Case Report

Sedation with dexmedetomidine in myotonic dystrophy patient case report

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ABSTRACT

15-year-old male patient, with Steinert's desease, was proposed for orthopedic surgery in left foot. He was sedated by continous perfusión of dexmedetomidine and an ultrasound-guided popliteal block was performed. The level of sedation achieved was adequate, without observing further adverse effects except of bradycardia with initial bolus. Dexmedetomidine was proved to be useful in this case; however, use of the drug should be carefully started at a low initial dose in patients with myotonic dystrophy.

Key Words: *Steinert's desease, Anesthesiologists, Myotonic dystrophy, Respiratory systems*

ABBREVIATION

DM1: Myotonic dystrophy type 1 or Steinert's disease; BMI: Body Max Index; ICU: Intensive Care Unit; ASA: American society of anesthesiologists; ECG: Electrocardiogram

INTRODUCTION

Type 1 myotonic dystrophy, known as Steinert's disease, is an autosomal dominant multisystemic disorder that affects the musculoskeletal, central nervous, gastrointestinal, endocrine, cardiac, and respiratory systems. DM1 has an incidence of 1 in every 8,000 births with a worldwide prevalence of 2-14: 100,000 [1,2].

It develops as a result of an expanded CTG triplet repeat of a non-coding DNA segment in the DMPK gene in chromosome 19q13.3. Although the gene is located on chromosome 19, there is no association with malignant hyperthermia [3,4].

Written consent for publication has been obtained from the patient guardians.

Classic symptoms of DM1

DM 1 can be classified depending on the time of onset symptoms [5]. There is congenital, infantile and adult form. Their development and characteristics are different although some symptoms may be similar. Classification and symptoms summary are shown in Table1.

CASE DESCRIPTION

15-year-old male patient, 65 kg, BMI 21, was programmed for surgery of plantar flexor tenotomies of the second to fifth toe of the left foot. He was previously diagnosed with congenital myotonic dystrophy type 1 (Steinert Syndrom) and had a brother with the same pathology.

A pre-anesthetic evaluation was performed prior to surgery where the latest reports from different medical specialists were collected. His periodic cardiology check-ups were normal. Neurologically, he presented moderate cognitive developmental delay and distal weakness in the hands and leg. The patient had a typical "myotonic facies" with a favorable airway. Obstructive sleep apnea syndrome was not suspected (but without recent polysomnography), no recent respiratory infections were present.

He was premedicated with bromazepam 0.5 mg orally 30 minutes prior to surgery and did not present anxiety during transfer to the operating room. Basic ASA monitoring was used (EKG, non-invasive blood pressure, heart rate and oxygen saturation). A peripheral venous catheter was placed in the left hand, where continuous perfusion of dexmedetomidine was connected, prior bolus administration of 1 mcg/kg for 10 minutes and continuing throughout the procedure at 0.5 mcg/kg/minutes. During the initial bolus, a decrease in the heart rate from 65 bpmto 44 bpm was observed. We administered 0.1 mg/kg of atropine and decreased the initial bolus dose to 0.8 mcg/kg, resolving the bradycardia. Subsequently, we proceed to perform an ultrasound-guided popliteal block administering 8 ml of lidocaine 1% and 8 ml of levobupivacaine 0.5%. Oxygen therapy was administered throughout the procedure with nasal dispositive at 2 bpm. The patient did not suffer any desaturations or airway obstruction.

The surgery went uneventful; we administered metamizole 0.4 mg/kg as an analgesic and dexamethasone 4mg to prevent postoperative nausea and vomiting.

The level of sedation achieved was adequate, the patient was comfortable, easily awakable and cooperated. After the surgery, the dexmedetomidine infusion was withdrawn without observing further adverse effects. The patient was transferred to the Intermediate Care Unit without agitation or anxiety. The patient lived close to the hospital and had good family support; hence we decided to discharge him home and followed up with phone calls in the evening and the following morning. The pain was controlled by oral NSAIDs.

