A CASE FOR EDUCATION

Mahaim-mediated tachycardia using at times the atrioventricular node and other times a left lateral accessory pathway

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Introduction

The diagnosis and management of patients with broad complex tachycardia (BCT) can be challenging and requires careful assessment. While the acute treatment is guided by the patient's immediate clinical status, invasive electrophysiological (EP) testing is of particular value in differentiating ventricular tachycardia from supraventricular tachycardia with aberrancy and for guiding the definitive treatment. We describe an unusual and challenging case of a patient presenting with BCT who underwent an EP study, revealing 3 distinct tachycardias utilizing a Mahaim and left lateral accessory pathway, both of which were successfully treated with catheter ablation. Mahaim fibers, originally described by Mahaim and Winston in 1941,[1] are rare accessory pathways with unique properties, usually found on the right side of the heart, that can present with an antidromic atrioventricular reentrant tachycardia (AVRT) with the appearance of typical left bundle branch block (LBBB) that may masquerade as aberrantly conducted supraventricular tachycardia or ventricular tachycardia. The unique EP properties of Mahaim pathways make their diagnosis and treatment particularly challenging. We describe the EP mechanisms of the 3 distinct tachycardias in our patient, and show how catheter ablation of the accessory pathways successfully treated the patient, with no recurrence of symptoms or arrhythmia at follow-up.

KEYWORDS Supraventricular tachycardia; Mahaim fibers; Accessory pathway; Catheter ablation; Broad complex tachycardia
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Case report

A previously fit and well 32-year-old woman presented to the emergency department with palpitation, dizziness, and hypotension. Her electrocardiogram (ECG) showed a regular BCT (Figure 1), and she was therefore promptly electrically cardioverted to restore sinus rhythm. She had no other previous medical, surgical, or psychiatric history, and no family history of cardiovascular disease or sudden cardiac death. Physical examination of her cardiovascular system was unremarkable. The 12-lead ECG following cardioversion to sinus rhythm did not show any obvious abnormalities and an echocardiogram revealed normal left ventricular and right ventricular (RV) size, wall thickness, and systolic function, with left ventricular ejection fraction of 65%. There were no resting wall motion abnormalities and no significant valve disease apart from mitral valve prolapse with mild mitral regurgitation. Further history from the patient revealed similar self-terminating episodes of palpitation over the previous 2 years. These palpitation episodes were described as regular, rapid heartbeats with sudden onset and offset, associated with dizziness but not related to exertion. A previous 12-lead ECG and Holter monitor performed to investigate these symptoms had not revealed any abnormalities.

She was admitted for further assessment including invasive EP testing. A standard 4-wire study was performed with catheters placed in the high right atrium, coronary sinus, His bundle region, and RV apex. The procedure was performed using conventional EP and fluoroscopic guidance without 3-D electroanatomical mapping. The baseline intracardiac recordings showed normal findings with concentric retrograde and antegrade conduction according to signals in the coronary sinus (CS) catheter. Cycle length (CL) in sinus

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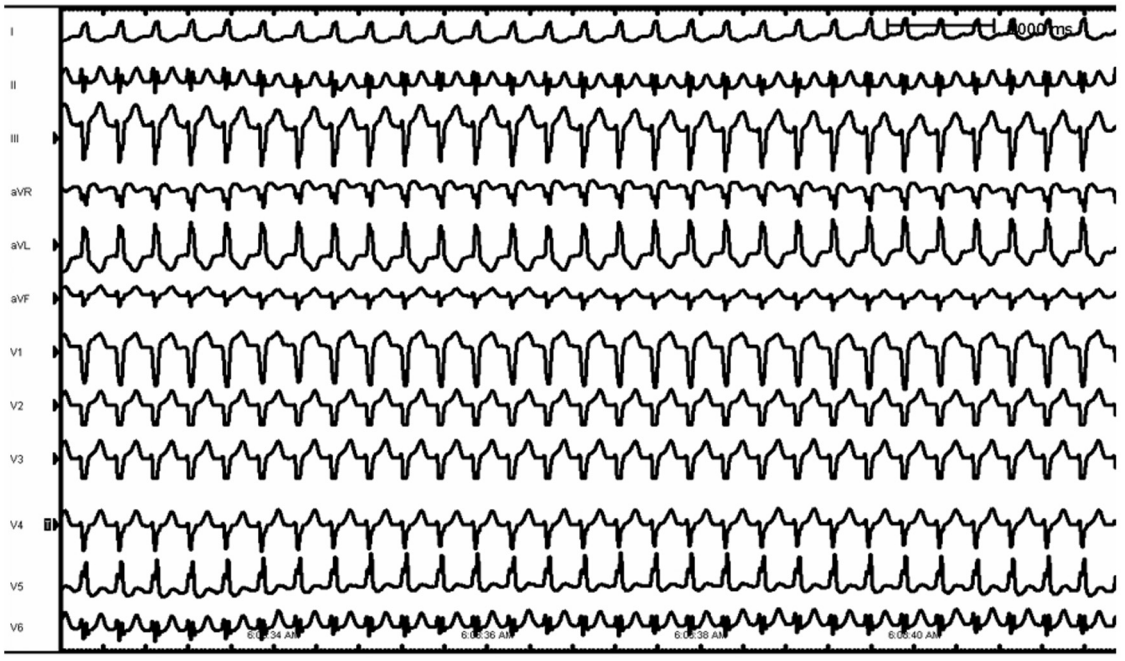

Figure 1 Electrocardiogram during tachycardia 1 showing regular broad complex tachycardia with typical left bundle branch block morphology.

rhythm was 536 ms with AH and HV intervals of 55 ms and 37 ms, and QRS duration of 100 ms. BCT was spontaneously induced with LBBB pattern, QRS transition at V_4, and a CL of 339 ms (Figure 2). The retrograde VA activation was eccentric, with the earliest atrial activation at CS 1-2 (CS distal), suggesting conduction via a left lateral accessory pathway. The antegrade limb of the tachycardia did not seem to be going through the AV node, with RV catheter activation earlier than His recording, where there was no apparent His signal. Pacing from the RV catheter entrained the tachycardia with V-A-V response and retrograde atrial activation sequence identical to the tachycardia, with a post-pacing interval minus tachycardia cycle length (PPI-TCL) of 40 ms (Figure 3), suggesting AVRT.

