Rev Port Cardiol. 2014;33(12):801.e1–801.e6

Revista Portuguesa de Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

CASE REPORT

Syncope in a febrile state: A case report of Brugada syndrome☆

Juliana Martins*, **Carlos Braga**, **Carina Arantes**, **Vítor Ramos**, **Alberto Salgado**, **Adília Rebelo**, **Adelino Correia**

Serviço de Cardiologia, Hospital de Braga, Braga, Portugal

Received 5 January 2014; accepted 10 July 2014
Available online 19 December 2014

KEYWORDS
Sudden death;
Brugada syndrome;
Fever

Abstract In 1992, Brugada and Brugada first described a new entity, which became known as Brugada syndrome, that is associated with a high risk of ventricular arrhythmias and sudden cardiac death in patients without structural heart disease. This syndrome is characterized by a distinct electrocardiographic phenotype, type 1 Brugada pattern, consisting of a coved ST-segment elevation (≥0.2 mV) followed by a negative T wave in more than one right precordial lead. This pattern is dynamic, and can be spontaneous or concealed, but is unmasked under certain circumstances, like febrile states.

The authors report a case in which the diagnosis of Brugada syndrome was made in the course of etiologic investigation of recurrent syncope in a febrile state.

PALAVRAS-CHAVE
Morte súbita;
Síndrome de Brugada;
Febre

Síncope em contexto febril – caso clínico de síndrome de Brugada

Resumo Em 1992, Brugada *et* Brugada descreveram pela primeira vez uma entidade, conhecida atualmente por síndrome de Brugada, associada a aumento do risco de arritmias ventriculares e morte súbita cardíaca em indivíduos sem cardiopatia estrutural. Esta síndrome caracteriza-se por um fenótipo eletrocardiográfico distinto, padrão de Brugada tipo 1, o qual consiste numa elevação (≥ 0,2 mV) «arqueada» do segmento ST seguida de uma onda T negativa em mais de uma derivação precordial direita. Este fenótipo é dinâmico, podendo ser

☆ Please cite this article as: Martins J, Braga C, Arantes C, et al. Síncope em contexto febril – caso clínico de síndrome de Brugada. Rev Port Cardiol. 2014;33:801.e1–801.e6.

* Corresponding author.
E-mail address: Ju_vpa@hotmail.com (J. Martins).

espontâneo ou encontrar-se oculto, sendo desmascarado em múltiplas circunstâncias, nomeadamente em contexto febril.

Os autores apresentam um caso clínico em que o diagnóstico de síndrome de Brugada é efetuado na sequência do estudo etiológico de síncopes recorrentes em contexto febril.

Case report

We report the case of a 40-year-old man who went to the emergency department (ED) with fever and productive cough for about two days. While in the ED he had two episodes of syncope preceded by palpitations, sweating and blurred vision, followed by rapid recovery of normal consciousness. He was not under electrocardiographic monitoring during either episode.

The patient reported recurrent syncope in febrile states since childhood, and a few months before this admission had had an episode of pre-syncope preceded by palpitations during the night. He had no other relevant medical history and was not taking any routine medication; on the day of admission he had taken only paracetamol. There was no family history of heart disease, sudden cardiac death (SCD) or recurrent syncope.

On physical examination the patient was normotensive, tachycardic, febrile, and slightly polypneic, with diminished breath sounds bilaterally and no alterations on cardiac auscultation.

The electrocardiogram (ECG) after the syncopal events (Figure 1A) showed a coved ST-segment elevation in V1 and V2, maximum 0.6 mV in V2, followed by a negative T wave; these alterations in ventricular repolarization are compatible with type 1 Brugada pattern.

Laboratory tests showed elevated inflammatory markers and D-dimers; serum potassium and myocardial necrosis biomarkers were within normal ranges. Following chest computed tomography angiography, which revealed no thromboembolism, the patient was transferred to the intermediate care unit with a diagnosis of community-acquired pneumonia complicated by type 1 respiratory failure. Empirical antibiotic therapy was begun with ceftriaxone and clarithromycin and he was kept under continuous electrocardiographic monitoring.

The patient's clinical course was favorable and his fever resolved within 24 hours. There were no further episodes of syncope or arrhythmia. ECG performed in apyrexia showed resolution of the typical Brugada alterations seen in a febrile state (Figure 1B). Transthoracic echocardiography revealed no structural heart disease.

Since the patient's setting was compatible with Brugada syndrome (BrS), with spontaneous type 1 Brugada pattern and a history of recurrent syncope, he received an implantable cardioverter-defibrillator (ICD) for primary prevention of SCD. He was discharged on the 11th day of hospitalization and advised to monitor and immediately treat any rise in body temperature, and was informed on the need to avoid drugs contraindicated in BrS.

Six months after discharge, the patient was asymptomatic and without arrhythmic events. Genetic screening of his family was underway.

Discussion

BrS is one of the hereditary conditions known as channelopathies, primary cardiac rhythm disturbances caused by ion channel anomalies that result in susceptibility to ventricular arrhythmias, and hence SCD, in individuals without structural heart disease.[1,2] It is estimated to be responsible for at least 4% of all sudden deaths and at least 20% of sudden deaths in patients with structurally normal hearts.[2]

BrS is a hereditary disease with autosomal dominant transmission. In 1998, Chen et al. identified the first mutation associated with the condition, in the *SCN5A* gene, which codes for the alpha subunit of the cardiac sodium channel.[3,4] Mutations in other genes, coding for calcium and potassium channels, were subsequently associated with BrS, reflecting its genetic heterogeneity. Mutations in *SCN5A* are the most common genotype (>70%), but are present in only 11–28% of patients.[5]

The actual prevalence of BrS is unknown. It is estimated to affect around 5/10 000 individuals, but this figure may be an underestimate due to the existence of concealed electrocardiographic forms.[6,7]

Despite its autosomal dominant pattern, BrS is 8–10 times more prevalent in males except in children, in whom the gender distribution is similar. Clinical presentation tends to be more severe in males; structural differences in ion currents and the influence of hormonal factors have been suggested as possible explanations for differences between the sexes.[8]

Although it may occur at any age, BrS is typically found in young adults, with a peak of incidence between the third and fourth decade of life.[2] One of the interesting aspects of the case presented is that symptoms appear to have been present since childhood. Although three of the eight patients in Brugada and Brugada's original description were children, knowledge of the condition at pediatric ages is limited[1] and presentation in this age-group is uncommon. Studies in Japanese populations estimate a prevalence