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BRUGADA Syndrome: Risk stratification and management, About a case

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ABSTRACT:

Background: BRUGADA syndrome is a rare hereditary channelopathy characterized by cardiac arrhythmia, nocturne epilepsy with/without genitosphincterian disturbance, the diagnosis remains a challenge as patients are often young and asymptomatic. Missing or delayed diagnosis can result in severe cardiac complications such as sudden death.

Medical case: here we report the case of a 56-year-old woman, with history of epilepsy and resuscitated death two years ago, who presented with recurrent nocturne syncope and fever, An ECG performed and showed an elevation of ST segment in v1 and v2 leads. The TTE and coronarography are without abnormalities, evoking a Brugada syndrome.

Conclusion: The early diagnosis of cardiac arrhythmia in patient with Brugada syndrome could help to prevent severe complications, reduce the morbidity and mortality, and improve the outcome of patients.

KEY WORDS: BRUGADA syndrome. Arrhythmia. Sudden cardiac death. Risk stratification.

LEARNING OBJECTIVE:

Through this case, we show the essential interest of the risk stratification of sudden death in the suspected Brugada patient and we share our management of this rare challenging disease, including diagnosis steeps and treatment tools.

INTRODUCTION:

BRUGADA syndrome is defined as a rare hereditary cardiac arrhythmia, characterized by convex elevation of the ST segment in the right precordial leads, and presenting an accumulated risk of sudden cardiac death, despite the absence of structural abnormalities. The diagnosis key and definition of BS is its characteristic ECG, which may be or not present spontaneously. The risk stratification in patients with Brugada syndrome is an essential step for an accurate prognosis and appropriate therapeutic decisions.

CLINICAL CASE:

We report the case of a 56-year-old woman, without cardiovascular risk factors, without familial sudden death history, who presented with recurrent syncope occurring at night in a febrile context, associated with seizures epilepsy, motivating a first consultation in the neurology department where a neurophysiological assessment was carried out returning normal (Cerebral CT-scan, EEG, Cerebral MRI). The patient underwent an antiepipleptic treatment with clinical stabilization. Two years later, the patient presented with syncope with generalized tonic-clonic epileptic crisis, followed by cardiorespiratory arrest requiring hospitalization in the intensive care unit where she was intubated and ventilated for 48 hours then extubated, the ECG showed a regular sinus rhythm, with convex elevation of the ST segment (2mm) in V1 and V2 leads (figure 1), the TTE was performed and is without particularities as well as the coronarography and cardiac MRI.

Our patient underwent a successful implantation of a defibrillator (figure 2) as a secondary prevention in presence of the typical BS type 1 ECG pattern and history of resuscitated sudden death.

DISCUSSION:

Brugada syndrome is a rare hereditary arrhythmogenic cardiovascular channelopathy with autosomal dominant transmission, associated in 15 to 25% of cases with a mutation in the SCN5A gene which codes for a subunit of the sodium channel involved in cardiac depolarization [1]. Despite the absence of identifiable structural heart disease, this pathology predisposes to the ventricular fibrillation (VF), responsible of sudden death. It is currently defined according to the ECG features: ST segment elevation > 0.2 mV, followed by a negative T-wave, recorded on at least 1 right precordial lead: V1 or V2 on the 2nd, 3rd or 4th intercostal space. This type 1 ECG pattern can be spontaneous or induced by the administration of a sodium blocker such as ajmaline or flécaïnide [2-3].

The BS prevalence is ranging from 1 to 5/10000 inhabitants with significant geographic disparities. It is now accepted that Brugada syndrome and unexplained sudden death syndrome which is endemic in Southeast Asia correspond to the same entity [4]. The average age of diagnosis or sudden death is 40 years, (two days to over 80 years). Rhythm disturbances in this syndrome are more common in men than women (M / F = 10/1) and are favored by fever [4].

Our patient is a 56-year-old in whom syncope occurs in a context of fever.

The pathophysiological mechanism could be explained by a decrease in the rapid sodium current and an increase in the transient potassium outgoing current, resulting in a loss of the action potential dome and premature repolarization of the cell concerned. The loss of the dome of the action potential in the epicardial regions of the right ventricle leads to dispersion of repolarization on the surface of the epicardium, thus constituting the arrhythmogenic substrate in this pathology.

The mechanism triggering Ventricular fibrillation(VF) is a phase 2 reentry. Furthermore, the increase in the amplitude of phase 1 and the exaggeration of the transmural electrical gradient at the level of the action potential plateau are responsible for the accentuation of the J wave and the ST segment elevation on the transmural electrogram [5].

