

Combined Spinal Epidural (CSE) Anesthesia for Labor Analgesia in a patient with Mast Cell Activation Disorder (MCAD)

John Rouck MD, Michael Kim MD, Suzanne Mankowitz MD: Columbia University Irving Medical Center

MCAD is an extremely rare family of disorders characterized by accumulation of pathologic mast cells and associated release of mast cell mediators.

Mediator release can be triggered by environmental allergens (pollens and pet dander), temperature swings, stress, food preservatives (sulfites, nitrites, benzoates), dyes (frequently food dye and radiocontrast dye), and medications including beta lactams and NSAIDs **(1, 2)**.

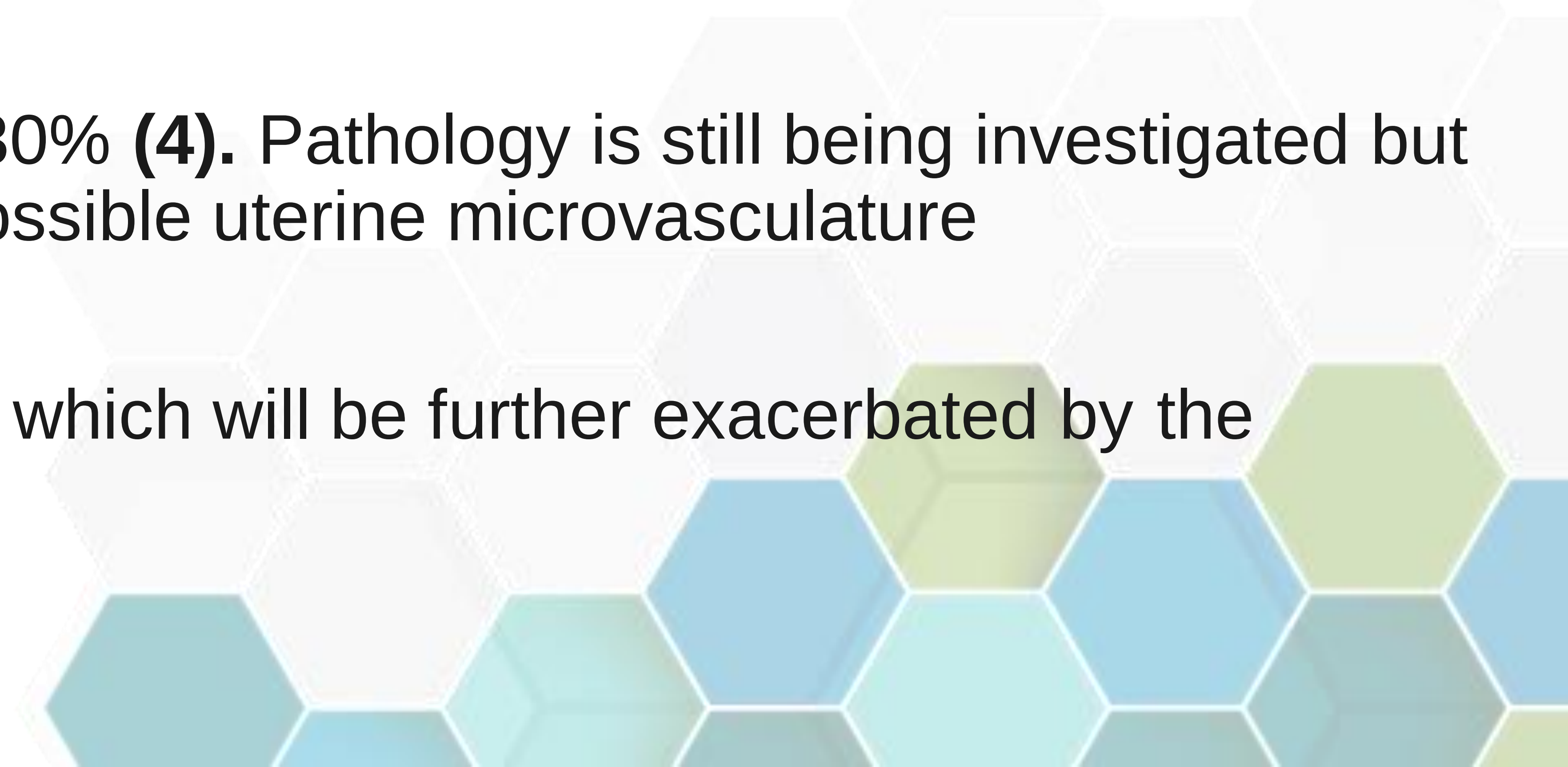
Symptoms include organomegaly, diarrhea/GI upset, severe osteoporosis, cutaneous involvement (flushing, angioedema, urticaria, nodule formation), respiratory effects (such as airway edema), tachycardia, blood pressure fluctuations (hypotension and lability), pain, and anaphylaxis **(1, 2)**.

