CLINICAL VIGNETTE

Multivalvular Rheumatic Heart Disease: Challenges in Nonsurgical Management

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Introduction

Rheumatic heart disease (RHD) is the most serious complication of rheumatic fever (RF). It is believed to result from autoimmune responses following an infection with group A beta-hemolytic streptococci[1]. RHD is relatively uncommon in the United States and other developed countries; however, it remains a major cause of cardiovascular disease with significant morbidity in the developing countries[2]. The disease process causes leaflet thickening, commissural fusion, chordal thickening, shortening and fusion [3].

Isolated mitral stenosis (MS) and combined MS and regurgitation are reportedly the most common valve involvement. Multivalve involvement has been reported in less than half of patients with MS and tends to occur more often in the aortic valve, as the accompanying valvular involvement. The tricuspid valve is affected in only a small percent of patients; the pulmonic valve is rarely affected[4]. Surgical intervention with double valve replacement or mitral valve repair with aortic valve replacement is considered the optimal treatment strategy in patients with concomitant aortic and mitral valve disease [5]. However, in patients who opt not to have the surgery the choice of alternative intervention remains more controversial and can be determine on a case by case basis.

Case presentation

A 50-year-old female of Middle Eastern origin with a past medical history of rheumatic fever at age seven, presented for consideration of percutaneous balloon mitral valvuloplasty (PBMV) versus valve replacement surgery. A diagnosis of mitral stenosis was made in her 20s, but was thought to be mild to moderate at that time without a need for intervention. The patient was able to have two children and go through pregnancies without any episodes of congestive heart failure. Over the past year prior to presentation, she gradually developed worsening dyspnea that significantly limited her ability to partake in dancing, which she had been easily doing throughout her life. At the time of presentation she denied orthopnea, paroxysmal nocturnal dyspnea, pre-syncope or syncope. She has never had atrial fibrillation and was not on anticoagulation.

On initial physical examination, she was found to have a 1/6 systolic murmur that best heard at right upper sternal border with radiation to bilateral carotids, as well as a 2/4 diastolic murmur appreciated in the same region in early diastole and a faint 1/6 diastolic murmur at the apical region. There was a short opening snap sound consistent with mitral stenosis.

Echocardiogram showed normal left ventricular size, wall thickness and systolic function with ejection fraction of 55-60%. The right ventricular size and function were normal. The left atrium was severely enlarged with left atrial volume index of 90 ml/m^2. The Mitral valve was found to have rheumatic deformity with doming of the anterior leaflet, moderate annular calcification, severe stenosis and mild regurgitation (Fig. 1A). The peak transmitral gradient was 18 millimeter of mercury (mmHg) with a mean transmitral gradient of 12 mmHg (Fig. 2A). The mitral valve area by pressure half-time was 1.50

square centimeters (cm^2). The Aortic valve was found to have rheumatic changes with moderately thickened and calcified cusps and restricted cusp opening (Fig. 1C). There was mild to moderate aortic valve stenosis and regurgitation with aortic valve area of 1.13 cm^2 ($0.75cm^2/m^2$) and the mean transaortic gradient of 16 mmHg (Fig. 1D). Tricuspid and pulmonic valves were both structurally normal with normal leaflet excursion. There was mild tricuspid regurgitation and trace pulmonic regurgitation. Pulmonary artery systolic pressure was elevated at 53 mmHg. The Wilkins score for the assessment of mitral stenosis was calculated as 5 (Table 1).

Table 1 **Assessment of mitral valve anatomy according to the Wilkins score**

Score	Leaflet mobility	Leaflet Thickening	Calcification	Subvalvular Thickening
1	Highly mobile, only leaflet tips restricted	Leaflets near normal in thickness (4-5 mm)	A single area of increased echo brightness	Minimal thickening below the leaflets
2	Normal mobility of Leaflet mid and base portions	Midleaflets normal, margins thickened (5-8 mm)	Scattered areas of brightness at leaflet margins	Thickened chordal structures < 1/3 of length
3	Valve still moves forward in diastole, mainly from the base	Entire leaflet thick (5-8mm)	Brightness extends into leaflet mid-portion	Thickening extended to distal 1/3 of chords
4	No/minimal forward movement of the leaflets in diastole	Entire leaflet very thick (>8-10mm)	Extensive brightness leaflet	Extensive thickening of all chordal structures, including papillaries

Electrocardiogram (ECG) revealed sinus bradycardia at 58 beats per minute, left atrial enlargement, and poor R-wave progression with nonspecific ST-T segment abnormalities without any significant Q wave.

Considering the patients' concomitant aortic and mitral valve disease, surgical intervention with double valve replacement or mitral valve repair with aortic valve replacement was indicated; however, the patient opted not to have the surgery. Given the finding of the favorable mitral valve morphology, the patient was considered to be a suitable candidate for PBMV. The procedure was performed uneventfully. The mean transmitral pressure gradient decreased from 12 to 7 mmHg with an increase in mitral regurgitation from mild to moderate (Fig. 2A, 2B). Doppler echocardiographic assessment of the tricuspid regurgitation (TR) jet demonstrated a drop in peak pressure gradient across the tricuspid valve from 50 to 28mmHg (Fig. 2C, 2D).

