility. Mild mitral and tricuspid regurgitation were present. The estimated right ventricular systolic pressure was 55 mmHg. She also had moderate pulmonary regurgitation with an end diastolic gradient of 13 mmHg. The aortic valve was normal. The septal mitral annular velocity was 6 cm/s. E wave deceleration was 100 ms. Inferior vena cava was 16 mm in diameter and was not collapsing with inspiration. There was a large round mobile left atrial mass of size 21 × 19 mm that was not attached to the left atrial wall or the mitral valve (Fig. 2b). The mass was freely mobile in different planes and was free floating. It was prolapsing into the mitral valve till the tips of the mitral valve leaflets from the left atrium but was not prolapsing across the mitral valve. There also was a left atrial appendage thrombus with a maximal dimension of 10 mm.

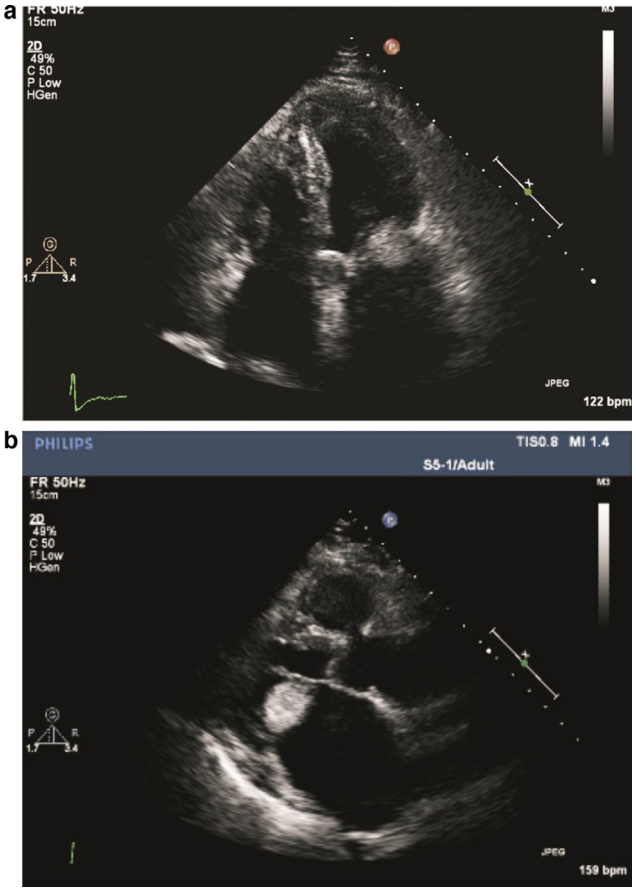

Fig. 2 – (a) Apical four chamber view showing biatrial enlargement and ball valve thrombus prolapsing into the mitral valve. (b) Parasternal long axis view showing a ball valve thrombus at mitral valve.

She was diagnosed to have restrictive cardiomyopathy in atrial fibrillation and left atrial ball valve thrombus. Restrictive cardiomyopathy was diagnosed on the basis of a predominant right heart failure with normal LV systolic function, biatrial enlargement, and Doppler features suggestive of restrictive physiology. She was advised immediate hospitalization but was not willing at that time. She was admitted the next day and given 1 dose of subcutaneous enoxaparin immediately. 2 h later, she had sudden onset left hemiplegia. Repeat echocardiography showed that the large mobile left atrial mass had disappeared but the left atrial appendage thrombus was persisting. Non-contrast CT scan of the head was normal and she was thrombolysed with alteplase approximately 100 min after the onset of hemiplegia. CT angiography done 90 min after thrombolysis showed a large thrombus in the right common carotid artery. She continued to have grade 0 power in both the left upper and lower limbs and was planned for immediate mechanical thrombolysis. However, the digital subtraction angiogram done prior to the mechanical thrombolysis was normal suggesting that the thrombus in the right common carotid artery had completely thrombolysed by then. Subsequent non-contrast CT scan of the head showed a small focal bleed in the infarcted segment that soon resolved.

Her investigations showed hemoglobin of 12.8 g%, serum creatinine 1.4 mg/dl, and ESR 3 mm fall in 1st hour. Liver function tests, blood sugar, lipid profile, urine examination, and thyroid function tests were normal. Serum troponin I was

negative. Serum electrophoresis showed mild hypergammaglobulinemia. M band was not seen. Urine Bence-Jones protein was negative. Fine needle aspiration cytology from abdominal fat pad was negative for amyloid. Genotyping was not carried out.

She was clinically diagnosed as having idiopathic restrictive cardiomyopathy and managed with anticoagulation, aspirin, and diuretics. Power in left lower limb has returned to near normal while in the left upper limb she has grade III/V power.

