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Case Report Open Access

A Case of Mitochondrial Myopathy with Cardiac Involvement: Management in the Same Day Surgery Setting

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Abstract

Leigh's disease, also known as Subacute Necrotizing Encephalomyelopathy, is an inherited myopathy causing degradation of motor skills with occasional cardiac involvement. Anesthesia experience for this disease has been rarely reported. Anesthetic can be safely preformed on a patient with Leigh's disease in ambulatory setting. Extreme caution is required if the patient presents with cardiomyopathy manifestations of the syndrome.

Keywords: Leigh's disease; Mitochondrial cardiomyopathy; Intraoperative bradyarrhythmia

Introduction

During the last decade, disorders of the respiratory chain, so-called mitochondrial disorders, have emerged as a major clinical entity. Though Leigh's disease, also known as Subacute Necrotizing Encephalomyelopathy, has been mostly reported in infancy and childhood, it is described in adults [1]. Leigh's disease is a myopathy causing degradation of motor skills with occasional cardiac involvement. Leigh's disease occurs either because of the two possibilities: mutations in mitochondrial DNA or deficiency of X linked enzyme called pyruvate dehydrogenase [2]. Acquired Leigh's disease as a result of spontaneous mutation also happens [3]. Reports of mitochondrial myopathies with cardiac involvement remain sparse [4].

Case Report

A 57 year old female was brought to the Endoscopy Suite from a long-term care facility for outpatient esophagogastroscopy and colonoscopy. Medical history was remarkable for late onset Leigh's myopathy, cardiomyopathy, chronic respiratory failure, ventilator dependence, frequent pneumonias, chronic bronchitis with asthmatic component and chronic upper abdominal pain. Patient had no associated congenital malformations or deformities. She had no information in regards to her family or genetic history. Her surgical history consisted of tracheostomy and gastric feeding tube placement. Patient appeared alert and oriented and, communicating via blinking and moving palms, reported no additional history. Blood pressure on the arrival to GI suite was 105/90; pulse was 102; and oxygen saturation via pulse oximeter was 96% on FiO_2 0.3 with controlled ventilation through existing tracheostomy. Bilateral rhonchi and distant heart tones were heard on auscultation. EKG was remarkable for ST-T changes, prolonged QT and PR intervals. Chest X-ray showed significant cardiomegaly. Her transthoracic echocardiography revealed 15% ventricular ejection fraction, dilated chambers and no significant valvular dysfunction. Perioperative plan was discussed with gastroenterologist and postprocedural admission was planned for cardiovascular monitoring. Monitored Anesthesia Care with sedation was planned. Consent was obtained directly from the patient. Appropriately set malignant hyperthermia (MH) cart was checked and available. Peripheral intravenous access was established and she was prehydrated with 300cc of normal saline. Sedation was achieved with divided doses of Etomidate to maintain maximum cardiovascular stability. Tracheostomy cuff inflation was confirmed and gastrostomy tube placed on intermittent suction to decreased aspiration risk. Patient was mechanically ventilated with 50% oxygen (50/50 O2/Air mixture). Judiciously divided doses of Etomidate were continuously given to maintain sedation and to assure patient's comfort. Gastroduodenoscopy was performed uneventfully. During colonoscopy there was a short episode of bradycardia with significantly prolonged QT intervals and brief hypotension with blood pressure of 75/40, which were treated effectively with 0.4 mg of Atropine and 200cc normal saline bolus. Vital signs remained stable throughout the rest of the procedure. No significant pathology except for chronic gastritis was noted by gastroenterologist. Upon completion of endoscopy she was admitted to PACU where no further cardiovascular or respiratory events were noted. Patient was transferred to the medical floor when appropriate discharge criteria were met. No anesthesia related complications were noted and next morning she was discharged to the chronic care facility.

Discussion

Leigh's Disease is a progressive neurometabolic disorder with a general onset in infancy or childhood, often after a viral infection, but can also occur in teens and adults. The classical form of Leigh's syndrome is inherited as an autosomal recessive trait and has been linked to a genetic defect in one of several different genes. These genes cause either a deficiency of the enzyme pyruvate dehydrogenase, or an abnormality in other enzymes that activate pyruvate dehydrogenase. It is characterized by necrotizing lesions on the brain, particularly in the midbrain and brainstem. Mitochondrial myopathy is caused by disturbance of the mitochondrial DNA and has the characteristics of functional impairment of primarily the brain, muscles and myocardium [5]. There is no cure for Leigh's Disease. Treatments generally involve variations of vitamin and supplement therapies, often in a "cocktail" combination, and are only partially effective. Various resource sites include the possible usage of: thiamine, coenzyme Q10, riboflavin,

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biotin, creatine, and succinate and in some cases, a special diet. The prognosis for Leigh's Disease is poor. Depending on the defect, individuals typically live anywhere from a few years to the mid-teens. Those diagnosed with Leigh-like syndrome or who did not display symptoms until adulthood tend to live longer [6]. Mitochondrial myopathy ultimately involves all organs and systems. Patients with this disorder often have hypertrophic cardiomyopathy and overgrowth of the fibrous membrane that divides the various chambers of the heart and creates asymmetric septal hypertrophy. No effective treatment has been reported for associated cardiomyopathy [7].

