Ablation of a symptomatic spontaneous automatic focus arising from an atriofascicular fiber

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Introduction

Atriofascicular accessory pathways (AP), often referred to as Mahaim pathways, are uncommon electrical connections that most often course along the lateral right atrium and insert within the distal right bundle branch (RBB). Pathognomonic electrophysiological characteristics are well described, including unidirectional anterograde conduction with AV node–like decremental properties. Reported targets for ablation include the site of the Mahaim potential along the atrial aspect of the tricuspid annulus or at the ventricular insertion site of the pathway.1 Spontaneous activity arising from an atriofascicular AP fiber have been described during ablation of its atrial insertion.2 Herein, we report the first case of ablation of symptomatic atriofascicular AP automatically mimicking apicolateral right ventricular (RV) ectopy, which manifested years after AP ablation for antidromic atrioventricular reentrant tachycardia (AVRT).

Case report

A 32-year-old woman with a structurally normal heart was referred to our institution for symptomatic premature ventricular contractions (PVCs) despite antiarrhythmic drug therapy. Sixteen years prior, she had antidromic AVRT via an atriofascicular AP that was successfully ablated at its proximal insertion site. She remained asymptomatic until recently, when brief but frequent palpitations emerged during fertility treatments that were different from her prior symptoms in the setting of AVRT. Twenty-four-hour Holter monitoring documented frequent monomorphic PVCs (18% of all heart beats). A 12-lead electrocardiogram (ECG), depicted in Figure 1A, revealed PVCs with a left bundle branch block (LBBB) morphology and superior axis. No sustained ventricular tachycardia was recorded. Physical examination and echocardiography were unremarkable.

As a result of persistent and debilitating symptoms, an electrophysiological study was performed under local anesthesia with conscious sedation (ie, intravenous midazolam). Standard multielectrode catheters were introduced through the femoral veins and positioned under fluoroscopic guidance in the high right atrium, coronary sinus, His-bundle region, and RV apex. Reconnection of the atriofascicular pathway was demonstrated as follows: (1) pre-excitation induced by high right atrial pacing with a maximally pre-excited ECG during atrial pacing having the same morphology as spontaneous PVCs (Figure 1B); (2) decremental conduction across the atriofascicular bypass tract with progressive AV and AH interval prolongation coupled with shortening of the HV interval and progressive widening of the QRS complex during atrial pacing; (3) progressive prolongation of the AV interval with progressive prematurity of atrial extrastimuli; and (4) lack of retrograde conduction over the AP. The proximal insertion site was localized by identifying the atriofascicular pathway potential (ie, M-potential) along the atrial aspect of the posterolateral tricuspid annulus. The distal insertion was considered to be fascicular, with an RBB potential present before each non-pre-excited QRS complex. Antegrade refractory periods of the AV node and atriofascicular fiber were 290 and 350 ms, respectively. No tachycardia was inducible with atrial or ventricular pacing either with or without an isoproterenol perfusion (up to 2 μg/min). Frequent symptomatic and spontaneous PVCs were recorded throughout the procedure. Activation mapping was performed during PVCs to identify the site of earliest activation relative to the onset of the QRS. The earliest bipolar signal was localized to the apicolateral RV wall and was preceded (20 ms) by a sharp Purkinje potential (Figure 2). Pace mapping at a site where small potentials were recorded (presumably the distal RBB) produced a QRS morphology identical to PVCs and pre-excited beats. Radiofrequency ablation performed during sinus rhythm targeting this RV apicolateral site produced automatic ventricular ectopy (Figure 3). The morphology of this rhythm was identical to the patient’s spontaneous PVCs. No recurrence of PVCs occurred after ablation. In addition, ablation at this site eliminated atriofascicular antegrade conduction. The ECG after ablation was normal, without RBB block.
follow-up of 2 years, the patient remains asymptomatic, with no PVCs recorded by multiple 24-hour Holter recordings.

**Discussion**

Spontaneous automatic activity originating from an atriofascicular AP appears to be exceedingly rare, and has been reported in a minority of patients with atriofascicular pathways. This case describes highly symptomatic PVCs with the same QRS morphology as pre-excitation across an atriofascicular pathway. The origin of these PVCs was mapped to the distal insertion of the atriofascicular fiber, where ablation resulted in the disappearance of PVCs and elimination of atriofascicular pathway conduction.

