Case Report

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BRUGADA SYNDROME: SUDDEN CARDIAC DEATH REVIEW OF LITERATURE AND CASE REPORT

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ABSTRACT:

Brugada Syndrome or Sudden Unexpected Death Syndrome was first discovered by P. Brugada and J. Brugada in 1992.[1] It is a rare genetic disorder characterised by ST segment elevation in V_1 – V_3 leads on ECG, ventricular fibrillation and ventricular arrhythmias which can cause sudden unexpected death in an otherwise normal patient. We recently treated a case of Brugada Syndrome (Type 3) with surgical extraction, and we wish to highlight the fact that with thorough pre-operative anaesthetic and cardiac evaluation these potentially life threatening patients can be effectively treated for minor oral surgical procedures using regional anaesthesia with lignocaine thereby avoiding general anaesthesia.

Key Words: Brugada Syndrome (BrS), sudden cardiac death (SCD), sudden unexpected death syndrome (SUDS)

INTRODUCTION:

"In 1992, Pedro and Josep Brugada for the first time introduced a new clinical entity with ST segment elevation in V_1 – V_3 leads and right bundle branch block (RBBB) pattern associated with a high incidence of ventricular tachycardia/ventricular fibrillation (VT/VF)".[1] This new entity was termed Brugada Syndrome (BrS) or Sudden Unexpected Death Syndrome (SUDS), occurring in structurally healthy hearts in young individuals, causing life threatening arrhythmias and sudden death. Most of the patients are between second and fourth decades of life however "the youngest patient clinically diagnosed with the syndrome is two days old and the oldest is 84 years old".[2] There is a male predilection, "due to the presence of more prominent I_{to} channels in males than in females"[3] and in many countries it is the second highest cause of death in younger men after vehicular accidents.

Signs and symptoms include presyncopal and syncopal attacks and cardiac arrest (many a times during sleep). Routine ECG shows ST segment elevation in leads V_1 – V_3. Fever may precede syncope or tachycardia.

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There are three types of Brugada ECG Patterns (Figure 1)

Type One: coved type, where ST segment elevation > 2 mm.

Type Two: saddle back type, where ST segment elevation > 2mm with positive 'T' wave.

Type Three: coved or saddle back type, where ST segment elevation < 1mm with inverted T wave.

Brugada Syndrome is inherited as an autosomal dominant trait. In 1998, the Syndrome was linked to mutations in SCN5A, the gene that encodes the alpha subunit for the sodium channel and since then over 300 mutations of SCN5A have been identified.[4] Mutations of gene SCN5A cause loss of expression of sodium channel protein which decreases the sodium current resulting in slow conduction in the heart. Bezzina et al presented evidence supporting the theory that an SCN5A promoter polymorphism, common in Asians modulates, variability in cardiac conduction and may contribute to the high prevalence of Brugada Syndrome in Asian population.[5]

CASE DETAIL:

A 27 year old male patient reported in the department of dentistry, with excruciating pain in lower right second molar and insisted on getting it extracted. Clinical examination and orthopantomogram revealed an extremely carious lower right second molar (Figure 2-A).The patient was a recently diagnosed case of Brugada Syndrome (Type 3) (Figure 3). He gave a history of chest pain about seven years ago, however, a couple of months ago he had persistent chest pain for which he was admitted in the intensive care unit for about ten days. On cardiac evaluation, ECG revealed an elevated ST segment in $V_1 – V_3$ leads and partial RBBB pattern but structurally normal heart valves with normal pericardium and absence of clots or vegetation. His left ventricular ejection fraction was 60%. CST was performed by Bruce protocol where patient walked for 30 minutes with 10.1 METS which showed no angina/arrhythmia. Basal ECG showed RBBB persisted throughout the test. However there were no significant ST segment changes during the test. Adequate chronotropic and ionotropic response was achieved. CST was negative for stress induced reversible ischaemia and for arrhytmia. There was no diagnosed case of Brugada Syndrome in the family. His past surgical history revealed an appendicectomy and septoplasty. He was a chronic smoker and occasionally consumed alcohol.