Primary dangers include the possibility of lactic acidosis and unfavorable metabolic manifestations of some common anesthetics, such as volatile agents and barbiturates. Acid-base status and normoventilation should be monitored throughout the procedure. Because lactic acidosis is a frequent manifestation of the syndrome, use of lactated Ringer solution is discouraged. Myocardial involvement is common and can be manifested as cardiomyopathy leading to congestive heart failure, arrhythmia and conduction abnormalities [8,9]. Given the rarity of this disorder, there are only few of previous reports of anesthetic care in such patients. There is very limited evidence-based data in the literature on which to determine the most appropriate anesthetic management for such patients [10]. Review of literature does not indicate an increased susceptibility to MH by patients diagnosed with mitochondrial myopathy [11,12]. It is best to avoid depolarizing muscle relaxants, although these have been used safely in many patients with mitochondrial diseases [13].

Though anesthesia may present unpredictable risks for certain mitochondrial disease patients, most operative and perioperative complications that have occurred have been both predictable and preventable. Nevertheless, it is important to recognize that even a brief procedure, such as upper endoscopy, can potentially lead to complications [14]. Patients with mitochondrial myopathies may have a prolonged recovery time after non-depolarizing neuromuscular blocking agents [15]. There were no complications in a cohort of 16 children with mitochondrial defects, one of them with Leigh disease, who received general anesthesia with sevoflurane in 100% oxygen, supplemented with fentanyl, during brief surgical procedures, mostly muscle biopsy [16]. In this case Etomidate was chosen for its known good cardiac profile and rapid recovery. Other choices, such as remifentanyl and dexmedetomidine were suggested as well [10]. Cardiac involvement including hypertrophic and dilated cardiomyopathies has been reported to be frequent in mitochondrial diseases [17] and should be carefully appreciated by anesthesiologist. Patients with mitochondrial cardiomyopathy have poor prognosis and quite often and develop congestive heart failure and life-threatening arrhythmias [18].

Conclusions

Leigh's Myopathy is caused by disturbance of the mitochondrial DNA and frequently associated with cardiomyopathy. Anesthesia experience for this disease has been rarely reported. A thorough preoperative assessment of the respiratory, cardiovascular, and renal functions is necessary, for the proper preparation of the patient with Leigh disease, prior to anesthetic care. Anesthetic can be safely preformed on a patient with Leigh's disease in ambulatory setting. Normocapnia, normothermia, and avoidance of lactate containing solutions are recommended. Sedation can be achieved with Etomidate to maintain cardiac stability and closed cardiovascular monitoring is needed. Extreme caution is required if the patient presents with cardiomyopathy manifestations of the syndrome. In such case postoperative admission, cardiac monitoring and observation are highly recommended.

References

1. Pandit L (2007) Adult onset Leigh Syndrome. Ann Ind Acad Neurol 10: 55-77.
2. Wong L (2012) Mitochondrial syndromes with leukoencephalopathies. Semin Neurol 32: 55-61.
3. Ronchi D, Bordoni A, Cosi A, Rizzuti M, Fassone E, et al. (2011) Unusual adult-onset Leigh syndrome presentation due to the mitochondrial m.9176T>C mutation. Biochem Biophys Res Commun 412: 245-248.
4. Gozal D, Goldin E, Shafran-Tikva S, Tal D, Wengrower D (2006) Leigh syndrome: anesthetic management in complicated anesthetic procedures. Paediatr Anaesth 16: 38-42.
5. Shipton EA, Prosser DO (2004) Mitochondrial myopathies and anaesthesia. Eur J Anaesthesiol 21: 173-178.
6. Rahman S, Blok RB, Dahl HH, Danks DM, Kirby DM, et al. (1996) Leigh syndrome: clinical features and biochemical and DNA abnormalities. Ann Neurol 39: 343-51.
7. Mattman A (2011) Mitochondrial disease clinical manifestations: An overview BCMJ 53: 183-187.
8. Van Hove JL, Shanske S, Ciacci F, Ballinger S, Shoffner JS, et al. (1994) Mitochondrial myopathy with anemia, cardiomyopathy, and lactic acidosis: A distinct late onset mitochondrial disorder. Am J Med Genet 52: 114-120.
9. Macaya A, Munell F, Burke RE, De Vivo DC (1993) Disorders of movement in Leigh syndrome. Neuropediatrics 24: 60-67.
10. Shear T, Tobias J (2004) Anesthetic implications of Leigh's syndrome. Paediatr Anaesth 14:792-797.
11. Driessen J, Willems S, Dercksen S, Giele J, van der Staak F, et al. (2007) Anesthesia-related morbidity and mortality after surgery for muscle biopsy in children with mitochondrial defects. Pediatric Anesthesia 17: 16-21.
12. Driessen JJ (2008) Neuromuscular and mitochondrial disorders: What is relevant to the anesthesiologist? Curr Opin Anaesthesiol 21: 350-355.
13. Wallace JJ, Perndt H, Skinner M (1998) Anaesthesia and mitochondrial disease. Pediatr Anesth 8: 249-254.
14. Maslow A, Lisbon A (1993) Anesthetic Considerations in Patients with Mitochondrial Dysfunction. Anesth Analg 76: 884-886.
15. Finsterer J, Stratil U, Bittner R, Sporn P (1998) Increased sensitivity to rocuronium and atracurium in mitochondrial myopathy. Can J Anaesth 45: 781-784.
16. Morgan PG, Hoppel CL, Sedensky MM (2002) Mitochondrial defects and anesthetic sensitivity. Anesthesiology 96: 1268-1270.
17. Anan R, Nakagawa M, Miyata M, Higuchi I, Nakao S, et al. (1995) Cardiac involvement in mitochondrial disease. A study on 17 patients with documented mitochondrial DNA defects. Circulation 91: 955-961.
18. Holmgren D, Wåhlander H, Eriksson BO, Oldfors A, Holme E, et al. (2003) Cardiomyopathy in children with mitochondrial disease; Clinical course and cardiological findings. Eur Heart J 24: 280-288.