Brugada Syndrome with Complete Right Bundle Branch Block Disclosed by a Febrile Illness

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Abstract

The characteristic of right-bundle branch block (RBBB) pattern in Brugada syndrome (BS) is an atypical pattern without a wide S wave in left leads. We present a case of a patient with BS who had a typical ECG pattern of complete RBBB (CRBBB) with a wide S wave in the left leads which was disclosed by a febrile illness. Our observations suggest that physicians should be careful to evaluate ECG change in response to a febrile state, even if the ECG shows a typical CRBBB pattern with deep and wide S waves in the left leads.

Key words: brugada syndrome, complete right-bundle branch block, fever, pilsicainide

Introduction

Brugada syndrome (BS) is a cardiac disorder associated with a high risk of sudden death. BS is characterized by an accentuated J wave and ST elevation in the right precordial leads, V1 to V3, on the surface electrocardiogram (ECG), with incomplete or complete right-bundle branch blocks (IRBBB or CRBBB, respectively) (1). An accentuated J wave stimulates an R’ and gives the appearance of RBBB. The R’ in BS is thought to be due to early repolarization of the right ventricular epicardium rather than the RBBB (2). The characteristic of RBBB pattern in BS is an inconstant and frequent pattern of IRBBB or atypical CRBBB (without a wide S wave in left leads) associated with the J point and ST segment elevation.

Herein we present a case of a patient with BS who had a typical ECG pattern of CRBBB with a wide S wave in the left leads which was disclosed by a febrile illness.

Case Report

A 64-year-old man was first diagnosed with CRBBB on a periodic checkup in 1996. He had a seizure while sleeping in 2004. Magnetic resonance imaging of the brain and electroencephalography examinations were each normal. His mother died suddenly when she was 51 years of age. He was examined by his personal physician for evaluation of fever and pharyngeal pain in August 2006 and was diagnosed with a common cold. The 12-lead ECG showed a coved-type ST segment elevation in V1 to V3, as in typical BS (Fig. 1A). After one week, he recovered from a febrile illness, and his ECG pattern returned to normal, with the exception of the CRBBB pattern (Fig. 1B). He had a typical RBBB ECG pattern, deep and wide S waves in the lateral leads, and the QRS duration was wide (176 ms). He was referred to our hospital. No structural abnormalities were detected by echocardiography or cardiac scintigraphy. A challenge test with pilsicainide, a sodium channel blocking drug, was performed. The ECG after intravenous injection of 0.5 mg/kg pilsicainide showed the typical coved-type ST segment elevation in V1 to V3, as observed during the febrile state (Fig. 2). Subsequently, an electrophysiologic study was performed in the fasting and non-sedated state. VF was induced by programmed ventricular stimulation from the right ventricular apex with S3 extrastimuli (Fig. 3). After written informed consent was obtained, an implantable cardioverter defibrillator (ICD) was implanted. During a one-year follow-up period, VF was not observed.

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**Discussion**

A number of case reports have described that fever triggers the clinical manifestations of BS (3-7). However, as far as we know, it has never been reported that an ECG pattern of BS has been evoked by a febrile illness in a patient with a CRBBB pattern. This is the first case report of a patient with BS in whom a typical CRBBB pattern was disclosed by a febrile illness.

The right bundle branch originates from the His bundle and crosses to right side of the interventricular septum, then extends along the right ventricular wall to the region of the anterolateral papillary muscle of the right ventricle. Eriksson et al (8) reported that the prevalence of bundle-branch block increases from 1% at age 50 years to 17% at age 80 years. The RBBB pattern in a patient with BS is not characterized by deep and wide S waves in the lateral leads, and the RBBB pattern has an almost normal QRS duration. The present BS patient had a typical RBBB ECG pattern with deep and wide S waves in the lateral leads and a wide QRS duration.

One of the current models explaining the ECG alterations observed in patients with BS is based on an imbalance between the depolarizing and repolarizing currents during the action potential phase 1, mainly in cells expressing a large transient outward (Ito) current as the epicardial cells of the right ventricle. BS is linked to different mutations in SCN5A, the gene encoding for the alpha subunit of the cardiac sodium channel (9). Dumaine et al (10) have reported a
definite link between high temperatures and alteration of sodium channel kinetics due to SCN5A gene mutations. Keller et al (11) screened the SCN5A mutant gene in four male patients with typical BS ST segment elevations in V1 to V3 or ventricular arrhythmias during fever. They reported that a pronounced acceleration of Na+ 1.5 activation and fast inactivation kinetics were confirmed with an increase in temperature to 42°C. They postulated that the effect of elevated temperature on the remaining INa, which is mainly or totally mediated by wild type channels, could be responsible for the ECG phenotype in patients with BS. Furthermore, they recently reported that a shift in activation and change in the slope factor at higher temperatures, mimicking fever, could reduce sodium current availability and trigger the manifestation of the BS phenotype (12). In the present case, a febrile illness disclosed the typical ECG pattern of BS in a man with CRBBB and the ECG abnormalities were reproduced with intravenous pilsicainide. This observation is in agreement with previous laboratory findings that have demonstrated a definite link between high temperatures and the alteration of sodium channel kinetics due to SCN5A gene mutations (10-12).

Our observations suggest that physicians should carefully evaluate any ECG change in response to a febrile state, even if the ECG shows a typical CRBBB pattern with deep and wide S waves in the left leads.

References


Figure 3. Intracardiac electrogram of induced VF in an electrophysiologic study. VF is induced by programmed ventricular stimulation from the right ventricular apex with S3 extrastimuli. HB: His bundle, CS: coronary sinus, px: proximal electrode, ds: distal electrode, RVOT: right ventricular outflow tract, RVA: right ventricular apex.