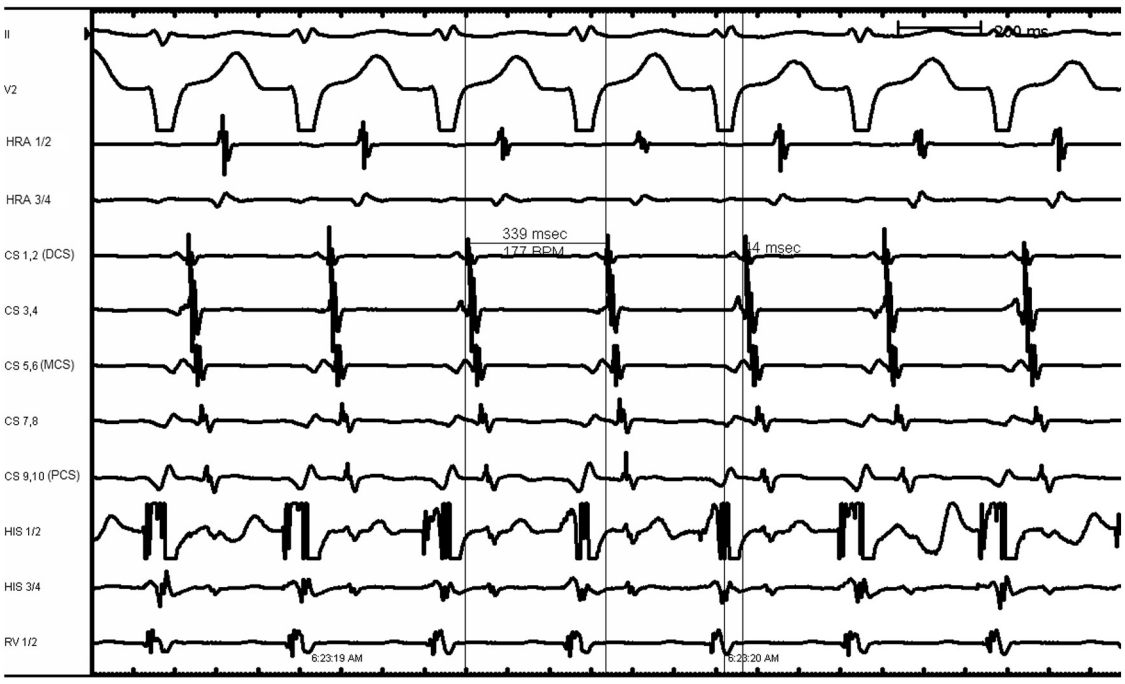

Figure 2 Baseline intracardiac signals recoded during tachycardia 1. Tachycardia cycle length is 339 ms. Retrograde atrial activation sequence is from distal to proximal coronary sinus (CS), with latest A signal in His.

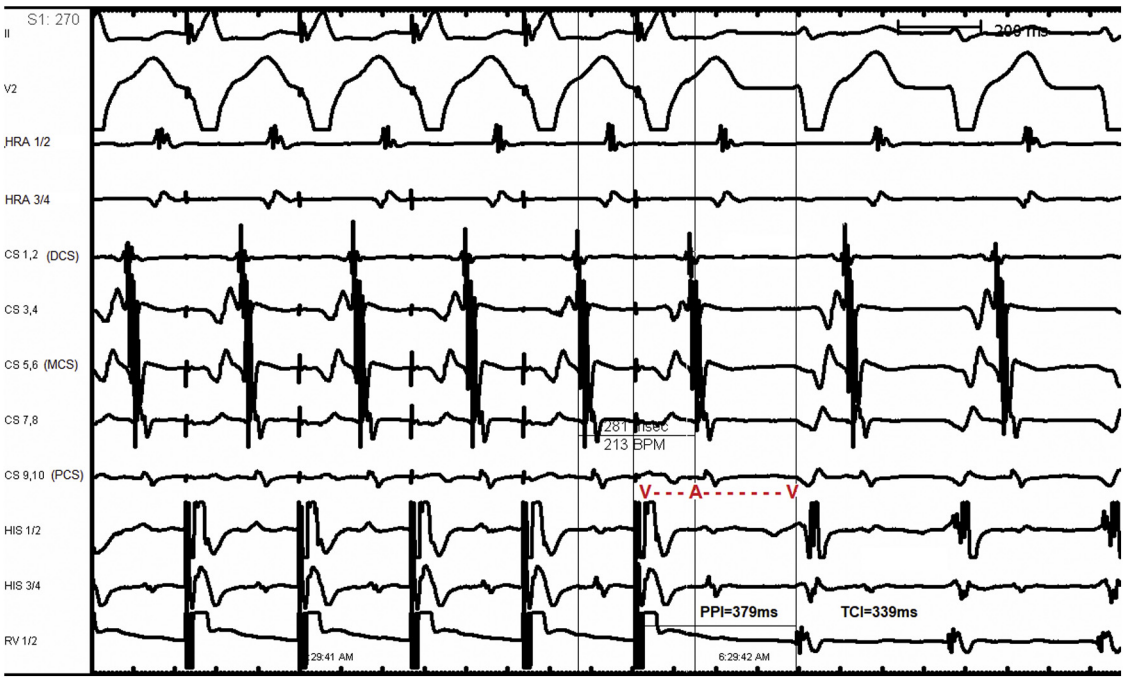

Figure 3 Entrainment of tachycardia 1 from the right ventricle, with a V-A-V response. Postpacing interval (PPI) is short, demonstrating ventricular involvement in the tachycardia circuit and therefore supporting a diagnosis of atrioventricular reentrant tachycardia.