Pathogenesis of pain thought to be due to mast cells releasing inflammatory mediators around nerve endings **(3)**.

Literature shows a complex relationship between MCAD and Pregnancy: Symptoms may improve or worsen. In one study 20-33% of women with mastocytosis reported increased symptoms during pregnancy, while in another 30% experienced symptom improvement during the first trimester **(4, 5)**.

Additionally, pregnant patients with mastocytosis were found to have a miscarriage rate of 25-30% **(4)**. Pathology is still being investigated but thought to be due to an increased inflammatory state caused by mast cells in the uterus and possible uterine microvasculature hypercoagulability **(6, 7)**.

MCAD patients with bone marrow involvement may have existing anemia or thrombocytopenia which will be further exacerbated by the increased circulating volume in pregnancy.



34 year old G3P0020 @38w2d with known MCAD presented for scheduled induction.

When symptomatic her symptoms included pruritic skin lesions, nausea, and diarrhea. She also had 3 syncopal episodes. She had a prior bone marrow biopsy without evidence of bone marrow involvement.

Symptom control was achieved through treatment with Omalizumab, Cetirizine, Ranitidine, and Montelukast. She was also taking Sertraline for anxiety and depression.

At time of presentation her symptoms were well controlled and she had not had a flare in 5 months.

Prior flares consisted of diarrhea, pruritic hives, and painful bumps on her tongue. Her flares would last for 2 days. These flares were triggered by alcohol, heat, stress, and certain perfumes. During flares she would take an additional Ranitidine dose. She never required epinephrine for symptoms.

She had a platelet count of 220 the day prior to her scheduled induction.

She took all of her daily medications as directed on day of induction of labor and continued them throughout admission.

Patient desired analgesia and **Combined Spinal Epidural Analgesia was performed.**

Patient was premedicated 1 hour prior to CSE with oral diphenhydramine 25 mg and oral prednisone 50 mg as suggested by her hematologist. This is thought to reduce chances of life threatening anaphylaxis but evidence is limited **(2)**.

CSE was performed without complication using spinal bupivacaine and fentanyl. Continued epidural analgesia was provided for approximately 31 hours via intermittent epidural bolus with bupivacaine 0.0625% mixed with fentanyl 2 mcg / mL and further supplemented with two physician boluses of bupivacaine and fentanyl.



Discussion and Takeaway

Patients should continue their ongoing controller medications (frequently consisting of anti-histamines, leukotriene inhibitors, and immunomodulators) at time of delivery. Antihistamines can be useful if patients start to develop flare symptoms **(2, 8)**.

Spontaneous anaphylactoid reactions can occur despite meticulous preventative measures. Epinephrine, antihistamines, and corticosteroids must be readily available at bedside.

Special attention must be paid to patients with known bone marrow involvement.

This case report adds to the growing body of literature that **neuraxial analgesia is safe for pregnant patient with mast cell disorders (2, 4, 6)**.

Triggers

- Stress
- Pain
- Temperature swings
- Medications
- Food Preservatives
- Dyes
- Environmental allergens

Preventative Measures

- Continue daily medications
- Early neuraxial analgesia: avoid pain and stress
- Post labor neuraxial for pain control
- Premedication prior to neuraxial with Antihistamines and Steroids
- Prepare to rapidly treat spontaneous anaphylaxis by keeping Epinephrine, steroids, anti-histamines at bedside.
- Communication with the patient's hematologist and or allergist can be beneficial.

1. Valent P, Sperr WR, Akin C. How I treat patients with advanced systemic mastocytosis. *Blood* 2010;116:5812-7

2. Bonadonna, P., et al. "Drug hypersensitivity in clonal mast cell disorders: ENDA/EAACI position paper." *Allergy* 70.7 (2015): 755-763..

3. Wirz, Stefan, and Gerhard J. Molderings. "A Practical Guide for Treatment of Pain in Patients with Systemic Mast Cell Activation Disease." *Pain Physician* 20.6 (2017): E849-E861.

4. Ciach K, Niedoszytko M, Abacjew-Chmylko A et al. Pregnancy and Delivery in Patients with Mastocytosis Treated at the Polish Center of the European Competence Network on Mastocytosis (ECNM). *PLoS One* 2016;11:e0146924

5. Watson KD, Arendt KW, Watson WJ, Volcheck GW. Systemic mastocytosis complicating pregnancy. *Obstet Gynecol* 2012;119:486-9.

6. Afrin, Lawrence B., et al. "Often seen, rarely recognized: mast cell activation disease—a guide to diagnosis and therapeutic options." *Annals of medicine* 48.3 (2016): 190-201.

7. Derbala, Youssef, et al. "Mast cell–induced immunopathology in recurrent pregnancy losses." *American Journal of Reproductive Immunology* 82.1 (2019): e13128..

8. Pardanani A. How I treat patients with indolent and smoldering mastocytosis (rare conditions but difficult to manage). *Blood* 2013;121:3085-94.