Latent Brugada syndrome leading to sudden cardiac arrest revealed by exercise electrocardiography: a case report

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Submitted: 22 October 2007
Accepted: 9 January 2008

Arch Med Sci 2008; 4, 4: 455–459

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Abstract

We present the case of a 45-year-old patient who was hospitalized after he had suffered from a sudden cardiac arrest (SCA) episode during ventricular fibrillation. This sudden cardiac arrest episode occurred in an otherwise healthy subject who previously had been under treatment only for arterial hypertension. In the course of the exercise test the patient was shown to have an ST-segment elevation in leads V_1-V_3, with a subsequent negative T-wave. A drug challenge was used, on the basis of which Brugada syndrome was identified as being the cause of the sudden cardiac arrest in this patient.

Key words: Brugada syndrome, sudden cardiac arrest, cardioverter-defibrillator implantation.

Introduction

We present a case of a 45-year-old male with past medical history of hypertension who was admitted to the hospital following an episode of ventricular fibrillation. No reversible cause was found at that time and the patient was implanted with a cardioverter-defibrillator (ICD). Later, during exercise stress test, ST-segment elevation with inverted T-waves was noted in leads V_1-V_3. Subsequently drug challenge with ajmaline revealed ST elevation typical of Brugada syndrome.

Case report

The patient is a 45-year-old male, hospitalized for the first time in 2001 for retrosternal pain that appeared in the course of upper respiratory tract infection. During that episode of chest pain ST-T elevations were noted in the inferior leads and, because of suspicion of an acute inferior myocardial infarction, the patient underwent coronary angiography. The angiography revealed subsequently normal coronary arteries. In the following years the patient was treated for hypertension and had recurrent retrosternal chest pains, that could not be clearly associated with exertion. In 2003, the patient was admitted again to the hospital but this time right bundle branch block (RBBB) was only seen on the EKG. Repeat EKG showed RBBB again and there was no elevation of biochemical cardiac necrosis markers. The patient was started on: Ca-blocker, statin, aspirin and proton pump inhibitor and subsequently discharged from the hospital. In December 2005

he was brought to the hospital following sudden cardiac arrest (SCA) at home. Upon emergency medical services arrival ventricular fibrillation was noted and he was successfully shocked to sinus rhythm in the field. Several days prior to the SCA episode, the patient again had symptoms of upper respiratory infection with fever. After admission to the Coronary Care Unit, elevated levels of myocardial necrosis markers were observed: creatinine kinase – max 5549 U/l, creatinine kinase myocardial fraction – max 21.5 U/l, troponin – max 0.49 ng/ml. Remaining lab work showed no

