

Adult Woman with Tachycardia Induced Cardiomyopathy

Conservative Treatment of Permanent Junctional Reciprocating Tachycardia

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Abstract

Permanent junctional reciprocating tachycardia (PJRT) is a rare form of atrioventricular reentrant tachycardia due to an accessory pathway characterized by slow and decremental retrograde conduction. The arrhythmia typically presents before adulthood with incessant tachycardia leading to cardiomyopathy and heart failure. We report the case of a woman with tachycardia induced cardiomyopathy due to PJRT detected at the age of 54 years. The patient refused to undergo catheter ablation and was successfully treated with a beta blocker.

Keywords: Permanent junctional reciprocating tachycardia; PJRT; antiarrhythmic therapy

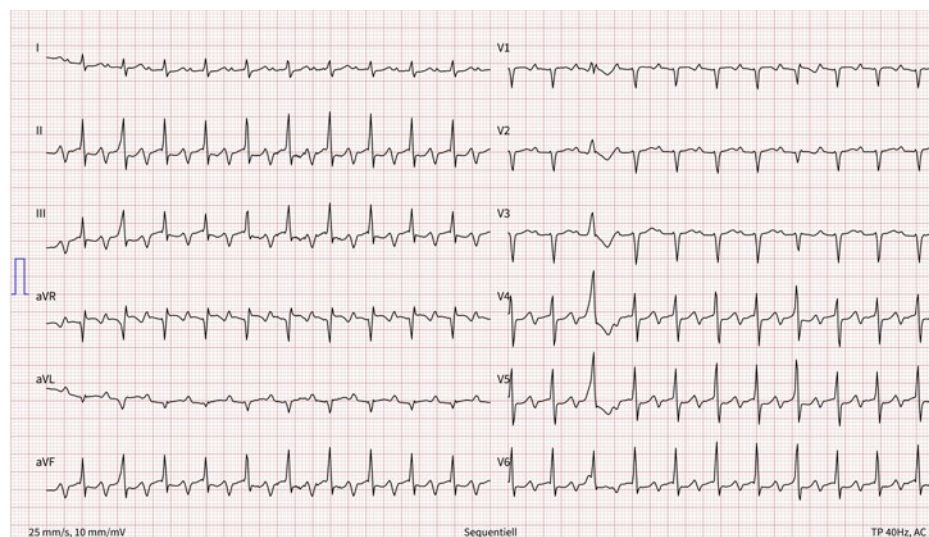


Figure 1: 12-lead ECG showing a narrow QRS complex tachycardia (HR 130 bpm) with long RP interval and negative P waves in II, III and aVF and positive P waves in V1.

Case Report

A 54-year-old woman was referred to our clinic because of regular, repetitive, fast palpitations occurring several times a day. After a few weeks, she developed marked fatigue and dyspnea.

The patient has had rare episodes of palpitation since her childhood never documented on an electrocardiogram (ECG).

The 12-lead ECG demonstrated a narrow QRS complex tachycardia with a long RP interval and prominent negative P waves in the inferior leads (fig. 1). The differential diagnosis included (1) typical atrial flutter, (2) low atrial tachycardia, (3) atypical fast/slow atrioventricular nodal reentrant tachycardia (AVNRT) and (4) orthodromic atrioventricular reentrant tachycardia (AVRT) using a slowly conducting accessory pathway known as permanent junctional reciprocating tachycardia (PJRT) [1]. Due to the P wave morphology like a sawtooth pattern, an initial diagnosis of atrial flutter was made, and the patient was started on anticoagulation, which was withheld after revision of the ECG diagnosis a few days later.

The transthoracic echocardiography revealed a normal-sized left ventricle with mildly impaired left ventricular systolic function (left ventricular ejection fraction [LVEF] 42%) and mildly dilated atrium (left atrial volume index 42 ml/m²).

Case Report

Holter ECG recording (fig. 3) demonstrated multiple episodes of tachycardia, consistently triggered by one or two premature ventricular contractions (PVC) (fig. 4) and ending with a P wave. These findings were compatible with an AV-reentrant tachycardia and almost certainly excluded a focal mechanism [1]. The history of palpitations since childhood and the incessant nature of the arrhythmia were both in favor of a PJRT [2].

Despite progressive palpitation and dyspnea due to systolic heart failure, the patient declined to undergo an electrophysiological study and catheter ablation. Therefore, she received antiarrhythmic and heart failure therapy including bisoprolol and ramipril. At six week follow-up, the patient reported a reduction of the frequency and duration of the palpitations and transthoracic echocardiography demonstrated a normalization of the systolic left ventricular function (LVEF 52%). The Holter ECG confirmed sinus rhythm without tachyarrhythmia. The rapid recovery of cardiac function achieved by effective rhythm control supported the diagnosis of tachycardia induced cardiomyopathy. Therefore, in accordance with the patient's explicit wish, no extension of the medical therapy nor additional imaging studies were performed.

Discussion

PJRT, also called Coumel tachycardia, is a rare form of an orthodromic AVRT using a concealed decremental accessory pathway for the retrograde activation of the atrium. The slow retrograde conduction properties of the accessory pathway facilitate the initiation and maintenance of the tachycardia. The arrhythmia, accounting for 1% of supraventricular tachycardias (SVTs) in children, typically presents at young age with incessant palpitation and heart failure symptoms (fatigue, exercise intolerance, dyspnea) due to tachycardia induced cardiomyopathy [2, 3, 7]. The paroxysmal form of PJRT appears to be more frequent in adults and less associated with heart failure. This feature may explain, why the diagnosis of PJRT may remain unrecognized until adulthood [5].