A minute later the tachycardia changed spontaneously, with a speeding up of the CL to 271 ms (Figure 4) but a similar broad complex LBBB surface ECG morphology as tachycardia 1. While the retrograde activation was now concentric, suggesting retrograde activation through the AV node, the antegrade limb remained the same with RV catheter leading. Earliest V signal was noted in the right ventricle (Figure 5) and earliest A electrogram was now at the His catheter with no apparent His signal. Entrainment from the right ventricle again demonstrated a V-A-V response and a short PPI-TCL of 48 ms (Figure 6). These findings supported a diagnosis of

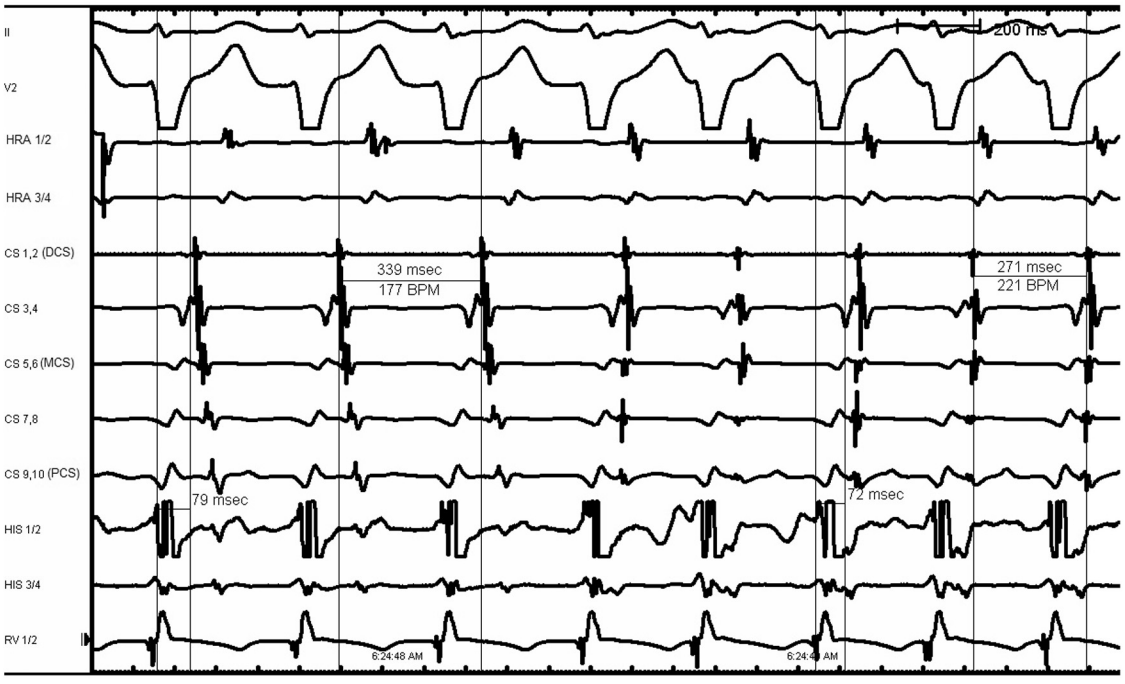

Figure 4 Transition from tachycardia 1 to tachycardia 2 showing the change in retrograde atrial activation sequence in the coronary sinus (CS) and His, suggesting retrograde conduction in tachycardia 2 is through the atrioventricular node.

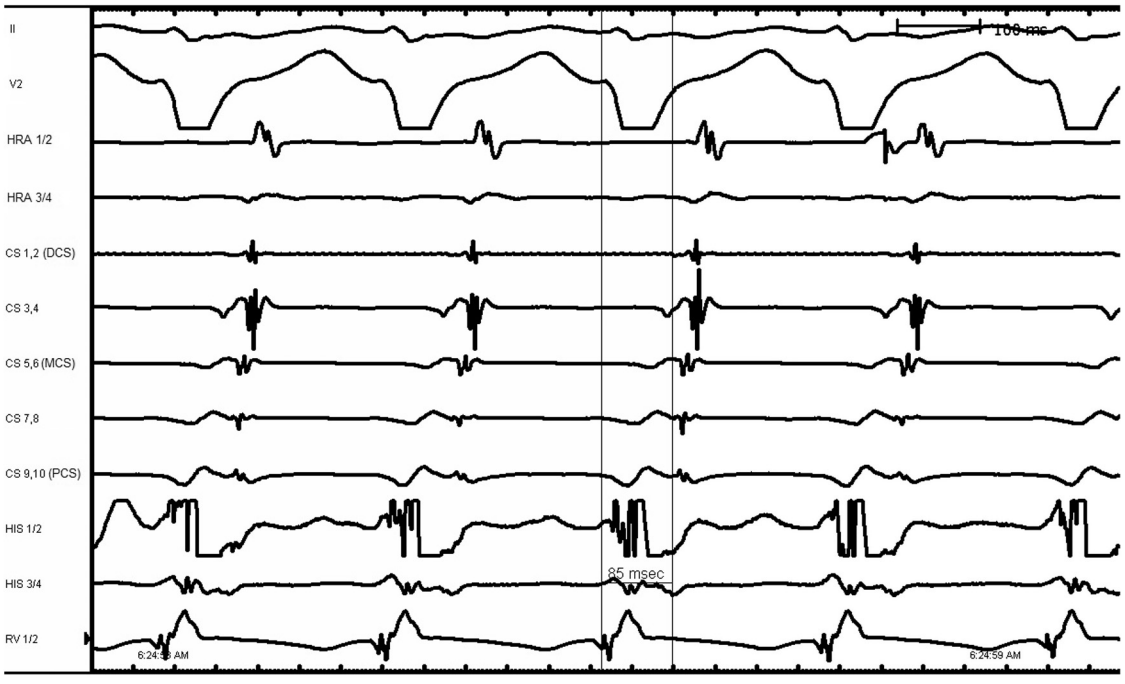

Figure 5 Earliest signal (right ventricular [RV] catheter close to right bundle branch) during tachycardia 2 on the RV catheter demonstrating ventricular preexcitation due to a right-sided accessory pathway.

