

BRUGADA SYNDROME ACCOMPANIED WITH CORONARY HEART DISEASE

DRAŽEN BEDEKOVIĆ, IVICA BOŠNJAK, MARIN VUČKOVIĆ, JERKO ARAMBAŠIĆ

Osijek University Hospital Center, Division of Cardiology, Osijek, Croatia

Brugada syndrome is a congenital disorder that can lead to sudden cardiac death. It is characterized by spontaneous or provoked typical ECG features and the occurrence of malignant ventricular arrhythmias, most commonly manifested by syncope or sudden death. The use of an implantable cardioverter-defibrillator is the only effective therapy for arrhythmic death prevention. The coexistence of Brugada syndrome and coronary heart disease is rarely described in the literature. We present a case of a patient with coexistence of two different heart conditions, symptomatic Brugada syndrome and coronary heart disease.

Key words: Brugada syndrome, syncope, coronary heart disease

Address for correspondence: Dražen Bedeković, MD
Osijek University Hospital Center
Division of Cardiology
31 000 Osijek, Croatia
E-mail: drbedekovic@yahoo.com

INTRODUCTION

Brugada syndrome is an inherited disorder of ion channels in cardiomyocytes, and most common are mutations in the SCN5A gene. It is characterized by specific features on ECG, can cause syncope and sudden cardiac death due to ventricular arrhythmias (1-3). Most patients have no structural changes in the heart or they are minimal, but cases of coexisting significant coronary heart disease, as well as ischemia-induced Brugada-like ECG features have been described (1,2,4,5). The diagnosis of Brugada syndrome is based on specific spontaneous ECG features or features occurring during test performance with sodium channel blockers application (e.g., ajmaline, procainamide, flecainide) (2). Brugada syndrome was first described more than 25 years ago. The definition and ECG diagnostic criteria have changed over time; today, we distinguish three types of ECG features, as follows: type 1 is diagnostic and consists of concave elevation of the ST-segment and J point by >2 mm at its peak, followed by negative T-wave with little or no isoelectric separation in one or more right precordial leads (V1 and V2); type 2 consists of high ST-segment elevation after J point (2 mm), which is followed by gradual descending ST-segment elevation (1 mm above the baseline), and positive or biphasic T wave, saddle configuration; and type 3 has a saddle or concave elevation of the ST segment of <1 mm in the right precordial leads (1).

The use of an implantable cardioverter defibrillator is effective therapeutic choice for prevention of sudden cardiac death due to arrhythmias (6). Although this syndrome typically manifests with arrhythmia and syncope episodes in younger age (1,2), we report a case of coexistence of two different heart conditions, Brugada syndrome and coronary heart disease in a 60-year-old patient.

CASE REPORT

A 60-year-old male was admitted to the coronary unit through the emergency department due to recurrent syncope. A few hours before admission while watching TV in a sitting position, the patient suddenly lost consciousness and spontaneously recovered. He had a similar event a few years before but did not report to the physician. He asserted to have occasional mild chest discomfort during physical activity. However, chest pain, shortness of breath, or any other symptom he did not report on the day of admission, but he consumed small amounts of alcoholic beverages the day before admission. He had well controlled arterial hypertension for several years, taking angiotensin-converting enzyme (ACE) inhibitor, diuretic and calcium channel blocker. His father died at the age of 57 by sudden death; he had a pacemaker implanted but the exact cause of death was uncertain.

The patient was generally in good health, afebrile, eupneic, normal neurological status, with audible murmur over the heart apex, intensity II/VI. The 12-channel ECG recorded showed a typical pattern for type 1 Brugada syndrome, i.e., right branch block and concave ST-segment elevation in V1 and V2 leads higher than 2 mm (Figure 1), without criteria for acute ischemia. All laboratory tests were within the reference values including repeated cardiac enzymes; echocardiography showed left ventricular ejection fraction of 53% and mild mitral regurgitation. Coronary angiography revealed the existence of hemodynamically significant stenosis (IFR 0.86) of middle segment left anterior descending (LAD) artery at bifurcation with first diagonal artery (D1), including D1 ostium with 75% lumen stenosis. Percutaneous coronary intervention was performed with dilatation of LAD and D1 and implantation of by drug-coated balloon (Figure 2). Diagnostic workup ruled out the existence of neurovascular disease as a cause of syncope. No ventricular or supraventricular arrhythmias, or atrioventricular conduction disturbances were observed by ECG monitoring, and existence of reflex syncope was also excluded. We implanted a single-chamber cardioverter defibrillator for prevention of sudden cardiac death due to possible ventricular arrhythmias associated with Brugada syndrome.

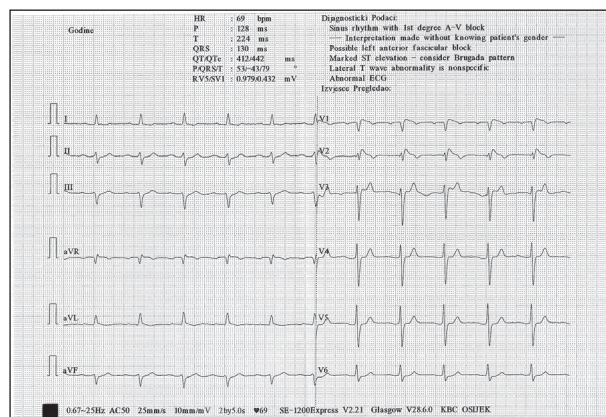


Fig. 1. ECG type I: Brugada – concave elevation of ST segment in V1-V2 leads.