DISCUSSION

Case discussion

There is limited literature about the use of dexmedetomidine in patients with DM1. Yoshino et al. [6] described the case of a 53-year-old woman with DM1 for a total abdominal hysterectomy with regional anesthesia and dexmedetomidine as a sedative agent. Airway obstruction was observed after the initial bolus at 2 µg/kg, therefore authors conclude that dexmedetomidine should be carefully started at a low initial dose in patients with DM1. Gaszynski T [7] report the anesthetic management using an opioid-free method of a patient with Steinert Syndrome under general anesthesia for laparoscopic cholecystectomy with a continuous infusion of dexmedetomidine as adjunct. Bolus dose was 0.6 µg/kg over 10 minutes followed by continuous infusion over 0.2 µ/kg/hour combined with propofol for maintenance of general anesthesia. In both cases the use of this drug was safe without adverse events reported. Our case is more similar to the one described by Yoshino et al. regional anesthesia associated with dexmedetomidine as a sedative agent. We did not observe airway obstruction but bradycardia presented with initial bolus. This may suggest that dexmedetomidine should be carefully started at lower initial dose in patients with DM1.

It is important to clarify that none of the above mentioned patients, including the one described in our case report, suffered from arrhythmias or serious respiratory problems.

Dexmedetomidine reduces heart rate and blood pressure through

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Table 1) Classification and Symptoms of DM [1].

	Congenital DM1	Childhood-onset DM1 (Symptoms of the adult form appear during the second decade)	Adult-onset DM1 (Classical form, first symptoms appear 15-35 years of age)
Facial abnormalities	Bilateral weakness	No "typical facies"	Cataracts "Typical facies": bifacial weakness, mild ptosis, progressive wasting of the muscles of mastication. Frontal balding.
	"Myopathic facies" (open,tent-shaped mouth)	Weakness of facial and neck muscles	
Central nervous system:	Psychomotor impairment	Mental handicap; difficulties in learning and speech	Psychosocial dysfunction and personality traits.
• Mental retardation	Delay in speech and motor development		Increased daytime sleepiness and obstructive sleep apnea
Endocrine dysfunction			Diabetes mellitus, hypogonadism, dysthyroidism
Gastrointestinal dysfunction:	Severe risk of aspiratorion	Could be present	Could be present, also constipation, gallstones, elevated G-GT
• Dysphagia			
• Squeletal muscles	Talipes equinovarus	Early motor development normal or moderately delayed	Distal weakness mild to severe.
Weakness	Other contractures could be present	Mild-moderate distal weakness	
• Myotonia	Mild to severe. Hypotonic and inmobile at birth.	Present mild-moderate myotonia	
	Not present before 3-5 years of age		
Cardiac dysfunction (arrhythmias, cardiomyopathy)	Often present but not symptomatic during childhood	Cardiac problems appear during the second decade	Always present: Arryhythmias: Atrioventricular block, atrial fibrillation,.. Dilated cardiomyopathy Sudden cardiac death
Prognosis	Mortality rate of 50% in the neonatal period and 25% in the first year.	Variable	During fifth-sixth decade are more common: chest infection by aspiration, respiratory failure, sudden cardiac death
	Death before 30 years of age (mainly cardiac causes)		

central sympatholysis but at higher concentrations it causes peripheral vasoconstriction leading to hypertension. It does not often cause deep sedation and patients can be easily awakened. Therefore, it is not suitable for patients who do not tolerate these effects, for example pediatric cases or patients with cognitive developmental delay. Fortunately, In the present case, the patient cooperated. We should have special caution when administering dexmedetomidine to patients with pre-existing bradycardia. Data on the effects of dexmedetomidine in patients with a heart rate <60 are very scarce and special care should be taken. Bradycardia does not normally require treatment, but it generally responds to anticholinergic medications or to dose reduction, as in the case of our patient [8,9].

