SUDDEN HEART FAILURE DUE TO ACUTE MITRAL REGURGITATION IN A PATIENT WITH ATRIAL SEPTAL DEFECT: CASE REPORT

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ABSTRACT

Introduction: The association between secundum atrial septal defect and mitral valve disease has been recognized for many years.

Case presentation: A female patient was admitted to the hospital with sudden onset of congestive heart failure symptoms. Diagnostic methods revealed an enlarged right ventricle, secundum atrial septal defect, tricuspid regurgitation and pulmonary hypertension, mitral regurgitation with chordal rupture of the mitral valve. Treatment involved replacement of the mitral valve with a mechanical prosthesis and closure of the atrial septal defect with sutures.

Conclusion: In a patient with asymptomatic atrial septal defect, a sudden increase in left to right cardiac shunting due to acute mitral regurgitation precipitated right heart failure.

Key words: secundum atrial septal defect, mitral valve disease, right heart failure.

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Introduction

Atrial septal defect (ASD) is the second most commonly diagnosed adult congenital heart disease (CHD) after bicuspid aortic valve(1). Although ASD is a frequent congenital anomaly its diagnosis is often made later in life in adult patients as symptoms may develop only gradually. As such, an ASD may remain unrecognized for years, with symptoms usually not presenting in patients until they reach their forties.

It is known that structural abnormality of the mitral valve (MV) may accompany an ASD. The incidence of this dual pathology ranges from 4% to 9.1% in patients with an ASD(2). The association between secundum ASD and mitral valve disease has been recognized for many years, first recorded by Lutembacher in 1916(3). At that time he described a patient with interatrial communication through a patent foramen ovale with an associated mitral stenosis (MS), presumably of rheumatic origin. Besides MS, mitral regurgitation (MR) may also exist as comorbidity in patients with ASD.

This report describes a patient with an ASD who suddenly developed MR and reviews the key clinical findings associated with this pathology and the eventual treatment of the patient.

Case presentation

A 44-year-old female, Caucasian orthodox, previously asymptomatic of cardiac disease, was admitted to the hospital with a sudden onset of congestive heart failure symptoms. She abruptly devel-
oped cough, shortness of breath with minimal physical activity and profound leg swelling. On review of her medical history, she admitted that she had experienced decreased effort tolerance over the past few years, but had ignored those symptoms. Auscultation discovered a pansystolic murmur at the cardiac apex with propagation to the axilla and splitting and accentuation of the pulmonary second sound. ECG showed sinus rhythm, right axis deviation and right ventricular hypertrophy. Transthoracic and transesophageal echocardiography revealed an enlarged right ventricle, large secundum ASD with left to right shunting, moderate tricuspid regurgitation and pulmonary hypertension (figure 1).

**Figure 1:** Transesophageal echocardiography. The presence of an atrial septal defect is detected.

The cause of severe eccentric mitral regurgitation appeared to be due to chordal rupture of the myxomatous anterior leaflet of the mitral valve, corresponding to the A3 segment (figure 2).

**Figure 2:** Transthoracic echocardiography. Color Doppler imaging shows an ASD type secundum with the presence of a Left-to-Right shunt and evidence of severe eccentric mitral regurgitation.

The patient’s left ventricle was of normal size and function. Cardiac catheterization was performed. The pulmonary artery systolic pressure was 62 mmHg, the diastolic pressure was 23 mmHg, and the mean pressure was 43 mmHg. The ratio of pulmonary to systemic blood flow was calculated at 2.7:1 and pulmonary vascular resistance was calculated to be 3.01 Wood units. Coronary angiography was normal. The patient was treated surgically, undergoing mitral valve replacement with a mechanical prosthesis and closure of the ASD with sutures.

**Discussion**

The association of secundum ASD and mitral regurgitation is not an uncommon finding. Factors both intrinsic to the atrial septal defect and extrinsic to the secundum ASD may be responsible for a finding of mitral regurgitation\(^{(2)}\). The extrinsic factors are typically related to mitral lesions coexisting with an ASD, such as rheumatic valvular disease, infective endocarditis with disruption of the chordae tendineae and traumatic valve rupture. Regarding factors that are intrinsic to the ASD, there are numerous reports describing how secundum ASD itself could give rise to mitral regurgitation.

