

Regional Anesthesia in a Patient with Familial Mediterranean Fever

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INTRODUCTION

- Familial Mediterranean Fever (FMF) is a rare hereditary condition characterized by recurrent episodes of painful inflammation that occur most commonly in the abdomen, but can also occur in the chest, joints, and rarely in the heart, membranes surrounding the brain and spinal cord, and testicles. These attacks can be accompanied by symptoms of fever, rash and headaches.
- Abdominal crisis occurs in 95% of cases, while patients with uncomplicated FMF are usually asymptomatic.
- Attacks develop within 1-2 hours following an event such as emotional stress, vary in intensity and usually last 12 to 72 hours.
- FMF predominantly affects people of Armenian, Arab, Turkish, or Jewish descent from the Mediterranean region with a prevalence of 1 in 200 to 1,000 people in these populations and less common in other populations.
- There exists documented case reports of patients experiencing episodes of serositis and fever triggered by the physical and emotional stress from surgery and anesthesia, specifically after the administration of spinal anesthesia.
- Little is known about the anesthetic considerations necessary to prevent and abort intraoperative pain episodes in patients with FMF.

CASE PRESENTATION

History of Present Illness:

- We present a case of a 53-year-old female patient with FMF who underwent monitored anesthesia care (MAC) and regional anesthesia for podiatry surgery (bunionectomy) at our institution. She has a history of FMF followed by rheumatology. This patient's FMF symptoms of fever and serositis were triggered by stress and anxiety but were controlled for the past six months on outpatient oral colchicine therapy 0.6 mg twice a day.

Physical Exam:

- Preoperative vital signs were within normal limits: temperature 36.2° C, heart rate 64 beats per minute, respiratory rate 20 breaths per minute, and blood pressure 114/56 mmHg. The patient was overweight measuring 167 cm and 72.8 kg, with a BMI of 26.1 kg/m². Preoperative airway exam demonstrated adequate mouth opening, a Mallampati class II airway, full cervical neck range of motion, midline trachea, and adequate thyromental distance. Physical exam, including cardiac and respiratory, was unremarkable.

Laboratory / Imaging:

- ECG (4/5/19) was only notable for flat t-waves in leads V3 and V4, but was present on prior reads. ECHO was unremarkable with trace mitral regurgitation and slightly thickened mitral valve leaflets with functional capacity of at least 4 METs. Laboratory was only notable for slight anemia with hemoglobin of 11.6 and leukopenia with WBC of 4.3.

Surgery / Anesthesia:

- Patient was instructed to be compliant with taking prophylactic colchicine 0.6 mg oral tablet twice a day, taken on day of surgery.
- In the operating room, the patient was given oral midazolam for anxiolysis and intravenous fentanyl for pain control prior to the performance of a left ankle peripheral nerve block.
- All five peripheral nerves of the left ankle were identified by anatomic landmarks and local anesthetic solution of ropivacaine 0.5% and lidocaine 1% mixture was used for the left ankle peripheral nerve block. The nerve block was uneventful.
- The patient's vital signs remained stable throughout the 3-hour uneventful procedure. At the conclusion of surgery, the patient was transported to the post-anesthesia care unit (PACU) with continued oxygen supplementation at 8 L/min via face mask. She continued to do well in the PACU and was later discharged home the same day.

DISCUSSION

Preoperative Management:

- Since the 1970's, colchicine has been identified as a lifelong treatment of choice for FMF, but only effective as prophylaxis and ineffective for acute attacks. The drug prevents attacks by controlling inflammation and halting the development of amyloidosis by specifically inhibiting neutrophil chemotaxis. It is ideal to optimize the dosing of the drug prior to proceeding with surgery based on a patient's age and their frequency and severity of symptoms.

Intraoperative Management:

- One documented report of an intraoperative FMF episode developed following administration of spinal anesthesia for a 20-year-old patient receiving pilonidal sinus surgery. Of note, he had been taking colchicine 2 mg/day for 3 years without exacerbation of attacks within the last year prior to surgery. The pain eventually resolved within 8 minutes after administration of intravenous fentanyl 100 mg and diclofenac 75 mg and surgery proceeded.
- We wish to draw attention to potential intraoperative problems in patients with FMF and provide potential promising measures to combat acute attacks:
 - Prior to the start of surgery, a patient's level of anxiety and stress should be assessed. Oral midazolam can be given for anxiolysis as needed.
 - Patient should be laid supine.
 - Deepening anesthesia level should be attempted to resolve abdominal pain.
 - IV fentanyl can be given for prophylactic pain control and maintain an adequate level of sedation prior to administration of a spinal or regional block.
 - Non-steroidal anti-inflammatory drugs can also be administered as another supportive therapy for pain and is recommended to be made available prior to the start of and during the surgery.

CONCLUSION

- Although FMF is a relatively rare hereditary condition in the general population, patients with such disorder may have serious complications of painful inflammation and fever. It is known that these pain attacks can be triggered by the administration of surgery or anesthesia, including spinal anesthesia.
- Our presentation of a 53-year-old female patient with FMF safely underwent MAC and uneventful regional anesthesia for podiatric surgery.
- However, given the potential for serious intraoperative pain episodes, it is important to perform careful anesthetic considerations with goals focusing on stress and anxiety reduction and prevention of acute attacks as well as becoming familiar with emergency management.

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