Case Report

Anesthetic management of a parturient with Kearns-Sayre syndrome, dual-chamber and VVI implantable defibrillator pacemaker/defibrillator, and preeclampsia for cesarean delivery: A case report and review of the literature

ABSTRACT

Kearns–Sayre syndrome (KSS), a rare form of mitochondrial myopathy, is a triad of chronic progressive external ophthalmoplegia, bilateral pigmentary retinopathy, and cardiac conduction abnormalities. In this report, we show how a combined spinal epidural anesthesia can be useful for cesarean delivery, as we illustrate in a dual-chamber and VVI implantable defibrillator pacemaker/defibrillator parturient with a KSS and preeclampsia.

Key words: Anesthesia; implantable defibrillator; Kearns–Sayre syndrome; pacemaker; parturient; preeclampsia

Introduction

Kearns–Sayre syndrome (KSS), a rare form of multisystem mitochondrial myopathy (1.171.6 cases/100,000 population),[1-2] is a triad of chronic progressive external ophthalmoplegia, bilateral pigmentary retinopathy, and cardiac conduction abnormalities that were first described in 1958 by Kearns and Sayre.[3] In addition, KSS may have other manifestations such as heart block, cerebellar ataxia, cerebrospinal fluid protein >100 mg/dL,[4] deafness, dementia, kidney dysfunction, muscle weakness, and endocrine abnormalities including growth retardation, short stature, or diabetes.[5] KSS is usually manifested before the age of 20 years with no racial or sex predilection, secondary to the deletions of muscle mitochondrial DNA and impaired mitochondrial function through decreased activities of some enzymes of the mitochondrial respiratory chain containing subunits encoded by mitochondrial DNA.[6]

Death is common in the third or fourth decade due to sudden cardiac death reported in up to 20%[7] or organ system failures. The use of permanent pacemakers has the potential to reduce the risk of sudden cardiac death.

Few published reports have addressed the perioperative anesthetic management and potential complications.[8-14] We review these and make recommendations for the anesthetic managements of this rare clinical syndrome.

To the best of our knowledge, this is the first report describing anesthetic management in a parturient with a

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pacemaker/defibrillator, KSS, and preeclampsia for cesarean delivery.

Case Report

A 40-year-old, 167 cm, 93 kg pregnant woman gravida 3, para 2, with a 20-year history of KSS, was scheduled for an elective cesarean delivery at 39 weeks' gestation, because of a history of a complicated vaginal delivery with tearing of the anal sphincter 4 years ago, which was successfully repaired.

Her KSS was manifested with external ophthalmoplegia, pigmentary retinopathy, and acquired left anterior hemiblock. Muscle biopsy demonstrated mitochondrial abnormalities. She presented with a high-grade Mobitz type II atrioventricular block (2:1) which required the establishment of a DAVID pacemaker/defibrillator (dual-chamber and VVI implantable defibrillator) 11 years earlier. She had undergone two normal vaginal deliveries.

Her pregnancy had been complicated with a progressive proximal muscle weakness in her extremities and preeclampsia.

On physical examination, respiratory rate) was 17/min, heart rate (HR) 80/min, blood pressure (BP) 165/95 mm Hg, and pulse oximetry oxygen saturation (SpO_2) 100% on room air. The patient's electrolytes, creatinine, and liver tests were normal. Hemoglobin concentration was 10.2 g/dL, and serum albumin level was 2.8 g/dL.

An electrocardiogram (ECG) showed a pacemaker rhythm at HR of 80/min [Figure 1]. Transthoracic echocardiography showed mild mitral and tricuspid regurgitations and left ventricular ejection fraction of 0.61.

In preparation for cesarean delivery, multidisciplinary discussions involving her obstetrician, cardiologist, anesthesiologists, and the family of the patient were conducted, emphasizing the possibility of hemodynamic or the DAVID compromise during anesthesia. The obstetrician was instructed to limit the use of a monopolar electrocautery during the operation to short intermittent bursts at the lowest possible energy. Combined spinal epidural (CSE) anesthesia was offered as the best option, and a written informed consent was obtained.

An external cardioverter/defibrillator unit with a pacing property to which she remained connected by external defibrillator pads, a magnet, and antiarrhythmic, vasopressor, and inotropic medications was on standby as rescue modality.

The patient was brought to the operating theater and volume loading with 500 ml of dextrose 5% in NaCl 0.9% solution was intravenously infused over 20 min, followed with an intravenous infusion of normal saline 0.9% at a rate of 5 mL/kg/h throughout the procedure. Monitoring was commenced with five-lead ECG with an active pacing mode [Figure 2], noninvasive blood pressure, and pulse oximetry. No sedative premedication was given. Oxygen (2 L/min) was delivered through a nasal cannula. Normothermia was maintained using a warming mattress.