antidromic AVRT with a right-sided accessory pathway. Following termination of the tachycardia, atrial pacing showed decremental AV conduction (Figure 7), and also initiated tachycardia 1, with clear indication of preexcitation and His signal disappearing into the ventricular signal with the antidromic tachycardia. RV pacing demonstrated retrograde concentric activation pattern and initiated tachycardia 2 (Figure 8), which, like tachycardia 1, was broad complex with LBBB morphology but with a faster CL suggesting a shorter circuit or fast conduction through one of the tachycardia components. The EP features demonstrated were consistent with Mahaim fiber atriofascicular accessory pathway. A third tachycardia with a narrow QRS complex and a faster CL of 264 ms (Figure 9)

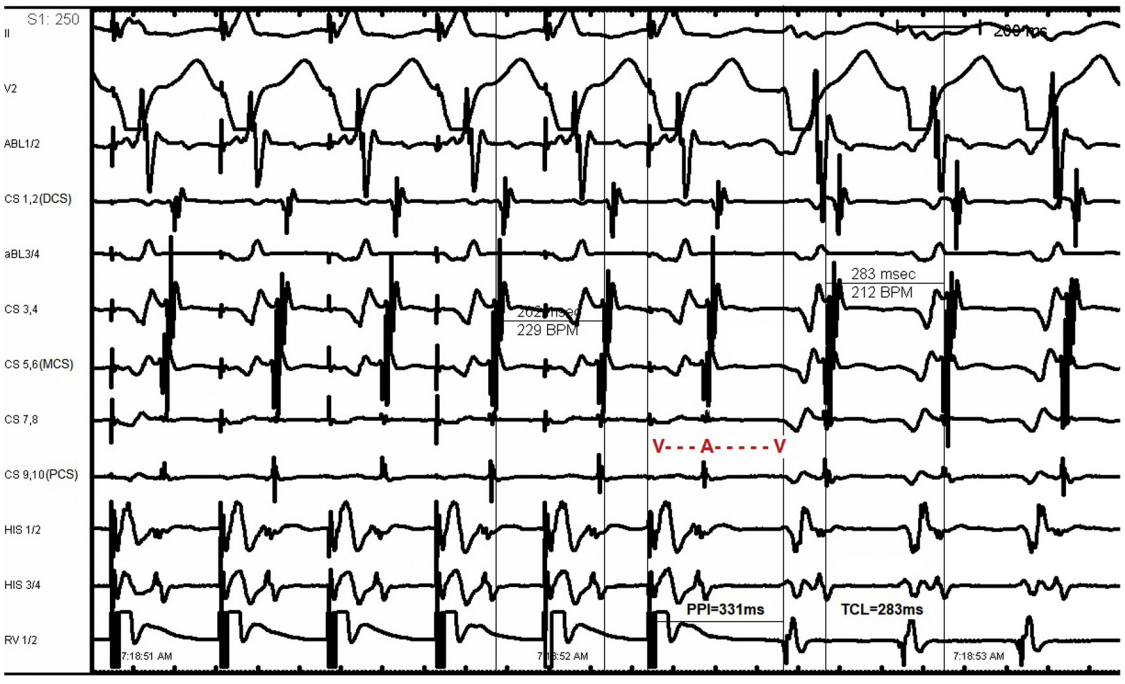

Figure 6 Entrainment of tachycardia 2 from the right ventricle showing a short postpacing interval (PPI) to suggest involvement of the ventricle in the tachycardia circuit consistent with atrioventricular reentrant tachycardia.

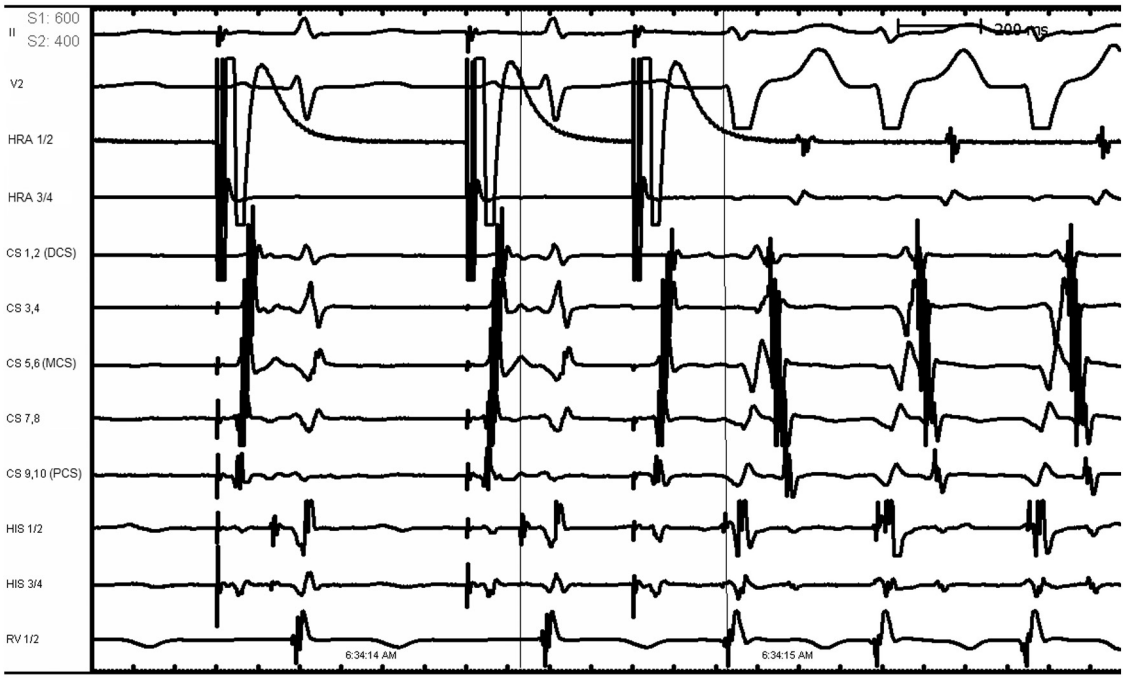

Figure 7 Atrial extrastimulus pacing with S1–S2 coupling interval of 600–400 ms, showing decremental atrioventricular (AV) conduction followed by initiation of tachycardia 1. The extrastimulus is conducted down the AV node to the ventricles, then retrograde via the left-sided accessory pathway, as demonstrated by the earliest signal in coronary sinus (CS) 1-2, to the high right atrium (ABL channel), and then back down to the ventricle via a right-sided accessory pathway, as demonstrated by the signal on the right ventricular catheter.

was also seen spontaneously. Antegrade conduction was through the AV node with eccentric retrograde atrial activation, earliest in the distal CS (Figure 10). This suggested an orthodromic AVRT with retrograde activation of the atrium via a left lateral accessory pathway.

