Brugada Syndrome: A Delayed and Atypical Presentation

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Abstract

Brugada syndrome is rare and life threatening cardiac channelopathy that can lead to sudden cardiac death. Fast recognition of a Brugada syndrome can be life saving. This case report presents an atypical case in which Brugada syndrome presented in electrical storm and was evident in both inferior and precordial leads.

Introduction

Brugada syndrome is a life threatening, hereditary, channelopathy that if left untreated, can often lead to sudden cardiac death. Since its illustration in 1992, it has become progressively more common. For this reason, it is becoming more important to recognize its pattern early to begin appropriate treatment and prevent disastrous consequences. At this point in time, there is little literature demonstrating that a Brugada pattern may be observed in inferior leads. This case report presents an atypical case in which Brugada syndrome presented in electrical storm and was evident in both inferior and precordial leads.

Case presentation

A 72-year-old man presented to the emergency department following an episode of generalized shaking and dizziness associated with vomiting and urinary and bowel incontinence. Patient had no prior cardiac history and was in his normal state of health until 2 days earlier when the patient noticed sudden onset of dizziness and headache. On the day of admission, his wife called Emergency Medical Service and he was found to be febrile at 104.9 F and in wide complex tachycardia with a rate of 160 bpm. He was administered IV Diltiazem, IV Amiodarone, and IV MgSO4. En route to the hospital, as well in the Emergency Room, this patient had to be defibrillated several times for episodes of ventricular fibrillation. There was no history of mouth frothing, tongue biting, loss of consciousness, or chest pain. No family history of sudden cardiac death or other cardiac issues.

Serial sets of chemistry panel and complete blood count were within reference ranges. Cardiac troponin I was elevated at 0.97. Urinalysis demonstrated nitrites, leukocyte esterase, and >50,000 white blood cells, blood cultures and urine cultures grew E. coli. Computed tomography of the head, abdomen, and pelvis, and chest X-ray were normal.

Serial 12-lead ECGs demonstrated findings, that when combined with history and presentation, suggested Brugada syndrome. Incomplete right bundle branch block, ST-elevation in lead III and VI and T-wave inversion (Figure 1). Patient was diagnosed with Gram-negative bacteremia with sepsis secondary to urinary tract infection with ECG features concerning for Brugada syndrome. He was managed with Tylenol, Meropenem, and Isoproterenol infusion initially which was then transitioned to PO Quinidine Gluconate.

On day 5, fever was well controlled and echocardiogram did not demonstrate any structural abnormalities. Patient underwent procedure for AICD placement successfully. On day 6, this patient underwent a coronary angiogram, which demonstrated normal coronary arteries. Quinidine was discontinued. After completing his course of IV meropenem and resolution of his gram-negative bacteremia on blood cultures, this patient was safely discharged home.

Discussion

Since becoming a clinical entity in 1992, cases of Brugada syndrome have increased drastically. Brugada syndrome is a relatively common cause of Sudden Cardiac Death (SCD) in a structurally normal heart. As cases are on the rise, it is clinically significant to recognize this syndrome early to avoid its devastating consequences. Two terms, distinguished by the presence or absence of symptoms, have been used to describe patients with the typical ECG findings of a pseudo-right bundle branch block and persistent ST segment elevation in leads V1 to V2:
1. Patients with typical ECG features who are asymptomatic and have no other clinical criteria are said to have the Brugada pattern (sometimes referred to as Brugada phenocopies).

2. Patients with typical ECG features who have experienced sudden cardiac death or a sustained ventricular tachyarrhythmia, or who have one or more of the other associated clinical criteria, are said to have the Brugada syndrome. Patients with ventricular premature beats or nonsustained VT, however, are generally not considered to have Brugada syndrome but only the Brugada pattern.

It is definitively diagnosed when ECG features are evident in the presence of a sodium channel blocker such as Flecainide. Brugada features can also be provoked by fever, electrolyte disturbances, and tricyclic antidepressants [1]. Once uncovered, Brugada features can quickly evolve into ventricular tachycardia and ventricular fibrillation that may lead to sudden cardiac death. The pathophysiology is thought to be due to a mutation in the SCN5A sodium channel in the epicardium and possibly the M cells. It is more common amongst younger, southeastern Asian populations [2]. Although not routinely described, Brugada syndrome may also present with ST-elevation in the inferior and lateral leads [3]. This report documents an uncommon presentation of Brugada syndrome with typical and atypical features. Initial ECG tracings demonstrate J point elevation in lead three as well as V2. This is a relatively rare finding in Brugada Syndrome. Although, the official diagnosis of Brugada syndrome does not mention inferior or lateral leads, there is some literature that reports it. This case emphasizes an additional occurrence where this phenomenon was visualized [4].

The patient’s presentation with over 14 defibrillator shocks is particularly notable. He is 72 years old, and being diagnosed for the first time. It is likely that this was triggered by the fever from bacteremia secondary to UTI. It is of interest to note that the patient was able to live to 72 years old without ever knowing or feeling cardiac abnormalities. This is the first occurrence of his lifetime, and significant enough to throw him into electrical VF storm.

Fever causes reduction in sodium current and changes to action potentials that may cause ventricular fibrillation [5] a major concern during his admission was that spiking a fever may lead to additional runs of tachyarrhythmia, therefore aggressive fever control with Tylenol was pursued. It is likely that if fever control was not pursued, patient may have continued to have runs of ventricular fibrillation, ultimately leading to sudden cardiac death.

This case highlights the importance of early recognition and features of Brugada pattern in a febrile patient. Toxic patients with supraventricular tachycardia may have an underlying Brugada syndrome, and it is imperative to have a broad differential including this. Quick administration of antiarrhythmics may have strong implications in the field when a patient is febrile and in electrical storm. Additionally, this article demonstrates that Brugada features in V1-V3 are not absolute, and there may also exist inferior lead evidence. With the increasing incidence in Brugada syndrome, unusual cases like this, support the need for further investigation into the physiological landscape of the epicardium in Brugada syndrome.

References