A case of incessant bundle branch reentry tachycardia occurring after phase 3 right bundle branch block as a first manifestation of left ventricular noncompaction cardiomyopathy in a patient with bicuspid aortic valve

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Introduction

Bundle branch reentrant tachycardia (BBRT) is a form of macroreentrant tachycardia encompassing both the left and right bundle branches. It usually occurs in the context of dilated cardiomyopathy, previous valve surgery, or other cardiac conditions that affect the His-Purkinje system. The baseline electrocardiogram (ECG) in sinus rhythm generally shows a left bundle branch block (LBBB) or right bundle branch block (RBBB). This case is a very unusual presentation of BBRT in a patient with subclinical left ventricular noncompaction and bicuspid aortic valve. Despite a normal resting ECG, the intracardiac electrograms showed a significant infrahisian conduction delay. The BBRT initiated after development of phase 3 RBBB as a result of fast antegrade atrioventricular (AV) conduction, possibly over an AV nodal slow pathway or owing to atrioventricular nodal reentry tachycardia (AVNRT).

KEY TEACHING POINTS

- Bundle branch reentry tachycardia could present as a first manifestation of subclinical left ventricular noncompaction.
- Bundle branch reentry tachycardia can develop after phase 3 bundle branch block with normal resting electrocardiogram (ECG).
- Some kind of infrahisian conduction disease is always present in patients with bundle branch reentry tachycardia, even with apparently normal ECG. These patients should undergo further investigations to look for the presence of an underlying structural heart disease.

Case report

A 49-year-old man presented to the emergency room with chest pain and palpitations. He described a known congenital valvular aortic stenosis but had no records of it. No family history of sudden cardiac death or unexplained syncope was known. The clinical examination showed a distressed sweating patient with a peripheral radial pulse of 240 beats/min, a rapidly deteriorating systolic blood pressure of 80 mm Hg, and a mild systolic murmur in the left second intercostal space and was otherwise unremarkable.

The ECG showed a broad complex tachycardia with RBBB morphology, right superior QRS (north-west) axis, and negative QRS complexes in the precordial leads V_2 to V_6 (Figure 1a).

At first an aberrantly conducted supraventricular tachycardia was suspected and adenosine was administered several times without affecting the tachycardia. Because of the threatening cardiogenic shock, several attempts to convert the rhythm electrically were then made. However, every time the tachycardia reinitiated very rapidly after several sinus beats, with the ECG showing severely depressed ST segments in almost all leads as well as ST-segment elevations in leads V_1 and aVR, possibly suggesting a subendocardial ischemia (Figure 1b). Finally amiodarone was administered and successfully suppressed the tachycardia. In total, 600 mg of amiodarone was given intravenously and was followed by 2 separate doses of 200 mg each orally. In subsequent ECGs the ST-segment changes returned to normal (Figure 1c). Laboratory findings showed a markedly elevated troponin level and were otherwise irrelevant. Echocardiographic imaging showed a normal ejection fraction and a bicuspid aortic valve with mild-to-moderate stenosis and regurgitation, as well as an aneurysm of the ascending aorta of 47 mm. Because of the acute ST-segment changes, a coronary angiogram was obtained and showed no coronary heart disease.

A diagnostic electrophysiology study was performed after a 24-hour amiodarone-free interval. At baseline, the patient