3. Discussion

Ball valve thrombus is uncommon and almost always seen in the setting of mitral valve disease – either native valve mitral stenosis or in the presence of a prosthetic mitral valve. In both these situations, the risk of peripheral embolism of the large ball valve thrombus is reduced due to a restricted mitral orifice. Ball valve thrombus with normal mitral valve is extremely rare. It has been reported in a case of hypertrophic cardiomyopathy[3] and a child with restrictive cardiomyopathy.[4] Our patient also had a normal mitral valve. On echocardiography, the free floating thrombus was repeatedly prolapsing into the mitral orifice but not embolizing since the mitral valve was restrictive relative to the thrombus size. A normal mitral valve, however, has a valve area of 4–6 cm^2 and there would always be a risk of peripheral embolism. This thrombus was obviously not attached to the left atrial wall since it was randomly moving around in different planes in different areas of the left atrium.

The ball valve thrombus was present in the left atrium at least a day before it embolized into the carotid artery. Why did this thrombus embolize? It had been repeatedly prolapsing into the mitral valve orifice repeatedly for at least 24 h without doing so. The exact reason for the embolization cannot be ascertained since an echocardiographic examination was not carried out in the minutes just prior to embolization. The likely reason is that the thrombus is a dynamic structure. There could be some lysis at the surface resulting in a slight reduction in size enabling its passage through the mitral valve. Fragmentation of thrombus is less likely since the CT angiography showed that the thrombus was restricted to the right common carotid artery. A thrombus that fragments is more likely to embolize to multiple arteries.

This case also provides an important insight into the formation of a ball valve thrombus. The ball valve thrombus must have initially started as an attachment to the left atrial wall likely the left atrial appendage. The thrombus has to be attached initially to the left atrial wall because a small free floating thrombus would have embolized in the presence of a normal widely opening mitral valve. The likely site of formation is the left atrial appendage since associated left atrial thrombus was present in this patient. The thrombus would then have gradually extended into the left atrial cavity. It would only have detached from the left atrial wall once it had attained a size large enough to prevent its embolization across the mitral valve.

This case also illustrates the fact that this thrombus was not a result of embolization from the pulmonary vein or the right atrium. A free floating thrombus coming from a pulmonary vein would be smaller and would thus embolize straight away. Earlier case reports, which have reported ball valve thrombi in the setting of mitral valve disease, could not exclude this possibility. This is because even smaller emboli may not be able to embolize and grow in size in the setting of a restrictive mitral orifice.

The fact that the ball valve thrombus is spherical indicates that the thrombus undergoes remodeling after it becomes a free floating structure. This is because a thrombus that has just detached from the left atrial appendage or the left atrial wall will not be spherical but will have at least some contours to match its attachment to the atrial wall. It has been proposed that this remodeling may be because of sculpting effect of multiple collisions with left atrial wall and mitral valve apparatus and centripetal force due to spinning motion of the thrombus.[5] This process of remodeling is also likely the cause of embolization since it may allow a thrombus that was not able to cross the mitral valve to change its shape and size and thus be able to embolize.

This case also highlights the possibility of other associated thrombi in patients with ball valve thrombus. Our patient also had a left atrial appendage thrombus.

This case also raises issues of management. There are two possible management approaches. The first approach is to surgically remove the thrombus using cardiopulmonary bypass along with ligation of the left atrial appendage. This would prevent a devastating systemic embolization. Subjecting a patient with restrictive cardiomyopathy to open heart surgery is fraught with problems. The surgery only removes the thrombus and the underlying heart disease still remains. The high filling pressures post-operatively can result in problems of persistent pleural effusions. The second approach of wait and watch is also risky. As seen in our patient, the large thrombus can embolize early resulting in life-threatening complications. Hence, despite all caveats, surgical removal of the thrombus on an urgent basis possibly remains the best approach.

This case also demonstrates the amenability of ball valve thrombus to systemic thrombolysis even after embolization, which has not been previously described. There is a single case report of successful treatment of free floating left atrial ball valve thrombus with anticoagulants.[6]

4. Conclusions

Left atrial ball valve thrombi are extremely rare in the absence of mitral valve disease. This case report clarifies a number of important issues on the formation and propagation of ball valve thrombi – facts that cannot be ascertained in patients with mitral valve disease and restricted mitral valve orifices. The thrombus formation in this patient is likely to have started in the left atrial appendage and with initial attachment to the left atrial wall. Ball valve thrombus would form when such a thrombus would get detached from the left atrial wall after attaining a large size.

Conflicts of interest

The authors have none to declare.

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