Unusual Incessant Ventricular Tachycardia
What Is the Underlying Cause and the Possible Mechanism?

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Ventricular tachycardia (VT) is defined as a tachycardia with a rate of ≥100 per minute, with ≥3 consecutive beats that originates from the ventricles, and is independent of atria or atrioventricular nodal conduction. Of all the VTs, 10% occurs in those with structurally normal heart and these are called as idiopathic VTs, and the rest 90% occurs in patients with structural heart disease. VTs may be classified based on the clinical characteristic (clinical VT, hemodynamically stable or unstable, repetitive, incessant, etc.), morphology (monomorphic, multiple morphologies, pleomorphic, polymorphic, bidirectional, etc.) or mechanisms (scar-related reentry, automaticity, and triggered activity). Incessant VT is defined as continuous sustained VT during several hours, which recurs promptly despite repeated intervention for termination. VT storm is considered if ≥3 separate episodes of sustained VT within 24 hours, each requiring termination by an intervention. Monomorphic VT has a uniformly similar QRS configuration from beat-to-beat, with either right or left bundle configuration depending on whether the dominant deflection in lead V1 is R or S wave. Multiple monomorphic VTs refer to ≥1 morphologically distinct monomorphic VT, occurring as different episodes, or induced at different times. Polymorphic VT has continuously changing QRS configuration from beat-to-beat indicating a changing ventricular activation sequence. Pleomorphic VT has ≥1 morphologically distinct QRS complex occurring during the same episode of VT, but the QRS is not continuously changing. Bidirectional VT (BVT) is characterized by beat-to-beat alternation of the QRS axis on the ECG and is usually regular.1

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Here, we describe unusual incessant VT in a young male and discuss evaluation, management, and the possible mechanism.

Case Report

A 33-year-old male, non-diabetic, non-hypertensive, presented with history of palpitation, chest discomfort, and fatigue of 6 hours duration. He had been otherwise healthy, not on any medications with no history of tobacco, alcohol, or drug use. At presentation, pulse rate was 144 beats per minute, irregular, blood pressure 96/60 mm Hg, and the rest of physical examination was unremarkable. Twelve-lead ECG revealed a wide complex regularly irregular tachycardia at a rate of 144 beats per minute, right bundle morphology, QRS duration of 200 to 220 ms, QRS axis alternating between +160° and −40°, QRS transition alternating from V3 to V6, and evidence of atrioventricular dissociation (Figure 1). Tachycardia was unresponsive to intravenous lignocaine, esmolol, amiodarone (300 mg during 1 hour), and 2 attempts of biphasic direct current (DC) cardioversion of 200 J. Because the tachycardia was incessant, patient was sedated, intubated, ventilated, and continued on intravenous amiodarone (900 mg/24 hours) with further 4 unsuccessful attempts at DC cardioversion. Tachycardia converted to monomorphic VT with occasional VT complexes of the 2nd morphology (Figure 2), which terminated 6 hours later while on infusion of amiodarone and lignocaine, and while he was being considered for radiofrequency catheter ablation. ECG in sinus rhythm was essentially normal (Figure 3), chest x-ray revealed superior mediastinal widening (Figure 4A), and echocardiogram showed normal left ventricular function. Cardiac MRI was performed, which revealed focal areas of thickening and increased signal intensity on T2-weighted images. Early gadolinium enhancement was seen in the mid interventricular septum, anterolateral and anteroinferior walls, with the lesions having a target appearance with peripheral slightly hyperintense rim and central hypointensity (Figure 4B). Delayed gadolinium enhancement was found predominantly in the midmyocardium and epicardial in basal and lateral segments of the left ventricle. 18F-fluoro-2-deoxy-D-glucose (FDG) positron emission tomography scan revealed basal and midanteroseptal perfusion defect with intense FDG uptake in these regions suggestive of myocardial inflammation along with evidence of inflammation in the paratracheal lymph nodes (Figure 4C). Serum biochemistry was normal including serum calcium, serum angiotensin-converting enzyme (ACE), digoxin, blood aconite levels (report received 2 weeks later), and IgG and IgM assays were negative for tuberculosis bacilli. Transbronchial lymph node biopsy revealed noncaseating granulomas suggestive of sarcoidosis (Figure 4D). Patient was initiated on 1 g of methylprednisolone (Solumedrol) per day for 3 days intravenously and amiodarone infusion of 1 g/d was continued for 3 days after which it was switched to oral therapy and patient was extubated after 36 hours. Later, oral prednisolone was initiated at 60 mg/d for 2 months and was tapered to a maintenance dose of 10 mg/d for 2 years and stopped after a normal repeat positron emission tomography FDG scan. A single chamber implantable defibrillator was implanted after 5 days of VT-free interval. Device-based testing was performed at a single drive train of 600 ms and ≤2 extrastimuli which did not induce the VT and a single shock at 14 J terminated induced VF.