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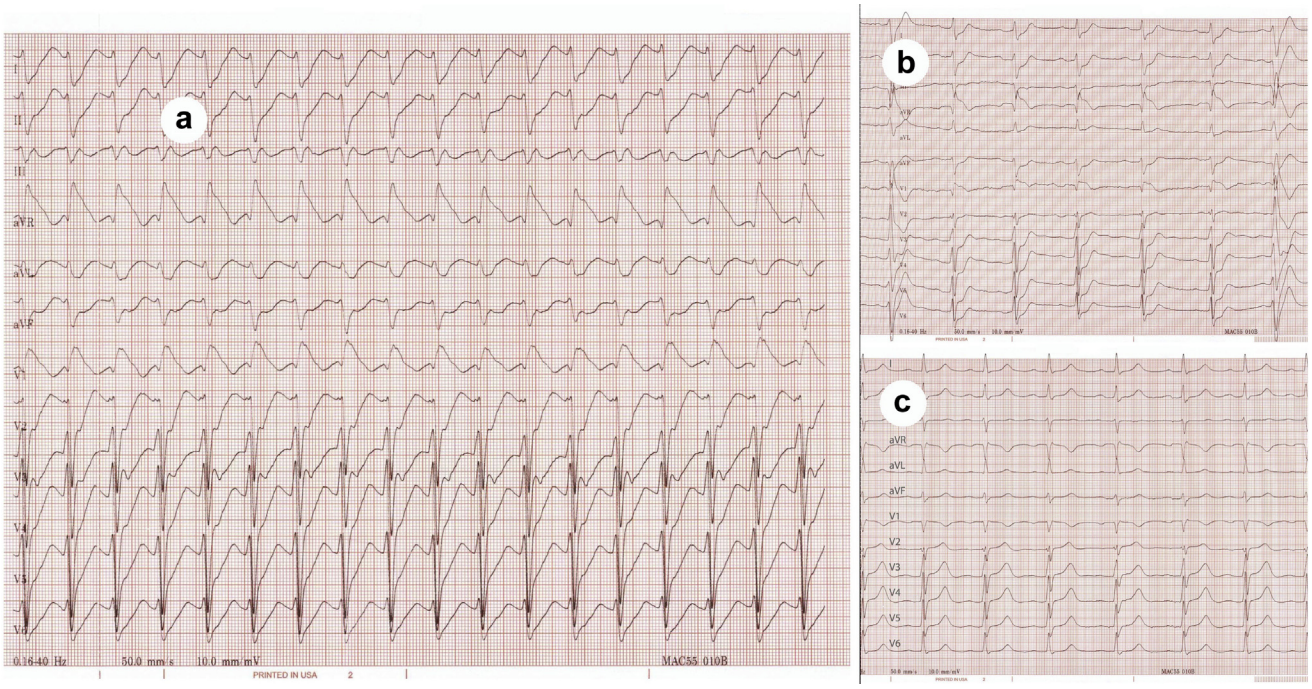

Figure 1 Manifestation electrocardiograms (ECGs). **a:** The 12-lead surface ECG of clinical tachycardia showing broad complex tachycardia with right bundle branch block morphology, right superior axis, and negative precordial QRS complexes in leads V_2 to V_6. **b:** The 12-lead surface ECG of the short-lived sinus rhythm between the cardioversion attempts showing a sinus rhythm with severely depressed ST segments in almost all leads as well as an ST elevation in V_1 and aVR. **c:** Normalization of the 12-lead surface ECG after successful suppression of the tachycardia under amiodarone.

was in sinus rhythm with a PR interval of 192 ms and no manifest preexcitation. The resting HV interval was significantly prolonged (77 ms) in the absence of a bundle branch block (Figure 2a). Retrograde ventriculoatrial conduction during ventricular pacing was not present. Atrial pacing with the tachycardia cycle length revealed a suprahisian as well as a pronounced infrahisian Wenckebach conduction pattern (Figure 2b). Programmed atrial stimulation produced an antegrade jump with repetitive typical AV nodal echo beats leading to a marked widening of the QRS complex in the form of a complete phase 3 RBBB (Figure 2c). The fact that those beats occurred right after an AV nodal jump and showed simultaneous atrial and ventricular activations distinguishes them from possible bundle branch reentrant beats. Also, no tachycardia was inducible at baseline. After repeated administration of orciprenaline and atropine, a stable AV 1:1 conduction in the tachycardia cycle length, possibly over the AV nodal slow pathway (SP), could be obtained. At the same time, the ECG showed a phase 3 RBBB, which was very similar to the clinical tachycardia (Figure 2d and e).

