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Papillary Muscle Rupture as a Complication of Barlow's Disease

Ile Kuzmanoski*, Aleksandra Georgieva, Ana Chelikikj, Valentina Andova

University Clinic of Cardiology, Skopje, Republic of North Macedonia

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***Correspondence:** Ile Kuzmanoski, University Clinic of Cardiology, Skopje, Republic of North Macedonia. E-mail: ile_kuzmanoski@yahoo.com
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Abstract

INTRODUCTION: Mitral regurgitation (MR) is the second most common valvulopathy worldwide, which can be divided into primary and secondary. According to Carpentier's classification, the primary MR is further divided into three types. Type II, which includes Barlow's disease, is described as excessive mobility of the mitral valve (MV) leaflets. Morbus Barlow is a common form of degenerative disease, with an incidence of 2–3% of the general population. Echocardiography plays an important role in its diagnosis. It is a usually benign condition, with only a few severe complications.

CASE REPORT: A 75-year-old male with a history of MR, for more than 10 years. On admission, the patient presented with severe fatigue and dyspnea with signs of heart failure and pleural effusion. On auscultation, a systolic murmur was noted, on all the precordium. The ECG revealed sinus rhythm with HR of 71/min and intermittent ventricular extrasystoles. An immediate transthoracic echocardiography (TTE) was performed showing myxomatous degeneration of both MV leaflets and a prolapse of the posterior leaflet. A severe MR was detected with a presumption of papillary muscle rupture (PMR). It also revealed enlarged left atrium and ventricle (LVEDd - 67 mm and LA - 46 mm), with preserved systolic function (EF~54%) and tricuspid regurgitation accompanied by pulmonary hypertension. The laboratory analyses were within normal ranges. The patient was transferred to a cardiovascular surgery clinic, where an immediate MV repair was performed.

CONCLUSION: Barlow's disease is a common echocardiography finding. Although a benign condition, it can rarely present with serious complications such as PMR, ventricular arrhythmias, and even sudden cardiac death. Echocardiography is the first imaging used for the detection of Barlow's disease and other MV diseases. Early recognition and confirmation with TTE or transesophageal echocardiography, plays also an appropriate treatment, play a key role in patient survival and overall prognosis.

Introduction

Mitral regurgitation (MR) is the second most common valvulopathy worldwide [1], which can be divided into primary and secondary. Carpentier's classification is a functional classification that divides the primary MR into three types. They are categorized by the mitral valve (MV) leaflets motion. Type I refers to MR with normal leaflet motion. Type II, which includes Barlow's disease, is described as excessive mobility of the MV leaflets. The most prevailing lesions found in type II dysfunction are chordae elongation or rupture and papillary muscle elongation or rupture [2]. Type three refers to restricted MV leaflet motion. The most common cause of MR in developed countries is degenerative MV disease. The three plausible etiologies for leaflet degeneration include Barlow's disease, Marfan's disease, and fibroelastic deficiency [2]. Morbus Barlow is a common form of degenerative disease, with an incidence of 2–3% of the general population [2], [9]. It is a long-term condition, which had been previously noted as a systolic murmur, of which the patient had been aware for many years. Echocardiography plays an important role in its diagnosis. It is a usually benign condition, with only a few severe complications.

Case Report

A 75-year-old male presented with a history of MR, for more than 10 years. On admission, the patient presented with severe fatigue and dyspnea with signs of heart failure and pleural effusion. On auscultation, a systolic murmur was noted, on all the precordia. The ECG revealed sinus rhythm with HR of 71/min and intermittent ventricular extrasystoles. Immediate transthoracic echocardiography (TTE) was performed showing myxomatous degeneration of both MV leaflets, thus confirming Barlow's disease (Figure 1).

