CLINICAL VIGNETTE

Management of a Patient with Myotonia Congenita Undergoing Patella ORIF

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Case

A 58-year old female with past medical history of myotonia congenita, anxiety, and gastroesophageal reflux disease, presented with a left patella fracture after suffering a ground-level fall. She was evaluated by the orthopedic service and scheduled for open reduction and internal fixation of her left patella. Preoperative vital signs, complete blood count, basic metabolic panel, and electrocardiogram were within normal limits. The history of myotonia congenita raised concern of increased anesthetic risk.

She was diagnosed with myotonia congenita 5 years previously with complaints of upper extremity muscle tightness predominantly in the mornings. She was prescribed mexiletine, a class 1b antiarrhythmic, but never took the medications as her symptoms improved without treatment. She denied previous surgeries or personal, or family history of anesthetic complications including malignant hyperthermia or muscle spasms.

On the day of surgery, the anesthesia machine was flushed with oxygen for 1 hour and halogenated anesthesia gas vaporizers were removed for malignant hyperthermia precautions in the event that general anesthesia was necessary. Anesthesia consent was obtained for a left femoral nerve block with intravenous sedation and general anesthesia if necessary. Preoperatively, patient received a single-shot ultrasound guided femoral nerve block with 35ml of 0.5% ropivacaine. Midazolam 2mg and fentanyl 50mcg IV was administered during the procedure for light sedation. Local anesthetic spread surrounding the femoral nerve was visualized on ultrasound imaging and no paresthesia or pain was noted during the procedure.

Patient was brought to the operating room and transferred to the operating room table with placement of standard anesthesia monitors and an upper body forced air warming blanket. Supplemental oxygen was provided with a simple face mask at 6 liters/minute. After routine surgical time out, patient was given cefazolin 2 grams IV for antibiotic prophylaxis and an additional midazolam 2 mg and fentanyl 50 mcg IV. Moderate sedation was maintained with propofol infused at 40 mcg/kg/min IV.

The surgery commenced with a midline incision overlying the patella. The patient had no reaction to incision. A simple transverse fracture of the patella was identified, manually reduced, and fixated with K-wires by the surgeons. After closure of incision and application of dressings, patient's knee was immobilized in a fully extended position with a hinged knee brace. Total surgery time was 1 hour and 52 minutes with an estimated blood loss of 25 ml. Lactated Ringer's 900 ml IV was administered during the case.

Patient was taken to the post-anesthesia care unit where she was awake and alert with no pain. She required no additional analgesia and was discharged home after 30 minutes of monitoring. Patient was seen in orthopedic clinic on post-operative day 9 for routine follow up. She was doing well overall with no evidence of hardware complication nor any numbness and tingling in the extremity.

Discussion

Myotonia congenital (MC) is a congenital form of muscular dystrophy characterized by uncontrolled temporary skeletal muscle excitability as a result of mutations in the muscle chloride channel gene (CLCN1). Two forms have been described - Type 1 is autosomal dominant and also known as Thomsen disease; Type 2 is inherited in a recessive fashion and is also known as Becker myotonia.[1] Thomsen disease is usually milder than the recessive form, in which myotonia may be associated with some weakness. It may be difficult to determine the pattern of inheritance, especially with sporadic cases, as members of affected families who carry the abnormal mutation may be asymptomatic or only mildly involved. A thorough evaluation of the family with appropriate genetic testing is usually required.[2] Patients classically exhibit a "warm up" phenomena where after resting, the muscles are initially stiff and difficult to move but normalize with continued exercise. Treatment with mexiletine is often beneficial.[3]

The main concerns for an Anesthesiologist caring for a patient with MC are the risk of malignant hyperthermia, masseter muscle spasm, aspiration, and prolonged postoperative weakness. Although there is no confirmed link between MC and malignant hyperthermia, it has been reported in, several case reports.[1] If general anesthesia is necessary, a non-triggering technique should be used by avoiding potent volatile anesthetics and succinylcholine. The anesthesia machine is typically prepared by prolonged flushing of the breathing system and removal of potent volatile anesthetic vaporizers.

In addition to potentially triggering malignant hyperthermia, succinylcholine is contraindicated as it can precipitate intense

myotonic contractions and trismus which may prevent opening of the mouth for intubation.[4] If muscle relaxation is required, judicious use of a non-depolarizing muscle relaxant should be used. Rocuronium may be the non-depolarizing muscle relaxant of choice as direct binding and reversal with suggamadex is possible. Reversal of non-depolarizing muscle relaxation with neostigmine may aggravate muscle contraction by facilitating depolarization of the neuromuscular junction.[5]

Given that shivering is also a trigger of myotonia, patients with MC should be kept normo-thermic through the perioperative period with the use of forced air warmers, IV fluid warmers, and other measures as indicated.

The literature suggests that local anesthetics are well tolerated. Of note, lidocaine is also a class 1b anti-arrhythmic similar to mexiletine which is used to treat MC. One caveat is that solutions containing epinephrine should be avoided as epinephrine can also serve as a trigger for MC.[1]

In the presented case, the patient was an ideal candidate for a peripheral nerve block, which allowed avoidance of general anesthesia and its potential issues with MC such as MH, muscle relaxants, hypothermia, and airway instrumentation. In addition, the nerve block resulted in excellent post-operative pain control without the need for systemic opioids, facilitating rapid recovery from anesthesia and discharge from the recovery room. Alternatively, neuraxial anesthesia could also be considered for patients with MC as a means to avoid general anesthesia. Several case reports describe successful use of neuraxial anesthesia for obstretric patients with MC undergoing cesarean section.[6,7] For this particular case, a peripheral nerve block was chosen over a neuraxial anesthesia due to its superior duration of post-operative analgesia and less likelihood of hypothermia and shivering.

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