Atrial Tachycardia in a Patient With Fabry's Disease

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Introduction

Both atrial and ventricular arrhythmias, in particular atrial fibrillation, are commonly observed among patients with Fabry's disease (FD). The manuscript describes a rare case of atypical atrial tachycardia (AT) in .a patient with FD and advanced structural heart disease.

Case report

A 60-year-old male subject with a 5-year history of FD and known stable coronary artery disease presented with NYHA class III heart failure (HF), peripheral edema, bilateral pleural effusions, and elevated plasma NT-proBNP level (11,004 ng/L) despite optimal pharmacologic therapy. Over 5 years, there had been a progression from severe concentric left ventricular (LV) hypertrophy and mild aortic stenosis (AS) with preserved ejection fraction (EF) to congestive HF with impaired LVEF (25%-30%), end-stage renal failure requiring renal transplant in 2013, and severe AS requiring transapical aortic valve implantation in 2014.

In 2011, a dual-chamber pacemaker was implanted owing to sinus node disease and intermittent complete atrioventricular block with subsequent upgrade to a cardiac resynchronization therapy pacemaker device because of a high degree of right ventricular pacing, left bundle branch block during intrinsic conduction, reduced LVEF, and NYHA class II HF symptoms. Cavotricuspid isthmus ablation was performed for typical right atrial (RA) flutter burden and amiodarone therapy was started for management of paroxysmal atrial arrhythmia.

In 2015, the patient presented with incessant monomorphic AT of 270 ms stable tachycardia cycle length (TCL) (Figure 1A) with an associated deterioration in NYHA class despite confirmed biventricular pacing (99%) at mode switch rate of 60 beats/min. Clear isoelectric intervals between successive P waves were registered simultaneously on all 12 leads of surface electrocardiogram, with P-wave morphology suggesting a focal RA origin.[1,2]

Under general anesthesia, a decapolar catheter was advanced into the coronary sinus (CS) and a CARTO 3 (Biosense Webster Inc) RA activation map performed with an open irrigated contact force radiofrequency ablation catheter (Thermocool SmartTouch; Biosense Webster Inc, South Diamond Bar, CA). RA activation mapping and entrainment from multiple sites as well as distal to proximal CS activation were consistent with a left atrial tachycardia. A normal (>0.5 mV) RA bipolar voltage map was registered (Figure 2). Following double transseptal access via a patent foramen ovale, a LA activation map revealed a macroreentrant circuit consistent with clockwise perimitral flutter (PMFl) and confirmed by entrainment from the anterior and posterior aspects of the mitral valve annulus (MA) (Figure 3). Low-amplitude, long-duration systolic fractionated electrograms (FEGMs) were observed throughout the left atrial anterior wall and septum, within a low (<0.5 mV) bipolar voltage zone. A unique diastolic FEGM accounting for 33% of the TCL was recorded halfway between anterior MA and right upper pulmonary vein (RUPV), exclusively spanning the electrocardiogram isoelectric interval (Figure 2). An anterior mitral line was performed between the MA and electrically silent RUPV to transect the low-voltage zone and incorporate the diastolic FEGM (Figure 3). Significant TCL prolongation at the ventricular aspect of the line was seen (from 270 ms to 330 ms) with subsequent termination during ablation at the area of the diastolic FEGM registration (Figure 1B). Conduction block was demonstrated by pacing lateral and septal to the ablation line with reversal of the CS activation sequence[3] from distal–proximal to proximal–distal, respectively, and modification of the stimulus to RA P wave time (Figure 1C). Bidirectional block was confirmed by differential pacing maneuvers. Burst atrial pacing failed to induce tachycardia following a 30-minute waiting period. Bidirectional block of the cavotricuspid isthmus was confirmed.

At 3 months follow-up, significant improvement in symptoms of HF and NYHA class were observed without arrhythmia recurrence.

KEYWORDS Atrial tachycardia; Perimitral flutter; Fabry's disease; Fractionated electrograms; Catheterablation

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KEY TEACHING POINTS

- Atypical atrial tachycardia may have a significant impact on the clinical presentation of Fabry's disease and the restoration of sinus rhythm may be beneficial.
- Identification of long-duration fractionated electrograms among widespread low-voltage atrial electrograms may help to identify the critical isthmus for maintenance of tachycardia.
- Interventional treatment of atrial arrhythmia can be safely and successfully performed despite advanced structural heart disease.

