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Myotonic Dystrophy and Cardiac Surgery: The Problem and the Panacea

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Abstract

Myotonic dystrophy is the most common muscular dystrophy seen in adults, which can affect the heart, brain, gastrointestinal tract, and other non-muscular organs. Patients with myotonic dystrophy can develop a range of cardiac abnormalities, including conduction abnormalities, dilated cardiomyopathy, arrhythmias, mitral valve prolapse, myocardial dysfunction, and ischemic heart disease. In this case report, we describe a patient with type 1 myotonic dystrophy, who underwent coronary artery bypass grafting and closure of atrial septal defect. We discuss the challenges experienced during the perioperative period and the management which led to a successful outcome.

Keywords: Myotonic Dystrophy; Cardiac Surgery; Coronary Artery Bypass Graft Surgery; Atrial Septal Defect

Abbreviations

ASD: Atrial Septal Defect; pH: Potential of Hydrogen; PaCO2: Partial Pressure of Arterial Carbon Dioxide; PaO2: Partial Pressure of Arterial Oxygen; SaO2: Oxygen Saturation of Arterial Blood; CT: Computed Tomography; TEE: Transesophageal Echocardiography; CPB: Cardiopulmonary Bypass; ICU: Intensive Care Unit; POD: Postoperative Day; PEG: Percutaneous Endoscopic Gastrostomy.

Introduction

Myotonic dystrophy is the most common muscular dystrophy in adults, which can affect the heart, brain, gastrointestinal tract, and other non-muscular organs. Genetically, two distinct types of myotonic dystrophies are described: type 1 and type 2, the former more common than the latter. Type 1 myotonic dystrophy is more frequently associated with bulbar weakness, cognitive dysfunction, hypersomnia, early cataract, facial weakness, diabetes mellitus, cardiac arrhythmias, and respiratory failure [1]. Patients with myotonic dystrophy can develop a range of cardiac abnormalities, including conduction abnormalities, dilated cardiomyopathy, supraventricular and ventricular arrhythmias, and myocardial ischemia [2,3]. Patients with myotonic dystrophy have increased sensitivity to drugs used in anesthesia, such as hypnotics, neuromuscular blocking agents, and opioids. The use of inhalational anesthetics might produce shivering that can precipitate myotonia. The use of intermediate or long-acting muscle relaxants might produce/aggravate prolonged muscle weakness. Weakness of pharyngeal muscles and a delayed gastric emptying time predispose these patients to aspiration. In addition, hypothermia used during cardiopulmonary bypass (CPB) can precipitate myotonia. In this case report, we describe a patient with type 1 myotonic dystrophy, who underwent

coronary artery bypass grafting and closure of atrial septal defect (ASD). We discuss the challenges encountered during the perioperative period and the management which ensured a successful outcome.

Case Presentation

A 57-year-old diabetic male, a known case of type 1 myotonic dystrophy for eight years, was referred to our hospital for acute coronary syndrome. He had a history of inferior wall myocardial infarction and percutaneous transluminal coronary angioplasty of the right coronary artery in 2007. The electrocardiogram was suggestive of old myocardial infarction, right bundle branch block with first-degree atrioventricular block (Figure 1). Troponin-I level was 2.34ng/ml. Echocardiography revealed regional wall motion abnormalities in inferior wall, posterior wall, and lateral wall with left ventricular ejection fraction of 40%, and an ostium secundum ASD of size 16 mm with a left to right shunt. Carotid Doppler, peripheral vascular Doppler, chest X-ray, and other laboratory investigations were normal. Coronary angiography showed ostial left main plaque and double vessel coronary artery disease.

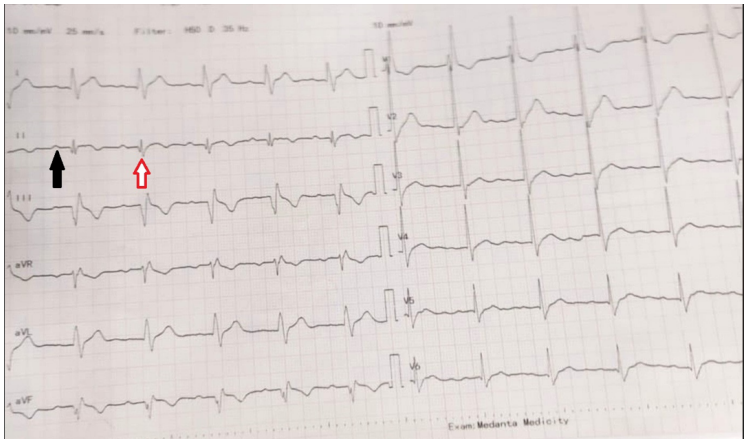

Figure 1: Electrocardiogram showing right bundle branch block (red arrow), first degree atrioventricular block (black arrow), and changes of old inferior wall myocardial infarction.

