

Rheumatic Mitral Stenosis Complicated with Underlying Wolff-Parkinson-White Syndrome

Saraschandra Vallabhajosyula^{1*}, Pranathi Rao Sundaragiri¹, Shashaank Vallabhajosyula²,
Saarwaani Vallabhajosyula²

¹Department of Internal Medicine, Alegant-Creighton University Medical Center, Creighton University School of Medicine, Omaha, NE, USA, ²Medical Student, Kasturba Medical College, Manipal University, Manipal, KA, India

ABSTRACT

Rheumatic mitral stenosis (MS) is an acquired valvular condition and is rarely associated with Wolff-Parkinson-White (WPW) syndrome. A 51-year-old Caucasian female with known MS presented with syncopal episodes, exertional fatigue and was noted to have atrial fibrillation (AF) with wide-QRS complexes. Old electrocardiogram revealed classical findings of WPW syndrome and wide-QRS was believed to be due to pre-excited AF. Transthoracic echocardiogram demonstrated moderate MS and preserved systolic function. Electrophysiological studies revealed left anterolateral accessory pathway (AP) that was successfully ablated. Pre-excited AF manifests as cardiomyopathy, hemodynamic compromise, multi-organ dysfunction, and ventricular fibrillation. Catheter ablation of AP with or without restoration of sinus rhythm is the recommended therapy, with mitral valve replacement in the same or subsequent procedure.

Keywords: Mitral stenosis, radiofrequency ablation, rheumatic heart disease, Wolff-Parkinson-White syndrome

INTRODUCTION

Rheumatic heart disease (RHD) is still a leading cause for pancarditis and valvular dysfunction globally with nearly 15.6 million existing cases and incidence of 282,000 new cases per year.^{1,2} Mitral valve (MV) involvement is noted in about 90% of the cases with mitral stenosis (MS) being the most common manifestation.³ Being an acquired condition, this valvular disease is rarely associated with congenital anomalies. We herein report a case from our center where underlying Wolff-Parkinson-White (WPW) syndrome posed a diagnostic challenge in a patient with MS.

CASE REPORT

A 51-year-old Caucasian female with a known history of RHD/MS presented with multiple syncopal episodes since two days. She developed dizziness with subsequent syncope on

minimal exertion, which lasted 2-3 minutes with spontaneous recovery with no residual weakness subsequently. At a local ER, she was noted to be in atrial fibrillation (AF) with multiple premature ventricular complexes on telemetry and referred to our center for further management.

Patient was asymptomatic at our center, but endorsed exertional dyspnea and fatigue in addition to her syncopal episodes. She denied chest discomfort, confusion, orthopnea, pedal edema, or palpitations. Past medical history was significant for RHD with multiple penicillin treatments during her childhood. She endorsed similar recurrent syncopal symptoms in the past that were attributed to hemodynamic compromise from her MS. Patient lived in rural Nebraska all her life and did not endorse any foreign travel or exposure to sick foreign contacts. Her RHD/MS with subsequent left atrial (LA) enlargement and permanent AF were being managed with beta-blockers, warfarin and digoxin. Family and personal-social histories were unremarkable.

Cardiac examination revealed hemodynamic stability (blood pressure 140/87 mmHg, heart rate 86/min) and irregularly irregular rhythm. Variable S₁, loud S₂, opening snap, and apical grade IV/VI mid-diastolic rumbling murmur with axillary radiation were appreciated. Laboratory parameters

*Corresponding address:

Dr. Saraschandra Vallabhajosyula, MD, Department of Internal Medicine, Alegant-Creighton University Medical Center, 601 N 30th Street, Suite #5850, Omaha, NE 68131, USA. Tel: (402) 690-4915, E-mail: SaraschandraVallabhajosyula1@creighton.edu

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were normal with the exception of normocytic anemia and prolongation of prothrombin time consistent with Warfarin use. Electrocardiogram (ECG) revealed AF and sagging ST-segment changes consistent with digitalization (Figure 1). Frequent wide-complex QRS changes were noted that was suspected to be secondary to Ashman phenomenon.

Transthoracic echocardiogram (TTE) demonstrated borderline concentric left ventricular (LV) hypertrophy and severe LA enlargement. MV showed moderate stenosis (valve area 1.7 cm²), peak and mean gradients of 22.3 and 9.1 mmHg, respectively and MS echo score index of 9.0. Review of old ECG from nearly 15 years ago showed the patient to be in normal sinus rhythm (NSR) with classical findings of WPW syndrome-short PR-segment, δ -wave (slurred up-stroke of QRS) and wide-QRS complexes (Figure 2). After detailed review of patient's ECG rhythms and prior holter recordings, the wide-complex QRS was deemed to be consistent with pre-excitation. Her current syncopal episodes were attributed to brief ventricular fibrillation (VF) due to antegrade conduction of AF through the accessory pathway (AP). A recent Holter recording provided for the syncopal episodes demonstrated alternating NSR and AF with second degree Mobitz type-I atrioventricular (AV) block. Intermittent aberrant conduction was noted with peak ventricular response of 226/min. Patient's beta-blockers were discontinued, and she was planned for electrophysiology (EP) studies.