A prolapse of the posterior leaflet was also noted. A severe MR was detected with a presumption of papillary muscle rupture (PMR) (Figure 1). It also revealed an enlarged left atrium (LA) and left ventricle end-diastolic dimension (LVEDd - 67 mm and LA - 46 mm), with preserved systolic function, ejection fraction (EF~54%), and tricuspid regurgitation accompanied by pulmonary hypertension. The coronary angiogram of our patient did not demonstrate any vessel obstruction or evidence of coronary artery disease, thus excluding myocardial infarction (MI) as a possible cause. The laboratory analyses were

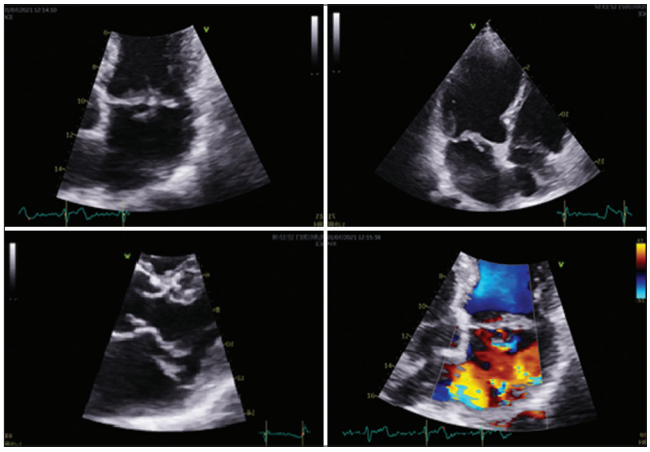

Figure 1: Echocardiographic findings on admission

within normal ranges. The patient was transferred to a cardiovascular surgery clinic, where an immediate MV repair was performed. The patient was discharged with an improved clinical condition, without any signs of heart failure. During the hospital stay, a new onset of atrial fibrillation was documented. The patient could not be converted into sinus rhythm and was discharged with long-term anticoagulant and antiarrhythmic therapy. On the regular 1-year follow-up, the patient was still in a stable clinical condition, without any symptoms or objective findings on the physical examination. A control echocardiography was performed, showing an improvement in the systolic function (EF~63%), and the dimensions of the left ventricle (LVEDd - 56 mm and LA - 46 mm) (Figure 2).

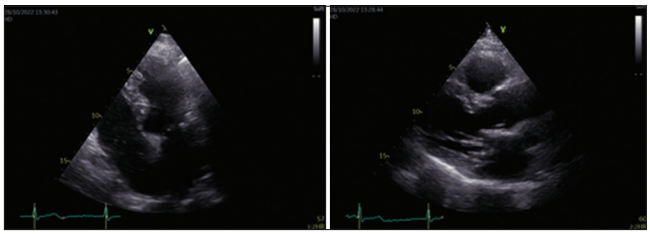

Figure 2: Echocardiographic findings on follow-up

Discussion

Spontaneous PMR in the absence of coronary artery stenosis is rarely reported in the literature. As a complication of MI, it appears with an incidence of 0.029%, however, there are only several case reports in lack of such etiology [3] and thus proving the rarity of this condition in the general population. Our patient's coronary angiography excluded MI as a cause of the rupture, thus making MV degeneration the most probable cause. Since it is a condition with a high mortality rate of 80% in the first 24 h if left untreated, making urgent surgical correction a necessity [4]. Acute rupture can present as a life-threatening condition, such as pulmonary edema or even cardiogenic shock. Even with proper treatment, the mortality rate ranges between 25 and 30% post-op within the first 30 days [3]. It is important to be noted that patients with degenerative MV disease and severe MR should undergo reconstructive surgical treatment before they become symptomatic, thus avoiding complications such as atrial fibrillation, pulmonary hypertension, LV dysfunction, and enlargement [2]. Our patient had this condition for many years before the acute event, which led him into having all the above-mentioned complications, and even PMR. On-time and appropriate treatment may result in improvement of echocardiographic findings and overall myocardial function after surgery and present with a long-term stable clinical condition of the patient. This improvement is confirmed by a study, showing that normalization of EF and reverse remodeling of the dimensions of the left ventricle is possible with immediate surgical correction of degenerative MR [5]. The transesophageal echocardiography (TOE) is also a great method for diagnostics of PMR, especially with the addition of color Doppler imaging, which helps in evaluating the severity of the MR [6], [7], [8]. It is a valuable diagnostic tool with 65-85% sensitivity in visualizing structural abnormalities [10]. This improvement is mostly seen in patients with preserved EF. This once again emphasizes the importance of on-time, proper management of such patients.

Conclusion

Barlow's disease is a common echocardiography finding. Although a benign condition, it can rarely present with serious complications such as PMR, ventricular arrhythmias, and even sudden cardiac death. Echocardiography is the first imaging used for the detection of Barlow's disease and other MV diseases. Early recognition and confirmation with TTE or TOE, plays also the appropriate treatment, play a key role in patient survival and overall prognosis.

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