Case Report

Disappearing Brugada Pattern : A Rare Presentation

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ABSTRACT

A 60 years old lady with no significant past medical history presented to our hospital with cellulitis right lower limb. She was in sepsis with multi organ dysfunction. On work up she had a WBC count of 57,300/cmm, with 95% polymorphs. Her hepatic and renal parameters were deranged. Her ECG was suggestive of Type I Brugada syndrome, Echocardiography was within normal limits. The patient was treated with higher antibiotics (Piperacillin / Tazobactum with Linezolid), chymoral forte and Mg sulphate dressing. Her fever and systemic dysfunction gradually improved. Since she had no cardiac symptoms, the ECG was done on alternate days. The Brugada manifestations gradually reduced and disappeared on 8th day. She was discharged on 14th day with oral antibiotics and dressing of right lower limb with normal ECG. She is kept in close follow up for fever induced Brugada syndrome.

Introduction :

Brugada syndrome is a distinct arrhythmogenic disorder widely recognized as a sudden cause of death in the young. It is identified by a classical ST segment elevation on electrocardiogram (ECG) that may be provoked in the context of a fever or vagal stimulation. The pathophysiology and genetic basis have been elucidated as an abnormality in ion channels. The management is centered around device therapy with the implantable cardioverter defibrillator (ICD), though pharmacological treatments are being actively pursued.

Case Report :

A 60 years old lady with no significant past medical history presented in April 2019 with chief complaints of fever with chills and body ache since 7 days. She had trauma to right foot followed by swelling, redness and fever since 7 days. On examination she had tachycardia, fever (temperature of 101^0 F), BP = 110/70 mm Hg, respiratory rate of 30/min, SPO_2 of 98% at room air and icterus. Local examination revealed right lower limb cellulitis below knee, systemic examination of CNS, RS, CVS were within normal limits and abdominal examination revealed hepatosplenomegaly. Her right lower limb had filariasis since 30 years. On work up she had a WBC count of 57,300/cmm with 95% polymorphs. Her hepatic and renal parameters were deranged. Her ECG was suggestive of Type I Brugada syndrome ***(Fig. 1)*** and echocardiography was within normal limits. The working diagnosis was that of right lower limb cellulitis with sepsis with multiorgan dysfunction with type I Brugada syndrome. The patient was treated with higher antibiotics (Piperacillin Tazobactum with Linezolid), chymoral forte and Mg sulphate dressing. Her fever and systemic dysfunction gradually improved. Since she had no cardiac symptoms ECG was performed on alternate days in which Brugada manifestations gradually reduced ***(Fig. 2,3)*** and disappeared on 11th day ***(Fig. 4,5).*** She was discharged on 14th day with oral antibiotics and dressing of right lower limb with normal ECG.

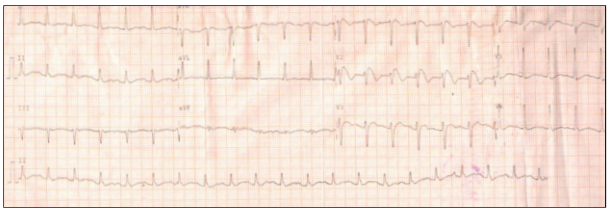

Fig. 1 : **ECG showing Type I Brugada Pattern in V1, V2, V3 (Day 1)**

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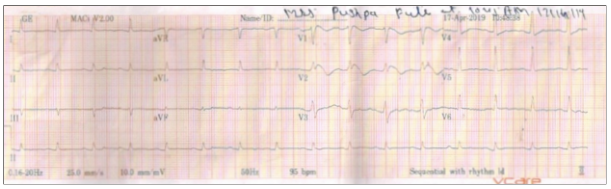