In addition to the cardiac location responsible for arrhythmia, channelopaties are associated with a dysfunction of other excitable tissues such as brain and muscle, thus explaining the other symptoms associated with BRUGADA syndrome. On the one hand and according to experimental models in animals, the SCN5A gene is particularly active in astrocytes, with a protein SCN5A particularly expressed in the Limbic region, which explains the recurrent epileptic activity at the slightest alteration. On the other hand, the channelopathy in the bladder muscle explains the urinary disorders found in BS. This syndrome which by definition considers the asymptomatic patients with nocturnal epilepsy and urinary incontinence [6].

In our case, the patient had generalized inaugural convulsive crisis, she underwent an antiepileptic treatment with good evolution.

The risk stratification of patients with Brugada syndrome is very important for an accurate prognosis and appropriate therapeutic decisions. Spontaneous elevation of the ST segment type 1 pattern is generally considered to be an independent risk factor for arrhythmic events and is an element of poor prognosis when associated with a positive electrophysiological stimulation [7]. Other risk factors include resuscitated sudden death, which presents the first manifestation of the disease in 25% of cases [8], male, syncope, and positive electro-physiological study.

Men have a higher risk of arrhythmic events than women, and within the male population, symptomatic patients have a significantly higher risk profile than asymptomatic patients [9].

In BS the patients with SCN5A (+) are at higher risk of arrhythmic events than SCN5A (-) patients, especially since they are symptomatic [10].

The real impact of this syndrome on sudden overall death remains uncertain, the only effective management to prevent the sudden death remains the implantation of a defibrillator, the decision to implant often remains difficult and controversial, and it is not totally safe, but can be associated with complications including the delivery of inappropriate shock, more frequently in symptomatic patients [8-11], others Less common complications have been reported such as ruptured catheter, infection, Clavian vein thrombosis, pericardial effusion and pneumothorax [8-11].

The implantation of the DAI is especially recommended in patients with a history of resuscitated sudden death, with a documented Ventricular tachycardia or VF (class I), it can be considered in patients with spontaneous ECG type 1 pattern and a history of syncope (Class IIa), medical treatment with quinidine has been proposed as a preventive treatment in patients with Brugada syndrome but there are no studies confirming its ability to reduce the risk of sudden death. Dietary and hygienic measures such as drug and excessive alcohol consumption avoidance, as well as heavy meals and early treatment of fever are recommended in patients diagnosed with BS [2].

Our patient underwent a successful implantation of a defibrillator as a secondary prevention in presence of the typical BS type 1 ECG pattern and history of resuscitated sudden death, she will be the subject of regular follow-up.

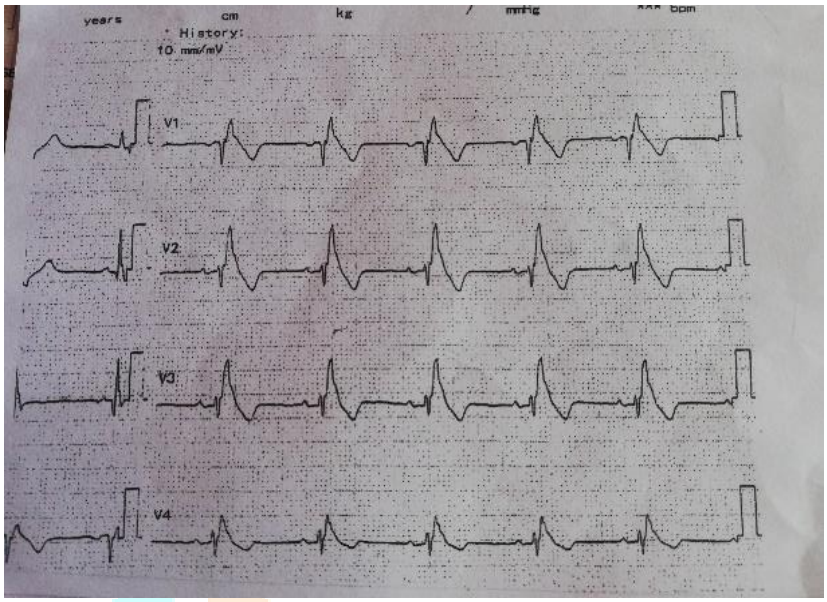

Figure 1: ST segment elevation > 0.2 mV, followed by a negative T-wave, recorded on at least 1 right precordial lead

