Journal of Clinical & Invasive Cardiology
Volume 1, Number 1, 2014

Case report

Cardiac arrhythmia masquerading as a seizure disorder

Meghan K. Borden Trojan, D.O.[1]; Sandeep M. Patel, M.D.[2]; Stephen J. D'Auria, M.D.[1]; Samir F. Saba, M.D.[3]

[1]Division of Internal Medicine, [2]Division of Cardiology, [3]Division of Electrophysiology

University of Pittsburgh Medical Center, Pittsburgh, USA

Abstract: A 53-year-old male with a history of seizures and hypertension presented for a routine evaluation. An in-office ECG revealed normal sinus rhythm with an incomplete right bundle branch block (RBBB) pattern and coved ST-segment elevation in V1-V2 with gradual descent ending in an inverted T wave. He had a family history of sudden cardiac death in his maternal grandmother at the age 41, and a maternal uncle suffered a heart attack in his late 20s. Physical examination, laboratory studies, transthoracic echocardiogram, and an exercise single-photon emission computed tomography stress test were all unremarkable. His ECG findings were typical for a Type I Brugada pattern. There was concern that the episodes of seizures were actually instances of arrhythmias. He was diagnosed with Brugada syndrome. The patient was deemed to be symptomatic, given that his seizure episodes were likely due to arrhythmias. The patient elected to proceed with implantation of a single-chamber AICD.
Key Words: Brudaga, arrhythmia, sodium channel disorder

Introduction

Brugada syndrome (BrS) is an inherited arrhythmogenic process in patients with structurally normal hearts.[1] It is associated with a greater risk of sudden cardiac death (SCD) and exhibits an autosomal dominant inheritance pattern with 20% of cases attributed to the *SCN5A* mutation in the inward sodium channel.[2] The following case explores the diagnostic work-up and management of a patient presenting with neurologic spells and unique electrocardiographic (ECG) findings.

Case presentation

A 53-year-old male with a history of seizures and hypertension presented to his primary care physician. An ECG revealed normal sinus rhythm with an incomplete RBBB pattern, and coved ST-segment elevation in V1-V2 with gradual descent ending in an inverted T wave (Figure 1). He was referred to the emergency department (ED).

The patient's history included two seizures characterized by flaccid posture and a blank stare. Previous computed tomography of his head was negative for any intracranial abnormalities. Electroencephalograms (EEG) were negative, and he was ultimately treated with Carbamazepine.

In the ED, he denied having any cardio-pulmonary symptoms. He admitted having had intermittent exertional jaw pain and substernal chest discomfort since he was 11 years old; however, these symptoms were unchanged. Family history (FHx) revealed SCD in his maternal grandmother at 41 years old, and his maternal uncle suffered a heart attack in his late 20s. On exam, he was hypertensive. The cardiovascular exam and chest x-ray were

Correspondence to: Samir F. Saba, University of Pittsburgh Medical Center, 200 Lothrop Street, Pittsburgh, PA PUH B535. E-mail: sabas@upmc.edu. Received 25 January 2014; Revised 4 February 2014; Accepted 14 May 2014; Published 12 July 2014.

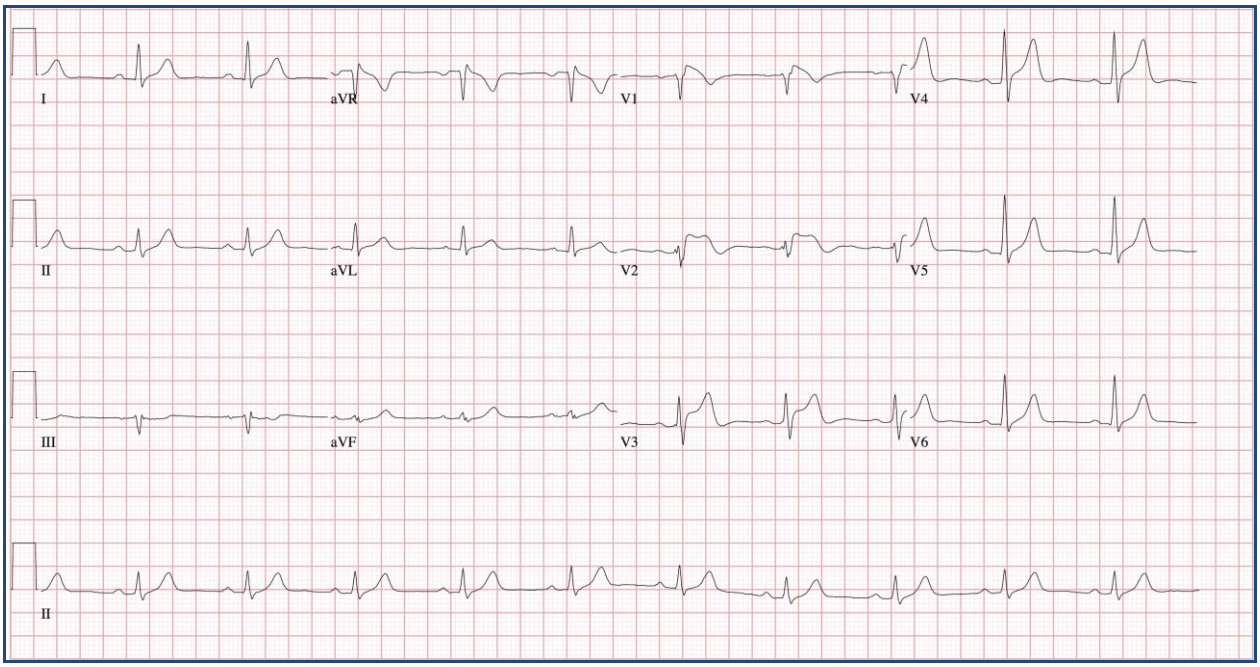

Figure 1: ECG performed at presentation.

unremarkable. His ECG is shown in Figure 1. Initial troponin-I level was <0.01 ng/dl (<0.01). Transthoracic echocardiography revealed an ejection fraction of 60–65% without other abnormalities. An exercise single-photon emission computed tomography stress test demonstrated a negative electrocardiographic response, and no perfusion defects at rest or with stress imply a low probability of ischemia or infarction. He was ruled out for acute coronary syndrome.