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Amiodarone was tapered and stopped after 6 months. Genetic testing for possible catecholaminergic polymorphic ventricular tachycardia with analysis for 4 genes: RYR2, CASQ2, ANK2, and KCNJ2 were negative (report received after 2 months). Four years later, patient has not had any device activation and has normal left ventricular function.

Discussion

This case describes an unusual presentation of isolated cardiac sarcoidosis with incessant regularly irregular BVT, normal LV function, and no conduction system involvement as the initial manifestation. Clinical manifestations of cardiac sarcoidosis are dependent on the location, extent, and activity of the disease. The 3 principal sequelae of cardiac sarcoidosis are (1) conduction abnormalities (2) ventricular arrhythmias, and (3) heart failure.2,3 Cardiac sarcoidosis usually presents with monomorphic VT or conduction system disturbances. Macroreentry around areas of granulomatous scar is the commonest mechanism of VT in sarcoidosis.4

BVT is a rare ventricular tachyarrhythmia. It is usually regular, demonstrates a beat-to-beat alternation of the QRS frontal axis varying between −20° to −30° and +110°. Tachycardia rate is typically between 140 and 180 beats per minute and the QRS is relatively narrow with duration of 120 to 150 ms. There are many causes of BVT described in the literature5–10 (Table). Although the most characteristic ECG pattern of BVT is right bundle branch block with an alternating QRS axis,9 other patterns such as alternating right bundle branch block and left bundle branch block, or alternating QRS axis with a narrow QRS,11 have also been observed. Few arrhythmias have been subject to such different hypothesis about the mechanism and it has fascinated electrocardiographers for many decades.

Figure 1. Twelve-lead ECG showing bidirectional ventricular tachycardia. Two morphologies of QRS complexes, namely A and B, with A to A interval being 72 beats per minute and B to B interval of 72 beats per minute, making up the tachycardia rate of 144 beats per minute. The relationship of A to B interval is constant at 0.56 s and B to A interval is constant at 0.32 s.

Postulated mechanisms have included a single focus in the proximal His bundle or bundle branches with alternating left fascicular block, or single or double foci in the distal His purkinje system and also reentry.10,12,13 Recently, Ping-Pong mechanism or reciprocating bigeminy in the distal his purkinje system because of delayed after depolarization has been proposed as the mechanism for BVT. It was shown that when the heart rate exceeded the threshold for bigeminy at the first site in the His–Purkinje system, ventricular bigeminy developed, causing the heart rate to accelerate and exceed the threshold for bigeminy at the second site. Thus, the triggered beat from the first site induced a triggered beat from the second site and vice versa.14

BVT in present case has many unusual features. The QRS duration is broad at 200 to 220 ms, unlike cases of in most BVT where it is relatively narrow at 120 to 150 ms. Wide QRS in this case possibly indicates intramyocardial/epicardial site of origin, unlike the usual BVT where the origin is supposed to be closer to the His purkinje system. In an elegant experiment using arterially perfused canine wedge preparations with transmural ECG and action potentials recorded simultaneously from epicardial, M, and endocardial cells epicardial origin of BVT has been demonstrated.13

The most intriguing feature is the regularly irregular rhythm as seen in Figure 1. A careful analysis of relationship between the 2 morphologies of QRS complexes namely A and B reveals that the A–A interval is 72 beats per minute (0.88 s) and B–B interval is 72 beats per minute (0.88 s), resulting in tachycardia rate of 144 beats per minute. The relationship of A to B interval is constant at 0.56 s, and B to A interval is constant at 0.32 s, and this relationship is constant throughout the tachycardia resulting in a regularly irregular rhythm. The broad QRS, with axis alternating between +160° and −40°, transition alternating from V3 to V6 in chest leads, possibly
indicates 2 independent foci. The original hypothesis of dual ventricular foci firing alternatively that was suggested as the mechanism was discarded because of the constant R–R interval that would presuppose a perfect alteration of the 2 foci. However, in this case, regularly irregular rhythm can be explained by automatic firing of 2 different foci at constant intervals and presence of entrance block in each of the foci akin to a dual parasystole.