Discussion

Patients with MS typically remain asymptomatic for many years or only mildly symptomatic during episodes of physiologic stress such as exercise, emotional stress, infection or pregnancy; however, they eventually develop gradually decreasing exercise tolerance, exertional dyspnea, palpitations and fatigue[6]. As the disease progresses, the increase in left atrial and pulmonary pressure results in tricuspid and pulmonary valve insufficiency, and secondary right heart failure.

Indications for intervention in patients with MS are based on clinical and echocardiographic characteristics. The available interventions are PBMV, surgical mitral commissurotomy, and mitral valve replacement (MVR). They can be considered according to established criteria and guidelines on the management of valvular heart disease (The Joint Task Force on the Management of Valvular Heart Disease of American Heart Association (AHA) and the American College of Cardiology (ACC)), as well as the patient's wishes, life expectancy, associated comorbidities and local expertise[5].

Wilkins score is the most commonly used echocardiographic parameter to evaluate mitral valve leaflet mobility, thickening, calcification, and subvalvular apparatus involvement[7]. A low score (less than or equal to 8) with no more than moderate mitral regurgitation is indicative of a favorable response to PBMV, whereas patients with higher scores are considered to be less fit and a surgical intervention would be a more preferred approach.

A decision for aortic valve replacement is mainly based on the presence or absence of symptoms. Surgical aortic valve replacement and transcatheter aortic valve replacement (TAVR) have become the standard options for severe AS.

Pulmonary hypertension (PH) is a major hemodynamic consequence of mitral stenosis and has significant prognostic value on the overall patient survival. The development of PH also considered an important indicator of outcome after surgical intervention; therefore, the relief of valve obstruction to decrease left atrial pressure and pulmonary vascular resistance is a well-known indication of corrective intervention.

Our patient was an otherwise healthy middle-aged female who presented with progressive dyspnea on exertion. She had concomitant aortic and mitral valve disease with severe mitral stenosis associated with mild regurgitation as well as mild to moderate aortic valve stenosis and regurgitation and moderate to severe pulmonary hypertension but mild tricuspid regurgitation which, taken all together, warranted a surgical intervention such as double valve replacement or aortic valve replacement combined with mitral valvuloplasty. However, she refused to undergo double valve replacement. Considering the finding of moderate to severe pulmonary hypertension and being symptomatic, the mechanical relief of MS was indicated. The patient had favorable mitral valve anatomic characteristics with a Wilkins score of 5 with no more than moderate mitral regurgitation, mild to moderate aortic stenosis and no evidence of left atrial or left atrial appendage thrombus; therefore, a PBMV was considered.

Furthermore the need for double valve replacement would be postponed for several years while improving her functional status and avoiding the disadvantages of a mechanical prosthetic valve replacement. The overall complication rate with this approach is relatively low and mainly determined by local expertise. In our patient, PBMV was uneventful and yielded a decrease in pulmonary pressure and improvements in transmitral gradient and mitral valve area with only a small progression of mitral regurgitation from mild to moderate grade without any further progression till last follow up.

The exact long term benefit of such a conservative approach in management of MS in selected patients with double valve disease and favorable mitral valve morphology however, yet to be determined