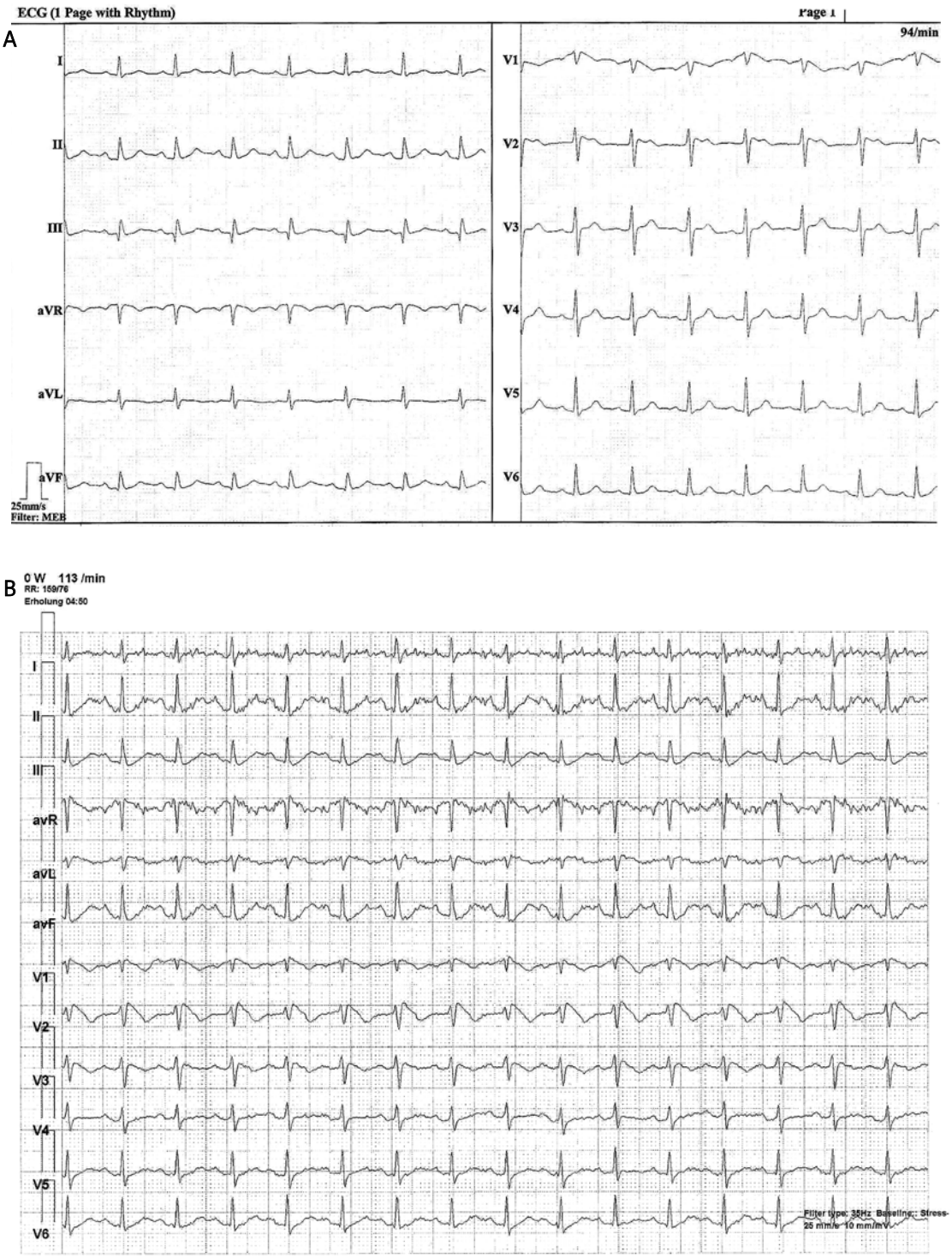

Figure 1. A – cardiac stress test registration – initial ECG, B – cardiac stress test registration – ECG in the 4th min and 50th s of the effort