Therefore, in summary, tachycardia 1 was proposed to be a duodromic AVRT utilizing the Mahaim pathway for antegrade conduction and a left lateral pathway for retrograde conduction. Tachycardia 2 was an antidromic AVRT utilizing the Mahaim pathway antegrade and AV node retrograde,

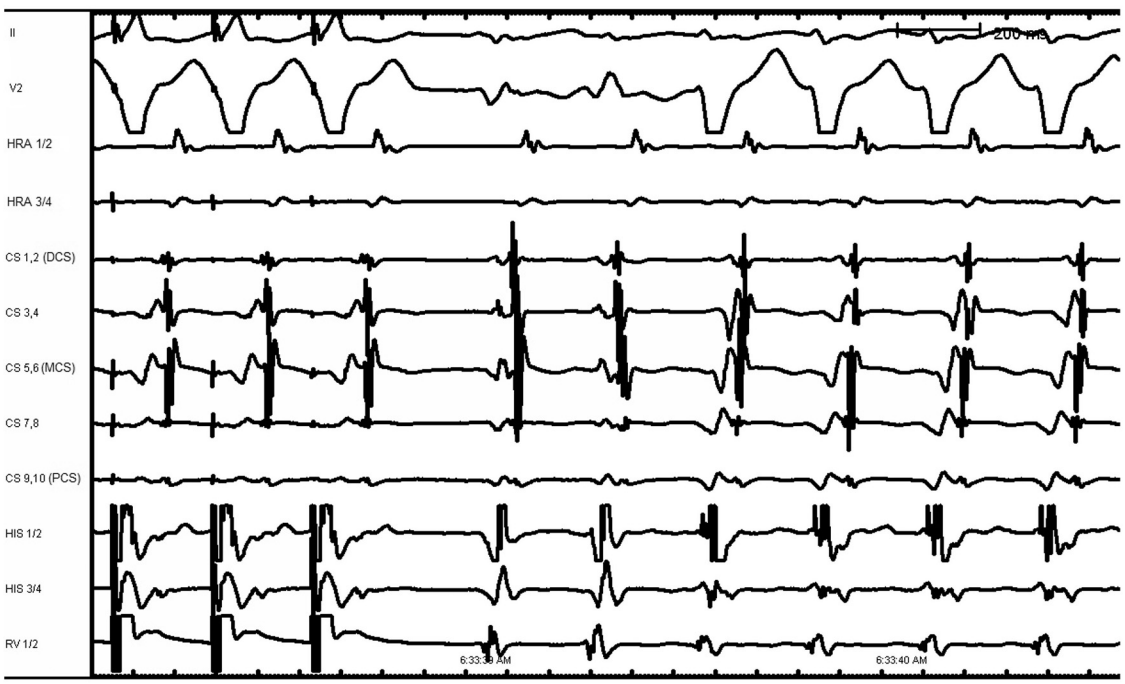

Figure 8 Tachycardia 2 was initiated by pacing from the right ventricle (RV). Owing to unidirectional (retrograde) block in the Mahaim pathway, RV pacing is able to induce an atrioventricular reentrant tachycardia circuit.

