A deadly mix - rheumatic mitral stenosis, preexcited atrial fibrillation, left atrial appendage thrombus and left atrial appendage accessory pathway

Kabilan S. Jagadheesan, Sasinthar Rangasamy, Raja J. Selvaraj

Department of Cardiology, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India

1. Case report

A 38-year-old female with rheumatic mitral stenosis on medical management presented with palpitations and presyncope since 10 days. Electrocardiogram showed an irregular rhythm at a rate of 160 bpm with preexcitation suggestive of atrial fibrillation with a left free wall accessory pathway (Fig. 1). Transthoracic echocardiography showed severe rheumatic mitral stenosis with a valve area of 1 cm², moderate mitral regurgitation and a mobile clot within the left atrial appendage (LAA). She was not on anticoagulation. Ventricular rate was between 160 and 180 beats per minute on monitoring.

Anticoagulation with heparin and warfarin was started immediately. Oral flecainide 50 mg bd was started along with oral metoprolol 25 mg bd after two days in an attempt to slow accessory pathway conduction. However, after three doses she developed recurrent polymorphic ventricular tachycardia and ventricular fibrillation requiring defibrillation. With no other obvious cause for the recurrent arrhythmias with QT prolongation during sinus rhythm, the drugs were stopped considering possible proarrhythmia. She was taken up for electrophysiology study with a plan of ablating the accessory pathway.

Diagnostic catheters were placed in the high right atrium, coronary sinus (CS), His region and right ventricle. Corona sinus catheter was pushed inside with distal CS bipole at 3'O clock. Atrial activation during ventricular pacing was eccentric with earliest atrial activation in the distal CS bipole (Fig. 2A). However, local VA interval was still long at the earliest site. Similarly during atrial pacing, ventricular activation was early in distal CS, but local AV interval was long (Fig. 2B). Accessory pathway effective refractory period was 600/260 ms. Mapping of atrial activation during ventricular pacing was done by a transseptal approach. Local VA remained widely separated all along the mitral annulus. Mapping away from the annulus showed significantly earlier atrial activation more medially and anteriorly (Fig. 2A). Echocardiography confirmed this location to be at the base of the LAA (Fig. 3, supplementary video). Ablation here resulted in change to central atrial activation and loss of preexcitation in sinus rhythm. Supplementary video related to this article can be found at http://dx.doi.org/10.1016/j.ipej.2017.09.001.

2. Discussion

Preexcited atrial fibrillation with a rapidly conducting accessory pathway is a medical emergency as ventricular fibrillation can result from the rapid excitation of the ventricle. This scenario has only rarely been described in rheumatic mitral stenosis [1–3]. This combination carries the additional problem of poor tolerance of rapid rates in the presence of significant mitral stenosis. Further complicating the scenario in this patient was the presence of a left atrial thrombus.

Left free wall accessory pathways can usually be ablated at the atrial or ventricular side along the mitral annulus. However, rarely the atrial insertion may be remote from the annulus. One of the common such locations at which atrial insertion may be seen is the LAA. In a study by Long et al., 5 patients were found to have accessory pathway insertion at LAA base [4]. Not identifying the LAA insertion can result in an unsuccessful ablation. Di Biase et al. reported finding LAA insertion of the accessory pathway in 4 patients with structurally normal heart and failed previous ablations [5].

Wide separation of the atrial and ventricular electrograms in CS...
recordings and during endocardial mapping along the annulus should alert one to this possibility. Although the ventricular insertion can be ablated from the tip of the appendage, the atrial insertion may be ablated from the base [6]. In our patient, presence of a thrombus made this more challenging, but we were able to safely complete the procedure by ablating at the base away from

**Fig. 1.** Electrocardiogram at presentation.
Twelve lead electrocardiogram showing wide complex irregular tachycardia at 160 beats per minute with small variations in QRS width suggestive of preexcited atrial fibrillation. Positive delta waves in leads V1 and inferior leads and negative delta waves in leads I and aVL are consistent with a left free wall pathway.

**Fig. 2.** Intracardiac electrograms during atrial and ventricular pacing.
Panel A shows the CS activation during ventricular pacing. Panel B shows ventricular activation during atrial pacing. The tracing in panel B was recorded just before successful ablation and the electrogram in the ablation catheter shows the signal at the successful site.
the thrombus. Ablation of a left sided pathway in the presence of left atrial thrombus has not been described before, but reports of ventricular tachycardia ablation in the presence of left ventricular thrombus [7] suggest that ablation in presence of thrombus may be safe and can be considered in a high risk setting. The patient recovered well post procedure and is scheduled for elective mitral valve replacement.

References

[1] Ikeshita M, Yamate N, Tanaka S, et al. A case report of simultaneous surgery for Wolff-Parkinson-White syndrome combined with haemolytic anemia and mitral stenosis. Jpn Circ J 1996;3:171–6.

[2] Ahmet TA, Hasdemir H, Akyoe A, et al. Wolff-parkinson white syndrome and rheumatic mitral stenosis: an uncommon coincidence that can cause severe hemodynamic disturbance. Indian Pacing Electrophysiol J 2008;8:304–7.

[3] Namboodiri N, Rajeev E, Dora SK, Tharakan JA, et al. Ebstein’s anomaly, Wolff-Parkinson-White syndrome and rheumatic mitral stenosis: role for combined electrophysiological and surgical management. Singap Med J 2007;5:133–5.

[4] Long DY, Dong JZ, Sang CH, Jiang CX, Tang RB, Yan Q, et al. Ablation of left sided accessory pathways with atrial insertion away from the mitral annulus using an electroanatomical mapping system. J Cardiovasc Electrophysiol 2013;24:788–92.

[5] Di Biase L, Schweikert RA, Saliba WI, Horton R, Hongo R, Beheiry S, et al. Left atrial appendage tip: an unusual site of successful ablation after failed endocardial and epicardial mapping and ablation. J Cardiovasc Electrophysiol 2010;21:203–6.

[6] Mollaayeh R, Eslami M, et al. Radiofrequency ablation of left atrial appendage accessory pathway. Europace 2016;18:667.

[7] Rao HB, Yu R, Chitnis N, DO D, Boyle NG, Shvinkumar K, Bradfield JS. Ventricular tachycardia ablation in the presence of left ventricular thrombus: safety and efficacy. J Cardiovasc Electrophysiol 2016 Apr;27(4):453–9.