The patient's history of intermittent chest discomfort since childhood, and FHx of SCD combined with the ECG findings, were compatible with a clinical diagnosis of Type 1 BrS. His seizure episodes were attributed to arrhythmias. The patient was evaluated for the *SCN5A* mutation, which was ultimately found to be negative. He received a single-chamber implantable cardioverter-defibrillator (ICD), and since then has had no documented events.

Discussion

BrS is an inherited arrhythmogenic process in patients with structurally normal hearts, initially presenting with a pseudo-RBBB pattern and persistent ST-segment elevation in leads V1-V3.[1,2] A normal RBBB pattern displays widened S-waves in the left lateral leads. This finding is absent in most patients with BrS.[3] In BrS Type 1, the ECG will reveal a cove-type pattern with ST-segment elevation (≥2mm) that descends with an upward convexity to an inverted T-wave.[1,4,5] Type 2 BrS, or the saddle back pattern, can be identified by a high r' takeoff (≥2mm) followed by ST-elevation.[2] The elevated ST-segment descends towards the baseline first, and then turns and rises to an upright or biphasic T-wave. Previously, type 2 BrS was divided into a type 2 and a type 3 pattern, where the ST-segment was elevated ≥1 mm in type 2 and <1mm in type 3.[4,5]

Clinical diagnostic criteria include FHx of SCD <45 years of age, coved type ECG patterns in family members, electrophysiological inducibility of the ECG patterns, documented ventricular fibrillation and/or self-limiting polymorphic ventricular tachycardia, and history of nocturnal agonal respiration and/or syncope. Importantly, if a patient only demonstrates ECG findings without symptoms, this is referred to as a Brugada pattern.[4]

In this case, the patient's wife awoke to the sound of him "gurgling." He was unresponsive and flaccid with his eyes open. This presentation provides an exercise regarding the differential diagnosis for neurologic "spells." The differential can be organized by category including cardiogenic, neurogenic, and metabolic. Cardiac etiologies can further be divided into rhythm disturbances, obstructive phenomena (pulmonary embolism, constrictive pericarditis/tamponade, and left ventricular outflow tract obstruction such as severe aortic stenosis or myxoma), ischemic (triple vessel or left main disease), or myopathic (hypertrophic obstructive cardiomyopathy, hypertensive heart disease with a small left ventricular cavity, and restrictive cardiomyopathy). Neurogenic etiologies include seizures, cerebrovascular accident, aneurysm, subdural/epidural hematomas, and subarachnoid hemorrhage. Metabolic etiologies include electrolyte abnormalities (hyper- or hyponatremia, hypercalcemia), hyper- or hypoglycemia, and acid/base disturbances can be placed into the metabolic category. Lastly, additional etiologies include bleeding, dehydration, vaso-vagal synope, toxins, medication overdose or withdrawal, or thyroid disorders. The lab work and neurological imaging were normal in the patient. There was no history of drug use or medications that could have been responsible. The episodes were not related to exertion and the TTE was normal, ruling out outflow tract obstruction, aortic stenosis, myopathy, or tamponade. His history did not fit that of a typical seizure. He denied any prodromal symptoms. His wife denied any jerking movements, loss of bowel or bladder control, oral or hand automatisms, or a post-ictal state making the diagnosis of a seizure much less likely. The episodes were estimated at forty minutes in duration, and the EEG did not reveal any epileptiform waveforms, all of which argue against a seizure as the etiology of his spells. Patients with BrS typically experience the arrhythmias during sleep, as in our patient.[1]

If BrS is suspected, referral to a cardiac electrophysiologist is warranted because patients may need to undergo pharmacologic or invasive electrophysiologic testing. A discussion regarding invasive electrophysiological testing is outside of the scope of this article. A supervised "drug challenge" may induce a Brugada-type pattern when one is not present on the surface ECG. Sodium channel blockers, such as procainamide in the US or aimaline in Europe, are used. These medications can transiently induce the characteristic Brugada type 1 ECG changes among patients with Bruada type 2 ECG pattern.[5]

Multiple studies have provided recommendations regarding the management of patients diagnosed with BrS. A European registry of BrS patients demonstrated that the only predictive factors of arrhythmic events are presence of initial symptoms and Type 1 ECG pattern.[6] In patients with BrS, the ECG pattern is more pronounced and the risk of ventricular tachycardia is greater with fever.[7] Therefore, the guidelines recommend the immediate treatment of fever as a class 1 recommendation.[8] Initial studies have shown that antiarrhythmic agents did not protect against SCD in affected patients.[9] These studies, however, did show benefits of an ICD in order to prevent SCD, which led to implantation for both primary and secondary prevention. The most recent guidelines state that ICD implantation is recommended in patients with a diagnosis of BrS who have survived a cardiac arrest and/or have documented spontaneous sustained VT, with or without syncope, as a class 1 recommendation.[8]

In conclusion, this case provides a unique exercise concerning the evaluation of a patient with neurologic spells. Determining the approach to the diagnostic work-up and management may be difficult; thus, physicians should avoid anchoring bias on previous diagnoses and keep a broad differential when the historical, physical, and diagnostic evidence do not support the working diagnosis.

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