Transesophageal echocardiogram (TEE) prior to EP study was unremarkable for LA thrombus (Figures 3 and 4). After direct-current cardioversion (DCCV) into NSR, right ventricular, high right atrial incremental and extra-stimulus

testing was performed. Ventricular pacing demonstrated 1:1 conduction over left-sided AP until 250 ms with retrograde effective refractory period (ERP) <230 ms when pacing at 450 ms. Atrial pacing demonstrated 1:1 conduction over the AP until 270 ms with antegrade ERP of 290 ms when pacing at 450 ms. The AP was localized to the left-anterolateral region, and this demonstrated orthodromic reciprocating tachycardia during atrial extra-stimulus testing. Under fluoroscopic and intra-cardiac echocardiographic (ICE) guidance, mapping radiofrequency ablation (RFA) of the atrial and ventricular insertion of the AP with insurance lesions was performed. Isoproterenol stimulation 1 h post-procedure did not demonstrate antegrade or retrograde conduction. Post-procedurally patient was restarted on beta-blockers with good hemodynamic response and no evidence of aberrant conduction. She reverted to AF on the post-procedure day 2 and was managed with rate control therapy and anticoagulation. She remained asymptomatic at discharge and subsequent 6-month follow-up.

DISCUSSION

WPW syndrome is a congenital disorder and exists in isolation in about 60-70% cases; however, atrial septal defect and Ebstein's anomaly are most frequently reported in conjunction.^{4,5} Rare reports of association with RHD/MS have been noted in literature complicating the clinical picture and subsequent diagnosis like in our case.⁵⁻⁸

AF in this setting is due to the dual individual influences from the underlying WPW syndrome resulting in atrial vulnerability and MS offering mechanical resistance to LA outflow.⁴ These acts synergistically in triggering AF as in our patient. Postulated etiopathogenic mechanisms

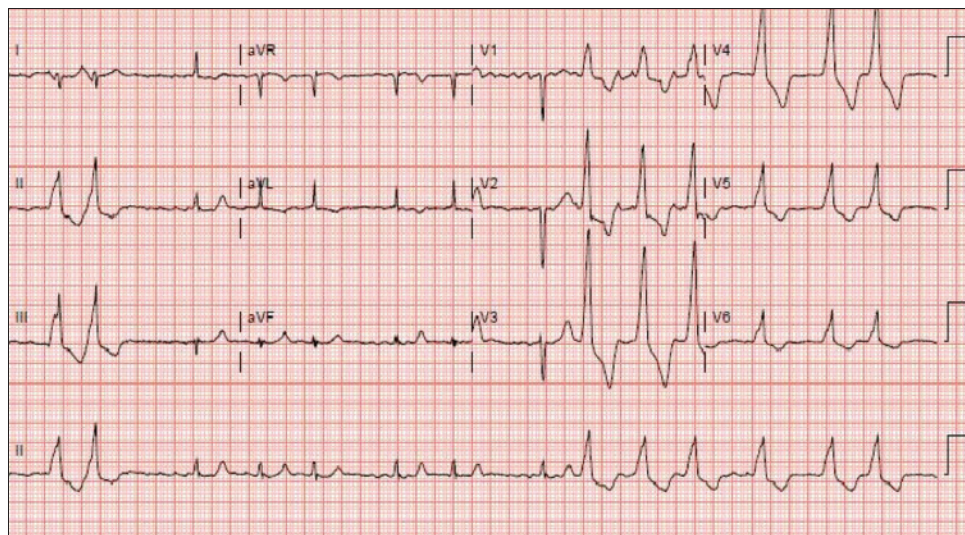


Figure 1. Electrocardiogram at presentation demonstrating atrial fibrillation and wide-complex QRS.



Figure 2. Electrocardiogram from 15-years ago demonstrating sinus rhythm, delta wave, short PR interval and wide-complex QRS.

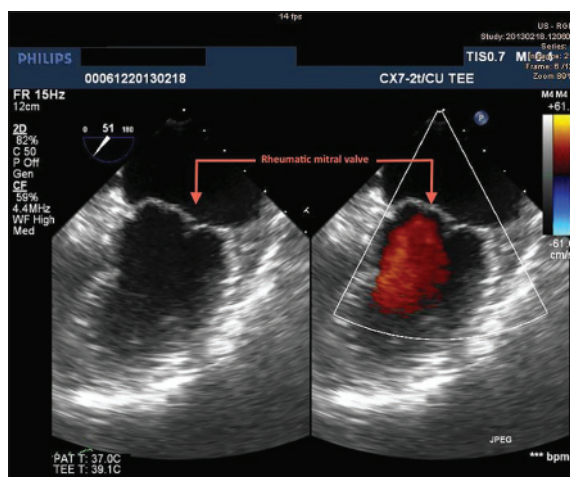


Figure 3. Transesophageal echocardiogram demonstrating rheumatic mitral stenosis.