For the last eight years, since the diagnosis of myotonic dystrophy, the patient was on treatment with phenytoin sodium and modafinil for muscle stiffness and somnolence respectively. The patient had nasal twang on speech. The patient was otherwise mobile and able to perform activities of daily living. In addition, there was history of on-and-off dysphagia. The patient had undergone surgery for bilateral cataracts in 2017. On examination, there was facial weakness (a long, narrow face, temporal muscle atrophy, bilateral ptosis, frontal balding) (Figure 2). The patient was not able to perform pulmonary function test. Arterial blood gas analysis while breathing room air showed a pH of 7.358, PaCO2 48.6mmHg, PaO2 71.8mmHg, bicarbonate 26.7mEq/L, and SaO2 94.8 %. A computed tomography (CT) scan of the head showed chronic microvascular ischemic changes in the brain parenchyma and minor atherosclerotic changes in the vertebral and internal carotid arteries. A multidisciplinary team of cardiologists, neurologists, and gastroenterologists were involved in the patient care. The possibility of prolonged postoperative ventilation was explained, and the risk of aspiration due to laryngeal and pharyngeal muscles' weakness and delayed gastric emptying time was anticipated.

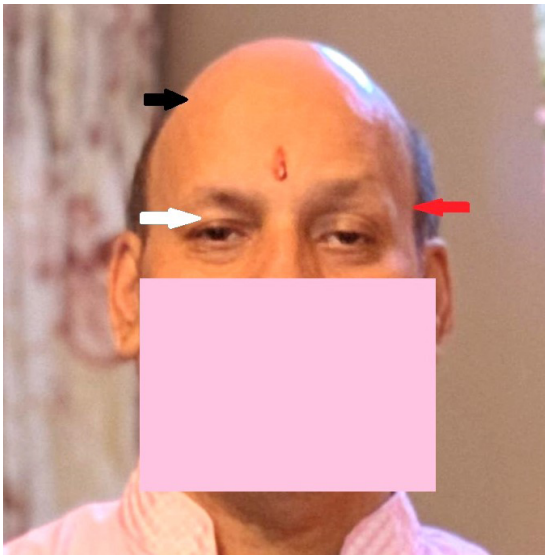

Figure 2: Photograph of the patient (taken before surgery) showing long, narrow face, temporal muscle atrophy (red arrow), ptosis (white arrow), and frontal balding (black arrow).

Following informed consent for the surgery and publication, the patient was taken up for surgery. Under standard cardiac monitoring including TEE and neuromuscular monitoring (train-of-four stimulus), a rapid sequence induction with cricoid pressure was performed. Intravenous induction consisted of midazolam (0.1mg/kg), fentanyl (3μg/kg), thiopentone sodium (3mg/kg), and rocuronium (1.2mg/kg). Tracheal intubation was performed after the disappearance of all four twitch responses, using a video-laryngoscope and bougie, given the Cormack Lehane grade of three. Anesthesia was maintained with isoflurane and intermittent doses of midazolam, fentanyl, and rocuronium. Intraoperative TEE confirmed preoperative echocardiography findings. Left internal mammary artery graft was used to bypass the left anterior descending artery, and a saphenous vein graft was used for the posterior descending artery. The ASD was closed with a pericardial patch using normothermic cardiopulmonary bypass (CPB) and cold, hyperkalemic cardioplegic arrest. The adequacy of ASD repair was confirmed by TEE.