Our patient was a Type 3 Brugada patient who was being regularly followed up by the cardiologist in view of his ECG pattern. The constant severe toothache was causing him a lot of stress and thus the definitive management indicated was extraction. Since it was a minor dental surgical procedure and given the patient's history, the tooth extraction was planned under local anaesthesia using lignocaine hydrochloride with adrenaline (1: 2,00,000) thereby avoiding general anaesthesia, and the various drugs used with it that could trigger ventricular

tachycardia in a BrS patient. Due to the patient's history of chest pain and diagnosis of Brugada Syndrome, patient was thoroughly evaluated by the anaesthetist and cardiologist prior to the dental treatment. A high risk fitness was obtained.

The patient was taken up in the intensive care unit. A ventilator and a defibrillator were kept standby. A 12 lead ECG was attached and was monitored continuously throughout the procedure. An I.V. line was secured (Figure 4-A). The anaesthetist and cardiologist along with the maxillofacial surgeons formed the surgical team. A right inferior alveolar nerve block was given using three ml lignocaine with adrenaline solution. Another one ml was used for intra-pulpal infiltration. After checking for subjective and objective signs the tooth was surgically extracted after sectioning the roots (Figure 4-B). The wound was closed using 3 – 0 vicryl. Patient tolerated the procedure well. Intra operatively patient was given four mg Dexamethasone along with injection Amoxycillin with Clavulanic acid 1.2 gm. Post operatively he was put on oral tablet Amoxycillin with Clavulanic acid 625 mg and tablet Paracetamol twice a day. Patient was discharged the same day and was followed up in the dental department (Figure 2-B).

DISCUSSION:

Brugada Syndrome is a major cause of sudden unexplained death syndrome (SUDS) and death is caused by ventricular tachycardia and fibrillation (a lethal arrhythmia) in the heart which appears with no warning. The diagnosis in Brugada Syndrome is based on the characteristic patterns on an electrocardiogram, which may be routinely precipitated by administration of certain drugs (ajmaline or flecainide). Brugada ECG pattern is very often hidden, but certain factors can unmask or trigger it like sodium channel blockers, febrile state, vagotonic agents, autonomic nervous system changes, excessive stress, tricyclic or tetracyclic antidepressants, first generation antihistamines (dimenhydrinate), a combination of glucose and insulin, hyperkalaemia, hypokalaemia, hypercalcaemia, alcohol toxicity, heavy meals at night just before sleeping, excessive vomiting, hot humid climatic conditions.[6]

According to Nademanee and Veerakul [6], north-eastern part of Thailand where SUDS is prevalent and where temperatures can soar to 41^{o}C a study is underway to gauge the climatic influences on occurrence of SUDS and they feel that physicians should factor in temperature as a cause of arrhythmogenesis in BrS. Several drugs could precipitate ventricular tachycardia and fibrillation which are listed in world Brugada registry in www.brugadadrugs.org (Accessibility verified July 01, 2015). All Brugada patients and their treating physicians should be aware of these precipitating drugs at all times. Type 1 is the only ECG diagnostic pattern of Brugada Syndrome while Types 2 and 3 are only considered to be suggestive of the diseases. The only line of treatment for type 1 Brugada patients is placement of an implantable

cardioverter device (ICD). Currently a spontaneous Type 1 ECG and presence of atleast 2 multi-parametric risk factors (including syncope, family history, and possible electrophysiologic study) are the only criteria for high risk patients requiring ICD placements (Class IIa in Japanese guideline). [2,6]

In contrast, asymptomatic patients with no family history of sudden cardiac death can be managed conservatively with close and regular follow up. Many Brugada patients are asymptomatic and classical pattern on ECG is picked up only by an experienced and trained physician.

Lignocaine is classified as class IIb drug in that "there is no clear evidence if its administration could cause lethal tachycardia in Brugada patients." However, if diluted with adrenaline and in small doses its use does seem to be safe for local anaesthesia (e.g. by dentists) (www.brugadadrugs.org). There are no randomised clinical studies in Brugada Syndrome patients therefore the level of evidence is mostly C (only consensus, opinion of experts, case studies or standard of care) and for some B (non randomised studies). "Lignocaine" displays rapid dissociation kinetics and produces little to no ST segment elevation in patients with congenital BrS.[7] Bupivacaine has been reported to unmask Brugada like ECG patterns when administered epidurally.[8] Hence we avoided bupivacaine and used lidocaine with adrenaline (1:2, 00,000 dilution) instead for our patient, which was well tolerated by him.

