Case Report

Anesthetic management of a young primigravida a case of symptomatic long QT syndrome with a permanent pacemaker *in-situ* undergoing lower segment cesarean section delivery: A case report

ABSTRACT

The patient with Long QT syndrome (LQTS) presents a unique challenge to the anesthesiologist. The anesthetic management of such patients requires a good knowledge of the pathophysiology of this rare disease. General anesthesia (GA) or combined spinal epidural anesthesia (CSEA) are the most preferred anesthetic techniques among anesthesiologists for such patients posted for the lower segment cesarean section delivery. In this report, we would like to share our experience of anesthetic management of a young primigravida, a case of symptomatic LQTS with a permanent pacemaker *in situ*. Anesthetic technique used for the lower segment cesarean section delivery was single-shot spinal anesthesia using a combination of a local anesthetic and strong opioid in the lumber subarachnoid space. Until recently, only one report in the literature has described this single-shot spinal technique for such cases. The advantages of single shot spinal over GA and CSEA can be utilized in a select group of these patients, which include fast onset, dense, reliable block with low local anesthetic toxicity, reduced catecholamine release, and relatively prolonged analgesia when combined with an opioid.

Key words: Dysrhythmia, long QT syndrome, lower segment cesarean section, management, pacemaker

Introduction

Long QT syndrome (LQTS) is a cardiac conduction disorder due to the abnormality of cardiac ion channels. It is manifested by a prolonged and increased dispersion of ventricular repolarization on a surface electrocardiogram (ECG). LQTS can have a congenital or acquired etiology and may be precipitated by some drugs or electrolyte imbalances. A re-entrant circuit may develop when abnormal repolarization is amplified by sympathetic activity. This can lead to torsade de pointes, syncope, seizures, and ventricular fibrillation.[1] The anesthetic technique used for LQTS patients must avoid any stimulus that might cause an arrhythmia, such as tachycardia, hypotension, or increased catecholamine release by pain or stress.[2-4] We managed a young primigravida at 38 + 2 weeks, a case of symptomatic LQTS with a permanent pacemaker *in situ*, with an eventful history in past, under single-shot spinal anesthesia successfully without any perioperative adverse events. In

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order to publish the case report in a medical journal, written consent was obtained from the patient on a consent form.

Case Description

Young primigravida in her twenties with a 38 + 2 week gestation period, a known case of LQTS with a permanent pacemaker in situ was posted for elective lower segment cesarean section (LSCS) delivery. She had a history of palpitation and recurrent sudden syncopal attacks before she landed up in the emergency department in an unconscious state and had also suffered cardiac arrest requiring cardiopulmonary resuscitation. After a cardiology evaluation, a diagnosis of LQTS was made and an automatic implantable cardioverter-defibrillator (AICD) was inserted and programmed to pace the heart at 80 beats/min. Along with AICD, metoprolol 75 mg twice daily (BD) per oral (PO) was also prescribed. There was no family history of similar complaints or history of any sudden death or diagnosis of the long QT interval. In the year 2010 (3 years later), AICD was removed due to malfunction and it was replaced with a St. Jude permanent pacemaker under general anesthesia. The patient was under regular follow-up at the cardiology clinic. The latest interrogation was done recently in December 2021 with the following details: pacing mode AAI, voltage 3.02 V, battery current 10 micro-A, a base rate of 90/min, the remaining capacity of elective replacement indicator >90%, and atrial tachycardia or atrial fibrillation-NIL. She had conceived naturally and on clinical examination, her functional level was class one of the New York Heart Association (NYHA) functional classification and had a functional capacity of more than four metabolic equivalents (METs >4). The cardiologist had reviewed the patient before surgery and recommended continuation of tablet metoprolol 100 mg BD, PO, and tablet aspirin 75 mg OD PO. Preoperative ECG showed normal sinus rhythm with intrinsic heart rate >90/min. Complete blood counts, routine liver and kidney functions, and electrolytes were all within the normal range.

