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Case Report

Parachute mitral valve with late presentation: rare case reports

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ABSTRACT

Congenital mitral stenosis involves the annulus, the zone immediately above and contiguous with the annulus, the leaflets, the chordae tendineae, and the papillary muscles. In a parachute mitral valve (PMV), all chordae tendineae which are usually shorter and thicker than normal type, inserted into this single papillary muscle. This condition restricts the motion of leaflets and obstructs the blood flow into the left ventricle during diastole. Here we present two cases of severe congenital mitral stenosis with severe pulmonary hypertension due to parachute mitral valve that allowed survival into adulthood without any specific treatment.

Keywords: Congenital mitral stenosis, Parachute mitral valve, Shone's complex

INTRODUCTION

The mitral valve is a functional complex that relies on normal morphology, geometrical relations and function of all its constituents: annulus, leaflets and the subvalvular apparatus including chordae tendineae and papillary muscles.[1] In a parachute mitral valve (PMV) all chordae, which are usually shorter and thicker, are inserted into a single papillary muscle.

This condition restricts the motion of leaflets and obstructs the blood flow into the left ventricle during diastole.[2] The incidence rate has been estimated at 0.6% of necropsy cases of congenital heart disease and 0.21% to 0.42% of clinical cases.[3] The average age at diagnosis is 1.5 years (range one day to 12.5 years) and the average age of onset of symptoms is 1.6 years.[3] Mean age of survival for PMV is 10 years. Here we present two cases of congenital mitral stenosis (PMV) with severe pulmonary arterial hypertension which were diagnosed at the age of 30 and 42 years respectively.

CASE REPORT

Case 1

The 30-year-old lady was referred in October 2017, presented with exertional dyspnea NYHA class II, which had begun 6 months ago and a recent hospitalization for breathlessness two weeks back. Her past medical history was noncontributory. Her general growth and development were within normal limits. Cardiovascular examination revealed a grade 3/4 diastolic murmur at the apex without radiation. ECG showed sinus rhythm with left atrial (LA) enlargement. Trans-thoracic echocardiography (TTE) revealed normal left ventricular size (LVEDD: 42mm) and enlarged left atrium (LA size: 80x64mm). Two papillary muscles were seen in parasternal short-axis view at mid-papillary level. However, all of the thickened and shortened chordae were seen converged into posteromedial papillary muscle. Anterolateral papillary muscle was rudimentary without any chordal attachments. Dysplastic chordae

tendineae and thickened mitral valve leaflets could be seen well in apical four-chamber view also. The mitral valve orifice was eccentrically located at the medial side with a mean pressure gradient of 12mmHg across the MV. No other mitral valve related anomaly was found. Estimated right ventricular systolic pressure was 64mmHg, suggestive of moderate to severe pulmonary hypertension. Biventricular systolic function was normal, LVEF: 60% (Figure 1). Aortic valve was bicuspid and interrogation of aortic arch and descending aorta did not reveal any Coarctation. Findings were confirmed on cardiac MRI (Figure 2). Patient underwent successful mitral valve replacement (MVR).

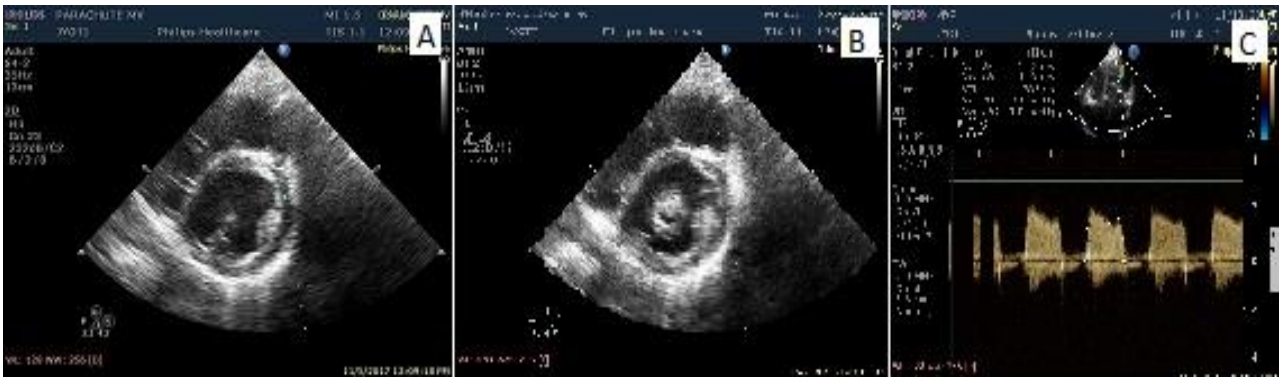

Figure 1: TTE images: A) Short axis view shows two closely placed papillary muscles B) Eccentrically placed mitral orifice as all chordaes inserting only on posteromedial papillary muscle C) Mean gradient of 12 mmHg across mitral valve.

