Tanzania Journal of Health Research
Volume 17, Number 3, July 2015
Doi: http://dx.doi.org/10.4314/thrb.v17i3.9

Brugada-like syndrome presenting with monomorphic ventricular tachycardia and Brugada-type electrocardiogram unmasked by fever in an infant: a case report

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Abstract
Brugada syndrome is an autosomal-dominant arythmogenic genetic disorder associated with mutation in the SCN5A gene. We report a case of 3-month-old Tanzanian male who was admitted at Muhimbili National Hospital in Dar es Salaam, Tanzania with severe pneumonia, high fever and monomorphic ventricular tachycardia. The patient was treated with intravenous Amiodarone. In addition, oxygen, parenteral antibiotics, antipyretics and intravenous fluids were also given. About 2 hours and 20 minute later the child stabilized. An ECG obtained shortly after termination of ventricular tachycardia showed the typical J-point and coved ST elevation typical of Brugada type I pattern. To the best of our knowledge, this is the first paediatric case with Brugada-type ECG to be reported in Sub-Saharan Africa. This case emphasizes the need to increase awareness among clinicians of clinical and genetic arythmogenic disorders. Multiple ECGs during and after febrile disorders should be performed in children who exhibit extreme tachycardia or signs of cardiac failure.

Keywords: Brugada, infant, ventricular tachycardia, electrocardiogram, Tanzania

Introduction

Brugada syndrome (BrS) is an autosomal-dominant arythmogenic genetic disorder associated with mutation in the SCN5A gene, which encodes the cardiac sodium-channel (Chen *et al*., 1998). The diagnosis relies on clinical criteria in the presence of Brugada-type Electrocardiogram (ECG) (Bayés de Luna *et al*., 2012). This ECG pattern has been predominantly reported among Asians and white populations with only a few cases reported in individuals of African origin, and none of among children. We report a case of 3-month-old Tanzanian male who was admitted at Muhimbili National Hospital in Dar es Salaam with severe pneumonia and high fever together with monomorphic ventricular tachycardia (VT) on the ECG. Following the drug cardiovesion, ECG converted to type I Brugada pattern which later disappeared after the fever had subsided.

Case report

A 3 month-old Tanzanian male was admitted to our hospital with the diagnosis of severe bronchopneumonia with fever, difficulty in breathing and inability to feed. His past medical history

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Doi: http://dx.doi.org/10.4314/thrb.v17i3.X

was unremarkable and there was no known exposure to tuberculosis. There was no family history of heart disease of any type or sudden cardiac death. The initial assessment revealed a respiratory rate of 58 cycles per minute, temperature of 39.8°C, heart rate of 238 beats per minute, haemoglobin oxygen saturation of 72% in room air, and blood glucose of 6.2mmol/l. Serum electrolytes (Na^+, K^+, Mg and Ca^+) and Thyroid function tests were all normal. An initial ECG showed monomorphic ventricular tachycardia at a rate of 233 beats per minute (Figure 1).