Anesthesia and myotonic dystrophy

Preoperative period: Preoperative evaluation of patients with DM should involve a multidisciplinary team including medical, neurology, cardiac, and anesthesiology specialties. Any preoperative weakness should be addressed and further evaluated. In patients with respiratory symptoms, we should consider pulmonary function testing. Patients may present with Cardiac rhythm management devices like pacemakers and defibrillators.

Patients suffering from DM are at higher risk of aspiration due to reduced gastric emptying and pharyngeal muscle dysfunction, preoperative administration of any sedatives should be avoided as it will further aggravate respiratory depression. Consideration should be given to preoperative prophylaxis with sodium citrate, metoclopramide, and H2 antagonists to prevent aspiration pneumonia.

Intraoperative period: Regional anesthesia with minimal sedation is the best option whenever possible. Spinal and epidural anesthesia have been reported to be successful and safe, either as a sole anesthetic or as a part of postoperative analgesia. As there are case reports of shivering and precipitation of myotonic crisis with uterine atony after epidural anesthesia for Cesarean section patients should be closely monitored during spinal or epidural anesthesia. During general anesthesia, muscle relaxants should be avoided. If muscle relaxants need to be given, succinylcholine should be avoided and complete muscular block reversal should be ensured. Sugammadex is preferred to neostigmine as muscle reversal. The fear that neurostimulation during the peripheral nerve block might precipitate myotonia may not be an issue these days as nerve stimulator can be avoided due to ultrasound availability. We should not forget that patients with DM are very sensitive to opiates and anesthetic agents since they have a higher risk of respiratory depression and postoperative ileus.

Apart from standard ASA monitors, neuromuscular block and temperature monitors should be applied. Rapid sequence induction should be the chosen as these patients are at higher risk of aspiration due to pharyngeal muscle weakness. DM can be precipitated intraoperatively by hypothermia, shivering, surgical or mechanical stimulation and electro cautery. Availability of temporary pacemakers and defibrillators should be ensured as well.

Postoperative period: Many cases of perioperative complications have been reported in patients with DM and most of them are respiratory continuous pulse oximetry and ECG monitoring is necessary for longer period depending on type of the surgery, drugs administered, surgical time, Furthermore, postoperative ventilation in patients with high risk of pulmonary complications should be considered. It is recommendable to restrict use of opioids and apply multimodal pain management after surgery. Pulmonary toilet with incentive spirometry, chest physiotherapy, and cough assistant devices play an important role in the sooner recovery of these patients.

There is no data of association with malignant hyperthermia, so it seems safe to use inhalation agents. The only case that presented intraoperative myotonic crisis was a hemorrhoidectomy performed under subarachnoid anesthesia. It was resolved by increasing sedation with propofol, so the causes of this complication could be the hypothermia in the operating room or surgical stress.

Respiratory complications are by far the most frequent in this type of patients

and most of them occur in cases of general anesthesia, even without the use of muscle relaxants. They are of great importance since some cases required mechanical ventilator support after surgery and consequent admission to the ICU. As previously mentioned, regional anesthesia, spinal or peripheral blocks, are probably safer options for these patients.

LIMITATIONS

In rare diseases like DM, most studies are based on reviews of clinical cases. There are no prospective randomized studies and it would be hard to develop them. Therefore, the sample of published cases is quite heterogeneous according to type of surgery, type of anesthesia, complications, characteristics and status of each patient, So the possibility of establishing statistical relationships is difficult and not completely reliable.

CONCLUSION

Rare multisystemic diseases are an anesthetic challenge. The evaluation of the case starts during preoperative assessment and an anesthesia plan should be formulated to avoid complications. When it comes to reducing adverse events, both intraoperative and postoperaive care is important. The postanesthetic vigilance varies depending on patient state, type of surgery, type of anesthesia, to minimize hospital stay.

FINANCIAL DISCLOSURES

None

CONFLICTS OF INTEREST

None

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