Rheumatic Mitral and Aortic Valve Disease- A Case Report from USA

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Abstract:

Although virtually eradicated in the United States, rheumatic heart disease still carries a sinister outcome causing significant mortality and morbidity in developing countries in Asia, Africa and the Pacific Islands where it still remains in endemic form.

Our case involves a 35-year-old female patient born and raised in India who recently migrated to the United States. Her echocardiogram revealed severe combined aortic and mitral valve stenosis and severe aortic regurgitation. We hereby analyze echocardiographic findings in concomitant rheumatic aortic and mitral valve disease with preserved left ventricular (LV) systolic function. We also discuss treatment implications with combined surgical replacements of both valves with mechanical prostheses.

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Introduction:

Rheumatic heart disease can be defined as an autoimmune reaction to pharyngeal infection by beta-hemolyticus Lansfield Group A streptoccus of rheumatogenic strain. These bacteria are engulfed by macrophages and presented to the immune system creating specific antistreptococcal antibodies resulting in bacterial elimination in 95-97% of patients.

In 2-3% of patients, due to antigenic mimicry the immune activation can cause multisystem inflammation including carditis.[1] In the acute phase pancarditis involving pericardium, myocardium and endocardium usually develop. Pericarditis and myocarditis have no long term sequela. However, valvulitis results in bead-like multiple vegetations along the line of closure of mitral and aortic valves and these represent early manifestations of rheumatic endocarditis which are easily detected by echocardiography. Subsequently, valvular fibrosis, commissural fusion and in the case of atrio-ventricular valves chordal thickening and shortening may be noted, leading to variable degrees of valve stenosis and regurgitation.[2,3] Two year follow-up data of the Global Rheumatic Heart Disease Registry (The REMEDY STUDY) was published recently which confirmed that rheumatic heart disease still has high morbidity and mortality in patients living in countries with suboptimal incomes.[4]

Although rheumatic heart disease used to be a frequent reason for surgical replacement of heart valves, this disease entity is currently uncommon in USA where calcific degeneration of the aortic valve leading to aortic stenosis, a congenitally bicuspid aortic valve, and mitral valve prolapse are much more common.[5] We describe an adult patient living in USA who presented with rheumatic involvement of both mitral and aortic valves. She had recently migrated from India where she was born and raised.

Case Report:

The patient was a 35-year-old female presenting with dyspnea on exertion (AHA Class III). She

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had a history of febrile illness with joint pain during childhood at 6 years old, had taken penicillin prophylaxis for rheumatic fever, and had discontinued therapy intermittently. She developed symptoms of heart failure such as exertional dyspnea and orthopnea at the age of 27 years and underwent a mitral balloon valvuloplasty for mitral stenosis 10 years ago in India. She is taking digoxin 0.25 mg once daily, furosemide 20 mg once daily and intramuscular benzathine penicillin G injections 1.2 million units every 3 weeks.

Her pulse rate was 64 bpm, regular and her blood pressure was 87/44 mmHg. Physical examination revealed a loud first heart sound, second heart sound with normal split. Opening snap or S3 was not audible. She had grade 3/6 mid-diastolic rumbling murmur at the apex suggestive of mitral stenosis, and an early diastolic murmur at the left sternal border most prominent in 3rd and 4th intercostal spaces suggestive of aortic regurgitation. She also had a crescendo-decrescendo mid-systolic murmur of aortic stenosis at the left 3rd intercostal space. Jugular venous pressure was normal and lung fields were clear to auscultation. Blood chemistry revealed normal findings and an anti-streptolysin O (ASO) titer was negative. ECG revealed sinus rhythm and LV hypertrophy and X-ray chest revealed LV enlargement and a "double density sign" of left atrial (LA) enlargement in the lower right cardiac border.

A recent two-dimensional transthoracic echocardiogram (2DTTE) was performed using a Siemens (St. Paul, MN,USA) transducer which showed mild LV dilatation, normal LV systolic function (LVEF=55%), severe aortic stenosis ; peak pressure gradient across the aortic valve 81 mmHg; mean pressure gradient 51 mmHg) and severe aortic regurgitation. It also showed severe mitral valve stenosis (mitral valve area=1.2 cm2 by pressure half time method) with thickening and calcification of both leaflets as well as commissural fusion. There was also significant mitral chordal thickening and shortening. The valvular lesions on echocardiography were all consistent with a rheumatic etiology. Trivial pericardial effusion was also noted (Figures 1 and 2). A live/real time two- and three-dimensional transesophageal echocardiogram (2DTEE,3DTEE) was also performed using the same equipment as 2DTTE and showed severe aortic stenosis (AVA=0.53 cm^2) with severe aortic regurgitation and severe mitral valve stenosis (MVA=0.66 cm^2,Figures 3A) Commissural fusion was better identified with 3D TTE and TEE.