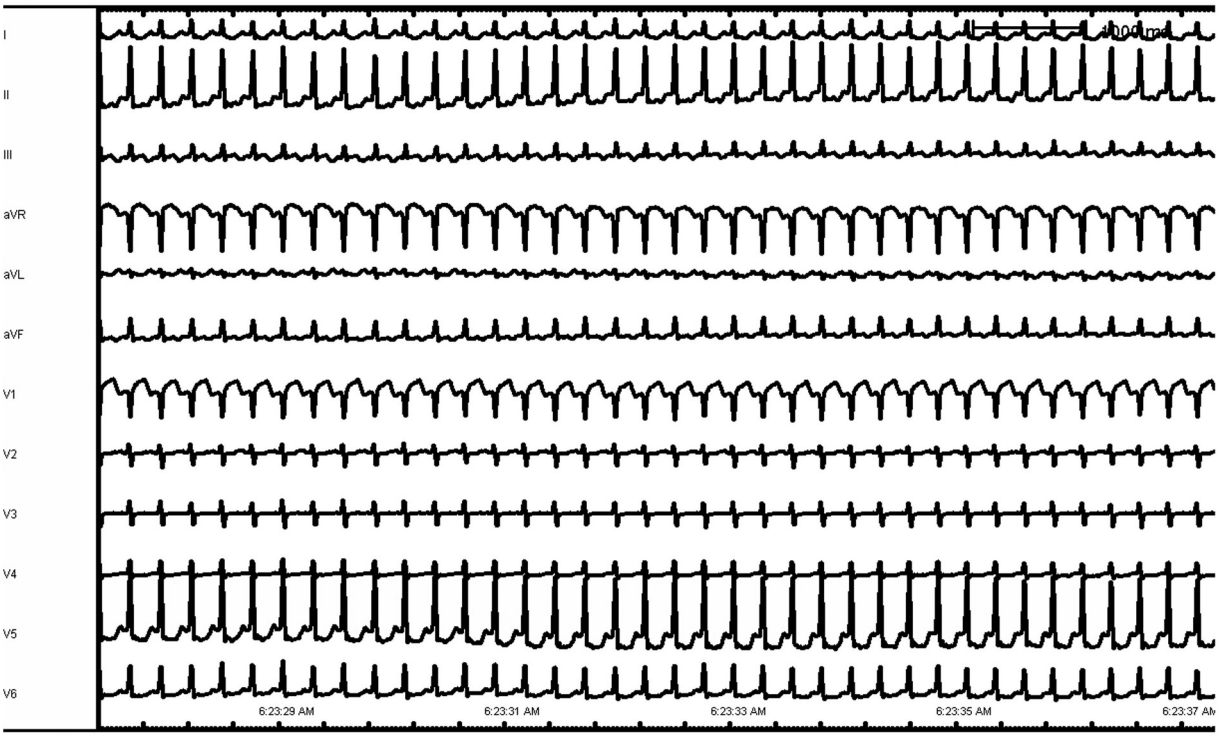

Figure 9 A 12-lead electrocardiogram of tachycardia 3 showing a narrow complex tachycardia.

and tachycardia 3 an orthodromic AVRT utilizing the AV node antegrade and left lateral pathway retrograde. These are illustrated in Figure 11. We proceeded to carry out catheter ablation of the left lateral accessory pathway, where ablation over a fused V-A signal resulted in immediate termination of the tachycardia with a V signal (Figure 12) and normalization of the retrograde atrial activation sequence to a concentric pattern. Tachycardia 2 was then reinduced and mapping was carried out around the tricuspid valve annulus, followed by around the ventricular insertion site at the region

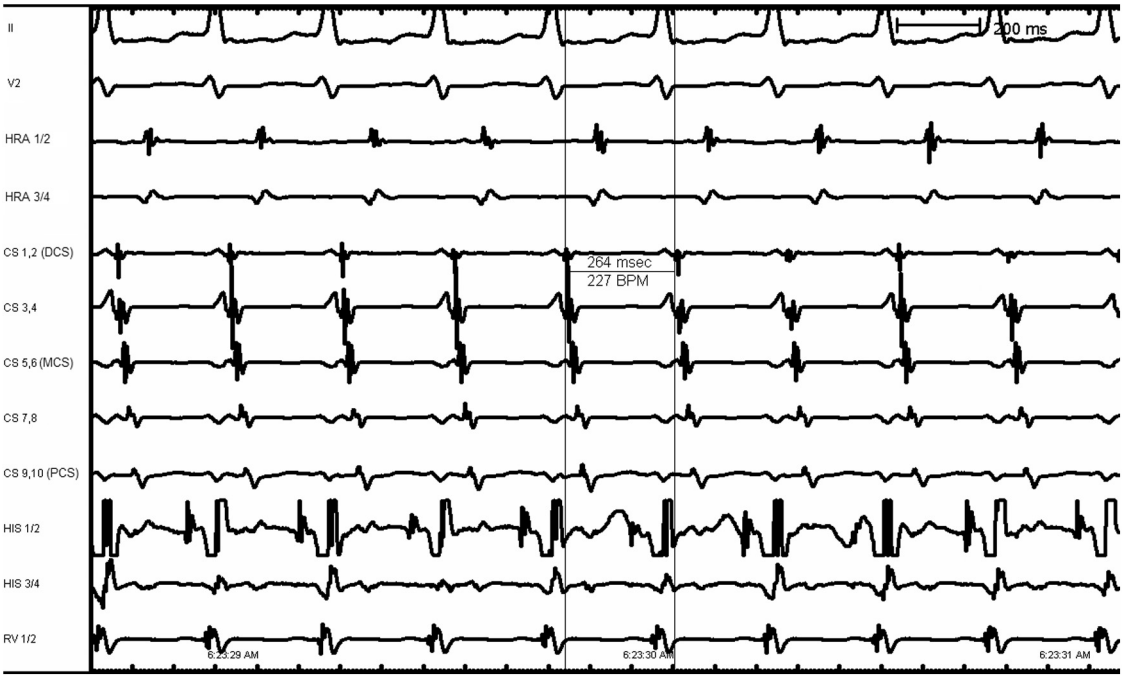

Figure 10 Intracardiac signals recorded during tachycardia 3. The antegrade conduction is through the His (atrioventricular [AV] node) to the ventricle, with eccentric retrograde activation, earliest in distal coronary sinus (CS), therefore demonstrating orthodromic AV reentrant tachycardia via a left lateral accessory pathway.

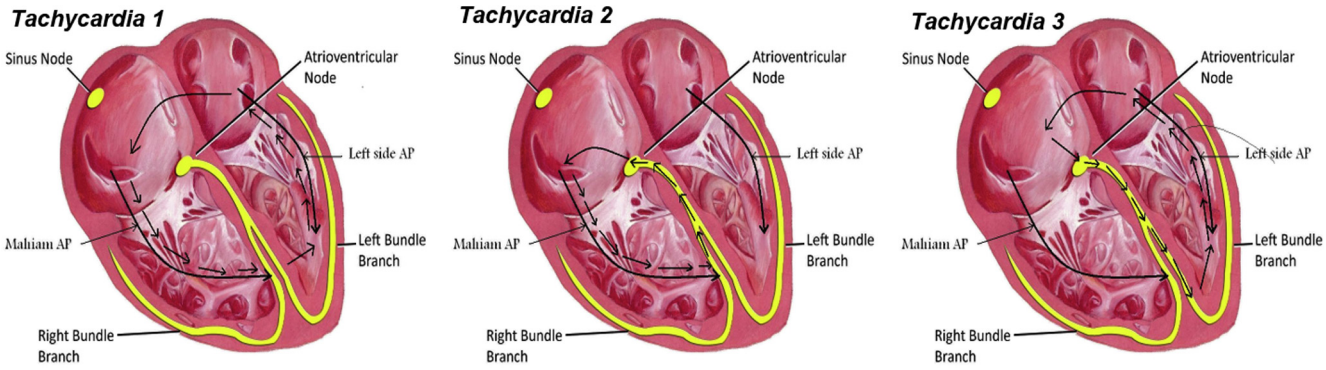

Figure 11 Schematic diagram of the proposed mechanism of the 3 tachycardias.

of the right bundle branch. Radiofrequency ablation was performed at this location where a signal 30 ms preceding surface QRS immediately terminated the tachycardia (Figure 13). All 3 tachycardias were rendered noninducible following ablation of these 2 pathways, with no recurrence at 12 months' follow-up.

