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Case Report

CASE REPORT

A good excuse for skipping the test: electrical storm in a teenager

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Abstract

We describe the case of a teenager with a structurally normal heart that presented with *torsades de pointes* and cardiac arrest. He had a history of epilepsy in childhood, mild cognitive impairment and cognitive visual dysfunction. The baseline electrocardiogram had prominent J waves and a marked early repolarization pattern in all the leads, with normal QT interval. We discuss the differential diagnosis for this interesting case, as well as the patient's management.

CASE REPORT

A 16-year-old male came to the emergency department after losing consciousness while taking a test in school. He had a brief prodromal period of dizziness and while he was unconscious he had involuntary movements, suggestive of a tonic-clonic seizure.

He had febrile seizures and parieto-occipital epilepsy in childhood, without recent seizures and no medication for over a year. He also had mild cognitive impairment and cognitive visual dysfunction. Family history was unremarkable.

While he was being examined, he had another syncope, with spontaneous recovery. Neurology was called, but a cardiologist that was passing by felt that his pulse was quite irregular and suggested an electrocardiogram (ECG).

The ECG (Fig. 1) showed sinus rhythm, J wave in all leads, normal QT interval and frequent monomorphic premature ventricular complexes (PVCs), with right bundle block morphology.

The patient was put on telemetry and immediately after that he had several episodes of frequent PVCs, followed by a polymorphic ventricular tachycardia (VT), *torsades de pointes* (TdP) and ventricular fibrillation (VF) (Fig. 1) requiring a total of six successful defibrillation shocks within the following hour.

After intravenous magnesium, potassium and labetalol infusion, there was a progressive decrease in PVC frequency and the J wave decreased.

Physical examination was unremarkable and body temperature was normal. Laboratorial investigations, including baseline electrolytes, acid-base analysis, thyroid function tests and drug screening, were negative, except for a small decrease in free carnitine and a slight increase in alanine levels (non-significant). Lactate and pyruvate levels were normal; the immune study and screening for metabolic diseases were also negative. Echocardiogram, cardiac magnetic resonance and cerebral magnetic resonance (including hypophysis assessment) were normal. No arrhythmias were seen during a treadmill stress test.

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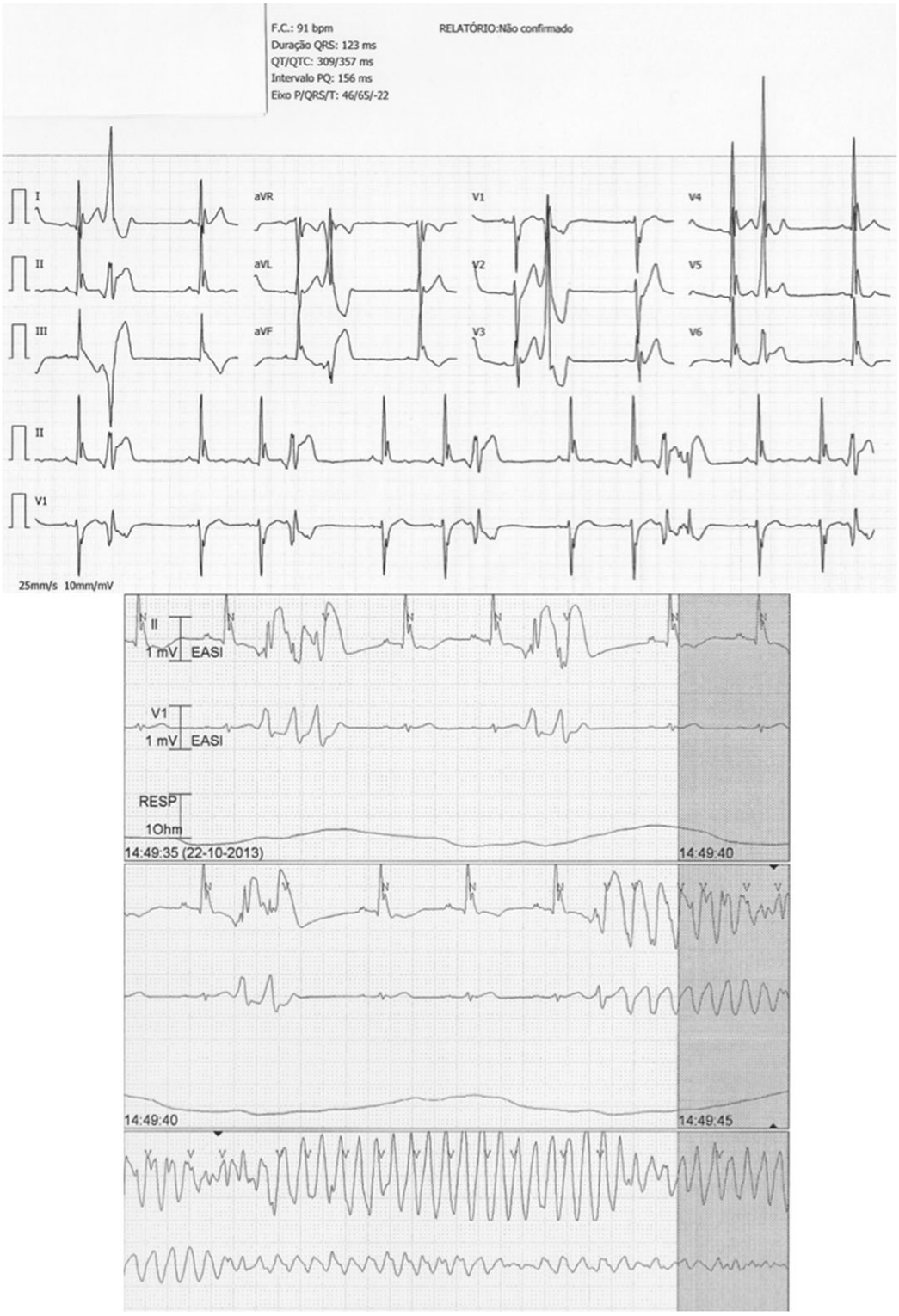