The initial anesthetic plan was to perform surgery under combined epidural and spinal anesthesia with a low-dose subarachnoid injection of local anesthetic. We anticipated difficulty in placing an epidural as the patient was overweight and had difficult anatomy of the spine. Also, we did not want to expose the patient to a painful procedure that might have required multiple attempts. We planned a single shot spinal anesthesia with local anesthetic and morphine as an adjuvant drug. We also formulated a backup plan of general anesthesia in case of failure of spinal anesthetic technique. The patient was informed and explained in detail about the spinal anesthetic techniques, all of her queries were addressed and reassured appropriately. General pre-anesthetic instructions were given with no premedications and fasting of 6 h for solid and 2 h for clear liquid was advised. The availability of the pacemaker technician was sought during surgery and the immediate postoperative period. On the day of surgery, the pacemaker was reprogrammed to asynchronous mode by the technician under ECG monitoring. After confirming all the necessary preparations including an external defibrillator, the patient was brought to the operating room. Monitors were attached, IV access was obtained, and preloading with a crystalloid solution was started. A baseline BP of 128/72 mm Hg, HR of 92/min, with regular rhythm on 5-lead ECG and SPO2-99% on room air was noted. A subarachnoid injection was performed with a 25G quincke's needle in a single attempt by an experienced anesthesiologist. The total drug volume given was 2.6 ml, that included 2.5 ml of heavy bupivacaine 0.5% and100 mcg of morphine. A sensory block of T4 was achieved within 5 to 6 min and surgery was started. Fortunately, no episode of significant hypotension requiring vasopressor therapy was encountered. Any drug implicated in prolonging of QT was avoided. Oxytocin 5 IU was given intravenously slowly over 5 min to avoid hypotension and/or bradycardia. The intraoperative period was uneventful. The patient delivered a healthy baby weighing 3.2 kg with Apgar scores of 9 at 1 min and 10 min, respectively. The patient was pain-free and shifted to the recovery room under continuous ECG monitoring. The pacemaker mode was reprogrammed back to AAI mode. Continuous ECG monitoring for 24 h and evaluation by the cardiologist was advised during the postoperative period. The patient remained asymptomatic with normal sinus rhythm on ECG during next 24 h postoperatively.

Discussion

The LQTS diagnosis is based mainly on ECG patterns, clinical symptoms, and genetic findings.[5] The prevalence of congenital LQTS is approximately 1:2000.[6] There are multiple subtypes of congenital LQTS ranging from LQT1 to LQT7, each one of these with specific features, triggers, and outcomes.[2-3] No genetic subtype was determined in our patient because genetic testing was not performed. Patients with LQTS are prone to develop dysrhythmias in the presence of certain triggers, such as adrenergic stimulation with intense exertion, emotional stress, fright, anger, or a sudden auditory stimulus;[2-4] although most episodes of dysrhythmias are transient, few might lead to torsade de pointes (Tdp), a fatal ventricular arrhythmia, and might result in sudden death.[7] Although the available information on the incidence of dysrhythmias during pregnancy in patients with LQTS is limited, there are several possible causes of the increase in dysrhythmias during pregnancy, including estrogen and progesterone, which increase adrenergic responses and affect mutant ion channel function.[4,7,8]

In the literature, concerning the anesthetic management technique, combined spinal epidural and general anesthesia have been preferred and described in pregnant patients.[2-4,7] It is noteworthy, however, that there are only a few case reports on the use of isolated spinal[9] or epidural[10] anesthesia. In clinical practice, most anesthesiologists would avoid subarachnoid injection of local anesthetic or spinal anesthesia in such patients fearing the rapid onset of hypotension following the sympathetic block which could precipitate ventricular arrhythmia and cardiac arrest in this group of patients. Due to the above concern and to ensure more safety and to reduce perioperative morbidity, most practitioners rely on epidural or general anesthesia. The epidural and general anesthetic techniques have their disadvantages as well. The epidural placement is a painful procedure especially if the patient's anatomy is challenging, which may lead to sympathetic activation, increase patient discomfort, slow onset or unreliable block, and risk of local anesthetic toxicity.[2-4] While administering general anesthesia, one should be cautious during the induction of general anesthesia. The sympathetic stimulation leads to increased catecholamine release during intubation and skin incision which may increase the risk of life-threatening dysrhythmia.[2-4] Also, many drugs used intraoperatively and inhalational anesthetic agents, halothane, enflurane, isoflurane, and sevoflurane have been known to prolong the QT interval. In select patients, spinal anesthesia may offer several advantages over epidural anesthesia or general anesthesia, including dense, reliable blocks, fast onset, low local anesthetic toxicity, and reduced catecholamine release.[2-4] Combining an appropriate dose of long-acting opioids with local anesthetic also prolongs postoperative pain relief. Because of anticipated difficulty in epidural placement due to certain patient factors and to avoid pain & discomfort to the patient, we decided to administer spinal anesthesia to our patient. Moreover, she was hemodynamically stable, asymptomatic, and had a good intrinsic sinus rhythm with no dysrhythmia.