Parasystole refers to those instances of double rhythm of the atria, atrioventricular junction or ventricles in which 1 pacemaker is protected from the impulses of the other. Parasystole requires an area of focal impulse formation which is surrounded by an area that protects (shield) the focus from external impulses thus resulting in an entry block but no exit block of the focus. Automaticity and protection are the hallmarks of parasystole. This feature lead to the diagnostic triad of classical parasystole,
that is, varying coupling intervals of the ectopic focus, mathematically related interectopic intervals and the presence of fusion beats. The original assumption of total independence (protection) of parasystolic focus from extraneous beats is no longer considered true. It is now evident that any pacemaker connected to the surrounding tissues by an area of depressed excitability producing entrance conduction disturbances while allowing the impulses to exit is subject to some degree of modulation by electrotonic depolarizations arising in the surrounding tissues, that is, depolarization of the surrounding field will induce a partial depolarization of the pacemaker cells. This results in the occurrence of modulated parasystole. Nonparasystolic beats falling close to the parasystolic beats can cause (1) maximal delay, (2) maximal acceleration, (3) lesser effects, (4) practically no effect, or (5) pacemaker annihilation of the parasystolic focus. Nonparasystolic beats falling during the first half of the parasystolic interval induce a prolongation of this interval, those occurring during the second half of the parasystolic cycle length cause an abbreviation of this interval. The electrotonic modulation (causing either prolongation or shortening of the parasystolic cycle length) is maximal at the middle point of the parasystolic interval. Thus, the coupling interval of the ectopic focus may not be constant in modulated parasystole and interectopic interval may also show variation. Modulation may also cause coupling intervals to be fixed, and the fixed coupling intervals may suggest a reciprocating bigeminy. It has been shown that if the impedance of the communication between the ectopic focus and the surrounding ventricle is low, locking of the ectopic pacemaker could be nearly fixed to give the appearance of a reciprocating bigeminy. This is substantiated by Figure 2 in our case, wherein after infusion of amiodarone there is partial suppression of focus A resulting in occasional ectopic beats during sustained monomorphic VT because of focus B. Measurement of B–B intervals shows that there is doubling of the rate from 0.88 s during BVT as seen in Figure 1, compared with 0.44 s in Figure 2 suggesting a parasystole as the mechanism of arrhythmia. The interlocking of the beat A to beat B still persists. But measurement of intervals between A–A in this strip does not show a mathematical relationship, which can be explained by modulation of parasystole focus A by the preceding beat (from focus B). El-Sherif et al have described concurrent discharge of a 2 to 6 parasystolic foci with a total of 24 parasystolic foci in the 6 patients with advanced heart disease. It is now accepted that irregularity of ectopic is the rule rather than an exception.

It is now evident that parasystole may give rise to various rhythm patterns. Parasystolic accelerated idioventricular rhythm—producing BVT has been described in digitalis toxicity. In the present case, dual parasystole is the most plausible explanation. The occurrence of only occasional beats of focus A (Figure 2) where the focus B is continuing in an accelerated rate excludes the possibility of a BVT because of a ping-pong effect as there is no consistency in occurrence of focus

Table. Causes of Bidirectional VT 5–10

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<th>Cause</th>
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<td>Digitalis toxicity</td>
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<td>Hypokalemic periodic paralysis</td>
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<td>Andersen–Tawil syndrome</td>
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<tr>
<td>Fulminant myocarditis</td>
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<tr>
<td>Catecholaminergic polymorphic ventricular tachycardia syndrome</td>
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<tr>
<td>Aconite poisoning</td>
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<td>Left ventricular noncompaction</td>
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VT indicates ventricular tachycardia.
A, which should have regularly alternated with focus B, if it was because of ping-pong effect.

Triggered activity and abnormal automaticity have been described secondary to myocardial inflammation in myocarditis. Incessant nature of the tachycardia, unresponsive to multiple DC cardioversion, antiarrhythmic agents such as β-blockers and lignocaine, suggests that the possible mechanism of VT is enhanced automaticity because arrhythmias due to reentry or because of delayed after depolarization typically are responsive to these maneuvers. It is possible that local inflammation around the automatic foci could create an entry block in cardiac sarcoidosis, resulting in parasystole. Multiple areas of granulomatous inflammation which is not uncommon in cardiac sarcoidosis could result in two or more parasystolic foci to coexist as in the present case. The mechanism of BVT is thought to be because of elevated intracellular calcium causing delayed after depolarization in anatomically separate parts of the conducting system. One possible explanation is that the inflammation causes cells from damaged areas to cause spontaneous release of Ca\(^{2+}\) from sarcoplasmic reticulum, which could generate waves of intracellular Ca\(^{2+}\) elevation and arrhythmias.

To the best of our knowledge in patients with cardiac sarcoidosis, BVT particularly irregular VT has not been described. This adds another differential diagnosis for this intriguing VT.

Conclusions

Cardiac sarcoidosis should be considered in the differential diagnosis of patients presenting with BVT, especially if the QRS is broad, the rhythm irregular and tachycardia incessant. Dual parasystole may be a possible mechanism in some cases of BVT.

Disclosures

None.

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