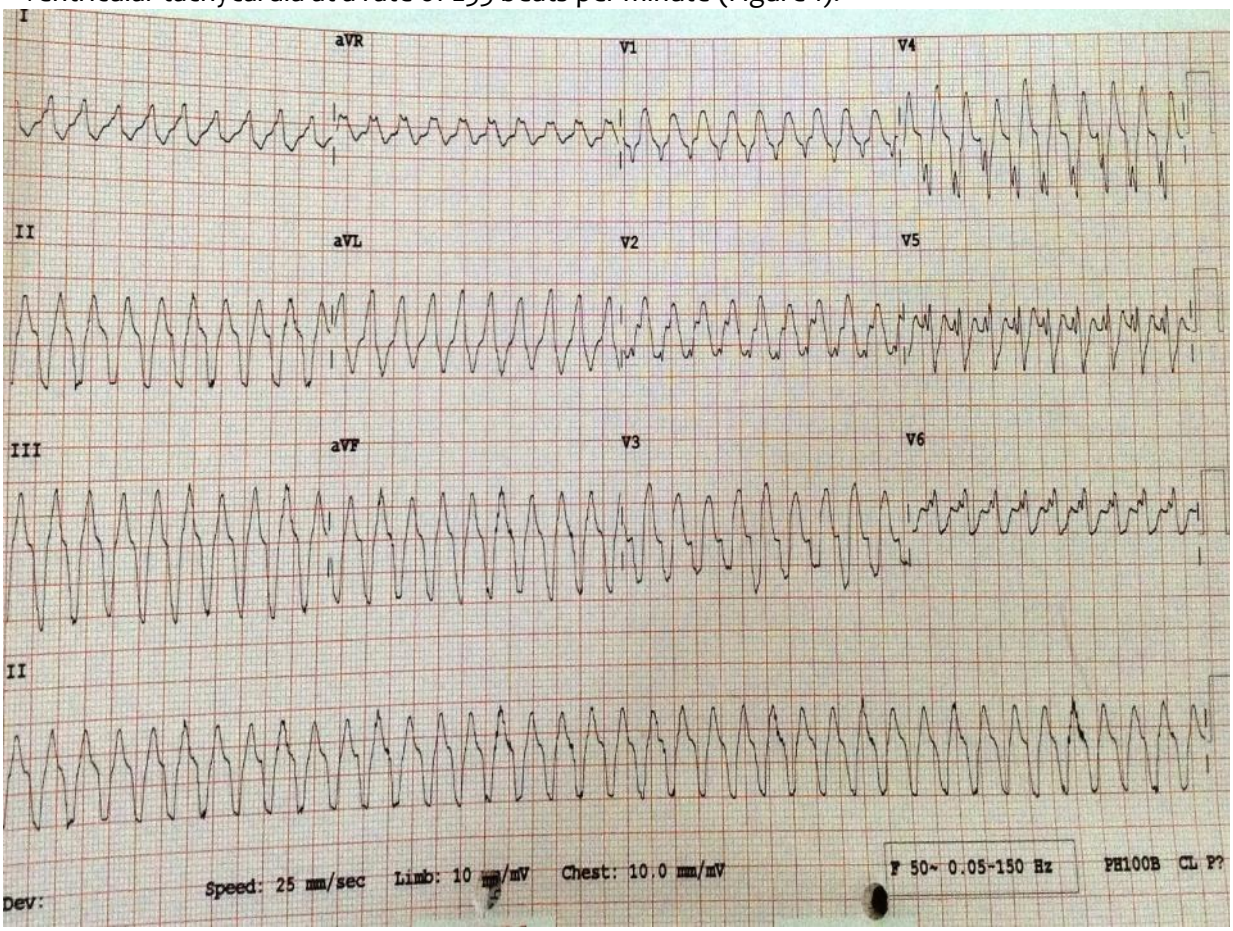

Figure 1: ECG showing monomorphic ventricular tachycardia with a heart rate of 233b/min

Therapy was initiated with intravenous Amiodarone according to available guidelines (Zipes *et al.*, 2006) at a dose of 5mg/kg IV slowly (20 minutes) in both attempts. In addition, oxygen, parenteral antibiotics, antipyretics and intravenous fluids were also given. Over the ensuing 2 hours and 20 minutes the child stabilized and although ventricular tachycardia persisted, there was progressive slowing of the heart rate until the occurrence of spontaneous conversion to a junctional tachycardia at a rate of 110 beats per minute. An ECG obtained shortly after termination of ventricular tachycardia showed the typical J-point and coved ST elevation typical of Brugada type I pattern in lead V2 (Figure 2).

The child improved tremendously and amiodarone was discontinued after six hours with no recurrence of ventricular tachycardia. An echocardiogram showed no evidence of structural heart disease. He was discharged home after five days in good condition. A pre-discharge ECG, when the child was afebrile, showed junctional and atrial ectopy with no right precordial J-point or ST

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elevation. The patient was doing well three months after discharge and a repeat echocardiogram and blood chemistry remains normal.

