Repellent narrow QRS tachycardia in a 61-year-old female patient with recent palpitations

András Vereckei1,*, László Gellér2

13rd Department of Medicine, Semmelweis University, Budapest, Hungary
2Heart and Vascular Center, Semmelweis University, Budapest, Hungary

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A 61-year-old female patient suffering from recent onset palpitations and dyspnea on exertion with hypertension and mitral valve prolapse in her past history came to our outpatient department. Echocardiography revealed a mild mitral valve prolapse, slightly decreased left ventricular (LV) function (LV ejection fraction: 51%) and a mild mitral regurgitation. Initial 12-lead ECG showed normal sinus rhythm (NSR) with left anterior hemiblock, upper normal limit (105–110 ms) QRS duration and left atrial abnormality (not shown). 24 hour ambulatory ECG monitoring (Figure 1) revealed a great variety of narrow QRS complex arrhythmias at first glance looking like sinus beats alternating either with aberrantly conducted atrial premature beats with hidden low amplitude P waves (Figure 1 first three rhythm strips) or with ventricular premature beats (Figure 1 upper rhythm strip) arranged in a bigeminal pattern. The third rhythm strip of Figure 1 may also be considered a narrow QRS complex tachycardia with alternating shorter and longer cycle lengths (CL) and the fourth rhythm strip of Figure 1 is a completely regular narrow QRS complex tachycardia with alternating narrow QRS complexes of slightly different morphology. These non-sustained narrow QRS tachycardias occurred repetitively separated by 1 or 2 sinus beats. After the 24 hour ECG monitoring another 12-lead ECG was recorded (Figure 2), which showed short, repetitive narrow QRS complex supraventricular tachycardia (SVT) runs separated by two sinus beats. The QRS morphology was identical to that of the initial 12-lead ECG.

After a closer look at the ECG demonstrated in Figure 2, the most striking feature of the repetitive narrow QRS complex arrhythmias is that there are distinct, regular sinus P waves throughout the whole ECG tracing with a normal sinus rate of 78–86 beats/min and the number of P waves are less than that of the QRS complexes. In fact, each sinus P wave is followed by two QRS complexes during the repetitive SVT runs. A hidden P wave in between the two visible sinus P waves is unlikely, because the T wave morphology of the first QRS complexes following each sinus P wave is identical to that of the sinus beats separating the repetitive SVT runs. Another important feature of the short, repetitive SVT runs, that a gradual prolongation of the PR intervals preceding the pauses followed by two sinus beats occurs, related to the gradual shortening of the RP intervals, thus an inverse RP-PR interval relationship is present, consistent with an atrioventricular (AV) Wenckebach periodicity.

The most likely explanation for the phenomenon, that each sinus P wave is followed by two QRS complexes during the repetitive SVT runs, is the presence of dual AV nodal pathways and 1:2 AV conduction, i.e., a single sinus beat elicits a double ventricular response due to double anterograde conduction via the fast and slow AV nodal pathways (also termed “double firing”) (Figure 3A). Actually, the baseline rhythm during the repetitive paroxysmal SVTs is NSR, which results in a nonreentrant AV nodal tachycardia with a high (double) ventricular rate due to 1:2 AV conduction called dual AV nodal nonreentrant tachycardia (DAVNNT). The ladder diagram (Figure 3A) also reveals that in addition to the visible progressive PR (PR1) interval prolongation reflecting fast pathway conduction, a gradual prolongation of the slow pathway PR interval (PR2) also occurs during the repetitive SVT runs, and the pause terminating the SVT is the consequence of blocking of the sinus impulse in both AV nodal pathways as a result of simultaneous Wenckebach sequences. This is due to the slightly shortened sinus CLs preceding the pause, which in itself and by prolonging slightly the effective refractory periods (ERP) of both AV nodal pathways, results in an encroachment of the sinus impulse on these ERPs. After the
Figure 1. Rhythm strips recorded during 24-h ECG monitoring.
anterograde block of the sinus impulse in both AV nodal pathways the next sinus beat is conducted solely through the fast pathway, its lack of conduction through the slow pathway to the ventricle might have two alternative explanations. First, this sinus impulse may be conducted through the slow pathway to the infra-His conduction system and blocked there or in the ventricles, because the conduction of the sinus impulse over the fast pathway right after the pause produces a long R-R CL, this preceding long R-R CL prolongs significantly the infra-His (ventricular) ERPs and therefore an infra-His block of the sinus impulse conducted through the slow pathway occurs. Alternatively, the unidirectional retrograde block in the slow pathway, which is a prerequisite of 1:2 AV conduction and occurs during normal sinus CL, is relieved temporarily during the long CL of the pause, and during the first beat following the pause a retrograde penetration of the sinus impulse conducted via the fast pathway to the slow pathway occurs, thereby blocking anterograde conduction in the slow pathway. While the first sinus P wave after the pause was conducted via the fast pathway but blocked at some level during slow pathway conduction to the ventricle the electrophysiological study did not reveal a detectable His potential during slow pathway conduction block, which ruled out the first explanation (infra-His block) and indirectly supported the second explanation (Figure 4).