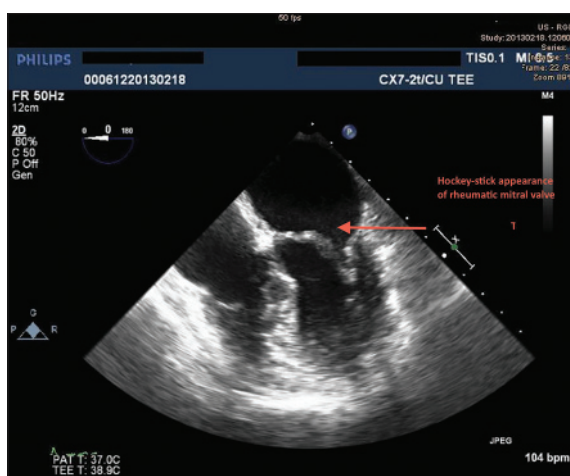


Figure 4. Transesophageal echocardiogram demonstrating characteristic hockey-stick appearance of rheumatic mitral stenosis.

for AF in WPW syndrome include atrial vulnerability and intrinsic atrial abnormality. Atrial vulnerability can be reversible and AP-dependent while 6-24% are intrinsic and AP-independent.⁹ Manifest antegrade AP have a

greater propensity to be associated with AF. Multiple wavelet-re-entry causes rapid, irregular atrial activity resulting in structural changes and further progression of AF. These patients have elevated P-wave dispersion and increased intra-atrial conduction times, which may or may not respond to AP ablation.⁴ Multiple indices of atrial vulnerability, including short atrial ERP, repetitive atrial firing, and longer atrial conduction delay have been proposed, but warrant further investigation.⁹

Pre-excited AF results in electro-mechanical dyssynchrony manifesting as cardiomyopathy, congestive cardiac failure, refractory AF, hemodynamic compromise and multi-organ dysfunction.^{5,7,8} Use of digoxin in these patients can paradoxically worsen heart failure due to shortening of the refractory period and diastolic duration. This can result in inadequate ventricular filling and cardiac output, causing hemodynamic instability.⁵ Like in all patients with WPW syndrome, AV node blocking agents are contraindicated. In addition, pre-excited AF can degenerate into incessant ventricular tachycardia and/or VF contributing to sudden cardiac death (SCD). The incidence of SCD in WPW syndrome is 1:1000, and is related to the shortest pre-excited R-R interval during AF.^{10,11} The risk of SCD is determined by antegrade refractory period, with recent literature advocating TEE and isoproterenol for risk stratification.^{10,12}

EP study forms the cornerstone of diagnosis of AF and WPW AP. Pre-operative EP study specifically in patients with WPW and MS with AF serves to accurately map and perform RFA of AP prior to surgical distortion of LA anatomy. If not amenable to ablation, it serves to provide precise mapping prior to surgical ablation.⁶ However, invasive EP studies carry the inherent risks of vascular injury, hematoma, embolic events, and cardiac perforation.¹⁰ Rapid ventricular rates due to AF conduction or atrial pacing through AP with low-refractory periods raise the pulmonary venous pressure in the presence of MS causing additional risk in these patients.⁶ Alternate imaging modalities such as tissue-Doppler imaging (measuring systolic velocity and strain pattern), TEE EP studies and chemical provocation with isoproterenol have been promoted in recent literature for further risk stratification for SCD in patients.^{10,12,13}

RFA of AP is the recommended first-line treatment with nearly 90% success-rates and minimal complications.¹⁴ However, in patients requiring concurrent surgical replacement of MV, surgical ablation might be recommended.⁵ MV replacement may be conducted simultaneously or in a different time frame, with both

methods reporting similar success rates.⁸ Even though conventional practice advocates restoration of NSR prior to AP ablation, recent evidence has commented on the complications of restoring NSR including obscuration of AP by drugs and alteration of patient compliance with deep sedation required for DCCV.¹⁴ RFA during pre-excitation has been shown to be equally efficacious and safe as RFA after restoration of NSR, including in patients with the combination of MS and WPW syndrome.^{7,14}

CONCLUSION

In summary, we believe that this case study will aid clinicians in recognizing the uncommon association between MS and WPW syndrome with emphasis on diagnostic and management considerations. Further randomized controlled studies are warranted to validate the treatment options discussed to develop an optimal management strategy.

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