The pre-anesthetic check-up should include a thorough, history, record of important events, clinical examination, and current medications. Complete detail of implanted electronic devices such as a pacemaker or AICD (detailed to be noted are; the indication for insertion, the type of device, the manufacturer, the date of last interrogation, the date of the last battery change, any clue of malfunction and the response to a magnet). The contact details of the manufacturer or the cardiologist who implanted it must be noted and should be contacted to obtain details about the management of the device in the perioperative period. The patient should be reviewed by a cardiologist well before the planned procedure to ensure the optimization of the disease. The medications especially beta-blockers should be continued perioperatively. Any electrolyte imbalance such as hypokalaemia, hypomagnesemia, and hypocalcemia should be corrected. The drugs that are implicated in prolonging the QT interval (antiarrhythmic agents class IA, IC, and III) should be avoided during the perioperative period. Reassurance and discussing the anesthetic plan with the patient might help in making the patient calm. During the intraoperative period, the drugs (thiopentone, succinylcholine, epinephrine, and norepinephrine), electrolyte disturbances (hypokalaemia, hypomagnesemia, and hypocalcemia), hypothermia, and sympathetic stimulation must be avoided as these might prolong QT interval and precipitate dysrhythmias.[2-4] The availability of an external pacemaker and defibrillator must be ensured before starting such a case. If any inadvertent episode of torsade de pointes occurs, it should be treated by cardioversion/defibrillation and magnesium sulfate injection (initial bolts of 30 mg/kg IV over 2 to 3 min followed by an infusion of 2 to 4 mg/min.[2-4] Intraoperative management protocol for patients with a pacemaker or AICD should be followed as per institutional guidelines. These guidelines must include the use of bipolar electrocautery, placing the grounding pad as far as possible from the device, and the availability of a pacemaker/ICD technician perioperatively. The patient should be monitored for at least 24 h postoperatively and the device must be interrogated by the treating cardiologist and reprogrammed if needed.

The disorder having a genetic predisposition puts the fetus also at risk.[7] There are several techniques to diagnose the LQTS in the fetus, indirectly with the evidence of sinus bradycardia on cardiotocography, non-invasively by magnetocardiography, and invasively by genotyping. Postnatal electrocardiography should be done to confirm a prolongation of QT interval as these findings might suggest a greater risk of sudden death in neonates as the same has been linked to sudden infant death syndrome. Regular ECG monitoring at certain intervals may be advised till the genetic test report is confirmed.[7]

Thus, from the above discussion and revisiting the literature, it can be concluded that preoperative optimization requires a thorough history and clinical examination, delving into previous hospital records, the current status of the disease, and recent information about the functioning, and interrogation of the implantable electronic devices. Patient counseling, formulation of a primary and backup anesthetic plan, and a crisis management plan including personnel, equipment, and drugs in advance are required. A careful and vigilant intraoperative management to prevent the precipitation of dysrhythmias by any stimuli, drugs, or

maneuver should be kept in mind. Post-operative cardiac monitoring and avoidance of events that might induce dysrhythmias are very important. Good communication and shared decision-making among the members of the multidisciplinary team comprising mainly an anesthesiologist, obstetrician, and cardiologist are very important for good clinical outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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