CSE anesthesia was administered to the patient in a sitting position using a loss of resistance to saline technique at the L3–L4 interspace using an 18-gauge Tuohy needle. Then, a 27-gauge, 127 mm pencil-point spinal needle was then passed through the epidural needle, and after confirming free flow of cerebrospinal fluid, hyperbaric bupivacaine 0.5% 12.5 mg with fentanyl 25 μg was injected into the intrathecal space. Next, a 20-gauge epidural catheter was advanced 5.0 cm into the epidural space and secured. The patient was positioned with left lateral tilt with the electrocautery current return pad placed on her right thigh in an effort to minimize possible interference with the pacing function of the DAVID unit. A sensory level up to T4 dermatome, as tested by pinprick, was achieved bilaterally.

Surgery commenced through a Pfannenstiel skin incision. Ten min after the induction of anesthesia, transient severe

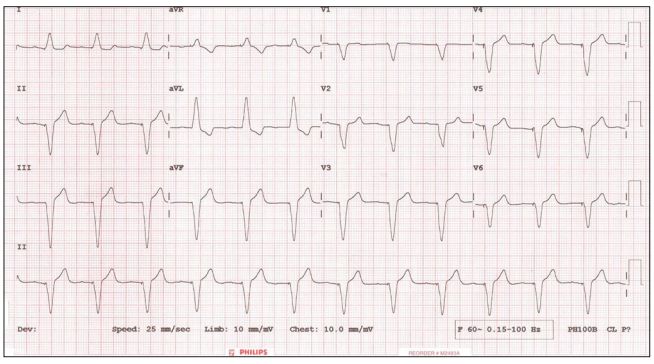

Figure 1: Twelve-lead electrocardiogram showing ventricular pacing rhythm at a heart rate of 80/min

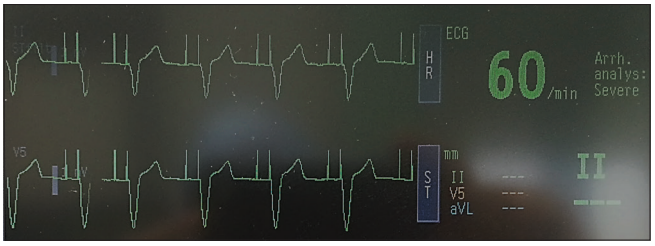

Figure 2: A snapshot from the intraoperative bedside monitor showing sequential atrioventricular pacing at heart rate of 60/min near the end of surgery

hypotension (BP was 67/33 mmHg that lasted for 10 min) was treated with administering a bolus of ephedrine (5 mg). A bolus of oxytocin 5 units was administered followed with an infusion of oxytocin 20 units in normal saline 500 mL immediately after delivery. A baby girl weighing 3210 g, with 1 and 5 min Apgar scores of 9 and 10, respectively, and a cord blood pH of 7.25, was born.

Intraoperative vital signs remained stable throughout the surgery, with the exception of a noted decreased HR to 60/min near the end of surgery. No significant electromagnetic interference (EMI) was noted intraoperatively. Total estimated blood loss during surgery was 500 mL. The patient's intraoperative blood glucose, arterial blood gases, electrolytes, and serum lactate levels showed no abnormalities. No local anesthetics were administered through the epidural catheter during surgery.

Postoperative analgesia was accomplished with a continuous infusion of bupivacaine 0.125% and fentanyl 2 μg/mL through the epidural catheter, lornoxicam (8 mg/12 h), and paracetamol (1 g/6 h). Her DAVID unit was interrogated by the manufacturer in the postpartum period. The postoperative course was uneventful, and the mother and the baby were discharged home on postpartum day 6, both in excellent condition.

Discussion

The mitochondrial respiratory chain consists of four multienzyme complexes located on the inner mitochondrial membrane. Enzyme defects in the mitochondrial respiratory chain result in exercise intolerance, fatigue, and muscle pain.[6]

We screened published articles evaluating the association of surgical patients with KSS and postoperative outcomes, with the help of two independent expert librarians familiar with the literature search. We searched the literature databases PubMed-MEDLINE (1950 to April 25, 2015). The search used the Medical Subject Heading keywords "anesthesia," "Kearns–Sayre syndrome," "muscle relaxant," "mitochondrial myopathy," "mitochondrial myelomyopathy," "complication," and "outcome." A citation search by manual review of references from primary articles also was performed. We recruited eight case reports demonstrating the anesthetic management of patients with KSS.[8-14] One report was excluded because of no available information on patient's details.[14]

Table 1 shows the patient's characteristics including age, gender, cardiac conduction abnormalities, associated manifestations echocardiographic data, type of surgery, anesthesia, analgesia, need for the admission to the Intensive Care Unit, time to hospital discharge after the procedure, and perioperative complications.