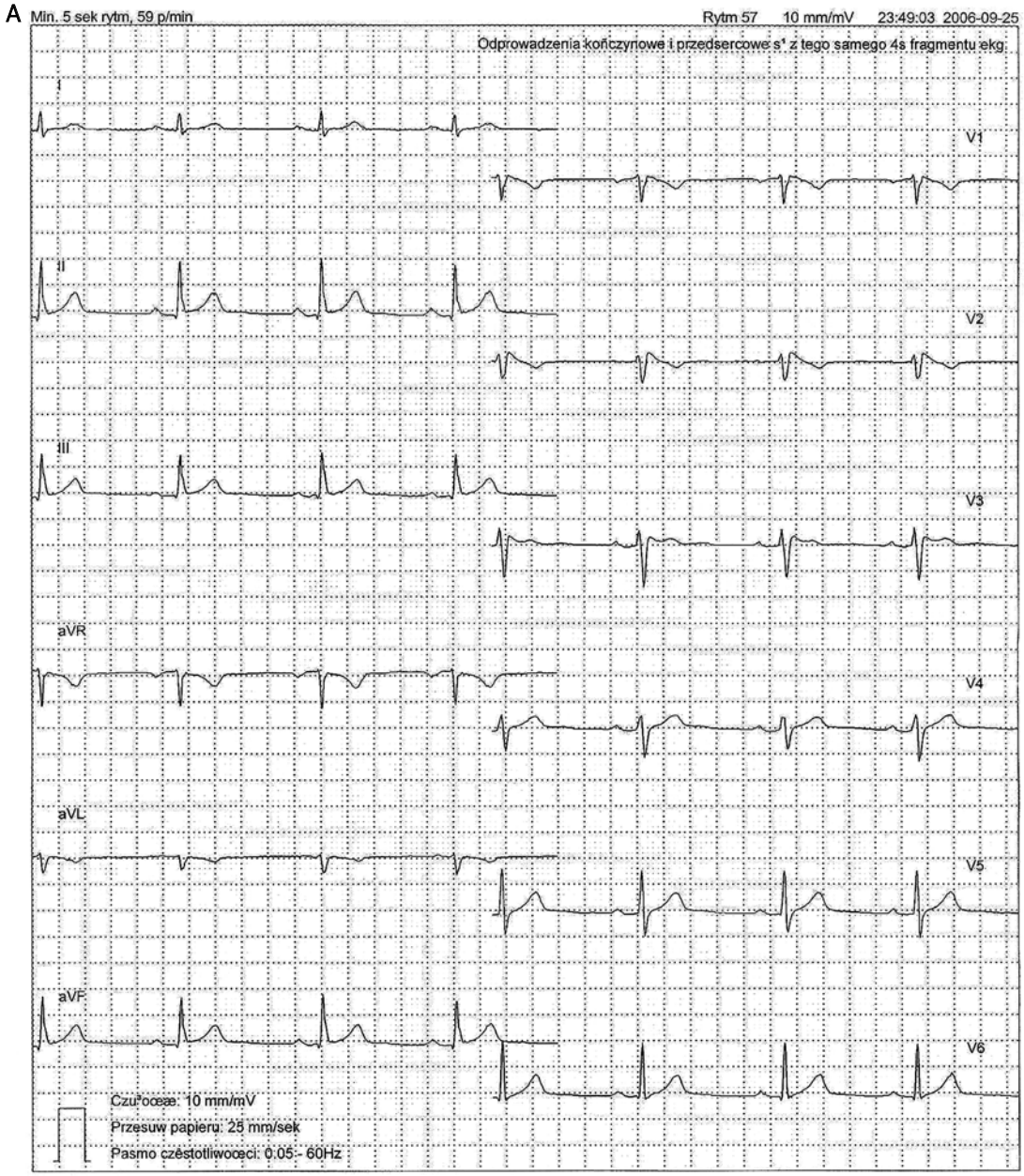

Figure 2. A – 12-lead resting ECG registration

electrolyte abnormalities, normal glucose concentration and normal complete blood count. Chest X-ray examination did not reveal any pathology. Computer tomography of the head did not show any evidence of bleeding or any other focal abnormalities. Abdominal ultrasound was unrevealing. Transthoracic echocardiogram showed no significant structural heart disease. His ejection fraction was estimated at 70%. Resting EKG showed sinus rhythm at rate of 89/min, intermittent right bundle branch block and QTc interval of 422 ms. It was then that, for the first time, the presence of intermittent right bundle branch block was noticed with a concave ST elevation in lead V_2. At that time the patient was transferred to our institution for further workup. An electrophysiology study was done. Sinoatrial node function and AH and HV intervals were normal. Programmed stimulation of the right atrium and right ventricle in the apex and in the outflow tract did not induce any arrhythmia. Since there were no reversible causes for the VF the patient underwent implantation of an ICD in March of 2006. In the course of further diagnostic testing T-wave