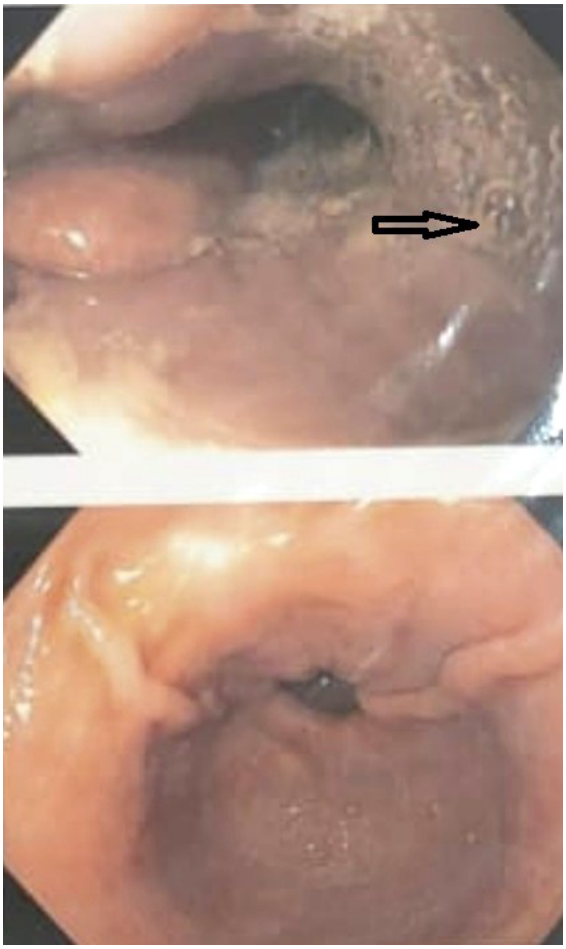

Figure 3: Upper gastrointestinal endoscopy showing residual food in the pyriform fossa sinuses (arrow).

Postoperatively, the patient was transferred to the cardiac intensive care unit (ICU). Following overnight ventilation, the weaning trial was attempted but the patient was agitated with jerky movements of the upper extremities. A repeat CT scan head and electroencephalogram were performed which showed no fresh changes. Intravenous levetiracetam (1gm loading dose, followed by 500mg 12 hourly) was started. After improvement of the neurologic status, the patient was extubated on postoperative day (POD) two. The early postoperative period was complicated by supraventricular arrhythmias (atrial fibrillation, atrioventricular block) which responded to intravenous amiodarone and pacing. On POD three, clear liquids followed by a soft diet were commenced, but the patient developed recurrent vomiting and dysphagia. An upper gastrointestinal endoscopy showed residual food in the hypopharynx and pyriform fossa sinuses (Figure 3). Oral feeding was stopped, Ryle's tube feeding was started, and the patient was transferred to the high-dependency unit.

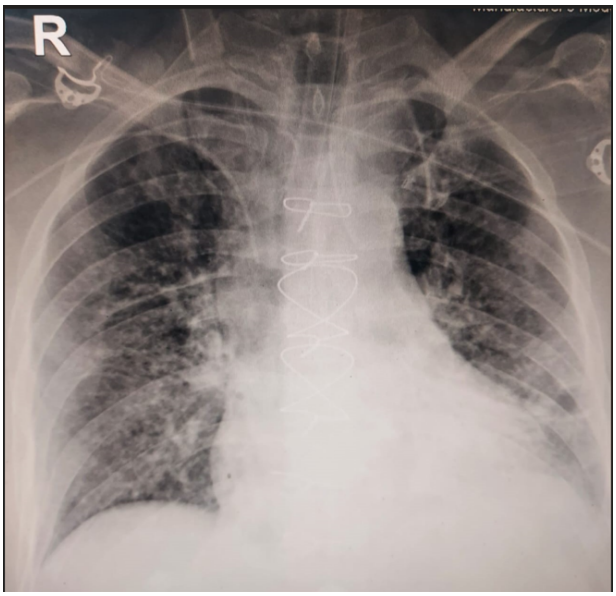

Figure 4: Postoperative chest X-ray showing features of aspiration pneumonitis.

In the high-dependency unit, the patient developed breathing difficulty associated with a decrease in oxygen saturation (87%). He was transferred back to the ICU with a suspicion of aspiration pneumonitis and required reintubation and mechanical ventilation (Figure 4). Bronchoscopy was done and the tracheal secretions showed *Klebsiella pneumoniae*; the antibiotics were escalated according to the sensitivity. After one week of ventilation and several failed trials of weaning, percutaneous tracheostomy was done. The patient was gradually weaned from the ventilator on POD 18. For hypersomnia, modafinil was restarted, and the patient appeared more alert. The ICU team worked closely with

the respiratory therapists to prevent further respiratory complications. Given the long-term need for enteral feeding, percutaneous endoscopic gastrostomy (PEG) was performed. The patient and the family were educated about PEG feeding. The patient was discharged from the hospital to domiciliary care on POD 27 with advice for a regular follow-up. One month after discharge, on a follow-up visit, the patient was taking oral feeds and vocalizing with a small tracheostomy tube in situ.