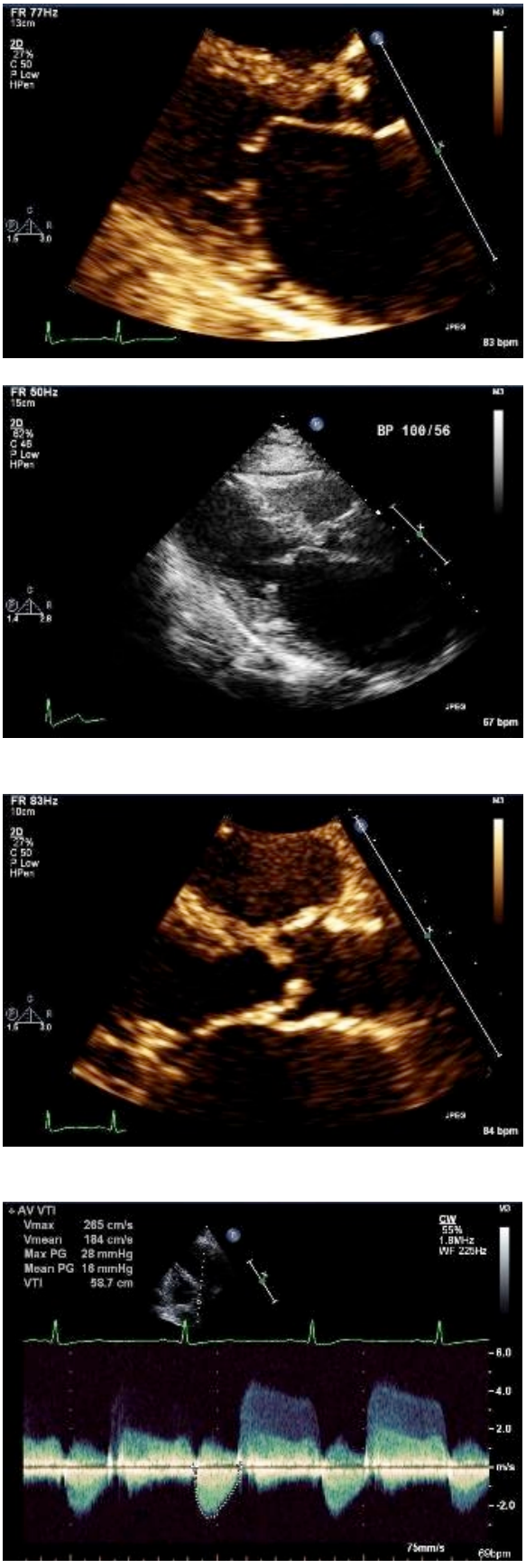
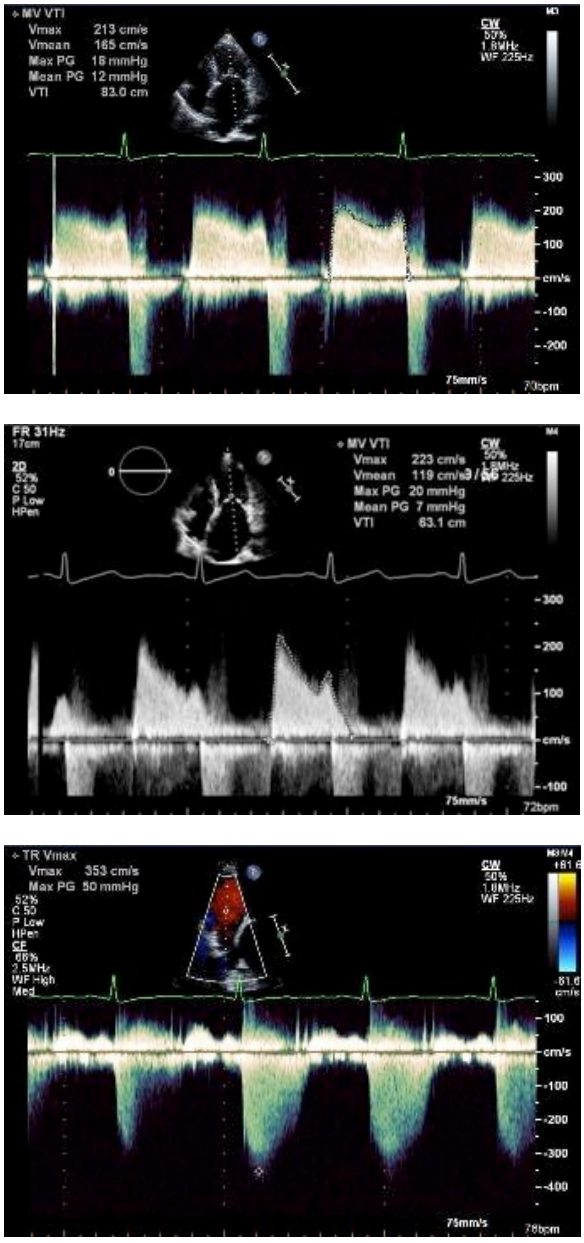
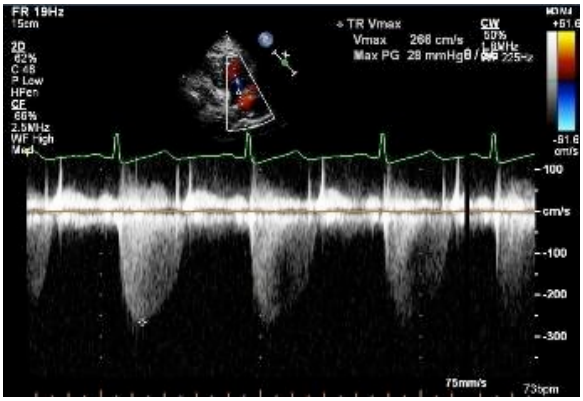


Figure 1- (A) Parasternal long-axis view, in diastole, demonstrating rheumatic mitral valve disease with classic “hockey stick” appearance, (B) Parasternal long-axis view, in diastole, after mitral valvuloplasty, (C) Parasternal long-axis view, in systole, revealing thickening of the aortic valve, most prominent at the tips, with restricted opening, consistent with rheumatic aortic valve involvement, (D) Continuous-wave Doppler of aortic valve, showing increased mean pressure gradient and jet velocity consistent with aortic stenosis and the regurgitant flow velocity pattern consistent with aortic regurgitation.

Figure 2- Continuous-wave Doppler of mitral valve, demonstrating mean pressure gradient and regurgitant flow velocity pattern, (A) before and (B) after mitral valvuloplasty; Continuous-wave Doppler of tricuspid valve, showing mean pressure gradient and jet velocity (C) before and (D) after mitral valvuloplasty.





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