Rare cases of spontaneous automaticity arising from atrioventricular APs have been reported. Spontaneous automaticity of Mahaim fibers has been infrequently observed, predominantly soon after ablation, sometimes competing with AV node escape rates. Sternick noted spontaneous automaticity in 12.5% of 40 patients with Mahaim fibers, with a spectrum of automatic rhythms.

**Figure 1** Pre-excitation induced by atrial pacing with the same morphology as the clinical premature ventricular contractions (PVCs). **A:** Recordings of the surface 12-lead electrocardiogram (ECG) with monomorphic spontaneous “PVCs.” **B:** Recordings of the surface 12-lead ECG during the electrophysiology study with incremental atrial pacing and an atrial premature pacing at 390 ms. The QRS morphology during maximum pre-excitation is similar to the spontaneous “PVC.”

| KEY TEACHING POINTS |
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| • Spontaneous automaticity of Mahaim fibers is rare and occurs predominantly soon after radiofrequency ablation. |
| • In this case, spontaneous automaticity occurred more than 15 years after radiofrequency ablation, preventing the patient from receiving fertility treatment. |
| • Electrophysiological study and careful mapping are essential in localizing the site of origin and identifying appropriate treatment. |

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ranging from asymptomatic slow rhythms to fast and repetitive bursts of tachycardia with an LBBB-like morphology. To our knowledge, no case of catheter ablation for symptomatic Mahaim automaticity recurring after prior AP ablation has previously been reported. In this case, symptoms emerged during fertility treatments, which raises the possibility that hormonal shifts were involved in the pathogenesis of automaticity. Cyclical arrhythmia patterns have previously been described, with estrogen providing antiarrhythmic effects. Estrogen-blocking drugs used to treat infertility can theoretically favor arrhythmias in susceptible women in a manner similar to the luteal phase of the menstrual cycle.

Mahaim pathways may be ablated along the atrial aspect of the tricuspid annulus (at the site of a high-frequency potential, shortest stimulus-to-pre-excitation interval, and/or greatest advancement of subsequent QRS complex by critically timed atrial extrastimuli during AVRT), or at the ventricular insertion site identified by activation mapping. In this case, the AP was initially ablated at its proximal insertion. Sixteen years later, antegrade conduction through the AP fiber persisted without recurrent antidromic AVRT. Ablation at the distal insertion of the atriofascicular pathway simultaneously eliminated the targeted PVCs and AP conduction. Haissaguerre et al previously explored the distal insertion site of decremental conducting APs in a series of patients. They found that the distal insertion of the Mahaim fiber may arborize 0.5–2.0 cm in diameter in some patients, explaining why ablation at this site may be challenging. In our case, application of radiofrequency energy at the distal insertion site produced transient acceleration of a ventricular rhythm with the same LBBB morphology. Heat-induced automaticity during radiofrequency current delivery has previously been reported. This type of automaticity has been likened to the junctional rhythm that arises during ablation of the AV nodal slow pathway. Complete elimination of automatic activity has been proposed to represent a hallmark for successful ablation of atriofascicular pathways. Some investigators have proposed that simultaneous elimination of AP conduction and automaticity by a single discrete lesion indicates that the same tissue is implicated in AP conduction and such an automatic response. In our case, disappearance of AP conduction, elimination of the spike potential, and the

Figure 2 Surface electrocardiogram, intracardiac electrograms, and electrode positions during sinus rhythm and premature ventricular contraction (PVC). Surface leads II, III, and V1 and intracardiac electrograms are shown at the successful ablation site. During the PVC, the distal unipolar electrogram recorded by the ablation catheter (MAP UNI) demonstrates a QS pattern and the bipolar recording indicates that the signal is 20 ms earlier than the surface QRS onset (blue calipers). The Purkinje signal is indicated by the arrow.
absence of induced RBB block suggest that the PVCs originated from the AP/insertion site rather than from the more distal His-Purkinje system.

**Conclusion**
Symptomatic PVCs may arise from the distal insertion of an atriofascicular AP. Catheter ablation at 1 discrete site can eliminate both PVCs and atriofascicular pathway conduction.