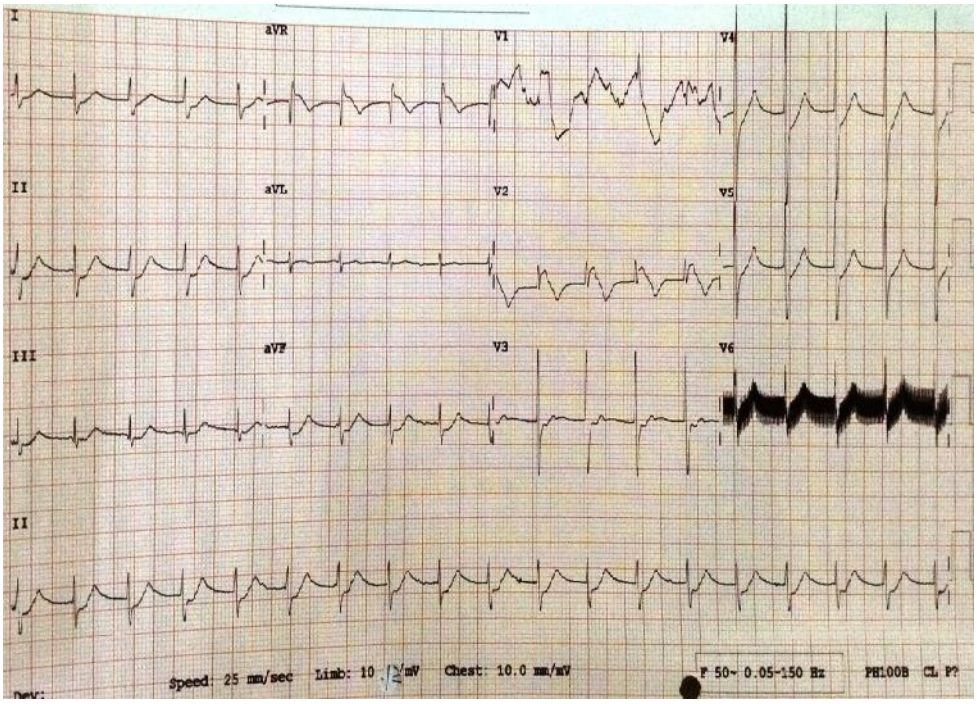

Figure 2: ECG showing J-point and coved ST elevation of Type I Brugada pattern in lead V2

Discussion

The prevalence of Brugada-type ECG in Africa is unknown although a few adult-cases of African-origin have been reported in Tunisia (Ouali *et al*., 2011), France (Bonny *et al*., 2008) and the United States of America (Suzuki, 2006). Although the condition is usually diagnosed among adults, paediatric cases have been reported in Europe (Priori *et al.*, 2000; Probst *et al.*, 2007), and Japan (Yamakawa *et al*., 2004). In the light of these reports, our assumption is that the condition is very rare in African children, particularly Tanzania. We also assume that most cases remain misdiagnosed or unreported due to lack of ECG machines in most of the health facilities in Tanzania and lack of awareness and knowledge on interpreting the ECGs among the clinicians. Our patient was only 3-months old. His younger age and the fact that he is black African, makes the case more interesting and unique.

In the presence of the Brugada-type ECG, BrS is diagnosed if one/more of the following clinical factors are present, according to the consensus report (Bayés de Luna *et al.*, 2012): (a) survivors of cardiac arrest, (b) presence of polymorphic ventricular tachycardia (VT), (c) history of non-vagal syncope, (d) familial antecedents of sudden death in patients younger than 45 years without acute coronary syndrome, and (e) or type 1 ST pattern in relatives.

Our patient's initial presentation was rapid monomorphic VT (Figure 1), and later coved ST elevation of Brugada-type I ECG in lead V2 after cardiovesion (Figure 2) while he was febrile. Since the monomorphic ECG pattern doesn't fulfil the usual diagnostic criteria for the BrS, there would be a necessity for electro-physiological studies including the administration of sodium- channel blockers for confirmation. A similar case has been reported in India, where the diagnosis of BrS was confirmed

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following induction by intravenous procainamide (Sastry *et a*l., 2001). We were unable to perform EPS to our patient. Febrile state, in conjunction with other factors has been shown to unmask the Brugada pattern on the ECG and trigger arrhythmias which may have initially be concealed (Wakita *et al*., 2004; Antzelevitch & Brugada, 2002; Adler *et al.*, 2013). Fever, in our patient may have led to the exposure of his Brugada-pattern ECG as the consequent ECG post febrile condition was normal.

Therapeutic recommendations include the use of an implantable cardiac defibrillator (ICD) in all symptomatic patients with Brugada-Type I ECG. EPS with sodium channel blockage is indicated in all asymptomatic patients with type 1 Brugada ECG and ICD should be inserted only if they are ventricular arrhythmias are inducible (Antzelevitch *et al.*, 2005). From these recommendations, it is clear that our patient would qualify for EPS. Genetic studies would also be important for the recognition of BrS mutations and differentiation from other Brugada phenocopies. Due to our limited capacity, none of these tests could be performed. Despite these diagnostic limitations, we remain concerned that our patient may be at increased risk of sudden death as it was observed in one long term follow-up study of asymptomatic individuals with Brugada-type I ECG pattern (Brugada *et al.*, 2002). To the best of our knowledge, this is the first paediatric case with Brugada-type ECG to be reported in Sub-Saharan Africa.

Conclusion

This case emphasizes the need to increase awareness among clinicians of clinical and genetic arythmogenic disorders. Electrocardiograms should be available to clinicians in primary care as well as referral settings. There should be more widespread instruction in basic electrocardiographic interpretation and arrhythmia interpretation. Multiple ECGs during and after febrile disorders should be performed in children who exhibit extreme tachycardia or signs of cardiac failure.

Competing interests

Authors declare that they have no competing interests.

Authors' contribution

JRM, FR and SH managed the patient and collected the clinical information. SBK was involved in literature search and manuscript writing. All authors critically revised the manuscript and approved the final draft.

Acknowledgements

We thank the staff of the Emergency Department and Cardiology Unit of the Muhimbili National Hospital for their cooperation. We also thank the legal guardian (Mother) of this patient for allowing us to share this case.

Consent

Written informed consent for publication of this case report and the accompanying images was obtained from the legal guardian of this patient (mother).

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