Cardiac catheterization showed normal coronaries and mild pulmonary hypertension with pulmonary artery systolic pressure in the range of 45 mmHg. Subsequently, the patient underwent surgery where all echocardiographic findings were confirmed. Both mitral and aortic valves were replaced with metallic prostheses.(St Jude Medical, MN,USA)

Postoperatively she has done well and her symptoms have gradually abated.

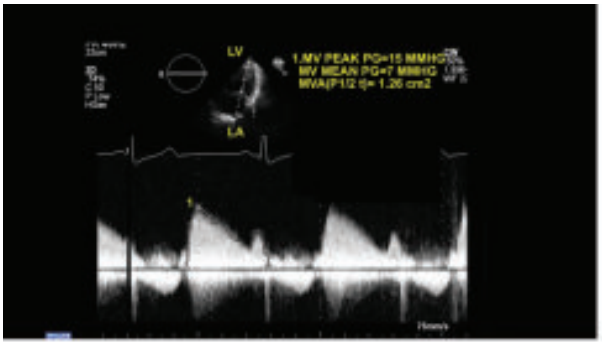

Fig.-1: *Two -dimensional transthoracic echocardiography. Mitral stenosis. Continuous wave Doppler of mitral valve (MV) shows peak and mean pressure gradients (PG) of 15 and 7 mmHg, respectively and mitral valve area (MVA) of 1.26 cm2 by pressure half time (P 1/2t) method consistent with moderate mitral stenosis.*

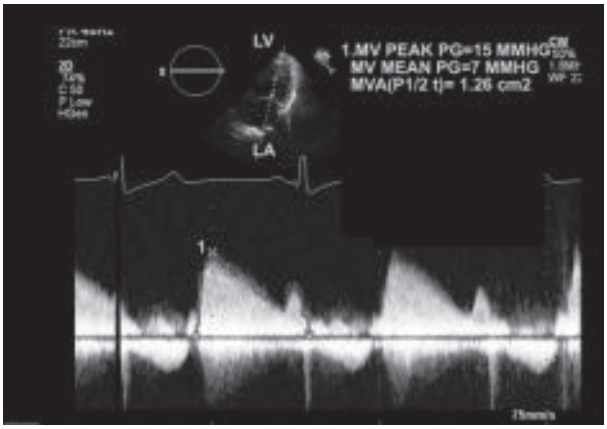

Fig.-2: *Two- dimensional transthoracic echocardiograms. Show a thickened MV valve.*

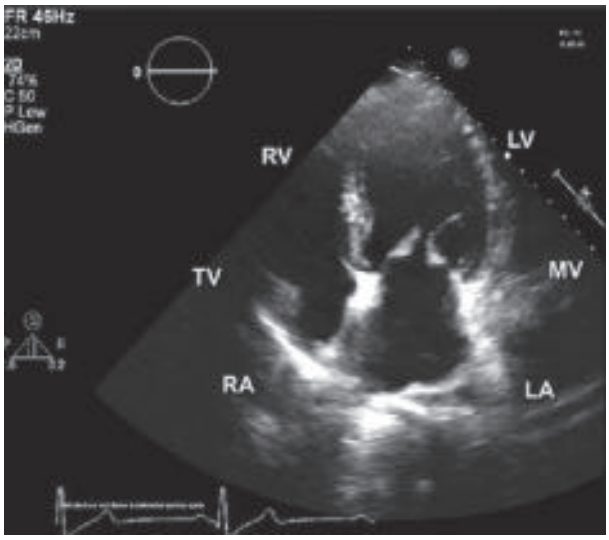

Fig.-3: *Two- dimensional transthoracic echocardiograms. Show a thickened MV valve.*