Eventually the tachycardia could be induced with fast atrial stimulation. An AV dissociation was present and the QRS complexes were always preceded by a His potential with an HV interval similar to that in sinus rhythm. Also V-V changes in the tachycardia cycle length were preceded by H-H changes (Figure 3a), which confirmed the diagnosis of a BBRT.[1] As a differential diagnosis it is also possible here to think about AVNRT with an upper common pathway block leading to AV dissociation. A possible way to differentiate between the two would have been entrainment from the right ventricular apex, which usually shows short postpacing intervals (<30 ms) in BBRT.[2] However, in our case no entrainment was possible owing to difficult tachycardia induction and its short duration. Nevertheless, the fact that the tachycardia was not affected by adenosine at the time of presentation makes an AVNRT extremely unlikely and strongly supports the diagnosis of BBRT.

The tachycardia was also not inducible with programmed right ventricular stimulation despite a very aggressive stimulation sequence. Only a very short run of 3 ventricular beats with LBBB morphology could be recorded and similarly showed a His potential preceding every QRS complex, albeit with a much longer HV interval (Figure 3b), suggesting a much more diseased right bundle branch than left bundle branch.

Since the HV interval was already significantly prolonged at baseline and because both bundle branches showed pronounced conduction abnormalities, no attempts at ablating the right or the left bundle branches were made owing to concerns of possible occurrence of a complete AV block with subsequent need for permanent RV stimulation. Nevertheless, owing to the fact that the BBRT was only inducible after the occurrence of phase 3 RBBB in high pacing frequencies from the atria, as well as that a slow pathway conduction seemed to be a prerequisite for a sustained AV conduction in those frequencies with the possibility of an AVNRT being a trigger for inducing the clinical tachycardia, we decided to perform a slow pathway modulation, after which the tachycardia became noninducible. However, because of

