

BRUGADA SYNDROME

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Introduction

Brugada syndrome describes the syndrome of sudden cardiac death in the setting of the following electrocardiography findings: Right Bundle Branch Block (RBBB) pattern with ST segment elevation in the right precordial leads^{1,2} (Fig 1). The RBBB may be incomplete while the ST segment elevation is minimal. The ECG findings need not be constant in all patients with Brugada syndrome and can have variation in individual patient itself³. Patients should be promptly referred for electrophysiological testing and treatment. Rapid referral and placement of an implantable cardioverter defibrillator (ICD) is associated with an excellent prognosis, whereas failure to diagnose this condition is associated with a high risk for sudden death.

Case report

Mr. S, 50-year-old male was advised for an ECG as a part of routine health check-up and it revealed features consistent with Brugada syndrome (Fig.2). On retrospective counseling he was found to have had syncopial attacks and paroxysmal palpitations. His chest x-ray and echocardiograph showed normal study. His father had a sudden death at the age of 48 and cause was not ascertained. Patient has three daughters and one son, all are asymptomatic. Screening, baseline ECG was taken for all and was normal.

A diagnosis of Brugada syndrome was made based on typical ECG pattern with history of syncope and self-terminating ventricular arrhythmia. The patient was referred to a higher center for an electrophysiological testing and to have an ICD implanted. Due to financial constraints patient could not afford it. His children were advised to undergo ajmaline challenge to unmask the ECG findings.

Discussion

Brugada syndrome is a familial disease, which shows an autosomal dominant mode of transmission, with male preponderance⁴. It is the most common cause of sudden cardiac death in individuals younger than 50 years in South East Asia with no underlying cardiac disease, with an estimated incidence of 5-66/10,000 subjects⁵. Higher prevalence in particular areas can be explained by its genetic nature.

Several mutations linked to this syndrome affecting the gene SCN5A that encodes for the cardiac sodium channel have been described, along with affects of some modifier genes⁶. Autopsy of patients had no structural abnormality in the heart unlike arrhythmogenic right ventricular dysplasia⁷.

Syncope or aborted sudden cardiac death is the only symptom in patients with Brugada syndrome¹. ECG abnormality is the hallmark of Brugada syndrome². Clinical spectrum varies from asymptomatic patients with normal ECG or typical ECG to symptomatic patients with or without typical ECG changes. Episodes of fast polymorphic VT-VF if terminated spontaneously the patient presents with syncopial attacks. When the episodes are sustained full-blown cardiac arrest and eventually sudden death occur⁸.

The ECG findings can be unmasked by administration of intravenous ajmaline (1mg./kg. In 5 mts.), flecainide (2mg./kg. In 10mts.) Or procainamide (10mg./kg. In 10 mts.). Because of potential induction of VT it should be performed under continuous medical surveillance with advanced life support facilities. Due to prognostic importance

all patients with suspected disease and family members should presently undergo drug testing⁹.

This syndrome has very poor prognosis when left untreated. Follow-up studies show development of new episode of polymorphic VT within 2years in 40% symptomatic patients, and also in 40% of asymptomatic patients with typical ECG patterns¹⁰.

As the anti-arrhythmic drugs do not protect against sudden cardiac death the only available treatment is the implantable cardioverter defibrillator (ICD). It effectively recognizes and treats the ventricular arrhythmias. 10-year follow-up has been associated with 0% mortality¹¹. All patients with Brugada syndrome except those asymptomatic patients who are not inducible for arrhythmias should be treated with ICD.

References

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Fig 1. Typical ECG findings of Brugada Syndrome

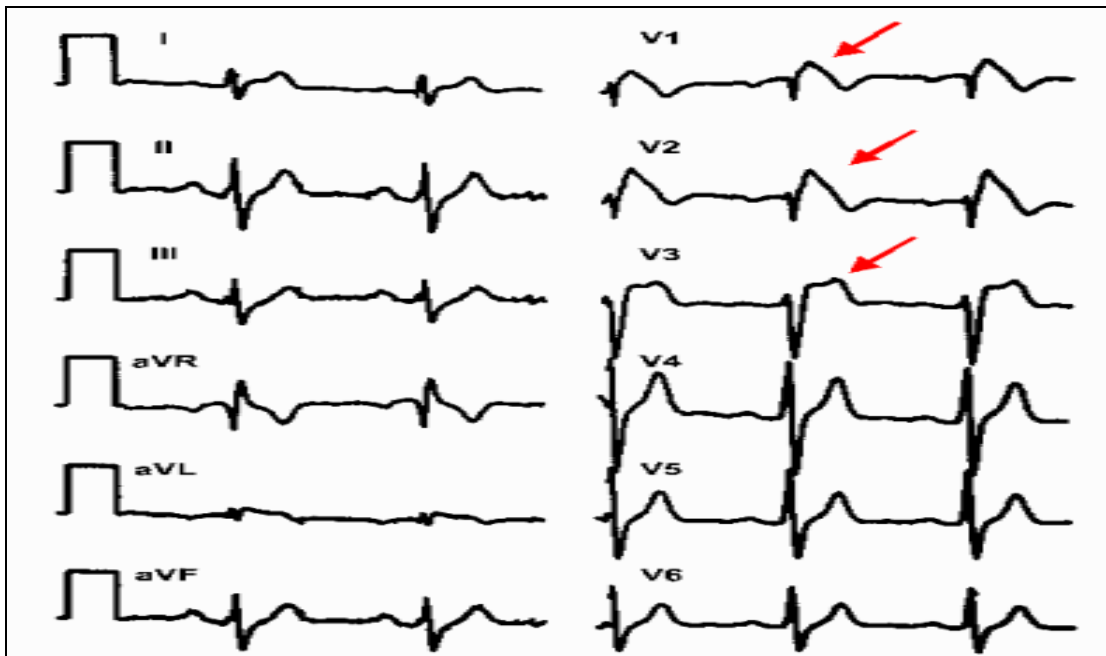


Fig.2. ECG of Mr S, showing features of Brugada syndrome

