Aborted sudden nocturnal cardiac death in a young man with structurally normal heart

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ABSTRACT

Sudden cardiac death is a devastating event, particularly when it occurs to young, otherwise healthy individuals. We report here a young Burmese male who survived sudden cardiac death with structurally normal heart. His electrocardiogram revealed features consistent with Brugada syndrome. He was referred for intra-cardiac defibrillator implantation. In this article, in addition to the case presentation, a review of Brugada syndrome medical literature is also presented.

Saudi Med J 2005; Vol. 26 (2): 323-325

Sudden unexpected death in the young is a cumbersome event that captures the mind of the relatives as well as healthcare givers. Victims of sudden cardiac death, in the vast majority of cases, are due to ventricular fibrillation (VF) and are associated with structural heart disease, particularly coronary artery disease (CAD). Sudden cardiac death (SCD) in the normal heart is an uncommon occurrence accounting for 5% of cases in an autopsy study.[1] It has been shown by another study that nearly 10-20% of patients dying suddenly or resuscitated from VF do not have demonstrable heart disease. These people are often young and tragically in some cases, sudden death is the first and only clinical event.[2] We report here a case of a young Burmese male who survived SCD, VF, and polymorphic ventricular tachycardia (VT) with apparently normal heart.

Case Report. A 28-year-old Burmese male laborer was found moaning in his bed during sleep by his room-mates. He was brought to the emergency department (ED) of Riyadh Medical Complex (RMC) when they could not wake him up. In the ED, he was found incoherent and confused. He was attached to an ECG monitor, which revealed VF, so he was immediately DC converted by 200 J/sec. On questioning, he gave no history of chest pain or previous similar episode, and he was not a known drug addict. There was no family history of sudden cardiac death or premature coronary heart disease (CHD). He had no known risk factors for CHD. His examination revealed an irregular heart rate of 72/min with a blood pressure of 90/60 mm Hg. He was tachypneic with a respiratory rate of 20/min, and he was afebrile. The rest of his systemic examination was unremarkable. He was drowsy, but no abnormal neurological findings were detected. Baseline ECG carried out in the ED showed an ST segment elevation in V1-V3, with a partial right bundle branch block (RBBB) and normally corrected QT (QTc) interval (**Figure 1**); thus, he was admitted to the RMC cardiac care unit (CCU), for further management. Upon arrival to the CCU, he had an episode of VT, which needed DC shock of 200 J/sec for conversion (**Figure 2**). He did not

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Received 17th July 2004. Accepted for publication in final form 24th October 2004.

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revert to sinus rhythm but was found to be in slow atrial fibrillation (AF) with the same ECG changes as stated above (**Figure 3)**. Investigations revealed a normal blood count with a normal platelet count and a normal coagulation profile. Urea, creatinine and electrolytes including magnesium and calcium were all normal as were his liver function tests. Serial creatinine kinase, creatinine kinase myocardial band, lactate dehydrogenase and troponin levels were as shown in **Table 1**. Serial ECG did not reveal any evolution of myocardial infarction. His chest x-ray was reported to be normal. Echocardiography revealed normal left ventricle (LV) in dimensions and function with ejection fraction of 65%. Regional LV wall motion was normal, with no LV thrombus seen. Aortic, mitral, tricuspid, and pulmonic valves were structurally as well as functionally normal. There was no pericardial effusion, and Doppler studies were normal. He was started on aspirin 100 mg, atenolol 50 mg as well as amiodarone 300 mg intravenous (IV) bolus, followed by maintenance IV infusion of 900 mg over 24 hours. He remained stable and had no pain except at the site of DC. During his stay in the CCU he had no further episodes of VF, but continued to have paroxysmal AF with a controlled ventricular rate, therefore, he was started on anticoagulation with intravenous heparin (according to his body weight) 5000 units bolus followed by 1000 units per hour continuous infusion to maintain his activated partial thromboplastin time 2 x control. With the clinical history of nocturnal moaning (nocturnal agonal respiration), VF on presentation, AF and typical ECG changes in V1-3 with pseudo-RBBB pattern and ST elevation of a saddle-shape, along with an Asian origin of the patient, a diagnosis of Brugada syndrome (BS) was made and patient was advised on further management in a tertiary care cardiac center for intra-cardiac defibrillator (ICD) implantation.