Discussion

Myotonic dystrophy is the most common and severe form of myotonic syndromes with an incidence of 1 in 8,000 newborns and a prevalence of 2-14 per 100,000 population [4]. It is caused by an abnormal expansion of a trinucleotide cytosine-thymine-guanine sequence found on chromosome 19q13.3. The classical clinical features of type 1 myotonic dystrophy (Steinert's disease) in this patient were related to cranio-bulbar weakness and consisted of change in voice (nasal twang), dysphagia, hypersomnia, facial weakness, and ptosis. The multisystem involvement included the eyes (cataract), endocrine system (diabetes mellitus), central nervous system (hypersomnia), gastrointestinal system (dysphagia), and the heart. Cardiac involvement in myotonic dystrophy is characterized by conduction system abnormalities, supraventricular and ventricular arrhythmias, mitral valve prolapse, and, less frequently, myocardial dysfunction and ischemic heart disease [5]. Signs of myocardial ischemia may not be apparent in these patients due to their limited physical activity. Cardiac fibrosis and fatty infiltration affect the conduction system at sinoatrial and atrioventricular nodes, His–Purkinje fibres, providing a substrate for conduction defects, ectopic activity, and re-entrant arrhythmias [6]. Only a few cases of myotonic dystrophy patients undergoing cardiac surgery are published in English literature [7,8].

In our case, during CPB, normothermia was instituted as hypothermia is known to aggravate myotonia. The membrane of the dystrophic myotonic muscle is extremely sensitive to changes in extracellular potassium concentration. Hyperkalemia in myotonic dystrophy patients initially causes muscle hypotonicity, followed by hypertonicity upon further elevation of the serum potassium level. However, there are case reports about the safe use of mild hypothermia and cold hyperkalemic cardioplegic arrest in patients with myotonic dystrophy undergoing cardiac surgery [7]. The anesthetic plan was tailored to minimize the risk of cardiac and respiratory complications. If a difficult airway is anticipated, a video-laryngoscope, bougie, and quick assistance must be immediately available. While using rocuronium, sugammadex should be quickly available in case of failed intubation, and a decision to awaken the patient is taken. Succinylcholine can trigger a myotonic crisis causing difficulties in tracheal intubation and ventilation. In patients with muscular deficit or weakness, nondepolarizing muscle relaxants can cause a marked delay in muscle strength recovery. Because of the marked sensitivity to acetylcholine, neostigmine should not be used to antagonize neuromuscular block in myotonic patients. For all these reasons, short-acting nondepolarizing muscle relaxants that do not need reversal are preferred. Anesthetic concerns are also related to the extreme sensitivity of these patients to sedatives, opioids, and anesthetic agents. The risk of malignant hyperthermia is controversial and total intravenous anesthesia has been used in some reports. However, inhalational agents have been used safely along with monitoring of the depth of anesthesia, neuromuscular function, and temperature in these patients [9]. Other factors such as hypothermia, shivering, or mechanical or electrical stimulation during surgery can precipitate a myotonic crisis.

Postoperatively, these patients require close monitoring and management of respiratory complications including respiratory infection; and there may be feeding issues due to the worsening of dysphagia [10]. Postoperative respiratory complications may occur due to weakness and atrophy of respiratory muscles, poor cough efforts, impaired response to hypoxia, and somnolence. Although direct involvement of pulmonary or bronchial tissues is not reported, myotonia in the pharynx and esophagus can cause difficulty in swallowing and tracheal aspiration. Evaluation of all organ systems and timely referral for physical, occupational, speech, and swallowing therapy may improve outcomes [11]. A multidisciplinary team should implement tools to identify the risks and specific critical issues in these patients using a proactive approach.

Conclusion

Patients with myotonic dystrophy can present with a range of symptoms including but not limited to cognitive dysfunction, cerebrovascular accidents, anxiety, sleep disorders, and cardiac disorders. Cardiac surgery in these patients can be challenging, but with careful planning and close monitoring, successful outcomes can be achieved. A multidisciplinary team consisting of cardiologists and neurologists is essential for the early diagnosis of the multiorgan disorders and to provide optimal care for the patient.

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Conflict of Interest: There are no financial interests or any conflict of interest.

Patient Consent Form

The permission to publish this case report was taken from

the patient (as described in the "Case Presentation" section).

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