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CASE REPORT

Brugada Syndrome – Report of Familial Occurrence Diagnosed in the Emergency Department

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ABSTRACT

Introduction: Brugada syndrome represents the clinical manifestation of a rare disease with genetic etiology. The syndrome is characterized by ventricular dysrhythmias associated with syncope or sudden cardiac death in the lack of any structural cardiac disease. The diagnosis of Brugada syndrome is established if a type 1 electrocardiographic (ECG) pattern of ST-segment and QRS morphology is present, in association with certain clinical manifestations and/or familial history. **Case presentation:** A 31-year-old male patient, without any medical history, presented in the emergency department (ED) of a clinical center. His only complaints consisted in palpitations, chest discomfort, and emotional stress related to the recent death of his wife. Earlier on the same day, his wife, a 25-year-old female was brought via emergency medical services (EMS) to the ED after presenting ventricular fibrillation. The female patient presented a long term history of chest pain and one year prior to this episode she presented idiopathic ventricular fibrillation, for which she had undergone implantation of an automated cardioverter defibrillator. As the couple were cousins, the EMS specialist suspected the presence of a familial cardiac disorder. The electrocardiogram of the male patient revealed a coved-type ST-segment elevation of 4 mm in leads V1–V3 compatible with type 1 Brugada syndrome. **Conclusion:** In case of Brugada syndrome, a genetic disorder associated with increased risk of SCD, the patient's first-degree relatives should be investigated as well, in order to identify the presence of the syndrome and to prevent SCD. As the sole established effective therapeutic measure for patients diagnosed with Brugada syndrome, ICD implantation should be considered in order to decrease the risk of syncope and SCD. This case is particular because a rare disease with familial etiology was identified in both husband and wife, who were cousins.

Keywords: Brugada syndrome, syncope, sudden death

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INTRODUCTION

Brugada syndrome represents the clinical manifestation of a sporadic disease with genetic etiology, characterized by ventricular dysrhythmias associated with syncope or sudden cardiac death in the lack of any structural heart disorders. The condition is caused by a genetic mutation of the sCn5a protein sequence that encrypts the alpha sub-

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TABLE 1. Electrocardiographic characteristics associated with three types of Brugada syndrome

	Type 1	Type 2	Type 3
ST elevation	Coved or saddle-back or both	Saddle-back pattern	Coved type
J-point elevation	Less than 2 mm or 0.2 mV	At least 2 mm or 0.2 mV	At least 2 mm or 0.2 mV
ST segment	Less than 1 mm elevation	At least 1 mm elevation	Gradually descending
T-wave	Positive	Positive or biphasic	Negative

unit of the cardiac sodium channel and is transmitted in an autosomal dominant pattern. Brugada syndrome is believed to trigger sudden cardiac death (SCD) in otherwise healthy young adults during sleep in Southeast Asia and Japan.[1–7]

Three ECG patterns of Brugada syndrome have been described in the literature. Type 1 features a prominent coved ST-segment elevation displaying J-point elevation or ST-segment elevation ≥2 mm (0.2 mV) followed by a negative T-wave, with little or no isoelectric separation. Similarly, type 2 is also characterized by prominent ST-segment elevation, but the J-point elevation (2 mm) gives rise to progressively descending ST-segment elevation, and the T-wave is positive or biphasic, leading to a saddle-back appearance. Type 3 features a right precordial ST-segment elevation of <1 mm, of saddle back type, coved type, or both (Table 1).[4]

Any patient with near sudden arrhythmic death and a Brugada pattern ECG requires hospital admission in order to evaluate the indication for intra-cardiac cardioverter-defibrillator (ICD) implantation. According to published data, even individuals with no history of cardiac arrest but an ECG pattern of Brugada syndrome are at an increased risk of SCD, presenting a 2-year fatality rate of 8%.[8]

CASE PRESENTATION

A 31-year-old male patient, without any significant medical history, presented to the emergency department (ED). His only complaints consisted in palpitations, chest discomfort, and emotional stress related to the recent death of his wife.

Earlier on the same day, his wife, a 25-year-old female was brought via emergency medical services (EMS) to the ED after presenting ventricular fibrillation (VF). Unfortunately, resuscitation procedures were unsuccessful, and the female patient had deceased. The female patient presented a long history of chest pain, and one year prior to this episode she presented idiopathic VF, for which she had undergone implantation of an automated cardioverter defibrillator.

Additional interviewing revealed that the couple were consanguineous (cousin marriage). Thus, the EMS specialist suspected the presence of a familial cardiac disorder. The electrocardiogram of the male patient displayed a coved-type ST segment elevation of 4 mm in leads V1–V3, a pattern highly suggestive for type 1 Brugada syndrome (Figure 1).