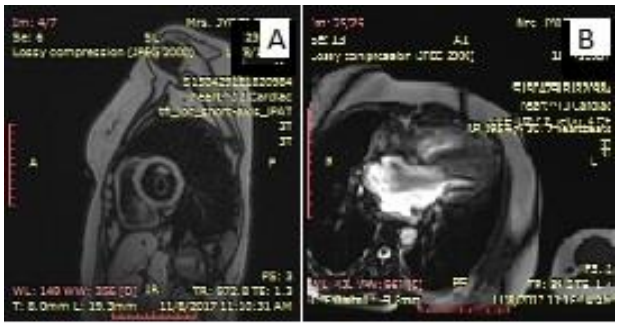

Figure 2: Cardiac MRI images A) Unattached anterolateral papillary muscle B) Attachment of all chordae to postermedial papillary muscle only.

Case 2

The 42-year-old male was referred to us for balloon mitral valvotomy (BMV) with a diagnosis of rheumatic heart disease with mitral stenosis. He had exertional dyspnea NYHA class II. His general growth and development were in normal condition. He had a localized ¾ mid diastolic murmur at the apex. ECG showed sinus rhythm with LA enlargement. TTE revealed normal left ventricular size (LVEDD: 44mm), enlarged left atrium (LA: 55x49mm). There was single papillary muscle seen at mid-papillary level at 4'O clock position in the parasternal short axis view.

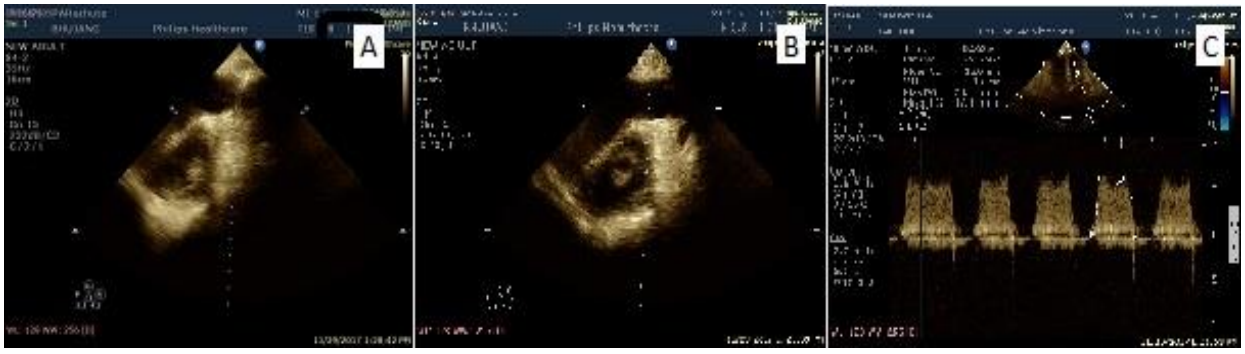

Figure 3: TTE images: A) Short axis view showing single laterally placed papillary muscle B) Eccentrically placed mitral orifice as all chordae inserting on single antero-lateral papillary muscle C) Mean gradient of 16 mmHg across mitral valve.

All thickened and shortened chordae converged into this single papillary muscle which was well visualized in apical four chamber view also.

There was no thickening of the leaflets, commissural fusion or restricted mobility of posterior mitral leaflet to suggest rheumatic etiology. The mitral valve orifice was eccentrically located at the lateral side with mean gradient of 16mmHg across the MV. Estimated RV systolic pressure was 84mmHg (Figure 3). Associated defects were ruled out. Thus, we diagnosed it as a case of congenital mitral stenosis, due to parachute mitral valve with severe PAH. Patient was referred for MVR.

DISCUSSION

Congenital mitral stenosis is an unusual congenital defect of the mitral valve most frequently accompanied by other abnormalities of the left heart such as aortic coarctation, valvular and subvalvular aortic stenosis and supravalvular mitral membrane, generally named as the complex of Shone.[2] Due to hemodynamic effects, diagnosis is usually made during infancy and childhood and remaining undiagnosed till adulthood is very uncommon. However, a number of incomplete forms have been detected in adults.[4-6] To best of our knowledge and according to the systematic review by Hakim et al, only nine cases of adults with PMV have ever been reported till 2010.[7] an isolated case of adult parachute mitral valve in a 29 year old female was reported in 2016 but none of those had severe mitral stenosis leading to severe pulmonary arterial hypertension.[8]

Incidence rate has been estimated at 0.6% of necropsy cases of congenital heart disease and 0.21% to 0.42% of clinical case.[9] Mean age at first hospitalization for cardiac disease was 2.1 years (range birth to 9.5 years) but it was not until a mean age of 4.5 years (range 21 days to 13 years) that the definitive diagnosis of congenital mitral stenosis was established. Despite occasional operative success in congenital MS, it remains a challenging lesion with a poor prognosis. Eighteen-year survival in all patients including operative management was 18%. Complete postoperative hemodynamic assessment is needed, sometimes serially, to evaluate long-term effects of operation as the children grow.[10]

CONCLUSION

This case reports highlight the need to familiarize clinicians about this uncommon entity i.e. parachute mitral valve and helps to differentiate it from other causes of mitral stenosis.

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