The various cardiac conduction abnormalities in KSS patients include third-degree AV block, complete and incomplete right bundle branch blocks, fascicular blocks, and supraventricular and ventricular arrhythmias [Table 1].

One patient showed manifestations of systemic toxicity from the local anesthetic articaine.[10] One patient required reintubation and mechanical ventilation after surgery that may be related to the incomplete reversal of the residual neuromuscular blockade from the administered 0.13 mg/kg of vecuronium defined as recovered T_1 value to 25% of the control value.[12] Thus, caution should be exerted to use short- or intermediate-acting neuromuscular blocking agents alongside close monitoring of train-of-four ratio.[9,11,12]

Mitochondria are a potential site of action of general and local anesthetics; thus, patients with mitochondrial myopathies may be sensitive to the effect of anesthesia. Literature shows that both general and regional anesthesia have been used successfully in patients with KSS [Table 1].[8-13] It is advisable to maintain adequate oxygen balance, stable cardiovascular function, and good gas exchange.[15] In addition, normothermia and normoglycemia should be maintained during the anesthetic management of patients with mitochondrial myopathies, to avoid metabolic stress-induced increased demand of energy production by the diseased mitochondria.[12]

Special anesthetic problems in patients with mitochondrial myopathies such as myocardial conduction disturbances, postoperative muscle hypotonia, and possible increased susceptibility to malignant hyperthermia can be avoided using regional blocks, when applicable.[8,10,13-14] Neuraxial analgesia has the potential to reduce the perioperative oxygen consumption during labor by 25%, particularly in parturients with KSS.[8]

Perioperative management of patients with cardiac pacemakers/defibrillators may be challenging because of the increasing sophistication of these devices. Thorough preoperative evaluation and prophylactic placement of temporary pacing or at least transcutaneous pacing are important for the avoidance and minimization of intraoperative complications in patients with pacemaker/defibrillator. In addition, close coordination between the anesthesiologist, cardiologist, obstetrician, and manufacturer

Table 1: The recruited date from the 6 published case reports[8-13]

Report	Age	Sex	Procedure	Conduction abnormalities	Associated manifestations	Echocardiography data	Anesthesia	Analgesia	ICU	LOS	Complications
Faris *et al*.[8]	28	Female	Cesarean delivery after failure of labor progression	Paroxysmal supraventricular tachycardia	Diabetes mellitus, hypothyroidism, depression, mild proximal muscle weakness	N/A	Epidural analgesia/ anesthesia	Epidural morphine	No	POD3	None
Calzavacca *et al*.[9]	40	Male	Laparoscopic cholecystectomy	Trifascicular block Occasional supraventricular ectopy		Mitral valve prolapse causing mild mitral regurgitation, LVEF 0.5	General anesthesia via an ETT using propofol, fentanyl, rocuronium, N_2O/O_2 mixture, sevoflurane	Paracetamol, Tramadol	Yes	POD5	None
Finsterer *et al*.[10]	28	Female	Tooth extraction	AV block III Pacemaker dependent	Frequent vomiting, growth retardation, myocarditis, muscle wasting, short stature, cerebellar atrophy	A slight tricuspid insufficiency	Left submandibular nerve block using 60 mg articaine and 0.009 mg epinephrine	N/A	No	POD1	A severe adverse reaction to articaine
Hara *et al*.[11]	29	Female	Implantable cardioverter-defibrillator implantation	Mobitz type II second-degree AV block with 2:1 conduction, first-degree AV block, right bundle-branch block, ventricular fibrillation	Muscle weakness in extremities, bulbar paralysis	No abnormalities were detected	General anesthesia via an LMA using propofol, N_2O/O_2 mixture	N/A	No	N/A	None
Kitoh *et al*.[12]	51	Male	Exploratory laparotomy for possible appendicitis	Complete atrioventricular block, paroxysmal atrial tachycardia, atrial fibrillation Pacemaker dependent	Peripheral edema, hepatomegaly, muscular weakness, bilateral sensorineural hearing loss	Biventricular enlargement, diffuse hypokinesia, LVEF 0.35	General anesthesia via an ETT using thiopental, fentanyl, vecuronium, N_2O/O_2 mixture, isoflurane	N/A	Yes		Postoperative reintubation and ventilation for the noted respiratory muscle weakness. Ventricular tachycardia followed with atrial fibrillation with ST-segment depression
Fritz *et al*.[13]	20	Male	Cataract extraction	AV block	MERRF syndrome	N/A	Local anesthesia and sedation	N/A	No	N/A	None