Kloesel et al[9] in 2011 did a literature search and compared results of previous reports with theirs regarding outcomes of patients with BrS who underwent surgeries and anaesthetic care and found 21 case reports and four case series. They collected data of 52 anaesthetics and 43 patients. In our literature search we found mention of only two patients of BrS who underwent surgeries in the maxillofacial region. 1) Plate fixation for mandibular fracture in 56 year old male. 2) Tooth extraction, incision and drainage of odontogenic infection in 55 year old male.[10] However both these patients were treated under general anaesthesia. We decided to avoid general anaesthesia thereby keeping the drugs to be used to the minimum.

By thorough pre-anaesthetic evaluation, proper patient counselling, intra-operative pain control using optimum amount of lignocaine, 12 lead ECG continuously monitored at all times during procedure, constant blood pressure monitoring, avoiding use of certain drugs like bupivacaine, keeping a defibrillator standby and by having a cardiologist and anaesthetist in your surgical team these patients can be successfully managed. It is mandatory that these patients are taken up in the ICU and post-operatively monitored for a minimum of four hours. There is a dearth of articles in the Maxillofacial and Dental literature regarding the management of these patients and we feel there is a need of more awareness of this not so rare cardiac condition among the dental and maxillofacial surgeons. With proper

planning these patients with potentially life threatening and unique cardiac conditions can be safely and efficiently managed by dental/maxillofacial surgeons.

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REFERENCES:

1. Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: A distinct clinical and electrocardiographic syndrome. A multicenter report. J. Am Coll Cardiology 1992; 20: 1391-1396.
2. Antzelvich C et al. Brugada Syndrome: Report of the second consensus conference. Endorsed by the Heart Rhythm Society and the European Heart Rhythm Association Circulation 2005; 111: 659-70.
3. Diego J M et al. Ionic and cellular basis for the predominance of the Brugada Syndrome phenotype in males. Circulation 2002; 106: 2004-11.
4. Chen Q et al. Genetic basis and molecular mechanism for idiopathic ventricular fibrillation. Nature 1998; 392: 293-296.
5. C R Bezzina et al. Common sodium channel promoter haplotype in Asian subjects underlies variability in cardiac conduction. Circulation 2006; 113: 338-344.
6. Gumpanart Veerakul M D, Koonlawee Nademanee M D. Brugada Syndrome: two decades of progress. Circ. Journal 2012; 76: 2713-2722.
7. Hideki Itoh et al. A paradoxical effect of lidocaine for the N406S mutation of *SCN5A* associated with Brugada syndrome. International Journal of Cardiology 2007; 121(3): 239-248.
8. Phillips N et al. Brugada type electrocardiographic pattern induced by epidural bupivacaine. Anaesthesia Analogue. 2003; 197: 264.
9. Benjamin Kloesel et al. Anaesthetic management of patients with Brugada Syndrome: A case series and literature review. Can Journal Anaesthesia / Can Anaes 2011; 58: 824-836.
10. Nicholas Theododu, Joseph E. Cillo. Brugada Syndrome (Sudden Unexpected Death Syndrome): Perioperative and Anaesthetic Management in Oral and Maxillofacial Surgery. J Oral Maxillofac Surg. 2009; 67(9): 20121-25.

FIGURES

Figure1. Types of Brugada ECG Patterns.