Discussion. Brugada syndrome is a primary electrical disease resulting in abnormal electrophysiological activity in the right ventricular myocardium. Recent genetic data linking Brugada syndrome to an ion channel gene mutation (SCN5A) provides further support for this hypothesis.[3,4] It is an autosomal dominant disease with varied

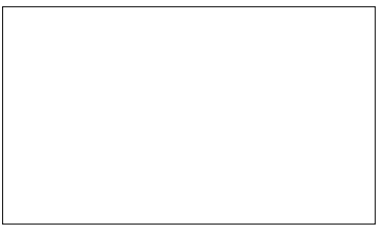

Figure 1 - Shows partial right bundle branch block (RBBB) with ST elevation in V1-V3, normal QTc (corrected QT) interval with no S wave in lead one or left chest leads typical of Brugada syndrome.

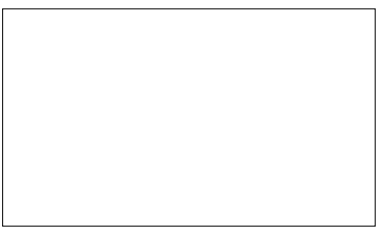

Figure 2 - Shows atrial fibrillation, with partial RBBB with ST elevation in V1-V3, normal QTc interval with no S wave in lead one or left chest leads typical of Brugada syndrome.

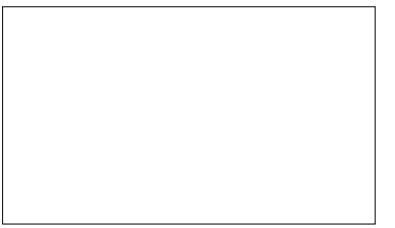

Figure 3 - Shows ventricular bigeminy and an episode of polymorphous ventricular tachycardia initiated by a premature ventricular contraction (PVC) with a short coupling interval.

Table 1 - Laboratory findings in conventional units (CU).

Test	Upon arrival	6 hours from arrival	12 hours from arrival	18 hours from arrival	24 hours from arrival	72 hours from arrival
EKG	VF	A Fib	SR	A Fib	A Fib	SR
CPK	181	147	290	337	478	154
CK-MB	0	2	5	4	6	0
LDH	266	451	228	800	790	650
Troponin-I	0.01	0.03	0.03	-	-	0.01

VF - ventricular fibrillation, A Fib - atrial fibrillation, SR - sinus rhythm, CPK - creatine phosphokinase, CK-MB - creatine kinase myocardial band, LDH - lactate dehydrogenase, EKG - electrocardiogram

expression. The diagnostic criteria, as proposed in a consensus report from the study group on the molecular basis of arrhythmias of the European Society of Cardiology, are appearance of ST segment elevation (coved type) in more than one right precordial lead (V1-V3) in the presence or absence of a sodium channel blocker, and one of the following; a) documented VF, b) self-terminating polymorphic tachycardia, c) family history of SCD at less than 45 years, d) type 1 ST segment elevation in family members, e) electrophysiological inducibility, f) syncope, and g) nocturnal agonal respiration.[5] The AF, which was present in our patient, is often described in Brugada syndrome.[6] Approximately 20% of SCD of patients with structurally normal heart is believed to be due to Brugada syndrome.[7] Moreover, previous studies have shown that asymptomatic subjects with this syndrome have an 8-38% annual risk of life-threatening ventricular arrhythmias and SCD.[8,9] Clearly, the recognition of the electrocardiographic features of BS is an easy task to an expert cardiologist, nonetheless its rare prevalence that has been reported to be in the range of 0.07-0.61% may result in overlooking the correct diagnosis.[10-13]

Our patient may have been having self-terminating episodes of polymorphous VT as confirmed one-time in CCU by arrhythmia monitoring memory. A prolonged QTc interval in right precordial leads has been mentioned as an additional diagnostic criterion.[3] The treatment of this condition includes implantation of an ICD in a selected group of patients, but amiodarone or beta-blockers have not been found helpful in preventing sudden death. One needs to be aware of Brugada syndrome as Saudi Arabia has a high expatriate population especially from Asia, namely, Burma and Bangladesh.

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