Brugada syndrome diagnosed after ventricular fibrillation with anamnesis of fever

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SÚHRN

Syndróm Brugadaových je genetická porucha charakterizovaná komorovými tachyarytmiami, ktoré môžu viesť k zástave srdca, synkope alebo náhlej srdcovej smrti. Tento syndróm je asociovaný s niekoľkými elektrokardiografickými obrazcami, ako sú napríklad elevácie ST v predných prekordálnych zvodoch a nekompletná blokáda pravého Tawarovho ramienka. Mnoho ľudí so syndrómom Brugadaových nemá žiadne príznaky, ak áno, môžu sa vyskytnúť kedykoľvek a niekedy môžu byť vyvolané horúčkou, alkoholom alebo dehydratáciou. Implantácia implantovateľného srdcového defibrilátora je jediná liečba, ktorá sa osvedčila ako účinná pri liečbe a prevencii náhlej srdcovej smrti u pacientov so syndrómom Brugadaových. Uvádzame zriedkavú kazuistiku syndrómu Brugadaových, ktorý sa manifestoval komorovou fibriláciou vyvolanou horúčkou.

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ABSTRACT

Brugada syndrome is a genetic disorder characterized by ventricular tachyarrhythmias that may lead to cardiac arrest, syncope or sudden cardiac death. This syndrome is associated with several electrocardiographic patterns such as ST elevations in the anterior precordial leads and incomplete right bundle branch block. Many people with Brugada syndrome do not have any symptoms if yes, they can occur at any time and are sometimes triggered by high temperature, alcohol or dehydration. The implantation of an automatic implantable cardiac defibrillator is the only treatment proven effective in treating and preventing sudden cardiac death in patients with Brugada syndrome. We present a rare case report of Brugada syndrome induced by fever which was manifested with ventricular fibrillation.

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Introduction

The syndrome affects between 1 and 30 per 10,000 people, it is 8–10 times more common in males than in females and in people from Asia aged 30–50 years.¹ It is also known as *Lai Tai* (Thailand), *Bangungot* (Philippines), and *Pokkuri* (Japan) and seems to be the most common cause of natural death in men younger than 50 years in this region.² It is a genetic disorder with alterations in the *SCN5A* gene (10–30% cases) and 300 mutations have been described.^{3,4} There could be other genes affected, which can cause a variant of Brugada syndrome, including the genes coding for alpha1- and beta2b-subunits of the L-type calcium channel (CACNA1C and CACNB2), which are thought to cause a syndrome of precordial ST elevation, sudden death, and short QT interval.^{5,6} The most common manifestations is syncope or cardiac arrest, which occur during sleep, but many patients remain asymptomatic. Sudden cardiac death can occur in family history. Approximately in 20% of patients atrial fibrillation is an associated arrhythmia.⁷ This rare syndrome is named after the Spanish cardiologists Pedro and Josep Brugada who described the condition in 1992.⁸

Case report

A 46-year-old male was hospitalized after successful cardiopulmonary resuscitation. Computed tomography angiography (CTA) and computed tomography scan (CT) of

head showed physiological findings. The medical history of the patient was without a serious illness but with an anamnesis of recent common fever and an unexplained syncope. There was no family history of arrhythmias or sudden unexplained death. The 12-lead electrocardiogram showed a sinus rhythm with non-specific changes in ST segment in chest leads. The cardiac specific markers were slightly elevated, coronarography showed no signs of coronary artery disease. Echocardiogram showed normal cardiac structures and function. 24-hour electrocardiogram Holter monitoring revealed multiple runs of asymptomatic non-sustained polymorphic ventricular tachycardia and non-specific ST elevation in chest leads which evoked concern for a Brugada-like pattern. The diagnosis of Brugada syndrome was established based on the characteristic Brugada type 2 electrocardiogram pattern (Fig. 1) in conjunction with ventricular fibrillation documented by emergency medical service. The patient was referred to cardiac centre and an implantable cardioverter defibrillator (ICD) was implanted. The elevated cardiac specific markers were elevated presumably because of the resuscitation and defibrillation. The patient is doing well and his neurological status had a good recovery.

Discussion

Each syncope should be performed with a 12-lead ECG and interpreted with an experienced physician or elec-

Fig. 1 – Electrocardiogram of a patient with Brugada syndrome

Table 1 – Diagnostic electrocardiogram patterns in Brugada syndrome			
Characteristic	Type 1	Type 2	Type 3
J wave amplitude	≥ 2 mm	≥ 2 mm	≥ 2 mm
T wave	Negative	Positive or biphasic	Positive
ST–T configuration	Cove-type	Saddleback	Saddleback
ST segment, terminal portion	Gradually descending	Elevated by ≥ 1 mm	Elevated by < 1 mm

trophysiologist. We should consider diagnostic three ECG patterns for Brugada syndrome (Table 1).^{9,10} Hypercalcemia, hyperkalemia and an acute coronary syndrome should be excluded which may have similar pattern on ECG to Brugada syndrome. A febrile state, hypokalemia, alcohol and cocaine intoxication, sodium channel blockers and heterocyclic antidepressants may unmask Brugada syndrome on ECG. Patients should be genetically tested for a mutation in *SCN5A*. Echocardiography or other examinations (MRI) should be performed to exclude arrhythmogenic right ventricular cardiomyopathy, myocardial injury or other cardiomyopathy. In some cases the intravenous administration of drugs that block sodium channels such as flecainide, procainamide, ajmaline and pilsicainide.¹¹ This may unmask the Brugada syndrome on ECG and it should be performed with continuous cardiac monitoring and in a setting equipped for resuscitation.

Conclusion

The Brugada syndrome is a channelopathy caused by an alteration in the transmembrane ion currents that together constitute the cardiac action potential. The exact mechanism underlying the ECG alterations and arrhythmogenesis in Brugada syndrome are not fully understood. Nowadays the only treatment proved effective in treating and preventing sudden death in patients with Brugada syndrome is an implantation of ICD. No pharmacological drugs or treatment have reduced the sudden cardiac death.¹²

Conflict of interest

The authors declare that there are no conflicts of interest.

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