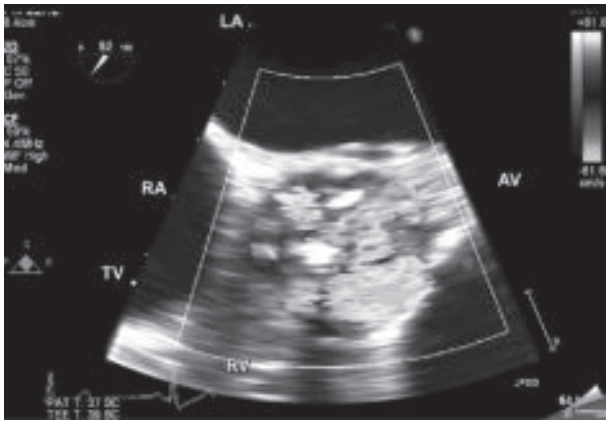

Fig.-4: *Two- dimensional transesophageal echocardiograms. Show a thickened AV valve.*

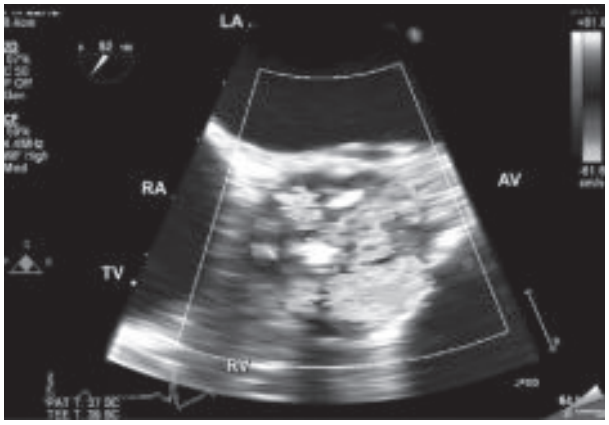

Fig.-5: *Color Doppler examination of AV valve in short axis view showing aortic regurgitation.*

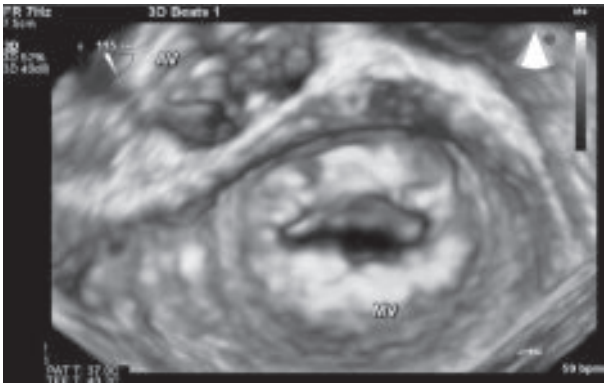

Fig.-6: *Three dimension echo showing mitral valve with stenosis.*

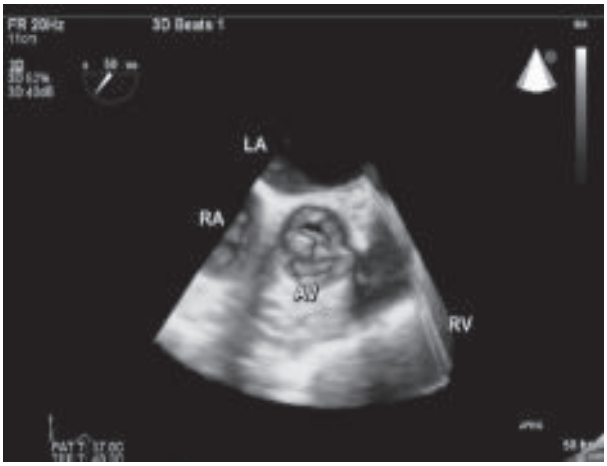

Fig.-7: *Three dimension echo showing aortic valve with stenosis.*

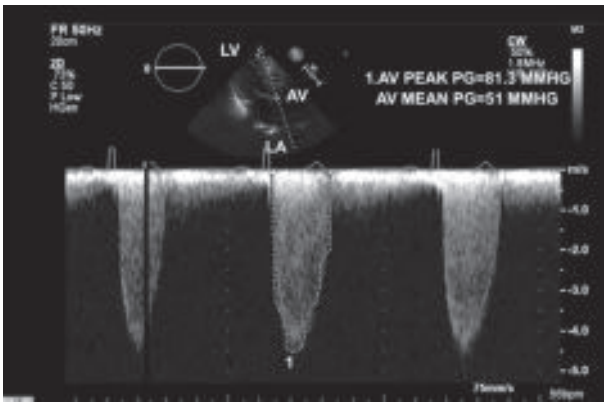

Fig.-8: *Two -dimensional transthoracic echocardiography. Aortic stenosis.Continuous wave Doppler through theAV shows peak and mean pressure gradients of 81 and 51 mmHg, respectively consistent with severe aortic stenosis.*