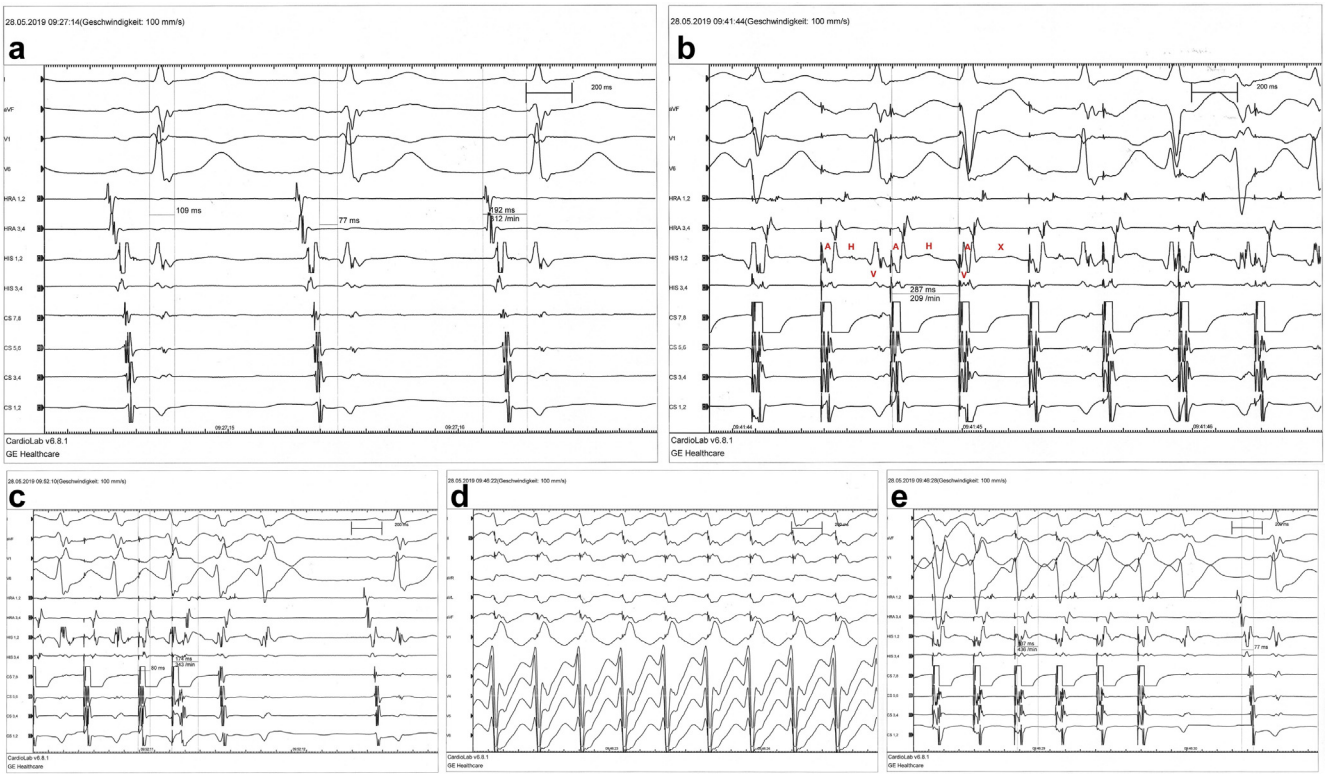

Figure 2 Electrophysiologic study. **a:** Normal PR and prolonged HV intervals in the absence of a bundle branch block. **b:** Atrial pacing with the tachycardia cycle length (TCL) revealed a suprahisian as well as a pronounced infrahisian Wenckebach conduction pattern. A = atrial activation; H = His bundle activation; V = ventricular activation; X = no atrioventricular conduction. **c:** Programmed atrial stimulation producing an antegrade jump with typical atrioventricular (AV) nodal echo beat showing simultaneous atrial and ventricular activations. A marked widening of the QRS complexes in the form of a complete right bundle branch block is also present. **d:** Under intravenous orciprenaline and atropine a stable AV 1:1 conduction in tachycardia cycle length could be obtained with the electrocardiogram showing a phase 3 right bundle branch block with a morphology very similar to the clinical tachycardia (owing to recording artifacts lead V_2 had to be removed from the tracing). **e:** Comparison of the atriohisian intervals during TCL atrial stimulation and sinus rhythm showing a possible slow pathway conduction during stimulation. CS = coronary sinus; His = His bundle; HRA = high right atrium.

the electrophysiologic findings with the above-mentioned conduction disturbances we decided to perform a cardiac magnetic resonance study to look for an underlying structural heart disease. This subsequently revealed a noncompacted left ventricle according to the criteria of Petersen and colleagues[3] (Figure 3c and d) and a preserved ejection fraction. After several discussions with the patient and because of the possibility of future disease progression with the risk of developing a spontaneous complete AV block and/or other ventricular tachycardias, a decision was made to implant a VDD-ICD system. A medical therapy with a beta blocker and an angiotensin-renin inhibitor was also initiated. No new arrhythmias have occurred since then. However, owing to progressing aortic stenosis and ascending aortic aneurysm a surgical repair was successfully undertaken several months after the initial presentation.

Discussion

This is an unusual case showing a rare form of BBRT with RBBB morphology induced after occurrence of a phase 3 bundle branch block with otherwise normal baseline ECG findings. Owing to the tachycardia induction with atrial stimulation with a stable AV slow pathway conduction and possibly with AVNRT, an SP modulation was performed. Subsequent cardiac magnetic resonance study revealed a subclinical noncompaction cardiomyopathy. A similar finding was reported by Barraa and colleagues.[4] In their case, however, there was a typical baseline LBBB with much longer HV interval. Our case could represent an early stage of conduction system disease in the course of left ventricular noncompaction. It also appeared in a patient with a bicuspid aortic valve. However, controversial results concerning the correlation between left ventricular noncompaction and bicuspid aortic valves were reported in previous publications.[5,6] Further investigations in larger patient populations might be warranted to determine a possible association between the two.