While normal during sinus rhythm, when in tachycardia, the ECG demonstrates a long RP interval with negative P waves in the inferior leads (fig. 3) [1]. This can be explained by the localization of the accessory pathway at the inferoposteroseptal aspect of the right atrium in close proximity to the coronary sinus (CS) ostium in about 80% of the patients [5].

The differential diagnosis includes other 1:1 SVTs with negative P waves in the inferior leads as well as focal atrial tachycardia originating in the region of the CS ostium or atypical (fast/slow) AV-nodal reentrant tachycardia.



Figure 2: Magnification of lead V1 and V5 during ongoing supraventricular tachycardia with slight variation of the tachycardia cycle length (450–460 ms). After a premature ventricular complex (asterisk) coming 70 ms before the anticipated QRS complex, a minimal prolongation of the PP interval to 470 ms can be observed. This may suggest the existence of an accessory pathway with decremental conduction properties. However, an atypical fast/slow AV nodal reentrant tachycardia (ANVRT) is not definitively excluded by this finding.

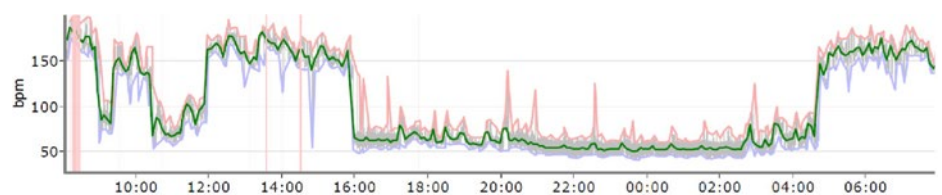


Figure 3: Holter ECG recording demonstrating multiple and long-lasting episodes of tachycardia with heart rates between 130 and 180 bpm.

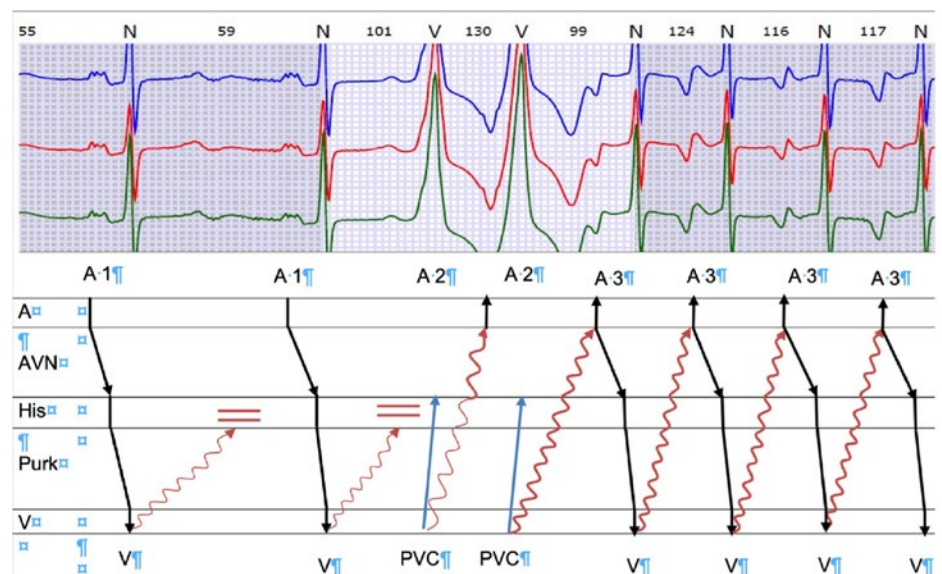


Figure 4: A critically timed premature ventricular contraction (PVC) finds the bundle branch-His-AV-nodal pathway refractory and therefore is conducted retrogradely to the atrium only over the accessory pathway followed by a negative P wave hidden in the T wave of the second PVC. After the second PVC, the retrograde activation of the atrium via the slow decremental accessory pathway occurs even later, finding an excitable AV-node-His-Purkinje system leading to anterograde activation of the ventricle. Perpetuation of this mechanism creates a stable reentrant circuit and results in a narrow complex tachycardia.

Typical atrial flutter with 2:1 AV-conduction may present with a similar ECG when every second negative sawtooth flutter wave is hidden within the QRS complex. Intravenous adenosine bolus can be helpful to clarify the diagnosis and terminate the tachycardia.

In our case, the repetitive induction of the tachycardia by one or two PVC and the constant termination with a retrograde P wave favored an AV-reentrant tachycardia.

Differentiating the PJRT from an atypical AVNRT is difficult. The mild prolongation of the PP interval after a premature ventricular complex might be explained by a prolonged retrograde conduction to the atrium via the accessory pathway with decremental conduction properties, or simply by unspecific changes in the tachycardia cycle length (fig. 2). Therefore, the ECG findings are not able to differentiate definitively between PJRT and atypical fast/slow AVNRT. However, the clinical presentation with repetitive occurrence of the tachycardia and development of systolic heart failure was highly suggestive for a PJRT [1–3].

Antiarrhythmic drugs can rarely suppress the arrhythmia. In general, beta blockers and digoxin are ineffective and calcium channel blockers are contraindicated in case of systolic heart failure. Class IC and III drugs may partially suppress the tachycardia. A trial with antiarrhythmic drugs can therefore be an option in selected patients with PJRT who are not able or willing to undergo an electrophysiological study and catheter ablation.

Conclusion

This case demonstrates the importance of the ECG in making the diagnosis of PJRT and how a reduction in arrhythmia burden with beta blockers in conjunction with heart failure therapy may lead to recovery of tachycardia induced cardiomyopathy. However, catheter ablation of the accessory pathway is highly effective and should be considered as the first choice treatment in adults with PJRT [5, 6].

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Ethics Statement

Written informed consent was obtained.

Conflict of Interest Statement

No financial support and no other potential conflict of interest was reported.

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