ICU: Intensive Care Unit; N/A: Unavailable; POD: Postoperative day; LVEF: Left ventricular ejection fraction; ETT: Endotracheal tube; AV: Atrioventricular; LMA: Laryngeal mask airway; LOS: Length of stay; MERRF: Myoclonus epilepsy with ragged red fibers

regarding peripartum management, prevention of increases in plasma catecholamines due to labor or operative pain, and finally, possible direct suppression of arrhythmias by pharmacologically active plasma levels of local anesthetic is of highest importance.[16] In general, the antitachyarrhythmia therapy of the implantable cardioverter defibrillator should be deactivated during surgery as they can result in inappropriate shocks or inhibition of pacing because of various extraneous factors such as shivering, or fasciculation induced by diathermy, and by false sensing as a result of sinus tachycardia or rapid atrial fibrillation. Alternatively, the use of bipolar electrocautery whenever possible or monopolar electrocautery in short bursts (<5 s at a time, with 10 s between use) far away from the site of the device is acceptable. However, careful monitoring of the arterial pressure or pulse oximetry is essential during electrocautery, as ECG monitoring can be affected by EMI.[17]

In conclusion, epidural anesthesia offers distinct advantages in parturients with KSS including easy conversion from labor analgesia to surgical anesthesia, preservation of fetal–maternal hemodynamics, prevention of increases in

energy production by the diseased mitochondria due to labor or operative pain, and finally, possible direct suppression of arrhythmias by pharmacologically active plasma levels of local anesthetic.

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 name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Remes AM, Majamaa-Voltti K, Kärppä M, Moilanen JS, Uimonen S, Helander H, *et al.* Prevalence of large-scale mitochondrial DNA deletions in an adult Finnish population. Neurology 2005;64:976-81.
2. Schaefer AM, McFarland R, Blakely EL, He L, Whittaker RG, Taylor RW, *et al.* Prevalence of mitochondrial DNA disease in adults. Ann Neurol 2008;63:35-9.
3. Kearns TP, Sayre GP. Retinitis pigmentosa, external ophthalmophegia, and complete heart block: Unusual syndrome with histologic study in one of two cases. AMA Arch Ophthalmol 1958;60:280-9.
4. Muravchick S, Levy RJ. Clinical implications of mitochondrial dysfunction. Anesthesiology 2006;105:819-37.
5. Harvey JN, Barnett D. Endocrine dysfunction in Kearns-Sayre syndrome. Clin Endocrinol (Oxf) 1992;37:97-103.
6. Moraes CT, DiMauro S, Zeviani M, Lombes A, Shanske S, Miranda AF, *et al.* Mitochondrial DNA deletions in progressive external ophthalmoplegia and Kearns-Sayre syndrome. N Engl J Med 1989;320:1293-9.
7. Roberts NK, Perloff JK, Kark RA. Cardiac conduction in the Kearns-Sayre syndrome (a neuromuscular disorder associated with progressive external ophthalmoplegia and pigmentary retinopathy). Report of 2 cases and review of 17 published cases. Am J Cardiol 1979;44:1396-400.
8. Faris AS, Tawfic QA, Jeyaraj L. Obstetric anesthesia considerations in Kearns-Sayre syndrome: A case report. Korean J Anesthesiol 2014;67:283-6.
9. Calzavacca P, Schmidt W, Guzzi M. General anaesthesia for laparoscopic cholecystectomy in a patient with the Kearns-Sayre syndrome. Case Rep Anesthesiol 2011;2011:806086.
10. Finsterer J, Haberler C, Schmiedel J. Deterioration of Kearns-Sayre syndrome following articaine administration for local anesthesia. Clin Neuropharmacol 2005;28:148-9.
11. Hara K, Sata T, Shigematsu A. Anesthetic management for cardioverter-defibrillator implantation in a patient with Kearns-Sayre syndrome. J Clin Anesth 2004;16:539-41.
12. Kitoh T, Mizuno K, Otagiri T, Ichinose A, Sasao J, Goto H, *et al.* Anesthetic management for a patient with Kearns-Sayre syndrome. Anesth Analg 1995;80:1240-2.
13. Fritz T, Wessel K, Weidle E, Lenz G, Peiffer J. Anesthesia for eye operations in mitochondrial encephalomyelopathy. Klin Monbl Augenheilkd 1988;193:174-8.
14. Klockgether-Radke A, Henze T, Braun U, Kettler D. General anesthesia in two patients with mitochondrial myopathy. Anaesthesist 1993;42:111-4.
15. Footitt EJ, Sinha MD, Raiman JA, Dhawan A, Moganasundram S, Champion MP, *et al.* Mitochondrial disorders and general anaesthesia: A case series and review. Br J Anaesth 2008;100:436-41.
16. Frost DA, Dolak JA. Cesarean section in a patient with familial cardiomyopathy and a cardioverter-defibrillator. Can J Anaesth 2006;53:478-81.
17. Dawes JC, Mahabir RC, Hillier K, Cassidy M, de Haas W, Gillis AM, *et al.* Electrosurgery in patients with pacemakers/implanted cardioverter defibrillators. Ann Plast Surg 2006;57:33-6.