Discussion

This case provides a unique demonstration of the mechanism of 3 distinct AVRTs utilizing Mahaim and left lateral accessory pathways. Mahaim conduction is usually used to describe accessory pathways with decremental conduction properties that usually only conduct antegrade and have long conduction times. The original description by Mahaim and Winston referred to fibers arising from the AV node or His-Purkinje system and terminating in the right ventricle. The AV node was assumed to be involved in the antegrade conduction associated with tachycardia involving these pathways.[2] However, since Klein and colleagues[3] reported on a patient with a Mahaim pathway that was mapped to the anterior lateral tricuspid annulus, the more accurate term "atriofascicular" has been widely adopted to describe this group of pathways. These accessory pathways represent an "accessory" AV conduction system consisting of a proximal component positioned at or above the tricuspid annulus, similar to the AV node, connecting to a distal component described as a "long" fiber to the right bundle branch without an intermediate insertion into the right ventricle. "Short" pathways have also been described, which insert into the ventricle and are termed atrioventricular rather than atriofascicular. Thus, pathways with Mahaim characteristics can be atriofascicular, atrioventricular, nodofascicular, or nodoventricular, depending on their variable proximal and distal insertions, and the term Mahaim is widely used as an umbrella term for pathways with these unique EP

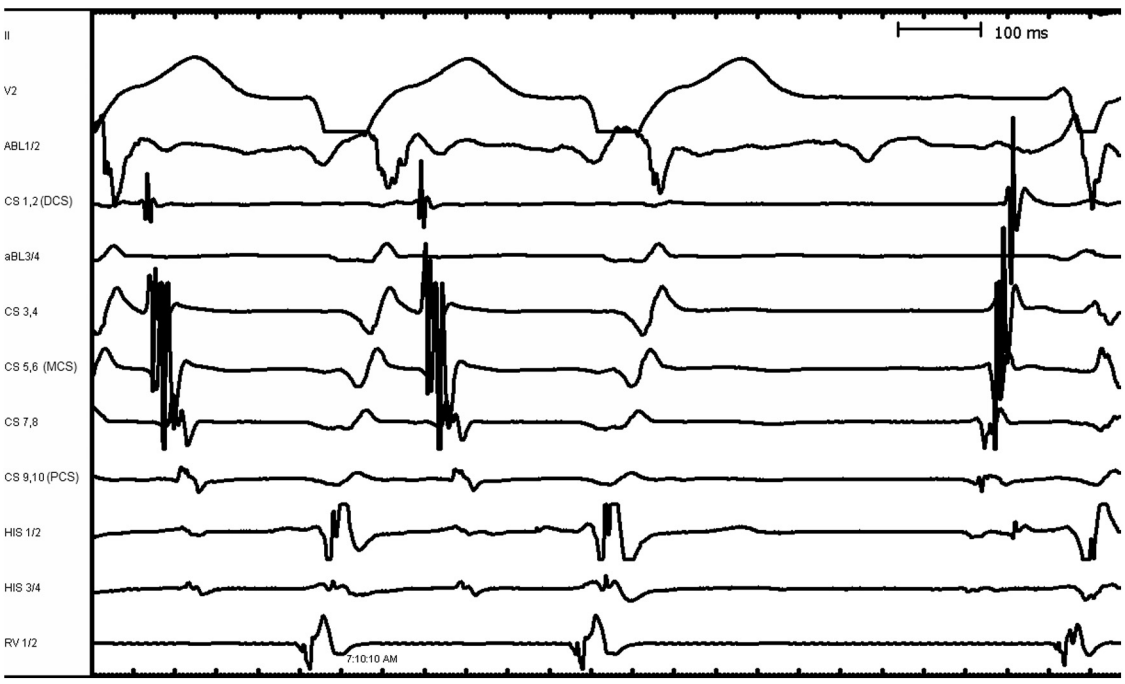

Figure 12 Signal from location of successful ablation at the left lateral mitral valve annulus. Fused A and V signal on ablation catheter (ABL) with immediate loss of accessory pathway conduction, with ablation terminating tachycardia on a V signal.