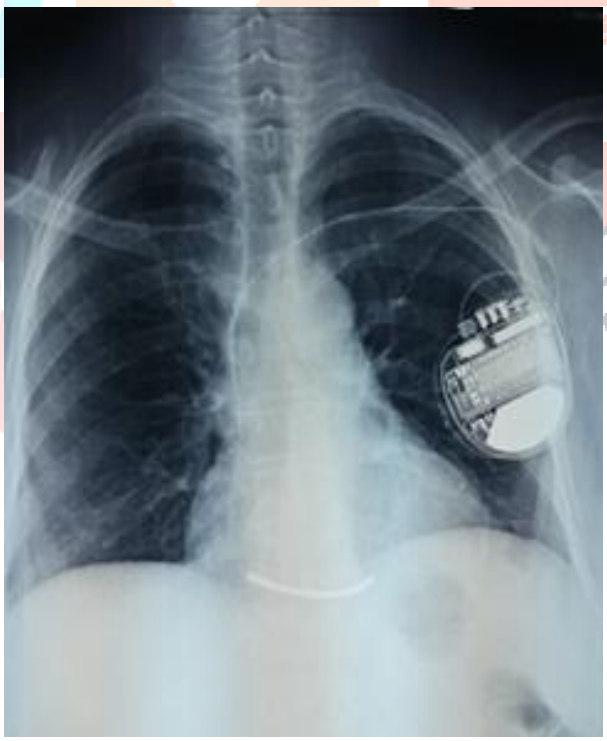

Figure 2: chest Xray after implantation of DAI

CONCLUSION

The risk stratification of the arrhythmia and sudden death in patients with Brugada syndrome is a very important step, because it makes it possible to draw up an accurate prognosis and to carry out an early and adequate management reducing the morbidity and mortality of this pathology.

Conflict of interest: there is no conflict of interest.

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Abbreviations list:

BS: Brugada syndrome

ECG: electrocardiogram

TTE: transthoracic echocardiography

MRI: magnetic resonance imagery

VF: ventricular fibrillation

VT: ventricular tachycardia

DAI: implantable automatic defibrillator

References:

[1] A. Maltret. Trouble du rythme et de la conduction chez l'enfant. J PED PUE 2016. Volume 29, Issue 4, Pages 191-210. Doi : 10.1016/S1166-4568(17)84533-2

[2] Priori SG, Blomström-Lundqvist C, Mazzanti A, Blom N, Borggrefe M, Camm J et al 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. *Eur Heart J* 2015;36(41):2793–867. doi: 10.1093/eurheartj/ehv316

[3] Priori SG, Wilde AA, Horie M, Cho Y, Behr ER, Berul C et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. *Heart Rhythm* 2013;10(12):1932–63. doi: 10.1016/j.hrthm.2013.05.014

[4] Antzelevitch C, Brugada P, Brugada J, Brugada R. Brugada syndrome: from cell to bedside. Curr Probl Cardiol. 2005 Jan;30(1):9-54. doi: 10.1016/j.cpcardiol.2004.04.005. PMID: 15627121; PMCID: PMC1475801.

[5] A Leenhardt, P Milliez, A Messali, M. Meddane, I. Denjoy, F. Extramiana. Brugada syndrome. Réanimation 2007 , volume16, issue 4 ,285–9. Doi : 10.1016/j.reaurg.2007.05.004

[6] Camacho Velásquez JL, Rivero Sanz E, Velazquez Benito A, Mauri Llerda JA. Epilepsy and Brugada syndrome. Neurologia. 2017 Jan-Feb;32(1):58-60. English, Spanish. doi: 10.1016/j.nrl.2015.03.010.

[7] Li X, Sacher F, Kusano KF, Barajas-Martinez H, Liu N, Li Y, et al . Pooled Analysis of Risk Stratification of Spontaneous Type 1 Brugada ECG: Focus on the Influence of Gender and EPS. Front Physiol. 2019 Jan 31; 9:1951. doi: 10.3389/fphys.2018.01951.

[8] McEnteggart S, Estes NAM 3rd. Prediction and Prevention of Sudden Death in the Brugada Syndrome. Am J Cardiol. 2019 Dec 1;124(11):1797-1802. doi: 10.1016/j.amjcard.2019.08.044.

[9] Yuan M, Tian C, Li X, Yang X, Wang X, Yang Y, et al. Gender Differences in Prognosis and Risk Stratification of Brugada Syndrome: A Pooled Analysis of 4,140 Patients from 24 Clinical Trials. Front Physiol. 2018 Aug 22;9:1127. doi: 10.3389/fphys.2018.01127.

[10] Yang Y, Hu D, Sacher F, Kusano KF, Li X, Barajas-Martinez H, et al. Meta-Analysis of Risk Stratification of SCN5A With Brugada Syndrome: Is SCN5A Always a Marker of Low Risk? Front Physiol. 2019 Feb 19;10:103. doi: 10.3389/fphys.2019.00103.

[11] El-Battrawy I, Roterberg G, Liebe V, Ansari U, Lang S, Zhou X, et al. Implantable cardioverter-defibrillator in Brugada syndrome: Long-term follow-up. Clin Cardiol. 2019 Oct;42(10):958-965. doi: 10.1002/clc.23247.