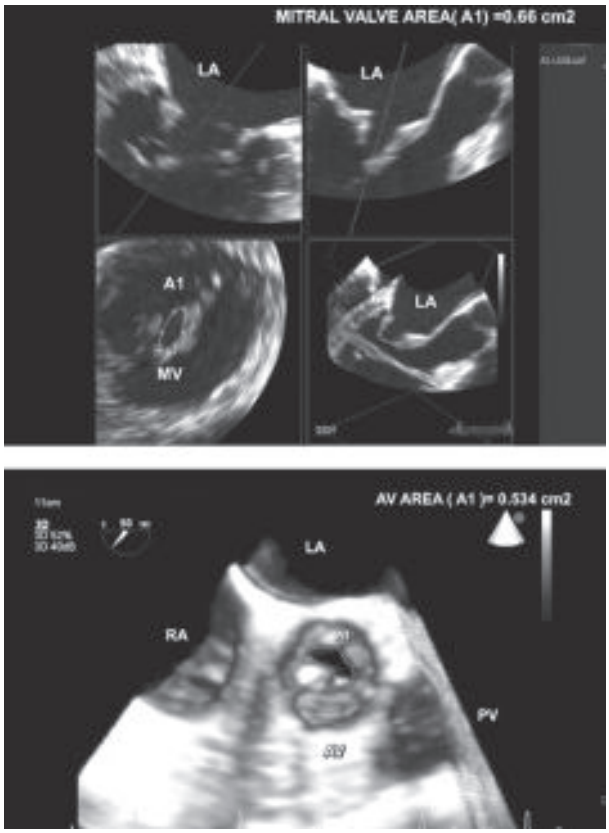

Fig.-9: *Three- dimensional transesophageal echocardiograms. MVA of 0.66 cm2 consistent with severe mitral stenosis. AV area of 0.534 cm2 indicative of severe aortic stenosis.*

Discussion:

We describe an unusual case of rheumatic involvement of both mitral and aortic valves with severe mitral stenosis, severe aortic stenosis and severe aortic regurgitation presenting in the USA. All echocardiographic findings were confirmed at surgery.

Our case report is unique in its presentation for the following reasons: (1) it elucidates rheumatic valvular heart disease which is currently very rare in United States, but is still a major health problem in developing countries.(2) In the United States, we encounter aortic valve stenosis which is mostly due to degenerative calcific disease or a bicuspid aortic valve. (3) Rheumatic aortic regurgitation is also very rare in United States, and is preceded in prevalence by aortic root disease or endocarditis. (4) According to the American Heart Association (AHA), most U.S. cases of mitral stenosis occur in older adults who had rheumatic fever before the advent of antibiotics to treat infection by Group A Streptococcus. It is also more likely to occur in immigrant population such as ours.[6]

Our case involves a young female patient of reproductive age, which had tremendous implications in the decision making process regarding the types of valves that were going to be suitable for a double valve replacement. Biologic valves were suggested as initial choice, as they do not require long term anticoagulation and are therefore safer for pregnancy. However, the durability of biologic valves is less than mechanical valves and the patient would most likely require re-operation later on in life. Mechanical valves are more durable but have implications for long term anticoagulation, specially for a patient of reproductive age.

After a thorough discussion with the cardiothoracic surgeon regarding the risks, benefits, and options facing her, the patient and her husband opted for mechanical valve replacements. Of note, our patient had a history of seizures and was on Phenytoin, which was switched to Levetiracetam to avoid possible interaction with Warfarin.

Recently published Two Year Follow-up of the Global Rheumatic Heart Disease Registry (The REMEDY Study) found that in developing countries rheumatic fever and its sequela still represented a challenge with adverse outcome and increased morbidity and mortality even in young patients. Early detection of rheumatic involvement of the heart with use of echocardiography may be helpful in this population.

Our patient was fortunate to have received care and treatment at a tertiary care center in the United States. This case serves to provide a greater sense of awareness regarding the healthcare disparities that still exist around the world.

Conflict of Interest - None.

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