Fig. 2 : **ECG showing Type I Brugada Pattern in V1, V2 (Day 3)**

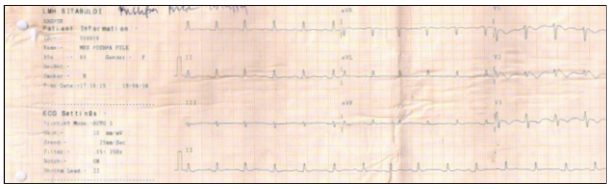

Fig. 3 : **ECG showing Type I Brugada Pattern in V1, V2, V3 (Day 5)**

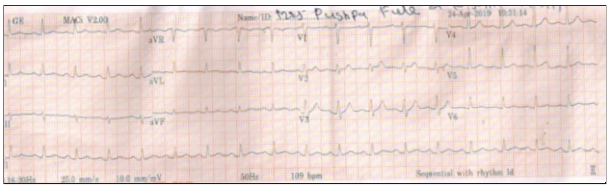

Fig. 4 : **ECG showing sinus tachycardia without any features of Brugada (Day 8)**

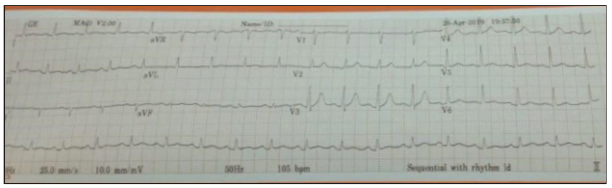

Fig. 5 : **ECG showing sinus tachycardia with no features of Brugada (Day 11)**

Discussion :

Brugada syndrome is a genetic disease presenting with a characteristic electrocardiogram (ECG) and a tendency to develop malignant polymorphic ventricular arrhythmias that may lead to syncope or cardiac arrest. The characteristic ECG, which includes a coved-type ST-segment elevation > 2 mm in the right precordial leads, is termed "type I" Brugada ECG pattern and is required to make the diagnosis of Brugada syndrome.[1] Too often, however, the ECGs of patients with Brugada syndrome have lesser degrees or different contours of ST-segment elevation ("saddleback" rather than "coved"), which are termed type II or type III Brugada pattern. These ECG patterns are suggestive, but not diagnostic, of this disease (as shown in ***Fig. 6***). Moreover, in a given patient with Brugada syndrome there are marked day-to-day changes in ECG morphology. In a large series of patients with documented Brugada syndrome who underwent repeated ECG recording over the years,[2] only every third ECG was diagnostic (i.e, showed the type I pattern) and every third ECG was completely normal, making the diagnosis of Brugada syndrome challenging.

The diagnostic criteria of Brugada syndrome are described in the ***Table No. 1.***[3]

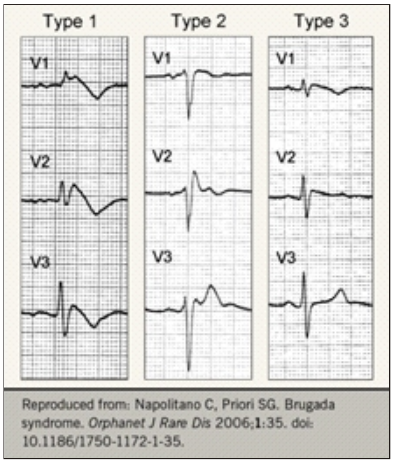

Fig. 6 : **ECG pattern of various types of Brugada syndrome.**

Coved ST-segment elevation (type 1) ≥2 mm in multiple precordial leads (V1–V3) **either** spontaneously **or** by following sodium blockade

And one of the below:

Ventricular arrhythmias

- Documented ventricular fibrillation
- Polymorphic ventricular tachycardia
- Inducibility of ventricular arrhythmias on electrophysiology studies

Family history

- Family history of sudden death before the age of 45 years
- Presence of coved-type electrocardiogram (ECG) in a family member

