CASE REPORT

Parachute-like mitral valve as a cause of mitral regurgitation

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Abstract
Background: Parachute mitral valve (PMV) is commonly associated with mitral valve stenosis but may occasionally be associated with mitral valve regurgitation.
Case report: We present a case of an asymptomatic 41-year-old female with a background of incomplete Shone's complex displaying a PMV regurgitation pattern.
Conclusion: Cardiovascular imaging with the use of transoesophageal echocardiogram and cardiac magnetic resonance were essential tools for the diagnosis and quantification of the severity of mitral valve regurgitation. Hippokratia 2016, 20(3): 238-240

Keywords: Parachute mitral valve, Shone's complex, mitral valve regurgitation

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Introduction

Parachute mitral valve (PMV) is a congenital valvular anomaly in which all the chordae tendineae of the mitral valve are attached to a single papillary muscle. Cardiac imaging with transoesophageal echocardiogram (TOE) and cardiac magnetic resonance imaging (CMR) can be used to clarify the complex left sided structural cardiac abnormalities and provide a precise estimation of the mechanism and severity of mitral valve dysfunction[1].

Case presentation

A 41-year-old asymptomatic female was referred to our department due to echocardiographic findings of PMV morphology and significant mitral valve regurgitation. Her personal medical history involved a surgical repair of aortic coarctation (ACoA) with end-to-end anastomosis at the age of six years. TOE showed a single papillary muscle centrally placed receiving chordae from both the anterior and posterior mitral valve leaflets (Figure 1A). The mitral valve orifice was mildly stenotic, and a moderate to severe mitral valve regurgitation due to the prolapse of the elongated chordae tendinae was noted (Figure 1B). The aortic valve was bicuspid with mild stenosis and regurgitation. Left ventricular outflow tract had a tunnel-like morphology with a subaortic membrane (Figure 1C) but without obstruction. A CMR showed normal left ventricular volumes and preserved systolic function. A hypertrophied anterolateral papillary muscle receiving chordae from both mitral valve leaflets (Figure 1D) and a hypoplastic posteromedial papillary muscle were identified (Figure 1E). The mitral valve regurgitation was assessed as moderate (regurgitation fraction 23%). Finally, only mild restenosis at the aortic isthmus was noted (Figure 1F). A decision was made to follow-up the patient closely for symptomatic decline as well as worsening of valvular function.

Discussion

PMV is an extremely rare congenital cardiac defect and in the majority of patients constitutes part of the Shone's complex. This complex was described more than 50 years ago, and it can be found in 0.6 % of all congenital heart defects. The components of the complex include subaortic stenosis, ACoA, supravalvular mitral ring and PMV[2]. The patient in this case report can be classified as an incomplete form of the complex, which is combined with a bicuspid aortic valve and subaortic membrane. The incomplete forms of Shone's complex constitute the majority of case reports encountered in the literature. In adulthood, Shone's complex displays a relatively low mortality but also a considerable morbidity which is related to repeated hospitalizations for heart failure and interventions for the left-sided cardiac valves[3].

True PMV requires the presence of a single papillary muscle whereas in parachute-like asymmetric mitral valve there are two papillary muscles with different embryologic development[4]. The dominant papillary muscle is hypertrophic and receives the total amount of chordae tendinae. On the other hand, there is a hypoplastic papil-

