GENERAL ANESTHESIA FOR A PATIENT WITH BRUGADA SYNDROME

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Abstract

Brugada Syndrome is a genetic cardiac disease characterized by electrocardiogram changes consisting of an incomplete right bundle branch block, and ST-segment elevations in right precordial leads V1-V3. These patients are at high risk for developing spontaneous arrhythmias that can be fatal. Many factors during general anesthesia, such as medications, temperature changes, and heart rate variations, could precipitate lethal arrhythmias in this patient population. This case report describes a case of general anesthesia for a patient with known Brugada syndrome.

Keywords: Brugada syndrome, sudden unexpected death syndrome (SUDS), sudden infant death syndrome (SIDS), sudden unexpected nocturnal death syndrome (SUNDS).

Introduction

Brugada Syndrome is a genetic cardiac disease characterized by electrocardiogram (ECG) changes consisting of an incomplete right bundle branch block, and ST-segment elevations in right precordial leads V1-V3. These patients are at high risk for developing spontaneous arrhythmias that can be fatal. First described in 1992 by Pedro Brugada[1], it is now proposed to be a major cause of sudden unexpected death syndrome (SUDS) in young patients with no known history of cardiac disease, and is second only to automobile accidents as a cause of death among young patients in some countries[2]. Clinical presentations can also include sudden infant death syndrome (SIDS) and the sudden unexpected nocturnal death syndrome (SUNDS), a typical presentation in young males from Southeast Asia. Many factors during general anesthesia, such as medications, temperature changes, and heart rate variations, could precipitate lethal arrhythmias in this patient population. Agents that are used regularly in the operating room, such as volatile anesthetics and anticholinesterases are known to cause prolongation of the QT interval, which in these patients can progress to ventricular tachycardia that is often refractory to medication. This case report describes a case of general anesthesia for a patient with known Brugada syndrome.

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Case Report

A 19 year old 58 kilogram Thai male with known Brugada Syndrome presented for closure and free flap of an anterior chest wall wound. He was first diagnosed with Brugada syndrome at age 12 after an episode of syncope. He had an automatic implantable cardioverter defibrillator (ICD) placed at that time and reported that it had discharged several times since being implanted. He had no other co-morbidities, did not take any medications, and had a history of having three prior successful general anesthetics, one for the ICD placement, one for repair of a rotator cuff tear, and one for resection of a dermatofibrosarcoma in his anterior chest wall. He had been otherwise healthy until he was diagnosed with the dermatofibrosarcoma. This tumor was successfully resected without any anesthetic complications and a vacuum assisted closure dressing was placed over his anterior chest. He then presented for closure of the wound and insertion of a microvascular free flap six days after resection of the dermatofibrosarcoma. Prior to this case, his ICD was deactivated to prevent inadvertent discharge of the ICD during intraoperative use of electrocautery. Preoperative ECG revealed an incomplete right bundle branch block and T-wave inversions in V1 and V2 (Figure) but no other abnormalities. Upon arrival into the operating room, external pads attached to a defibrillator were placed on the patient.

Fig.
Preoperative ECG
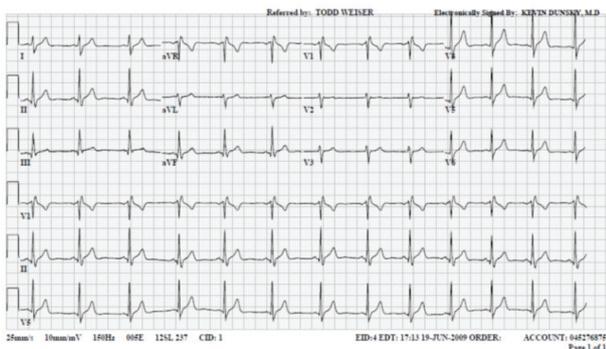
Once in the operating room and prior to pre-oxygenation, standard ASA monitors and the bispectral index (BIS) monitor were placed on the patient. He was given 2mg of midazolam and 100mcg of fentanyl. After adequate pre-oxygenation, anesthesia was induced with 140mg of propofol, followed by 8mg of vecuronium for paralysis. The trachea was intubated, and anesthesia was maintained with desflurane and fentanyl. Maintenance of neuromuscular relaxation was achieved with vecuronium (total intraoperative dose of 20mg.) Immediately following endotracheal intubation, a left radial artery cannula was inserted. Hemodynamics were stable throughout the surgery, which lasted 4.5 hours. Additionally, the patient's heart rate was maintained between 50 and 70 beats per minute and his temperature was maintained between 35 and 36 degrees Centigrade. There was no occurrence of ST segment elevations or ventricular arrhythmias throughout the surgery. Following the surgery, the patient was brought to the post anesthesia care unit (PACU) with the trachea still intubated, on monitors, and with the external defibrillator and pads attached. The ICD was reactivated in the PACU. The patient was extubated once he regained adequate strength and was transferred to a telemetry ward where continuous ECG monitoring showed no post-operative arrhythmias. He was discharged home on post-operative day five.