Limitations

The fact that the tachycardia was difficult to induce during the procedure may pose a limitation to our case, since noninducibility after SP modulation may not reliably confirm the elimination of the tachycardia if it was not easily induced at baseline. However, subsequent follow-up of the patient for more than 6 months up to the writing of this publication showed no recurrences of the tachycardia, which supports the claim that a

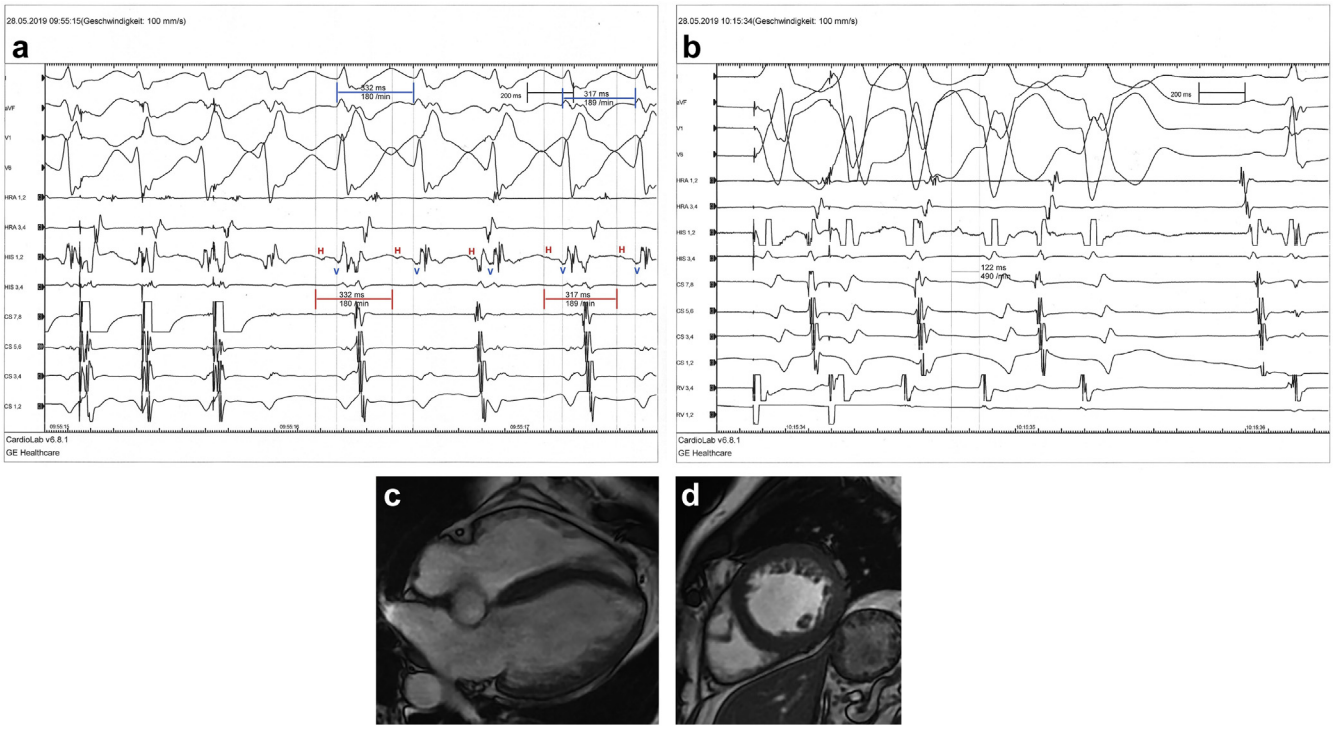

Figure 3 Diagnostic findings. **a:** The tachycardia was induced with atrial stimulation. An atrioventricular dissociation was present and the QRS complexes were always preceded by a His potential with an HV interval similar to that in sinus rhythm. Also V-V changes in the tachycardia cycle length were preceded by H-H changes. **b:** An induced short run of 3 ventricular beats with left bundle branch block morphology showed a His potential preceding every QRS complex with much longer HV interval. **c,d:** Cardiac magnetic resonance images of the patient showing a noncompacted left ventricle. CS = coronary sinus; His = His bundle; HRA = high right atrium; RV = right ventricle.

1:1 AV conduction over an SP was necessary for initiating the BBRT, since it provided the needed functional phase 3 RBBB.

Conclusion

BBRT can appear in seemingly normal individuals. However, it still indicates the presence of an underlying structural heart disease affecting the conduction properties of the His-Purkinje system and should warrant a thorough cardiac investigation.

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