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MINI-FOCUS ISSUE: VALVULAR HEART DISEASE

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Blown Wide Open

An Unusual Case of Torrential Mitral Regurgitation Resolving With a Cough

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ABSTRACT

Mitral regurgitation can have varying hemodynamic parameters dependent on factors such as pressure gradients, exercise, and/or provocative maneuvers. We present a case of unusual dynamic mitral regurgitation resolved by coughing in a patient with hypertrophic cardiomyopathy. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:668–71)

HISTORY OF PRESENTATION

A 65-year-old man with known hypertrophic cardiomyopathy without left ventricular outflow tract obstruction (LVOTO) presented with syncope to the emergency department. Upon presentation, he had an unwitnessed syncopal attack going up a flight of stairs to the gym. There was no chest pain, shortness of breath, palpitations, or neurological symptoms. There was dizziness before the syncopal episode. He also described a 2-week history of increasing presyncope with exertion with less than ordinary activity. His vital signs were normal at rest, and physical examination revealed a 2/6 pansystolic murmur at the apex radiating to the axilla.

LEARNING OBJECTIVES

- To recognize dynamic valve lesions in patients presenting with syncope so that appropriate management can be implemented.
- To recognize the limitations of exercise echocardiography in detecting some forms of dynamic MR in structural heart disease.

MEDICAL HISTORY

The patient had a history of hypertrophic cardiomyopathy with no LVOTO, moderate mitral regurgitation (MR), severe pulmonary hypertension, implantable cardioverter-defibrillator placement, paroxysmal atrial fibrillation, and dyslipidemia. Previous stress echocardiography did not demonstrate systolic anterior motion (SAM) of the mitral valve (MV), LVOTO, or any inducible ischemia or wall motion abnormalities.

DIFFERENTIAL DIAGNOSIS

Given the patient's nonspecific history and medical history of structure heart disease, the differential diagnosis for syncope in this case was quite broad. This included vasovagal syncope, syncope due to structural heart disease, tachyarrhythmia or heart block, orthostatic hypotension, and dehydration.

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INVESTIGATIONS

Investigations included complete blood count and electrolytes, which were normal. Creatinine level was mildly elevated at 127 μmol/l. Serial troponin T levels were 91 and 85 ng/l. Initial electrocardiography showed an atrial-paced rhythm with a normal QT interval and a left ventricular (LV) strain pattern. Device interrogation did not reveal any episodes of ventricular tachycardia, ventricular fibrillation, heart block, or pauses. There were no recorded shock therapies delivered.

Transthoracic echocardiography revealed normal LV cavity size and function. There was asymmetrical LV septal hypertrophy. There were dynamic changes in the degree of SAM and MR. During the first one-half of the study, the anterior MV leaflet was open throughout systole and diastole, with a large visible coaptation defect and torrential MR (Videos 1a to 1d). Following a cough, witnessed by the sonographer, the anterior leaflet coapted normally and the degree of MR become only trivial (Video 2). The posterior valve was tethered and restricted in its closure. Severe pulmonary hypertension with right ventricular systolic pressure of 123 mm Hg was also noted.

Right heart cardiac catheterization was completed, which showed moderate pulmonary arterial hypertension with mean pulmonary artery pressure of 37 mm Hg and normal pulmonary capillary wedge pressure (5 mm Hg). Nitric oxide challenge resulted in normalization of pulmonary hypertension. Left heart cardiac catheterization showed minor wall disease in the coronary arteries, with no critical stenosis. On the ward, the patient had episodes of transient hypotension at rest with blood pressures of 80s/50s mm Hg and transient hypoxia, which initially responded to fluids and vasopressors. He decompensated in the coronary care unit on post-admission day 3, requiring intubation and ventilation.

MANAGEMENT

The patient underwent septal myomectomy (3 × 2 × 1 cm) and Alfieri repair of the MV between the A2 and P2 segments. Intraoperative transesophageal echocardiography showed no SAM of the MV, mild mitral stenosis (mean gradient 2 mm Hg), and trivial MR (Videos 3a and 3b). The patient was discharged from the hospital 12 days after surgery without any significant complications.