Discussion

Brugada Syndrome was first described in 1992 by Pedro and Josep Brugada in patients with right bundle branch block, persistent ST segment elevation and sudden cardiac death[1]. That paper described eight patients without demonstrable structural heart disease who were resuscitated from cardiac arrest. Subsequently, much has been elucidated about the syndrome. Although Brugada syndrome presents primarily in adulthood, it can present at any age and is now postulated to be one cause of SIDS, although the mean age of sudden death is forty years. The majority of affected individuals are Asian, with a particularly high prevalence in Japan. Brugada syndrome is thought to account for a minimum of 4 percent of all SUDS cases, and a minimum of 20% of cases occurring in persons with structurally normal hearts[2]. A genetic basis for the syndrome has been linked to mutations in SCN5A, the gene that encodes the alpha subunit of the cardiac sodium channel gene. The gene locus is on chromosome 3p21-24 and it is inherited in an autosomal dominant pattern, with a variable penetrance[3]. The genetic mutation results in loss of

function of the sodium channel, which leads to both accelerated and prolonged refractory periods, which can cause reentrant arrhythmias[2]. The syndrome occurs more commonly in men than in woman. Additionally, there is a higher incidence of ventricular fibrillation and sudden death in Brugada patients at night, which may be related to the circadian variation in sympathetic and parasympathetic tone[2].

The diagnosis of Brugada Syndrome is made by ECG analysis and/or genetic testing. There are three types of ECG patterns that are often seen in patients with Brugada Syndrome. In the classic Type 1 ECG, the J wave is elevated > 2 mm and the ST-T segment descends with an upward convexity to an inverted T wave. This is known as a "coved type" Brugada pattern[2]. Type 2 and Type 3 Brugada ECG patterns both have elevation of the J wave > 2 mm. However, instead of a "coved type" convexity as in Type I, the ST-T wave has a "saddle-back" configuration in which the elevated ST segment descends towards the baseline, and then rises again to an upright or biphasic T wave[2]. In some patients, the characteristic ECG changes are transient, and the appearance of the ST segment elevations may vary. The diagnosis of Brugada Syndrome can be established in an individual with a Type I EKG and a mutation in SCN5A if at least one of the following are present: documented ventricular fibrillation, self-terminating polymorphic ventricular tachycardia, a family history of sudden cardiac death, coved-type EKG's in family members, electrophysiological inducibility, syncope, or nocturnal agonal respiration[2]. Brugada syndrome can be strongly considered in patients with Type 2 or Type 3 EKG's who also have at least one of the symptoms listed above for diagnosis. Patients with the characteristic ECG pattern but no other symptoms are said to have the Brugada pattern but not the syndrome[2].

An ICD is indicated if the patient has had a prior family history of sudden cardiac death, and/or if the patient qualifies for one based on electrophysiological testing. A resting ECG compatible with Brugada syndrome places the patient at a higher risk of sudden cardiac death. If Brugada syndrome is strongly suspected by the patient's clinical history, or there is a family history of Brugada, a sodium channel blocker may be administered in an attempt to induce the Brugada pattern on ECG to determine if placement of an ICD is indicated.

Any factor that creates an alteration in balance between parasympathetic and sympathetic systems can precipitate a lethal arrhythmia in Brugada patients. Several factors during surgery can precipitate a ventricular arrhythmia in patients with Brugada syndrome, such as fever, vagotonic agents, alpha-adrenergic agonists, beta-adrenergic antagonists, local anesthetics, electrolyte imbalances (hypokalemia, hyperkalemia, and hypercalcemia), class 1C and 1A antiarrhythmic drugs, anticholinesterases, and even volatile anesthetics[2]. For our patient, to avoid fluctuations in temperature, a forced air warming blanket was used to help maintain normothermia. Electrolytes were also monitored using arterial blood gas analysis, and no electrolyte supplementation was needed intra-operatively. Although all volatile anesthetics have been implicated in prolongation of the QT interval[4], we administered desflurane for maintenance of anesthesia due to its low blood gas solubility. Throughout the surgery, the patient was given intermittent boluses of fentanyl to both attenuate the QT prolongation which could occur with the sympathetic stimulation from laryngoscopy and intubation, and to provide intraoperative analgesia. In a recent study, pretreatment with fentanyl 2 micrograms per kilogram significantly lessened the QTc prolongation associated with laryngoscopy and tracheal intubation following propofol induction[5]. Fentanyl can also blunt the sympathetic response to surgical stimulation, which is an important consideration because a tachycardia could develop into an unstable arrhythmia requiring emergency cardioversion or defibrillation. An external defibrillator should be immediately available. At the end of the operation, we did not reverse the muscular paralysis with neostigmine and glycopyrrolate in order to avoid the potential tachycardic response associated with glycopyrrolate. He was transported to the post anesthesia care unit (PACU) still tracheally intubated, sedated, monitored, and with the external defibrillator still attached. In the PACU, he was given additional midazolam and fentanyl for sedation and once the patient showed adequate strength by demonstrating sustained head lift, he was extubated and his ICD was reactivated. He was then placed on a patient controlled analgesic pump to provide post-operative pain control

and was discharged five days post-operatively. Although the anesthetic management of this patient is consistent with routine practice, the monitoring, liberal use of fentanyl, and decision not to reverse the muscular paralysis at the conclusion of the surgery reflects a management strategy for patients with prolonged QT syndromes or a known genetic cardiac conduction abnormality such as Brugada Syndrome.

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