Figure 1: In the upper part of the picture, baseline ECG shows sinus rhythm, exuberant widespread J waves, inverted T waves in II and aVF and biphasic in V6, normal QT interval and frequent monomorphic PVCs, with right bundle block morphology; in the bottom part of the picture, the telemetry tracing is seen, with frequent ventricular ectopics that degenerated into torsades de pointes.

Differential diagnosis comprehended the following conditions:

(i) Mitochondrial cytopathy—considering the combination of epilepsy, visual changes, mild cognitive impairment, low carnitine and high piruvate levels. However, the clinical course would be quite unusual and genetic testing did not show any pathologic mutations usually present in these diseases.

(ii) Idiopathic VT/fibrillation—namely, the short-coupled variant of TdP [1], which is initiated by a short coupling

interval (<300 ms), mimicking the R-on-T phenomenon. However, we found only one described case in which this coexisted with J waves [2]. Moreover, he does not exactly meet the criteria for short-coupled variant of TdP, even though he could fit in another form of idiopathic VF if everything else is excluded.

(iii) J wave syndromes—the fact that this patient has very prominent J waves (also called Osborn waves) in the inferior and lateral leads in the baseline ECG, fitting an exuberant early repolarization (ER) pattern, is hard to overlook. Even though ER was usually considered benign, more recently it has also been associated with cardiac arrest (the so-called Haïssaguerre syndrome) [3].

(iv) Catecholaminergic polymorphic VT—the first episode happened in a stressful situation, but the blood and urine measurements of catecholamines and metanephrines were normal.

(v) A variant of a QT syndrome—mostly because of the occurrence of TdP, but none of the ECGs showed a markedly abnormal QT interval.

During hospital stay, beta blockers seemed to suppress the PVCs and the patient did not have any other episodes of VT. An implanted cardiac defibrillator (ICD) was placed for secondary prevention and the patient was discharged on bisoprolol 5 mg daily.

After a few months, he was readmitted after appropriate ICD shocks (and he had several episodes of non-sustained VT previously), while he was resting. A trial of verapamil was done, but he maintained very frequent ventricular ectopics. Again, he was given intravenous beta blockers, that suppressed the PVCs, and afterwards switched to nadolol tablets (80 mg bid).

The patient was sent for an electrophysiology study, but at the time of the study he did not have enough PVCs for performing activation mapping and ablation. Flecainide provocation test was negative.

The results of the genetic study for catecholaminergic polymorphic VT, QT syndromes and Brugada came back negative.

The patient did not have any other significant events so far —more than 2 years after the last electrical storm. He is usually in sinus bradycardia at rest, without ventricular pacing (programmed at 50 bpm).

DISCUSSION

After extensive discussion of the case, we concluded that this patient has idiopathic VF associated with ER. This occurs in young adults, usually males, that present with syncope or cardiac arrest, in the absence of structural heart disease or identifiable channelopathies, without prolonged QT intervals. Etiology is unknown and they have a poor response to verapamil—noteworthy because verapamil was the first line treatment originally proposed for short-coupled variant of TdP (essentially indistinguishable from idiopathic VF) [4]. Unfortunately, we do not know exactly how to distinguish 'arrhythmogenic' from 'normal' J waves. However, our patient had markedly abnormal ECG features: notched J-point elevation >0.2 mV and showing a transient increase, present in all the leads including the inferolateral, and dowslopping ST segment.

This is an entity of which little is known so far, namely its cause or triggering factors. There may be an underlying channelopathy—or more than one—that has not been identified yet.

ICD implantation for secondary prevention is mandatory. Other cases of J wave syndromes have been shown to respond well to long-term quinidine treatment, and to isoprenaline infusion in the acute approach of an electrical storm. However, when we observed the patient in the emergency department he was already in TdP, so we administered drugs used for other cases of *torsades* (usually associated with long QT). Since we do not have commercially available quinidine and the patient improved with beta blockers, we pursued this strategy.

Finally, this case reminds us that monitoring the heart rate and rhythm during an apparent generalized seizure can unravel surprising arrhythmias. Since this patient also had a history of epilepsy, this was a particularly challenging case.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

None.

ETHICAL APPROVAL

No ethical issues posed by the Ethical Committee.

CONSENT

Patient is now an adult and gave consent for publishing his case.

GUARANTOR

Dr Patrícia Rodrigues.

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