DISCUSSION

The MV complex comprises the mitral annulus, anterior and posterior mitral leaflets, chordae tendineae, papillary muscles, and LV myocardium. Primary MR refers to an abnormality of the MV apparatus, while secondary MR refers to a normal MV apparatus in the presence of ventricular and/or atrial disease (1).

MR and its dynamic nature are well recognized in published research and can be seen in primary or secondary MR (2). The MV apparatus is dependent on multiple complex factors, such as pressure gradients between the left ventricle and left atrium, which are altered by preload and afterload, LV geometry, and contractility (3). Furthermore, exercise and dynamic maneuvers such as Valsalva can alter the severity of MR. Exercise echocardiography is a useful tool in evaluating dynamic MR, especially in patients with symptoms during exertion or equivocal symptoms (4).

In patients with hypertrophic cardiomyopathy, MR is common due to SAM of the MV secondary to LVOTO (5).

Traditional SAM is caused by a Venturi effect "sucking" the anterior MV leaflet into the LV outflow tract (LVOT). In our case, we instead observed the anterior MV taking on a shape that allowed it to be "pushed" into the LVOT as the flow was exerted on the atrial surface of the anterior MV leaflet instead of the ventricular surface, thereby directing its motion away from the posterior leaflet and allowing torrential MR. The cough in this case appears to have resulted in conformational dynamic change in the anterior mitral leaflet's, favoring increased flow from ventricular contraction to push the valve closed in the normal fashion (**Figure 1**).

The best analogy to explain these findings would be a phenomenon affecting a sail called "backwind." In the initial phase (**Figure 1**, top right), the anterior MV leaflet is anteriorly displaced by the net forces created by systolic output and obstructs the LVOT, which leads to lack of coaptation and severe MR. This is similar to the jib of sailboat being pushed outward by backwind (**Figure 1**, top left). Following the cough, the anterior MV leaflet returns to its normal position and is able to attain adequate coaptation. This results in a lesser degree of obstruction and reduction of MR. Similarly, once the jib is maneuvered, the wind catches the sail and bows it to an appropriate position.

In this particular case, careful review of previous transthoracic echocardiograms did reveal some evidence of intermittent severe MR, which, because of its chronicity, was believed to be the cause of the pulmonary hypertension despite wedge pressures' being relatively normal when obtained at a time when