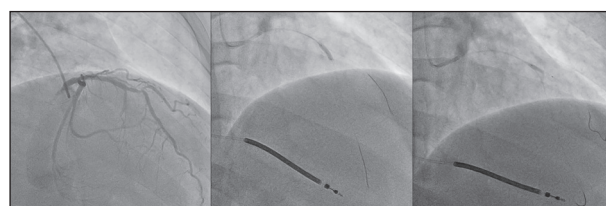


Fig. 2. Percutaneous coronary intervention on LAD and D1 arteries. Left image: coronary angiography showing significant bifurcation LAD/D1 stenosis; middle image: drug-coated balloon dilatation in LAD covering bifurcation with D1; right image: drug-coated balloon dilatation in proximal D1.

Three months after implantation of the cardioverter-defibrillator, the patient was in good health, had no syncope or recorded arrhythmias.

DISCUSSION

The incidence of Brugada syndrome has been estimated to five cases *per* 10,000 people; it is the cause of 4%-12% of total cardiac deaths and 20% of sudden cardiac deaths (2,4,7,8). Only a few published studies and case reports describe its coexistence with coronary heart disease. One smaller study including 55 cases with Brugada type ECG has reported significant coronary disease in 5 cases, yielding a 9.1% prevalence (10). Another retrospective analysis of 200 Brugada type ECG cases has reported significant coronary disease in 20% and coronary vasospasm in 11% of cases, with higher association of coronary disease in type 1 Brugada ECG pattern (11). The diagnosis of Brugada syndrome was based on a typical type 1 ECG pattern followed by recurrent syncope (6), although we did not prove the occurrence of ventricular arrhythmias by ECG monitoring. Type 1 ECG pattern does not require an arrhythmia provocation test with sodium channel blockers for diagnosis confirmation, whereas provocation test confirmation is necessary for types 2 and 3 (6). In our hospital, electrophysiological study was not available. Thorough clinical examination and tests failed to find another explanation for syncope. Coronary angiography showed significant coronary heart disease on LAD/D1 and successful revascularization was performed on those vessels. Although the patient reported occasional angina symptoms, acute coronary syndrome was not diagnosed at hospital admission (absence of symptoms or typical ischemia related ECG features on the day of admission, and normal troponin according to the 0-3 protocol), and the ECG remained unchanged after revascularization. We consider coronary heart disease in this patient to have been a concomitant accidental finding, which was not the cause of syncope or Brugada related ECG pattern. Diagnosis of acute coronary syndrome in Brugada type ECG and *vice versa* can be challenging due to ECG pattern overlap. In several published case reports, Brugada type ECG was masking anterior wall infarction (LAD and LMCA occlusion), as well as conus branch occlusion inducing ventricular arrhythmia in Brugada type ECG pattern (12-14). Diseases such as myoprecarditis can also mimic Brugada type ECG, and structural heart abnormalities can coexist with Brugada syndrome. We have previously reported on right ventricular structural abnormalities in a patient with Brugada type ECG induced with propafenone testing (15). Coexistence of Brugada syndrome and coronary spasm was found in 17% of Brugada cases in a small Japanese study (16).

Episodes of both entities usually occurred during the night and it was concluded that coronary spasm represented a risk factor for cardiac events in Brugada patients (16). Due to the increased risk of sudden arrhythmic death, syncope, type I ECG pattern and male gender, the patient received implantable cardioverter defibrillator (ICD). ICD is the only effective therapy to prevent sudden arrhythmic death for Brugada syndrome (2,6), and even with strong suspicion of ventricular arrhythmia existence we consider ICD implanting justified. In our opinion, considering recurrent syncope only as a symptom of successfully threatened coronary disease and discharging patient without ICD implantation could endanger patient life. Although our patient's father died of sudden death at the age of 57, this fact itself does not support the existence of Brugada syndrome in the family (6). The ECG sample analysis of the patient's relatives did not detect any specific patterns associated with Brugada syndrome, while genetic analyses were not available. The question of the hereditary component in this case remained unanswered.

In conclusion, we believe that there were two different diseases in this case, mildly symptomatic/asymptomatic coronary disease and symptomatic Brugada syndrome, both recognized and successfully treated. Although the assumed malignant rhythm disorder can occur at an age when it is usually associated with ischemic heart disease, we consider that Brugada syndrome most likely was the cause of the malignant rhythm disorder in this case.

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SAŽETAK

BRUGADIN SINDROM UDRUŽEN S KORONARNOM SRČANOM BOLESTI

D. BEDEKOVIĆ, I. BOŠNJAK, M. VUČKOVIĆ, J. ARAMBAŠIĆ

Klinički bolnički centar Osijek, Zavod za kardiologiju, Osijek, Hrvatska

Brugadin sindrom je nasljedni poremećaj koji može dovesti do iznenadne srčane smrti. Obilježen je spontanom ili provociranim tipičnim promjenama u EKG-u te pojavama malignih ventrikulskih aritmija koje se najčešće manifestiraju sinkopom ili iznenadnom smrću. Primjena implantabilnog kardioverter defibrilatora je jedina učinkovita terapija za sprječavanje aritmijske smrti. Koegzistencija Brugadina sindroma i koronarne bolesti je vrlo rijetko opisana u literaturi. Donosimo prikaz bolesnika s koegzistencijom dviju srčanih bolesti: simptomatskog Brugadina sindroma i koronarne bolesti.

Ključne riječi: Brugadin sindrom, sinkopa, koronarna srčana bolest