

Introduction

Mast Cell Activation Syndrome is a diverse, multisystem disease characterized by the accumulation of pathological mast cells in multiple organs with the release and degranulation of their mediators. Symptoms include facial flushing, itching, abdominal cramping, hypotension and anaphylaxis. In this case, the parturient patient presented to us with a history of MCAS. It is important to understand that the physical pain and psychological stress from pregnancy can induce flare-ups for a parturient with MCAS. Patients with MCAS have a higher risk of developing anaphylaxis after encountering a trigger. Therefore, it is important to have a proper anesthetic plan for the optimal well-being of the parturient.

Case

Our patient is a 25-year-old G1P0 with a history of moderate persistent asthma, anxiety, depression, allergic rhinitis, multiple allergies, chronic hypertension, and Mast Cell Activation Syndrome (MCAS). She experiences symptoms about three times per month, including facial and hand redness along with abdominal pain. She takes dupilumab, famotidine, cetirizine, and cromolyn sodium daily in order to suppress this mast cell activity.

Once patient arrives to L&D in labor (as patient is currently 32 weeks gestation during writing of this case report), a labor epidural should be done early in order to prevent stress from intrapartum pain which could cause an MCAS flare. Commonly used amide local anesthetics are safe to use (ester local anesthetics' metabolites can cause MCAS activation). Duramorph, however, is contraindicated due to its histamine release and instead, fentanyl should be used.

All her home medications were advised to be taken the day of admission and throughout her stay. She has been advised to take iron due to increased risk of postpartum hemorrhage in patients with MCAS. If an MCAS flare were to occur during labor, fibrinogen levels will be monitored and treated with either TXA or cryoprecipitate in order to prevent fibrinolysis and a type and cross will be ordered once a patient is admitted with two units of blood on hold.

In the event that the patient was to develop an anaphylactoid or anaphylaxis reaction, the anesthesiologist on duty will be prepared to have epinephrine, H1 blockers, and steroids. If an induction of general anesthesia were to occur, commonly used anesthetics such as propofol, midazolam, and ketamine would be safe to use except for etomidate. As for paralytics, vecuronium and cisatracurium will be used. The safety of succinylcholine and rocuronium in a patient with MCAS are unknown, however. The use of volatile agents is considered safe for maintenance of anesthesia as well as anti-emetics. For pressor support, phenylephrine and ephedrine are considered safe to use.

Lastly, cardiovascular agents such as atropine and beta-blockers will not be used as they can exacerbate mast cell activation. In order to treat the patient's hypertension during labor, the patient will be treated with hydralazine or methyldopa (since the patient is allergic to nifedipine). Although vaginal delivery can cause genitourinary mast cell activation, it is preferred over a cesarean section, which poses more surgical stress for the patient and can cause mast cell activation.

Discussion

Our patient's MCAS was idiopathic since there was no proliferation of abnormal mast cells seen (primary) as well as no identifiable trigger (secondary) that could account for her MCAS. MCAS may be triggered by allergen-specific immunoglobulin E, physical factors, and medications. The diagnosis of MCAS is made if the patient meets 1.) episodic signs and symptoms involving at least two organ systems (skin, upper or lower respiratory tract, gastrointestinal, or cardiovascular), 2.) increases of baseline serum tryptase level greater than 1.2 x baseline +2ng/ml after an episode (or an increase of more than 100% of patient's baseline of other mediators such as 2,3-dinner-11-beta-PGF2-alpha, N-methyl-histamine or LTE4 in a spot urine sample), and 3.) response to medications that stabilize mast cells, reduce production, release, and action of mast cell mediators.

The key difference between systemic/cutaneous mastocytosis and MCAS is that skin findings of urticaria pigmentosa (UP) are not seen in MCAS as they are unique to SM/CM only. Another distinguishing feature of SM is baseline tryptase level >20 ng/ml, whereas MCAS baseline is normal or slightly elevated.

It is imperative to understand the triggers that can cause an MCAS flare during labor, including the stress of labor and surgical stressor with a cesarean section. Careful consideration should be made to prevent any mast cell degranulation with proper use of premedications including antihistamines (preferably H2 blockers, helpful to reduce abdominal pain and nausea), leukotriene receptor antagonists, and corticosteroids prior to receiving any invasive procedures requiring anesthesia.

It is also important for the patient to receive an epidural early on during her labor and avoiding cesarean section as well as certain medications that cause mast cell mediator release including morphine, ester local anesthetics, NSAIDs, induction medications such as etomidate, certain paralytics, as well as atropine and beta-blockers. Extremes of temperature should also be avoided.

One should anticipate a higher risk of postpartum hemorrhage with an MCAS flare (which causes fibrinolysis) and an increased risk of anaphylaxis with prompt treatment of epinephrine, fluids, and airway management. After delivery, patient should be sent to a step-down floor for closer observation in case of MCAS flare as the patient can continue to have activation of triggers during postpartum period as well as for several months after delivery. In certain literature, it has been reported that omalizumab, cyto-reductive and immunomodulating therapies has been shown to prevent symptoms and reduce adverse events.

It is therefore prudent in creating a preoperative plan from the start with thorough history taking from the patient, excellent communication between the anesthesia and OB staff, avoidance of known and potential triggers, and management of perioperative crises with a parturient patient with MCAS.

References

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