Idiopathic Ventricular Fibrillation or Ischemic Ventricular Fibrillation?

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PATIENT DESCRIPTION

A 42-year-old healthy man collapsed suddenly in the street while walking. The patient received 2 minutes of basic life support until an automatic external defibrillator was brought and detected ventricular fibrillation (VF), which was successfully terminated by a single shock. The patient regained consciousness and was transferred to the hospital.

The patient's physical examination was normal with no neurologic deficit. Blood pressure was 147/102 mmHg. Brain computed tomography showed normal findings. The first troponin I measurement within 1 hour of the event was in the normal range (19.6 ng/L, normal < 20 ng/L) and rose to 99.9 ng/L after 3 hours.

The patient's medical history was notable for untreated dyslipidemia, heavy smoking, and anxiety. In addition, his father was diagnosed with ischemic heart disease at 46 years of age. The patient had never complained of chest pain.

The 12-lead electrocardiogram performed in the emergency department is shown in Figure 1A. Sinus tachycardia (rate 101 beats/min) showed a normal QRS pattern and duration (84 msec). The QTc interval was normal (387 msec). An early repolarization pattern including an ST elevation of 1 mm was observed in inferior and lateral leads including a notched J-wave. A single premature ventricular complex (PVC) with short coupling interval (240 msec) and a morphology of left bundle branch block (LBBB) with left QRS axis (≈-60°) was noted. There were no electrocardiogram signs of ischemia.

After admission to the intensive cardiac care unit, the patient exhibited another VF episode treated with cardioversion [Figure 1B]. Again, a short-coupled PVC (coupling interval of 280 ms) was noted preceding polymorphic ventricular tachycardia (VT)/VF. No change in sinus cycle length was observed prior to the arrhythmia. The patient was given amiodarone (300 mg intravenous/30 minutes). Bedside echocardiogram showed normal cardiac size and function, no left ventricular hypertrophy, valvular dysfunction, or mitral valve prolapse. Coronary angiography revealed a tight lesion at the first ostial diagonal bifurcation [Figure 1C], which was treated with stenting. During the procedure, a third VF occurred, which required cardioversion.

One hour after returning to the ward, another VF occurred. Treatment with quinidine (200 mg four times daily) was initiated and subsequently an automatic defibrillator was implanted. The patient was also advised to seek genetic counselling.

He remained asymptomatic without any documented arrhythmia at a follow-up 2 months later. Quinidine therapy was well tolerated.

COMMENT

A 42-year-old man presented with an out-of-hospital cardiac arrest with documented VF. His electrocardiogram was remarkable for ultra-short-coupled PVCs in isolation or preceding VF initiation, an early repolarization pattern, and a significant coronary lesion without electrocardiogram signs of myocardial ischemia.

The most common etiology for cardiac arrest in a middle-aged man is coronary artery disease (CAD), es-

pecially if, as in our case, the patient has multiple atherosclerotic risk factors. In this setting, the arrhythmia usually begins with a short-coupled PVC at maximal ischemic ST-segment elevation [1]. In our patient, such events were not observed during repeated electrocardiograms and at the onset of VF. Moreover, recurrent VF occurred 1 hour after successful angioplasty. Some patients with CAD and polymorphic VT may progress to arrhythmic storms refractory to conventional antiarrhythmic drug therapy and revascularization yet respond to quinidine therapy [2]. Most have a prior myocardial infarction and arrythmia occurs 3 days post-revascularization [2].

The current clinical presentation raises the suspicion for a non-ischemic origin of the arrhythmic episodes. The absence of any type of cardiomyopathy or mitral valve prolapse on the patient's echocardiogram suggests a likelihood of idiopathic VF or an inherited arrhythmia [1,3].

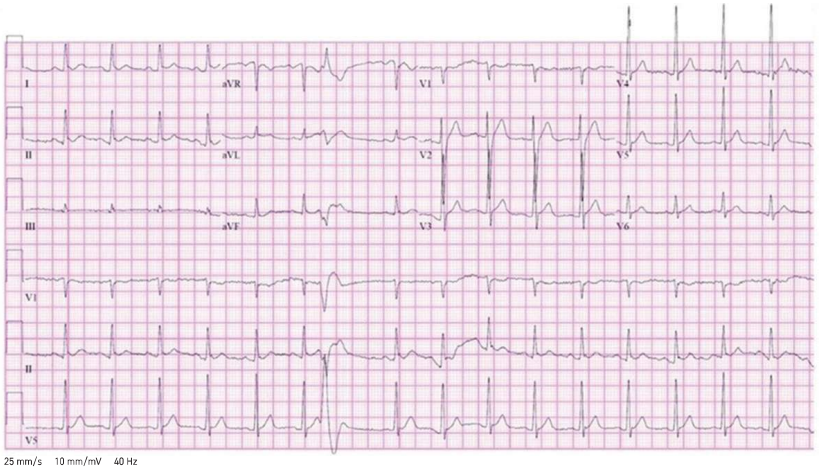
Among the inherited arrhythmia syndromes, two can be easily ruled out: long QT syndrome (LQTS) and catecholaminergic polymorphic VT (CPVT). The normal baseline QTc interval, the short-coupled PVC initiating polymorphic VT/VF, and the response to quinidine is hardly conciliable with LQTS [1]. In addition, CPVT is very unlikely to begin at rest or after a short-coupled PVC [1]. Therefore, we are left with Brugada syndrome, early repolarization syndrome (ERS), and short QT syndrome (SQTS), which all share the tendency to develop VF triggered by short-coupled PVCs and to respond to quinidine treatment [1].