**References**
1. Kothari S, Gupta AK, Lokhandwala YY, Vora AM, Kerkar PG, Thakur RK. Atriofascicular pathways: where to ablate? Pacing Clin Electrophysiol 2006;29:1226–1233.
2. Sternick EB, Sosa EA, Timmermans C, Cruz Filho FE, Rodriguez LM, Gerken LM, Scanavacca MI, Fagundes ML, Bueno SC, Veandecic MO, Wellens HJ. Automaticity in Mahaim fibers. J Cardiovasc Electrophysiol 2004;15:738–744.
3. Belhassen B, Ilan M, Glick A. Wide QRS rhythm in a young woman with recurrent palpitations: what is the diagnosis? J Cardiovasc Electrophysiol 2003;14:1376–1378.
4. Macle L, Shah DC, Jais P, Haissaguerre M. Accessory pathway automaticity after radiofrequency ablation. J Cardiovasc Electrophysiol 2002;13:285–287.
5. Ma FS, Ma J, Chu JM, Fang PH, Wang FZ, Chen X, Zhang S. The automaticity of Mahaim fibre and its response to effective ablation. Chin Med J (Engl) 2004;117:1768–1771.
6. Dora SK, Tharakan JA, Valaparambil A, Namboodiri N, Nair K, Peter T. Spontaneous automaticity of an atriofascicular accessory pathway. Europace 2006;8:140–143.
7. Strohner B, Scherenthiner C, Hwang C. Spontaneous automaticity arising from a successfully ablated Mahaim fiber. J Interv Card Electrophysiol 2007;20:25–28.
8. Sternick EB. Automaticity in a Mahaim fiber. Heart Rhythm 2005;2:453; author reply 453.
9. Yarnoz MJ, Curtis AB. More reasons why men and women are not the same (gender differences in electrophysiology and arrhythmias). Am J Cardiol 2008;101:1291–1296.
10. Chen CC, Lin CC, Lee TM. 17beta-estradiol decreases vulnerability to ventricular arrhythmias by preserving connexin43 protein in infarcted rats. Eur J Pharmacol 2010;629:73–81.
11. Savergnini SQ, Reis AM, Santos RA, Santos PE, Ferreira AJ, Almeida AP. Effects of short-term administration of estradiol on reperfusion arrhythmias in rats of different ages. Braz J Med Biol Res 2012;45:1248–1254.
12. Gowd BM, Thompson PD. Effect of female sex on cardiac arrhythmias. Cardiol Rev 2012;20:297–303.

**Figure 3**
A: Surface 12-lead electrocardiogram showing the automatic rhythm induced during radiofrequency ablation at the site of earliest premature ventricular contraction (PVC) activation, leading to an automatic irregular rhythm with the same left bundle branch block morphology. B: Intracardiac recordings from the coronary sinus (CS) and MAP catheters show AV dissociation during ventricular automaticity induced by radiofrequency ablation. C: Electroanatomic activation map of PVCs in the anteroposterior view. The earliest site of activation during PVCs is located in the apicolateral region of the right ventricle. Radiofrequency ablation was performed during sinus rhythm at this site (blue circle).
13. Klein LS, Hackett FK, Zipes DP, Miles WM. Radiofrequency catheter ablation of Mahaim fibers at the tricuspid annulus. Circulation 1993;87:738–747.

14. Haissaguerre M, Cauchemez B, Marcus F, Le Metayer P, Lauribe P, Poquet F, Gencel L, Clementy J. Characteristics of the ventricular insertion sites of accessory pathways with anterograde decremental conduction properties. Circulation 1995;91:1077–1085.

15. Morita N, Kobayashi Y, Katoh T, Takano T. Anatomic and electrophysiologic evaluation of a right lateral atrioventricular mahaim fiber. Pacing Clin Electrophysiol 2005;28:1138–1141.

16. Ducceschi V, Vitale R, Sokola EA, Ottaviano L, Santinuolo R, Gregorio G. Ablating the ventricular insertion of atrio-fascicular Mahaim fiber: Could be performed safely? Indian Pacing Electrophysiol J 2009;9:119–124.

17. Sternick EB, Gerken LM, Vrancedic M. Appraisal of "Mahaim" automatic tachycardia. J Cardiovasc Electrophysiol 2002;13:244–249.

18. Deam AG, Burton ME, Walter PF, Langberg JJ. Wide complex tachycardia due to automaticity in an accessory pathway. Pacing Clin Electrophysiol 1995;18:2106–2108.

19. Giudici MC, Jackman WM. Ectopy and wide QRS tachycardia: what is the mechanism? Pacing Clin Electrophysiol 2004;27:1424–1427.