Symptoms suggestive of arrhythmia

- Unexplained syncope
- Nocturnal agonal respiration

Table No. 1 : **Diagnostic criteria of Brugada syndrome**

Fever-induced Brugada is the term used to describe the aggravation of clinical and/or ECG characteristics of this syndrome during febrile states in susceptible individuals. The prevalence of type I Brugada ECG in patients with fever is 20 times higher than in afebrile patients, emphasizing the potency of fever in uncovering this ECG phenomenon. These findings also may imply that the number of asymptomatic Brugada patients diagnosed today is only the tip of the iceberg, as many more would have been discovered if their ECGs were recorded during febrile illnesses. All patients with fever-induced type I Brugada pattern were asymptomatic and remained so during 30 months of follow-up.[4] In 1999, Dumaine et al[5] reported the results of functional expression studies of a genetic mutation (T1620M) identified in patients with Brugada syndrome, showing that the loss of function of sodium channel current was accentuated at higher temperatures. The authors suggested the possibility that a febrile state may unmask the Brugada syndrome. Indeed, fever was the precipitating factor of arrhythmias in 18% of patients presenting with cardiac arrest in a large series of patients with symptomatic Brugada syndrome.[6] Thus, the concept of "fever-induced Brugada syndrome" is well accepted. In our case, patient had ECG features of Type I Brugada syndrome on day 1 when she had fever with sepsis. As gradually she responded to antiobiotics, fever subsided and so the features of Brugada. Hence our case is one of the rare type of fever induced Type I Brugada syndrome.

Treatment for Brugada syndrome is essentially aimed at correcting potentially life-threatening ventricular arrhythmias as they occur. There is no way of preventing arrhythmias from occurring in the first place. The single most effective and proven treatment is the ICD. Patients are electrically shocked, i.e. cardioverted into a normal sinus rhythm as a life-saving measure. These devices are the mainstay of therapy and constantly monitor the heart rhythm. Symptomatic Brugada patients should always be offered an ICD. This recommendation was borne out of a consensus conference held in September 2003.[7] Electrophysiology studies are useful in risk stratifying asymptomatic patients. An ICD should then be implanted where VF is inducible in the context of an incidental type-1 ECG. Alternatively, an ICD is appropriate where a sodium-channel blocker-induced ECG is precipitated in a subject who has a positive family history of sudden death. Those patients who have no family history and are symptom free but develop a type-1 ECG only after drug challenge should be kept under tight surveillance.[7] The flowchart in ***Fig. 7*** serves as a useful algorithm for management.[3] In our case since patient did not have any cardiac symptoms and her ECG at the time of discharge was normal, she is kept in close follow up.

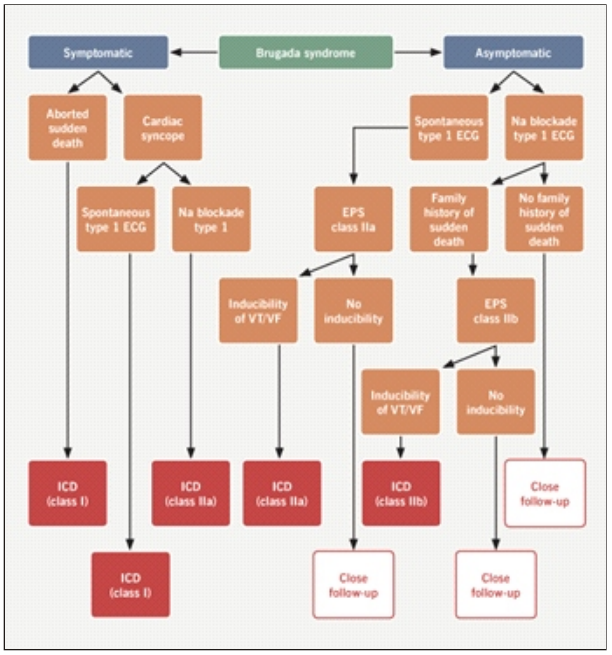

Figure 7 : **Flow chart for management of Brugada syndrome**

Conclusion :

Brugada syndrome is an inherited sodium channelopathy of cardiac myocytes. It is a major cause of sudden death in young people who have structurally normal hearts. Electrocardiography (ECG) is the investigation from which Bruagada patients are identified, though clinical features and modulating factors have a role to play. The implantable cardioverter-defibrillator is the single most effective and proven treatment in controlling

life-threatening ventricular arrhythmias. Fever is an important trigger and the onset of pyrexia (such as that seen in our patient) may unmask abnormal ECG morphology and raise suspicion of Brugada syndrome.

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