By angiographic and echocardiographic studies, Ballester et al.\(^{(4)}\) proposed that prolapse of the mitral valve in patients with secundum ASD may be related to the distorted left ventricular shape and small left ventricular volume. Pathological dissection demonstrated thickening of the medial half of the anterior leaflet and some fusion of adjacent chordae, leading to the conclusion that changes in the mitral valve might be caused by abnormal leaflet movement related to increased flow and altered left ventricular geometry in patients with secundum ASD. The authors found abnormal fibrous thickening at the medial half of mitral valve at which leakage of the valve occurred. They proposed a specific pathophysiology called “ASD- MR complex” and postulated that friction between anterior and posterior mitral leaflets due to abnormal left ventricular motion secondary to right ventricular volume overload might be responsible to the genesis of this lesion. Therefore, the pathogenesis of mitral regurgitation in ASD could be due to abnormal hemodynamic change caused by the ASD itself, similar to how aortic regurgitation may be caused by ventricle septal defect.

To summarize MR associated with secundum ASD could be due to a co-existent lesion in the valve or as a result of hemodynamic change resulting from the secundum atrial septal defect itself. Recognition of the co-existence of MR and ASD is important because the clinical course of mitral
regurgitation is altered by the presence of an ASD. Some patients with severe MR may not manifest symptoms of mitral regurgitation because the ASD unloads the left atrium.

In the case under review, patient had abrupt clinical deterioration caused by acute mitral valve incompetence. The onset of a large regurgitant volume led to a sudden volume and pressure overload of the left atrium and to retrograde pulmonary congestion, which was actually less than expected due to the left atrial unloading through the ASD. However, in this patient, the sudden increase in the left to right shunt further burdened an already overloaded right ventricle and precipitated right heart failure. Possible explanation for chordal rupture of the myxomatous anterior mitral leaflet in this patient could be due to the previously described changes in mitral valve function due to mechanical stress resulting from secundum ASD, perhaps related to a change in left ventricular geometry associated with right ventricular volume overload, as described in theory by Hammers et al.\(^5\).

Recent advance in medical and surgical treatment as well as in diagnosis has led to survival of more than 80% of individuals with congenital heart disease into adulthood.\(^6\) The aim of treatment in these patients is to maintain hemodynamic stability and to prevent the development of complications such as heart failure and irreversible pulmonary vascular diseases. Many of the abnormalities associated with congenital heart diseases can be surgically repaired with satisfactory results. For patients who undergo ASD closure, if residual mitral regurgitation remains significantly increased, elevated left atrial and pulmonary venous pressure may develop and may lead to increased symptoms of congestive heart failure.\(^7,8\) In some cases, closure of an atrial septal defect without repair of mitral regurgitation may lead to increased postoperative morbidity and mortality rates.

There are differences in opinion regarding the surgical treatment of mitral valve disease due to disrupted chordae tendineae—some authors recommend prosthetic valve replacement, whereas others emphasize the value of plastic reconstruction. For some patients, the ultimate decision regarding surgical technique may be difficult to determine until the time of operation.

In summary, we describe a 44 year-old woman with an unrecognized atrial septal defect who experienced a sudden prolapse of her mitral valve due to chordal rupture of a myxomatous anterior mitral leaflet. That led to acute severe mitral regurgitation that caused retrograde pulmonary congestion and increased shunting across the ASD. As a result, the patient developed pulmonary hypertension and progressed to right heart failure. It is likely that the thickening and fibrosis of her mitral valve were secondary to the chronic hemodynamic stress related to her ongoing left to-right shunt across her ASD. This mechanical stress ultimately may have caused the patient’s chordal rupture of the anterior mitral leaflet, which then precipitated her decompensate state.

Clinicians should be aware of the possible adverse consequences of undiagnosed and untreated congenital heart defects such as ASD. In addition, our case describes the clinical findings and evaluations that are necessary to confirm a diagnosis and provides an overview of the types of treatments that may be considered based on a patient’s hemodynamic status.

References

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