Discussion

Cardiac involvement in FD is common and results from the accumulation of glycoproteins within the myocytes, vascular endothelium, and valvular and conduction tissue. Although 60% of patients have some cardiovascular manifestation, of which the most common is arrhythmia, the burden of complications in this patient is unusual.[4] To the best of our knowledge, there are no data on the background and treatment of atypical AT in a patient with FD.[5]

The presented data support the hypothesis that PMFl was dependent on a relatively wide isthmus in the anterior LA, bounded by the anterior MA and an area of a scar anterior to the RUPV ostium. Ablation within this isthmus targeting multiple systolic FEGMs resulted in progressive lengthening of the TCL and arrhythmia termination during ablation at the diastolic FEGM site. Although an entrainment maneuver was not performed to avoid inadvertent arrhythmia termination, this suggests that PMFl was using that site as the critical narrow isthmus. We believe that all registered FEGMs coincided with the slow conduction zones but the critical slowing of the tachycardia wavefront was marked by multicomponent diastolic FEGM. Slow-conduction diastolic isthmuses have been reported to be critical for maintaining the reentry circuit and therefore represent a target site for ablation.[2,6] Ablation transecting the FEGMs zone may help to prevent other atypical ATs using these zones of slow conduction.

AT is well described in patients with a history of structural heart disease; however, occurrences of those arrhythmias in healthy individuals with low-voltage areas (LVAs) have also been described. Moreover, a correlation between low-voltage areas in the LA anterior wall and the LA–aorta contact area on magnetic resonance imaging scans has been found.[7] The anterior LA wall near the vestibule of the MA and just posterior to the aorta is exceptionally thin.[8] Where the anterior and septal LA wall are in close proximity to the aortic root, the LA myocardium may be potentially affected by an inflammatory response of the valve apparatus in the course of AS. In turn, this may explain the observed predisposition of the anteroseptal LA to scarring, which is not usually seen elsewhere within the atria and may have occurred in the presented case, where the critical zone of slow conduction in the anterior LA was located behind the transapical aortic valve implantation valve. On the other hand, the structural atrial remodeling and fibrosis caused directly by glycoprotein accumulation or indirectly by LA