Laboratory investigations of serum cardiac troponin-I, CK-MB, and serum potassium were within normal levels,

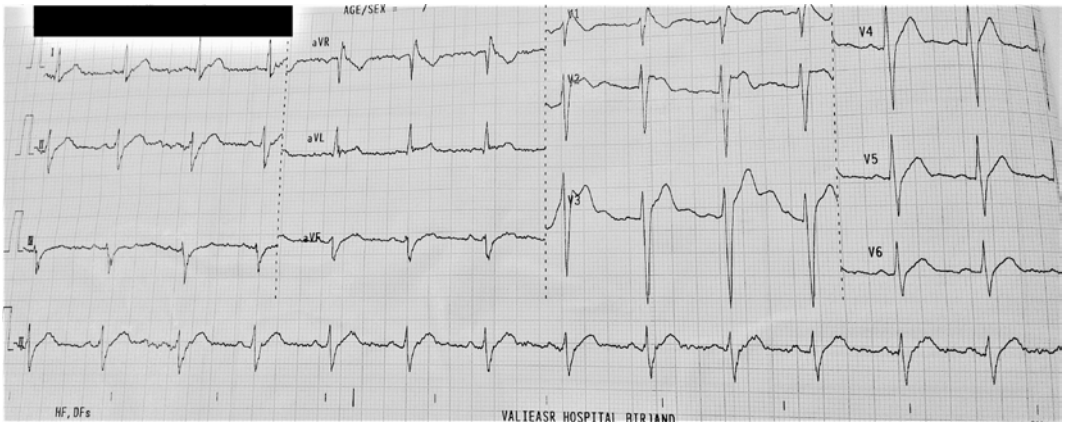

FIGURE 1. Twelve-lead electrocardiogram obtained from the male patient showing ST-segment elevation >2 mm at its peak followed by a negative T-wave (Brugada type 1)

and there was no sign of inflammation. The diagnosis of type 1 Brugada syndrome was confirmed based on ECG findings associated with a familial history of SCD at young age (a cousin and an uncle).

The patient was considered at high risk for SCD, as he presented a pre-syncopal condition associated with palpitations. Hence, he was admitted to the Cardiology Care Unit for implantation of an ICD. He was also asked to inform his first-degree relatives about the probability of the disease running within the family and the importance of medical investigations. The publication of this case was agreed by the patient and the clinical institution.

DISCUSSION

It is well known that patients with an ECG pattern highly suggestive for Brugada syndrome and no history of cardiac arrest present an increased risk for SCD, with a 2-year fatality rate of approximately 8%.[8] At the same time, a 2-year follow-up study of patients with Brugada syndrome indicated that a previous history of syncope, while having a spontaneously abnormal ECG and inducible sustained arrhythmias during programmed electric stimulation, has the highest incidence (27.2%) of SCD.[8]

Because of the genetic origin of the disease and the risk of SCD, family members (especially first-degree relatives) should be investigated through ECG and careful anamnesis focused on the history of symptoms. In our case, based on the consanguinity marriage condition and familial history of SCD at young age, the wife of the patient was also highly suspected for Brugada syndrome.

The particularity of this case is that a rare disease with familial etiology was identified in both husband and wife, who were cousins. Important to note is that the woman was diagnosed with a potentially lethal ventricular arrhythmia and received an ICD implantation in the previous year.

According to the second Consensus Conference of the American and European heart rhythm societies, the diagnosis of Brugada syndrome may be established once a type 1 ECG pattern is observed along with one criteria from the following: VF, polymorphic ventricular tachycardia (VT), a familial history of SCD before 45 years of age, coved-type ST-segment elevation in relatives, inducibility of VT with programmed electrical stimulation, syncope, or nocturnal agonal respiration.[6] In the present case report, the patient fulfils at least three of the mentioned criteria.

Literature data recommends the implementation of an ICD as the sole established effective therapeutic method for patients diagnosed with Brugada syndrome; other suggested therapies, such as ablation or cryosurgery and pacemaker implantation, are yet a matter of controversy.[6]

CONCLUSION

As the single established effective treatment for patients diagnosed with Brugada syndrome, the implantation of an ICD should be considered in order to decrease the risk of syncope and SCD. In case of Brugada syndrome, a genetic disease associated with increased risk of SCD, the patient's first-degree relatives should be investigated as well in order to identify the presence of the syndrome and to prevent SCD.

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CONFLICT OF INTEREST

Nothing to declare.

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