DAVNNT should be considered: (1) in the case of irregular paroxysmal SVT occurring in a patient showing sudden significant changes in the PR interval during NSR; (2) in the case of a regularly irregular SVT associated with sustained alternating R-R CLs; and (3) when during the SVT the number of P waves is half of that of the QRS complexes. The single exception to sustained CL alternans tachycardia during DAVNNT is the rare occasion, when the difference in the conduction times through the slow and fast pathways (the R-R CL) is exactly half of the sinus CL. In this case a completely regular SVT can occur, which was also occasionally present in our patient (Figure 1 4th rhythm strip, Figure 3B). The rhythm strips of Figure 1 demonstrate arrhythmias of alternating shorter and longer CLs due to 1:2 AV conduction with every 2nd QRS after the sinus P wave showing different degrees of aberrant conduction. AV nodal reentrant tachycardia (AVNRT) is the most common clinical manifestation of dual AV nodal pathway, abrupt changes in PR intervals during NSR are less common and the least common manifestation is DAVNNT with “double firing”, because certain special conditions should be present simultaneously for DAVNNT to occur. The following special conditions are required for the maintenance of DAVNNT: (1) Markedly delayed conduction through the slow pathway; (2) the ERPs of the AV node distal common pathway and the His-Purkinje system must be shorter than the difference between the conduction times of the fast and slow pathways; (3) a sufficiently long sinus CL to allow the slow conduction
Figure 3. The elucidation of the possible mechanisms of arrhythmias shown in Figure 2 and Figure 1 4th rhythm strip. (A): Ladder diagram elucidating the mechanism of the arrhythmia shown in Figure 2. Stars denote sinus P waves. Conduction through the fast pathway is denoted with continuous and through the slow pathway with dashed lines. Black rectangles demonstrate infra-His (ventricular) ERP, blank rectangles bordered with continuous line represent fast pathway, blank rectangles bordered with dashed line denote slow pathway ERPs. RP from SP QRS = RP interval calculated from the QRS complex resulting from conduction of the sinus impulse through the slow pathway, RP from FP QRS = RP interval calculated from the QRS complex resulting from conduction of the sinus impulse through the fast pathway. (B) Rhythm strips recorded during 24-h ECG monitoring. The lower rhythm strip is identical to the 4th rhythm strip shown in Figure 1. The upper rhythm strip shows the initiation of the regular SVT shown in the lower rhythm strip. For further explanation see text. A: atrium; AV: atrio-ventricular node; CL: cycle lengths; ERP: effective refractory period; FP: fast pathway; PR1: progressive PR; PR2: pathway PR interval; SP: slow pathway; SVT: supraventricular tachycardia; V: ventricle.

of the impulse through the slow pathway to the His-Purkinje system; and (4) the retrograde ventriculoatrial conduction should be absent or poor. In contrast to that, retrograde conduction through either the slow or the fast pathway or both should be present for AVNRT to occur.[1–3]

The differential diagnosis of DAVNNT with sustained 1:2 conduction resulting in alternating CLs includes junctional premature beats arranged in a bigeminal pattern and junctional parasystole. Several features of our patient’s arrhythmia argue against sporadic junctional premature beats:
Figure 4. Intracardiac electrogram demonstrating the end of a short, repetitive DAVNNT run with sustained alternating CLs with every 2nd QRS complex showing slightly aberrant conduction. (A): The 1st sinus impulse shows 1:2 conduction, the 2nd sinus impulse is blocked without a following His potential, indicating AV nodal block in both AV nodal pathways, resulting in a pause. The first sinus impulse after the pause (3rd sinus impulse in Panel A and 2nd sinus impulse in Panel B) is conducted via the fast pathway, but blocks without a following His potential (Panel B) in the slow pathway. AV: atrioventricular node; CLs: cycle lengths; DAVNNT: dual AV nodal nonreentrant tachycardia.

(1) the coupling intervals (CI) of the 2nd QRS complexes to the 1st QRS complexes after the P waves during 1:2 conduction were the same within a certain time period, but varied significantly at different time points during the 24 hour ambulatory ECG monitoring (sporadic junctional premature beats are rather associated with changing CIs at the same time period); (2) the lack of retrograde P waves; and (3) AV Wenckebach periodicity. Junctional parasystole is also unlikely, because it has a stable CL, however the CL between the 2nd QRS complexes after the sinus P waves changed significantly over time during the 24 hour ECG monitoring.

DAVNNT is a very rare arrhythmia (approximately 70 cases were reported so far), which is recognized with in-
creasing frequency. Palpitations are the most common, dyspnea, syncope, dizziness, fatigue and chest pain are less common symptoms. Usually there is a significant delay until the correct diagnosis is established, and DAVNNT is frequently misdiagnosed as nonspecific SVT, atrial fibrillation, 1st or 2nd degree AV block, ventricular tachycardia (VT) (in the case of aberrant conduction) and consequently patients are referred to unnecessary procedures or treatments, such as pulmonary vein isolation, transesophageal echocardiography, electrical cardioversion, anticoagulation, pacemaker or implantable cardioverter defibrillator (ICD) implantation. A major complication of DAVNNT may be tachycardiomyopathy, a slight form of which was present in our patient. DAVNNT might also result in inappropriate ICD shocks due to the presence of more QRS complexes than P waves, as current ICD algorithms might be unable to discriminate it from VT. The medical treatment of DAVNNT is usually ineffective. The definitive treatment is slow pathway ablation, which eliminates DAVNNT. The success rate of slow pathway ablation/modulation for DAVNNT treatment seems to be comparable but slightly lower than for AVNRT (> 90%). The electrophysiological study confirmed the diagnosis of DAVNNT, slow pathway ablation was performed in our patient in all possible locations positioning the ablation catheter gradually from low in the triangle of Koch to more superior areas, including areas, that were attempted from the aortic root, which either could not terminate the arrhythmia or terminated it only temporarily. At the final ablation persistent 3rd degree AV block appeared with wide QRS complex escape rhythm, therefore a DDDR pacemaker was implanted. Though the final outcome of slow pathway ablation was less than optimal, ultimately the arrhythmia and palpitations were eliminated and the patient is in a good condition and symptom-free.

References

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