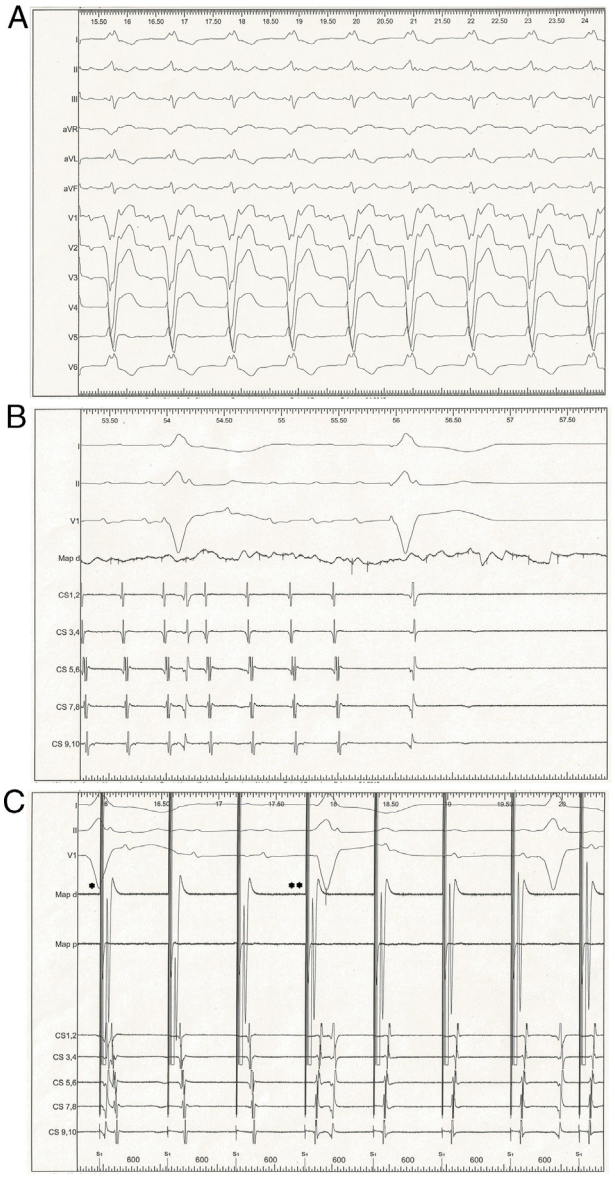

Figure 1 **A:** Monomorphic atrial tachycardia at 270 ms stable cycle length. Isoelectric intervals between successive P waves and biphasic P-wave morphology (positive, then negative) in lead V_1 suggesting focal right atrial arrhythmia origin. 25 mm/s paper sweep speed. **B:** Arrhythmia termination during radiofrequency application. I, II, V_1: standard electrocardiogram leads; CS 1,2→9,10: distal-to-proximal pairs of electrodes of the decapolar coronary sinus catheter; Map p: the proximal pair of electrodes of the mapping catheter; Map d: the distal pair of electrodes of the mapping catheter. 50 mm/s paper sweep speed. **C:** Demonstration of mitral line conduction block by pacing lateral (*) and septal (**) to the ablation line with reversal of the CS activation sequence from distal–proximal to proximal–distal, respectively, and modification of the stimulus to right atrial P wave time. 50 mm/s paper sweep speed.

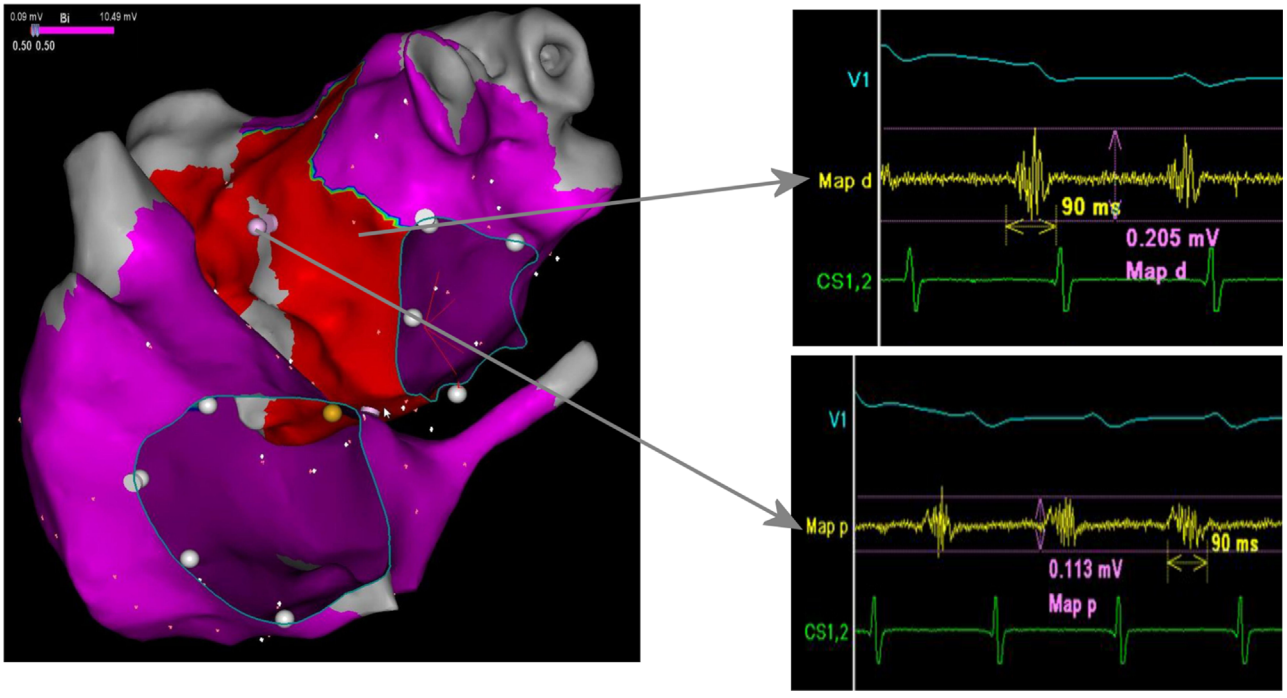

Figure 2 CARTO 3 right and left atrial (LA) bipolar voltage maps in left anterior oblique projection. Normal (>0.5 mV, coded in pink) right atrial while low (<0.5 mV, coded in red) LA voltage zone throughout anterior wall and septum registered. Low-amplitude, long-duration systolic fractionated electrograms (FEGMs) observed within low LA voltage zone. A unique diastolic FEGM (*pink dot*) accounting for 33% of the tachycardia cycle length recorded halfway between anterior mitral annulus and right upper pulmonary vein, exclusively spanning electrocardiogram isoelectric interval.