Brugada syndrome is typically suspected in the presence of a right

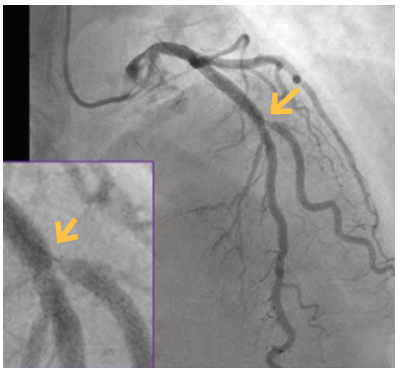
Figure 1. Electrocardiogram and coronary angiography at admission

PVC = premature ventricular complex, VF = ventricular fibrillation, VT = ventricular fibrillation

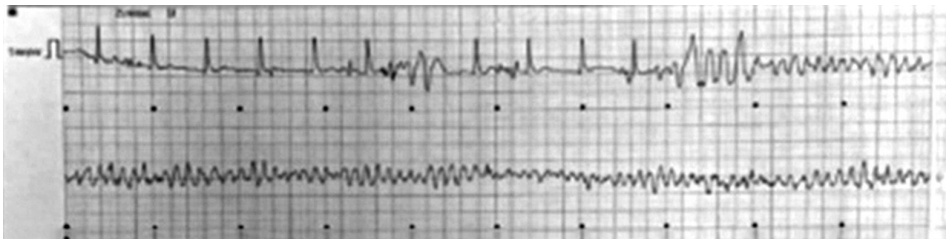
[A] Electrocardiogram at admission showing sinus tachycardia (rate 101 beats/min) with normal QRS pattern and duration (84 msec); QTc interval is 387 msec; an early repolarization pattern including an ST elevation of 1 mm observed in inferior and lateral electrocardiogram leads including a notched J-wave; single PVC with morphology of left bundle branch block and left QRS axis (≈-60°) with short coupling interval of 240 msec



[C] Coronary angiography showing tight lesion at the first ostial diagonal bifurcation (yellow arrow)



[B] VF episode, precipitated by a PVC with a short coupling interval of 280 msec and polymorphic VT deteriorating to VF



bundle-branch block with marked ST-segment elevation in the right precordial leads, especially at the time of VF onset [1], like in our patient. A sodium channel blocker can unmask a concealed Brugada electrocardiogram; however, our patient had no family history of Brugada syndrome or sudden cardiac death and no suggestion of the disease on his baseline electrocardiogram. Furthermore, in Brugada syndrome, the coupling interval of the PVC initiating VF is rarely as short as in our case [1].

SQTS is a rare genetic disorder characterized by a QTc ≤ 360 msec. In our patient the QTc was within normal limits (387 msec).

ERS is diagnosed in a patient resuscitated from polymorphic VT/VF without any heart disease and having J-point elevation ≥ 1 mm in ≥ 2 adjacent inferior and/or lateral electrocardiogram leads [1]. The early repolarization pattern is accentuated before the onset of arrhythmia [4]. Our patient had only 1 mm J-point elevation with no obvious increase before the short-coupled PVC and VF initiation.

Idiopathic VF was highly suspected despite the significant accompanying coronary lesion. We considered that the elevated troponin levels were probably a consequence of the patient's ventricular arrhythmias and shocks delivered to terminate them more than an acute coronary syndrome, as the first troponin obtained was in the normal range and rose after 3 hours. Elevated troponin levels have a variety of differential diagnoses and should always be considered in context.

No single parameter distinguishes idiopathic VF; however, the combination of a non-pause dependent ultrashort-coupled PVC [1], an LBBB morphology, and left axis suspected to originate from the right moderator band in a male patient strongly suggested the diagnosis of idiopathic VF.

CLINICAL IMPLICATIONS

Three important clinical implications can be drawn from the present case:

Although in its classic definition, the diagnosis of idiopathic VF is made by exclusion of all possible causes of VF [1]. Our case suggests that idiopathic VF and CAD may coexist in the same patient. Attributing the VF episodes to the latter diagnosis alone may have dramatic consequences, such as the recurrence of the arrhythmias despite successful coronary revascularization.

In a patient with no obvious heart disease presenting with syncope or aborted cardiac arrest, the occurrence of short-coupled PVCs may suggest an association with a malignant ventricular tachyarrhythmia in the setting of idiopathic VF [1,3,5].

Documentation of CAD during post-mortem examination in a patient who exhibited sudden cardiac death should not rule out the possibility that death may be unrelated to CAD.

CONCLUSIONS

We demonstrated how to appropriately diagnose the mechanism of VF in a young patient who exhibited various characteristics that could be caused by multiple potential underlying conditions.

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Remember, we all stumble, every one of us. That's why it's a comfort to go hand in hand.
Emily Kimbrough (1899–1989), American author and journalist

The bitterest tears shed over graves are for words left unsaid and deeds left undone.
Harriet Beecher Stowe (1811–1896), abolitionist and novelist