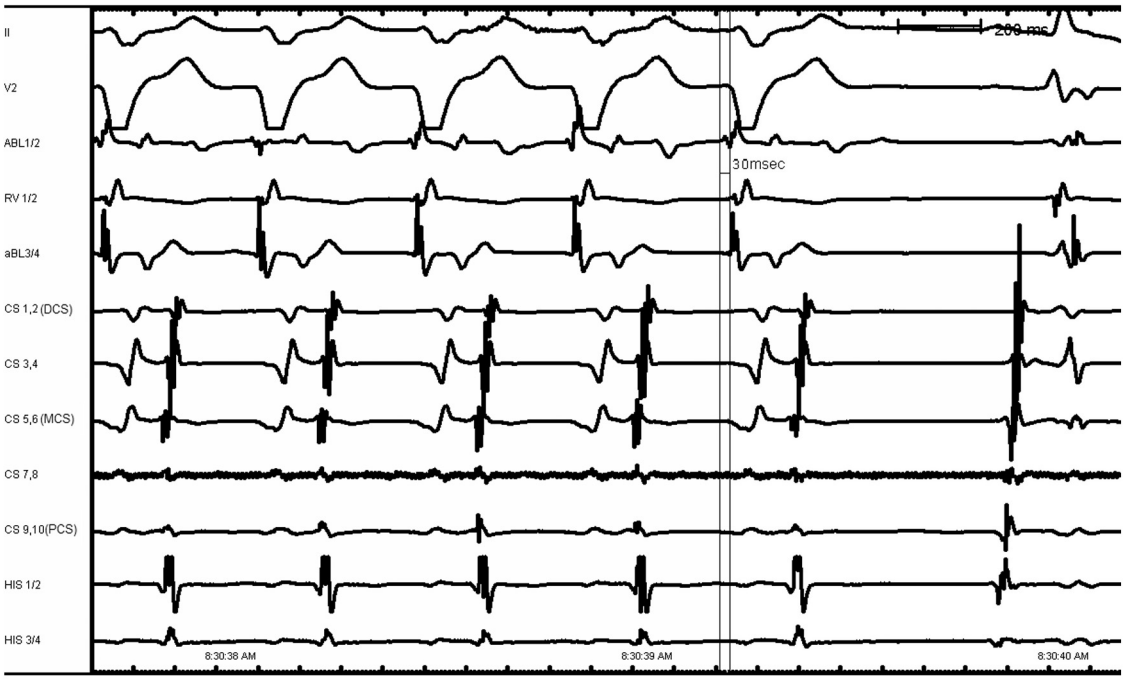

Figure 13 Site of successful ablation of the Mahaim pathway. Earliest signal seen pre–QRS onset on the ablation catheter.

characteristics. Location of Mahaim pathways in the left heart has also been reported.[4]

Histologic examination of the proximal component shows morphologically similar cells to those in the AV node, explaining some of the unique EP properties, which make their diagnosis and treatment particularly challenging. A key differentiating feature of atriofascicular Mahaim pathways is that during antegrade pre-excitation or reentrant tachycardia, the right bundle electrogram precedes His bundle activation.[5] To induce tachycardia a critically timed atrial premature beat when there is conduction block in the AV node may be needed, but this may not initiate tachycardia if the AV node has better antegrade conduction than the pathway. In this circumstance, ventricular premature beats can be used, as the Mahaim pathways typically block in retrograde direction.[6] In our case, initiation of tachycardia was relatively easy, possibly owing to the presence of an additional pathway on the left side. In Figure 7 we show initiation of tachycardia 1 (duodromic AVRT) with an atrial extrastimulus; and in Figure 8, tachycardia 2 (antidromic AVRT) initiated with ventricular pacing.

Demonstrating the presence of multiple accessory pathways and proving their involvement in the tachycardia circuit can be challenging. Analysis of the retrograde atrial activation sequence, changes in CL, and the use of pacing maneuvers can be helpful in making these distinctions. In our case, Figure 4 demonstrates shortening of the tachycardia CL with a change in retrograde atrial activation from eccentric to concentric as tachycardia 1 changes to tachycardia 2. This is highly suggestive of a left-sided accessory pathway critical to tachycardia 1. Advancing and resetting the tachycardia through delivery of an atrial stimulus at the lateral right atrium near the atrial insertion of the Mahaim pathway at a time when the AV node is refractory (septal-refractory atrial stimulus) may help to prove involvement of the right-sided Mahaim pathway in the tachycardia rather than as a bystander. This was not performed in our EP study and is a limitation of the case we have presented. Furthermore, while the eccentric retrograde atrial activation during tachycardia 3 is highly suggestive of orthodromic AVRT using a left-sided accessory pathway, this does not prove involvement of the pathway in the tachycardia circuit, and additional maneuvers, such as delivery of a His-refractory PVC, may be helpful to prove involvement of the left-sided pathway if the tachycardia is advanced and reset. Long CL pacing from the right atrium and left atrium can sometimes identify an atriofascicular pathway along the right atrial free wall, as ventricular activation from the right atrium can bypass the tricuspid annulus and pre-excite the ventricles independent of the AV node, whereas stimulation from the left atrium would not. However, in cases such as ours, where there was also a coexisting left-sided accessory pathway, this maneuver may not be helpful. Pacing from the left ventricle close to the left lateral accessory pathway was not attempted during this case; it may be useful to perform during orthodromic AVRT involving this pathway, as there will be evidence of fusion and shorter PPI-TCL, compared to pacing from the right ventricle, where a stimulus needs to be delivered early enough in order to advance and reset the tachycardia and thereby produce a paced QRS complex.[7]

Radiofrequency catheter ablation of accessory pathways is an established first-line treatment approach. Mapping of atriofascicular accessory pathways can be at the origin, which is usually around the lateral, anterolateral, or posterolateral

tricuspid annulus, or the distal insertion at the apical region of the RV free wall. In 80% of cases successful ablation is from the atrial side; however, in 20% of cases ablation needs to be carried out at the ventricular insertion.[6] The absence of retrograde conduction makes it more difficult to locate the atrial insertion site, and thus recording an "M" Mahaim potential may aid in identification of the pathway. Ablation at the ventricular insertion site should be carried out proximal to the pathway's first connection with the ventricle or right bundle to avoid prolonging VA conduction time and increasing episodes of AVRT.[8] The occurrence, during radiofrequency ablation at the target area, of a slow automatic rhythm similar in morphology to the fully pre-excited QRS complex has been identified as a positive sign of successful elimination of the accessory pathway and may provide further evidence of AV node–like EP properties, as it mirrors the junctional rhythm seen during slow pathway ablation.[9]

The ECG findings of patients with atriofascicular pathways is usually subtle, with no or very minimal preexcitation. The only indication may be the absence of septal Q waves in V_5 or V_6 and rS pattern in lead III. However, during tachycardia the QRS morphology is maximally preexcited with LBBB morphology and leftward superior axis, as seen in Figure 1. Adenosine is widely used as a diagnostic test to reveal accessory pathway conduction. In Mahaim fiber pathways, adenosine produces further conduction delay, and a transient increase in pre-excitation followed by conduction block, in most cases.[10]

Our case is a rare description of coexisting Mahaim and left lateral accessory pathways, both of which were involved in tachycardia. Some patients with Mahaim pathways may also have dual AV node physiology, atrial tachycardia, or ventricular tachycardia, and the pathway may function as an innocent bystander. Therefore, careful assessment is critical to prove involvement of the pathway in the tachycardia circuit and avoid overly aggressive treatment that may pose a risk of inadvertent harm to the patient.

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