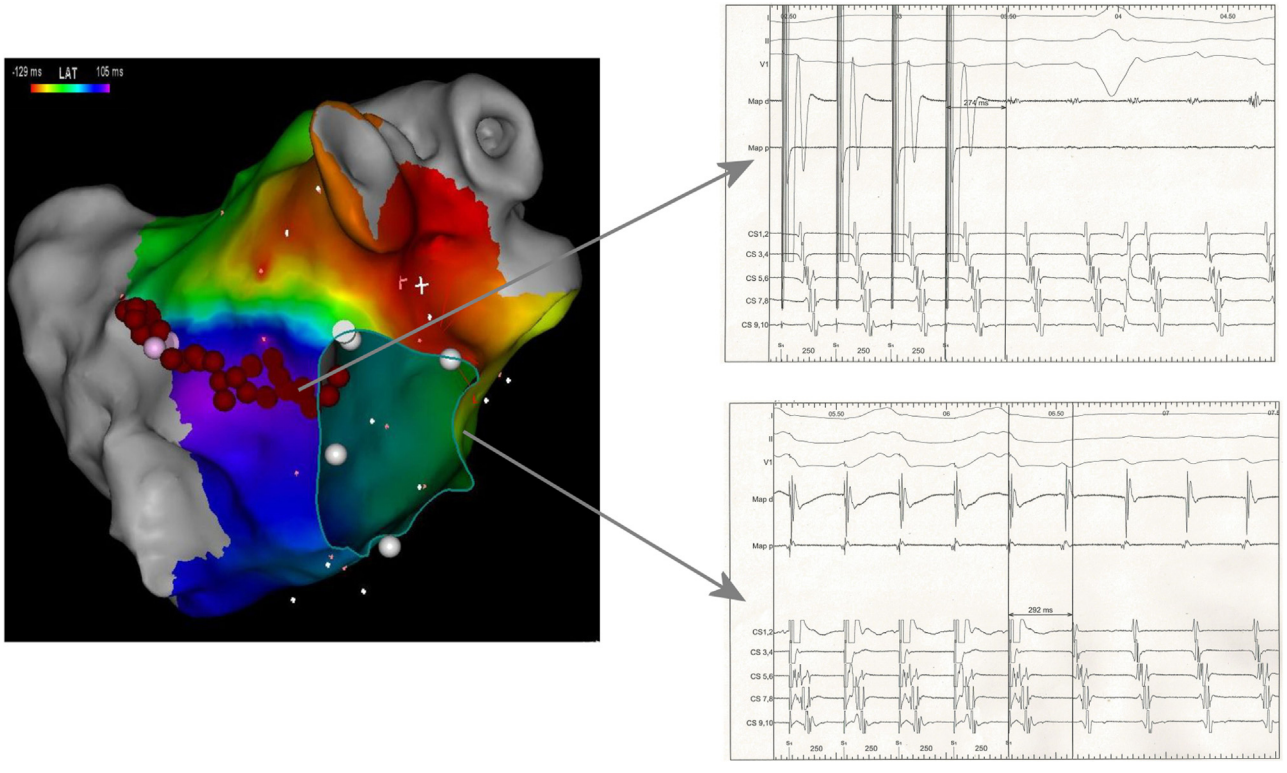

Figure 3 CARTO 3 left atrial activation map in left anterior oblique projection. Activation around mitral valve annulus (MA) covering 90% of the tachycardia cycle length consistent with clockwise perimitral flutter confirmed by entrainment from its anterior and posterior aspect (*white panels*). Ablation line between anterior MA and right upper pulmonary vein done (*red dotted line*) transecting the area of the diastolic fractionated electrogram registration (*pink dot*) where radiofrequency application terminated the tachycardia.

pressure overload and dilatation in the course of AS is expected to be a highly diffuse, rather than a localized, process. Therefore, multifactorial predisposition to AT among FD patients is likely.

Conclusion

This paper demonstrates that left AT may have a significant impact on the clinical presentation of FD and that restoration of sinus rhythm may be beneficial. Identification of long-duration FEGMs among widespread low-voltage atrial electrograms may help to identify the critical isthmus for maintenance of tachycardia. Finally, the interventional treatment of atrial arrhythmia can be safely and successfully performed despite advanced structural heart disease.

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