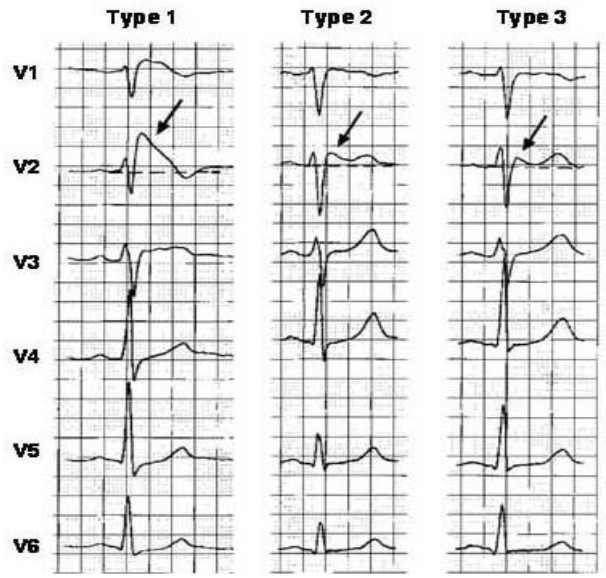

Figure 3. Patient's ECG showing Brugada pattern Type 3

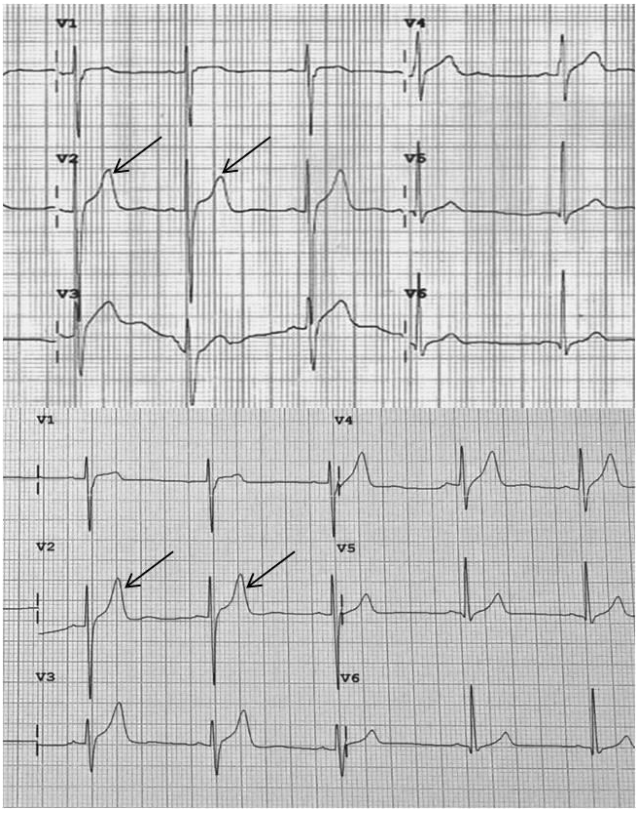

Figure 2.

A – Orthopantomogram pre-operative.

B – Orthopantomogram post-operative

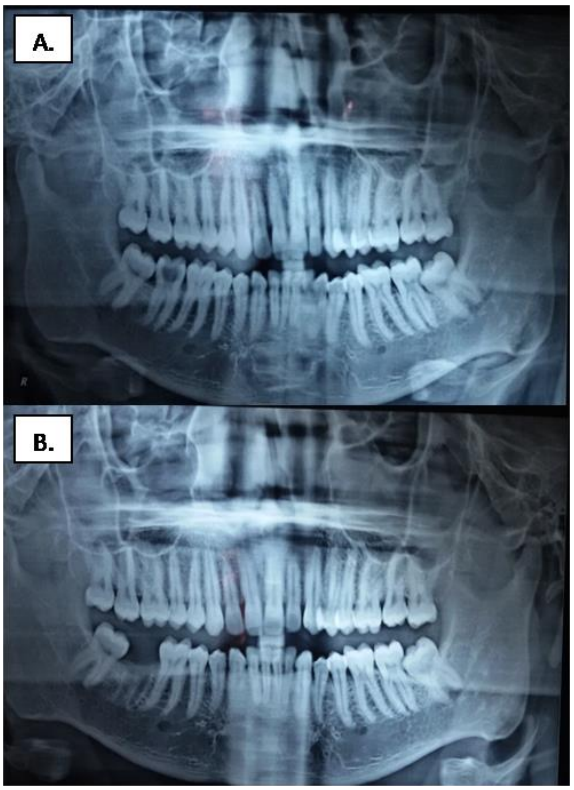

Figure 4. . A – Patient in ICU on the day surgery, with 12 lead ECG attached. B – Carious second molar surgically extracted.