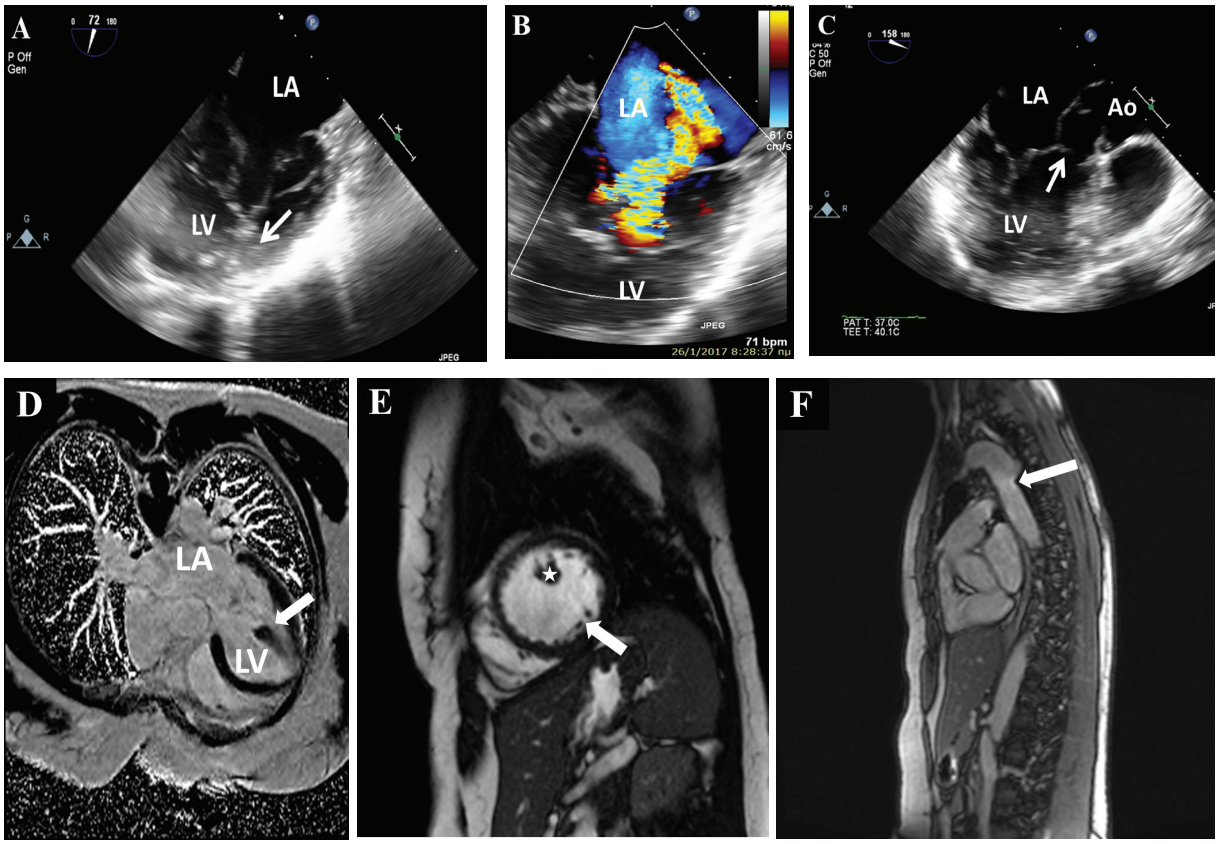

Figure 1: Transoesophageal echocardiogram (TOE) and cardiac magnetic resonance (CMR) images of the patient with incomplete Shone's complex. **A**) TOE mid-esophageal 72° view displaying the mitral valve leaflets and the tendineae chordae inserted to a single papillary muscle (arrow). **B**) TOE mid-esophageal 74° color-Doppler view showing the mitral valve regurgitation pattern. **C**) TOE mid-esophageal 158° view showing the subaortic membrane (arrow). **D**) CMR view displaying the chordae tendineae attached to the dominant anterolateral papillary muscle (arrow). **E**) CMR short axis view displaying the left ventricle with the hypertrophic anterolateral papillary muscle (asterisk) and the hypoplastic posteromedial papillary muscle (arrow) **F**) CMR view of the aorta displaying the mild stenosis at the isthmus level (arrow).
Ao: aorta, LA: left atrium, LV: left ventricle.

lary muscle that does not have any chordae attachments and could even elude diagnosis. This chordae orientation causes the asymmetry and the eccentric location of the mitral valve orifice. Mitral valve stenosis due to the PMV can commonly be diagnosed at childhood whereas the less common mitral valve regurgitation can progress even silently to the adulthood[5]. The mechanism for the mitral valve stenosis is the reduced mobility of the leaflets due to the short and thick chordae[6]. In contrast, the mitral valve regurgitation in PMV is usually the result of the prolapse of the elongated chordae into the left atrium[7].

In the reported patient mitral valve regurgitation was assessed as moderate to severe with echocardiography, while CMR graded the regurgitation as moderate. This discordance between the two imaging modalities was highlighted in a recent prospective multicenter trial[8] which suggested that CMR was more accurate than echocardiography in assessing the severity of mitral valve regurgitation. Consequently, patients with nonsevere mitral valve regurgitation who are incorrectly diagnosed as having severe mitral valve regurgitation could undergo inappropriate surgery. Thus, CMR could be applied especially in cases where echocardiography might provide suboptimal quantification data, such as patients with complex anatomy of the mitral apparatus (e.g. congenital heart disease) or patients with poor acoustic echocardiographic windows (e.g. obesity, prior sternotomy, lung disease)[9].

Another important issue is that the patient did not report any symptoms. It is known that adults with congenital heart disease may underestimate the degree of their functional limitation, as this is present since infancy[10]. Exercise testing is especially useful in these cases to unmask the objective occurrence of symptoms in patients who claim to be asymptomatic or have doubtful symptoms[11].

In conclusion, a comprehensive assessment should include the combination of thorough clinical assessment of the exercise capacity along with advanced imaging (TOE and CMR) in order to reliably define the mechanism and severity of the mitral valve dysfunction and guide the decision for surgical intervention.

Conflict of interest

Authors declare no conflict of interest.

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