Brugada Syndrome, A case Report

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Introduction:
Brugada syndrome is a clinical and electrocardiographic diagnosis based on syncopal sudden death episodes in patients with a structurally normal heart and characteristic ECG pattern composed of right bundle branch block (RBBB) and a specific shape ST-segment elevation in V1 to V3.

The first report on this syndrome was published in 1992, although some reports on a similar condition has been reported since 1989, since 1992 there has been an exponential increase in the number of patients recognized all over the world. Its incidence and prevalence are difficult to estimate, however asymptomatic subjects with Brugada type ST-segment shift were present at a rate of 0.14% in the general Japanese population.

Key words: Brugada Syndrome, Ventricular arrhythmia, sudden cardiac death.

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Patient characteristics:
A 15 years old Iraqi boy presented with history of recurrent syncopal attacks of seven years duration. He gave a strong family history of sudden death; two of his brothers died suddenly without previous complaints, one at the age of 16 years and the other at the age of 12 years. No one of his remaining family (two sisters, three brothers and his father) is symptomatic. His mother died due to obstetric problem.

Thorough examination of the patient was essentially normal. His ECG showed sinus rhythm, PR interval of 230 msec., RBBB pattern with a QRS width of 84 msec. and a specific Brugada type ST-segment elevation in V1-V3 (Figure 1). Holter monitor showed a varying ST segment shift over the 24 hours of the recording (Figure 2).

Echocardiogram was normal. EEG, complete blood count, blood biochemistry, serum electrolyte and chest X-ray are all within normal.

A diagnosis of Brugada syndrome was made and an Implantable Cardioverter Defibrillator (ICD) Ventack Prizm II VR, Guidant, St Paul, MN. With a defibrillator lead: Endotak reliance from Guidant was implanted.

An ECG screening for the rest of the family showed no similar abnormalities in any other member.

Discussion:
Brugada syndrome is usually identified as sporadic cases. However 50% of individuals who have this syndrome have a family history of the disease.

Genetic mutations composed of abnormalities in SCN 5 A gene results in abnormalities in the cardiac sodium channels and those patients are predisposed to rapid polymorphic ventricular tachycardia (VT) or ventricular fibrillation (VF).

In our patient the 12 lead ECG (Figure 1) showed a prolonged PR interval, which has been reported in 18% of patients with Brugada syndrome.
5. It also showed a RBBB pattern with QRS width of 84msc. and a specific shaped ST-segment elevation in V1-V3.

The Holter study showed a long short cycle length, which could precede the occurrence of polymorphic VT in few cases of Brugada syndrome. The diurnal variation in the degree of ST-segment elevation in the same lead has been noticed in our patient 1.

We proceeded to ICD implantation directly without EPS because a negative EPS will not change our decision for ICD implantation in this high risk patient for sudden cardiac death (SCD) as asymptomatic Brugada patients require protective treatment from SCD even when the VT are not inducible during EPS 6.

In this type of symptomatic patient with syncope and classical ECG pattern the risk of new arrhythmic event is estimated to be 19% within 54 months 7.

The rest of the family of this patient were asymptomatic and the 12 lead ECG for all of them showed no abnormal ECG pattern, procainamide challenge 8 for the family members were done and no abnormality appeared in the ECG after challenge.

In this syndrome there are few predictors of events occurrence:
1. A spontaneously abnormal ECG is a marker of possible sudden arrhythmic death in comparison to those who had abnormal ECG after drug challenge 8.
2. Male sex is considered as a risk factor for SCD as compared to female 1.
3. The inducibility of sustained VT during EPS, which may be the strongest marker of prognosis 1.
4. Symptomatic patients have unacceptably high rate of arrhythmic events which are more frequent in patients who present with aborted SCD compared to patients who present with repetitive syncopal episodes 9.

No effective antiarrhythmic drug is available 1. 9. ICD is indicated in symptomatic patients, however the group of asymptomatic individuals in whom the abnormal ECG was recognized only after drug challenge and they have very low event rate during follow up warrants no treatment 9.

As far as we know this is the first case of Brugada syndrome diagnosed in Iraq, we hope that this case report will make physicians and cardiologist oriented about this condition.

References:


FIGURE 1. 12 LEAD ECG SHOWING TYPICAL COVED ST ELEVATION IN V1,V2,V3.
FIGURE 2. HOLTER SHOWING SHORT-LONG CYCLE LENGTHS.