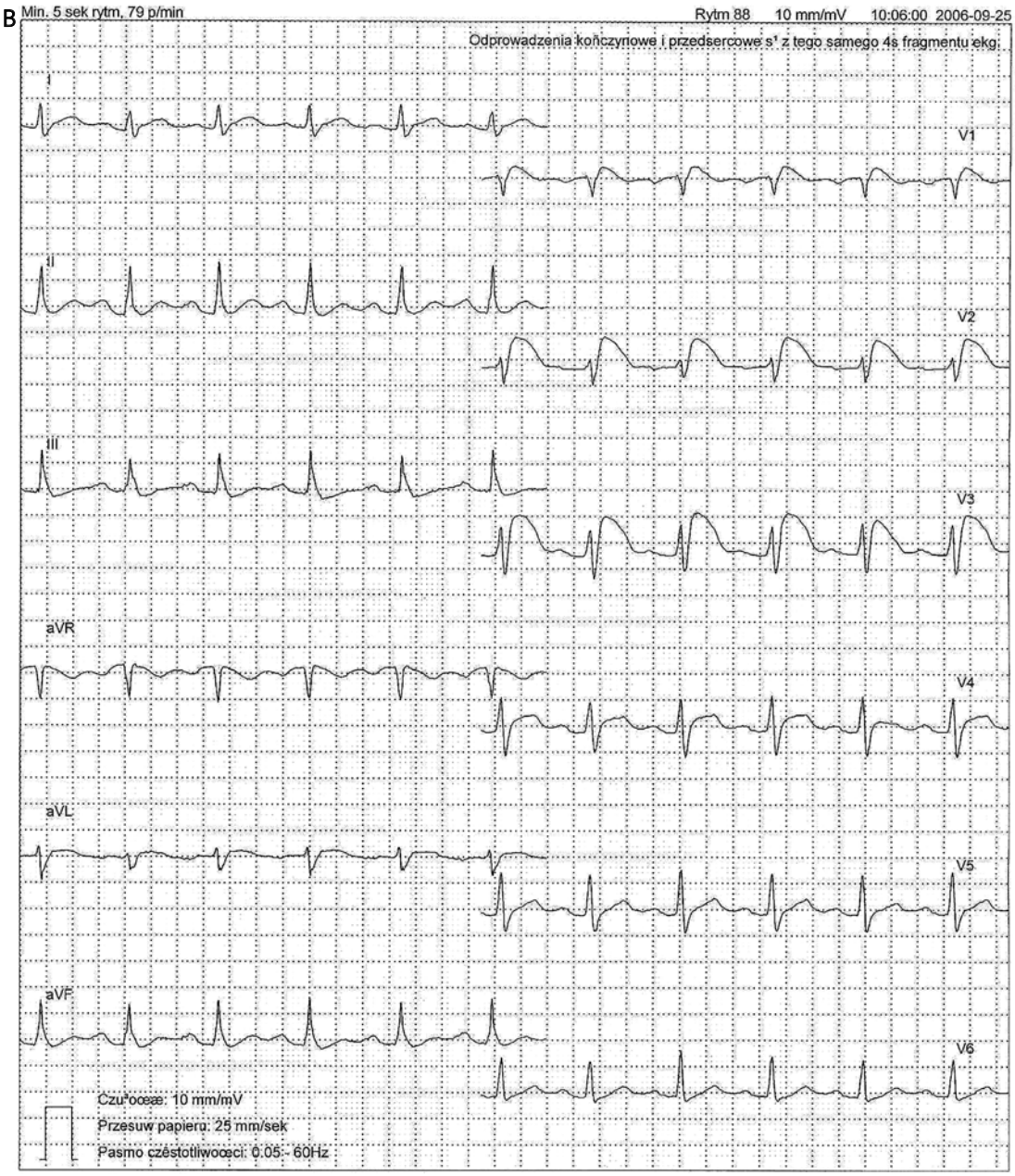

Figure 2. B – 12-lead ECG registration after an intravenous administration of ajmaline with dosis of 1 mg/kg of body mass

alternans (TWA) was performed as well as a stress test. TWA results were negative, but during the exercise stress test concave ST-T elevations as well as T-wave inversions in leads V_1-V_3 (J-point elevation by 1 mm) were seen (Figure 1). Since the patient was suspected of having Brugada syndrome he underwent drug challenge. Five minutes after a 1 mg/kg dose of ajmaline was administered, typical of Brugada syndrome, a concave ST-segment and J-point elevations as well as inverted T-waves were observed (Figure 2). The diagnosis, the nature of the disease and the risk to his relatives were discussed with the patient. Further evaluation has been offered to the patient's relatives but as of yet none of them have agreed to proceed with testing. During the routine follow-up there has been no cardioverter discharge and in the cardioverter memory no ventricular arrhythmias have been noted.

Discussion

The case described in this report is an example of a still underestimated disease, Brugada syndrome. Brugada syndrome constitutes a deadly threat that often remains latent for many years only

to manifest itself in a lethal arrhythmia in persons considered to be otherwise healthy [1, 2]. Present diagnostic criteria of the syndrome were established during the second consensus conference in 2005 [2]. It seems that the main reason for delayed diagnosis of Brugada syndrome is the periodic normalization of electrocardiographic features of the syndrome [3], which is related to incomplete penetration of the gene responsible for the syndrome. Moreover, changes in EKG may be subject to influences of different factors, such as body temperature [4] or autonomic system tone [5, 6]. Studies looking at the influence of autonomic system tone on ECG changes occurring in Brugada syndrome [6, 7] have demonstrated that α-adrenergic stimulation and β-receptor blockade intensify ST-segment elevations in subjects with Brugada syndrome, whereas β-adrenergic stimulation and α-receptor blockade reduce ST-segment elevations. Further surveys have also demonstrated that first generation antihistamine drugs, cocaine and tricyclic antidepressants may intensify EKG changes typical of Brugada syndrome [6]. It has been demonstrated that in such cases the use of class I antiarrhythmic drugs in a drug challenge test allows silent EKG features of Brugada syndrome to be revealed [7].

If we look closer at the case described in this report, the EKG changes were consistently seen during febrile illness. The manifestation of typical electrocardiographic features of Brugada syndrome during the increase of body temperature has been well documented [4, 8] and was present in this patient. On the other hand presence of EKG changes during the exercise stress test is atypical. In publications so far, attention has been drawn to the fact that during exercise stress test typical EKG changes of Brugada syndrome have a tendency to disappear. Electrocardiographic changes manifest themselves far more frequently at night, together with increased activity of the parasympathetic nervous system. In our case we have observed ST elevations in the right precordial leads during performance of the cardiac stress test. It is possible then that the function of the different parts of the autonomic nervous system in Brugada syndrome pathogenesis is thus not completely understood.

The current understanding is that patients with clinically and electrocardiographically symptomatic BS are characterized by a high risk of SCD, and should be implanted with an ICD [9].

In our case the decision to implant a cardioverter-defibrillator was taken before BS was diagnosed and was based on published guidelines for secondary prevention of SCD alone [10]. Nevertheless, in the end, it was the most appropriate therapeutic intervention for this patient.

In conclusion, any episode of ventricular fibrillation or SCD in a patient without any apparent structural heart pathology should arouse suspicion of Brugada syndrome – in particular in young subjects. Bearing in mind the serious consequences of the syndrome, one should therefore consider whether diagnostic manoeuvres to reveal EKG Brugada pattern ought to be integrated into a routine workup of patients who have suffered SCA.

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