ABBREVIATIONS AND ACRONYMS

LV = left ventricular

LVOT = left ventricular outflow tract

LVOTO = left ventricular outflow tract obstruction

MR = mitral regurgitation

MV = mitral valve

SAM = systolic anterior motion

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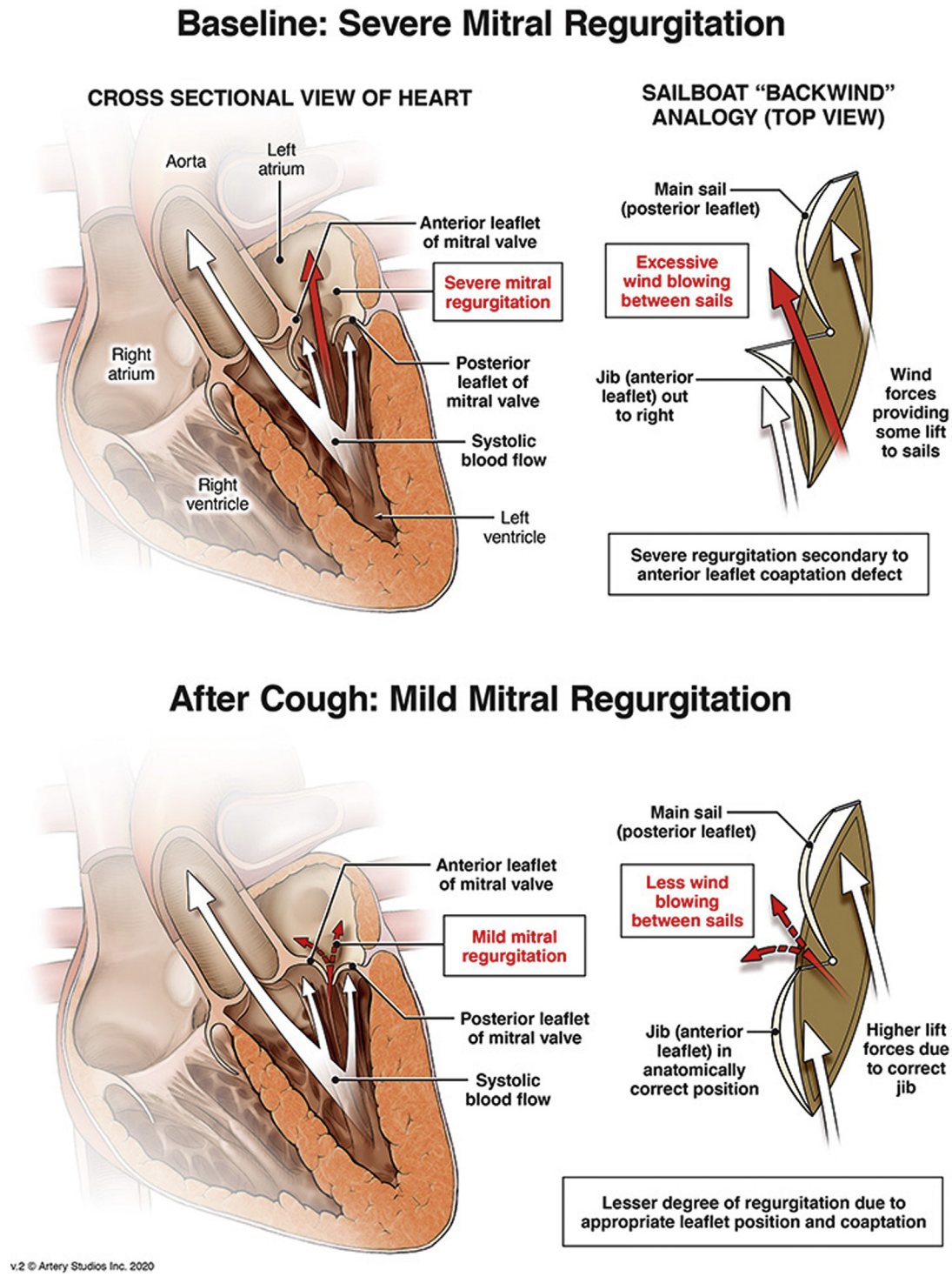
FIGURE 1 Central Illustration



(Top) Baseline severe mitral regurgitation with sailboat analogy of backwind. **(Bottom)** After cough, severe mitral regurgitation becomes mild with sailboat analogy.

MR was not severe. An Alfieri repair was thought to be the simplest fix, as the MV leaflets were relatively normal in structure and the leaflet conformation change responsible for the presentation would be prevented by ensuring that the atrial surface of the anterior mitral leaflet would remain atrial facing. This highlights that there are multiple reasonable strategies to accomplish MV repair in the setting of a floppy anterior leaflet.

FOLLOW-UP

At most recent follow-up approximately 7 years post-operatively, the patient has moderate MR with excellent functional capacity and is able to carry out all activities and exercise without any symptoms of chest pain, shortness of breath, palpitations, or syncope. A recent transthoracic echocardiographic examination showed preserved LV systolic function, mild to moderate MR that was not dynamic, and mild mitral stenosis (mean gradient 3 mm Hg).

CONCLUSIONS

This case illustrates an extremely rare and unusual dynamic MR captured by traditional transthoracic echocardiography. This shows that clinically significant valve disease may not always be detected by exercise echocardiography, and a high index of suspicion is needed to detect dynamic valvular lesions in patients with underlying structural heart disease. An excellent clinical outcome was achieved by septal myomectomy and Alfieri repair.

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KEY WORDS hemodynamics, hypertrophic cardiomyopathy, mitral regurgitation

APPENDIX For supplemental videos, please see the online version of this paper.