Mastocytosis in Our Pediatric Patients: A Rare Disease with Specific Anesthetic Challenges and Considerations

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Introduction

- **Definition**: Mastocytosis consists of a variety of clinical features characterized by increased mast cells in systemic organs.
- **Presentation**: Patients present with either cutaneous mastocytosis [(CM) - typically in childhood] or systemic mastocytosis (SM).
- **Incidence:** Estimated at 1/150,000[2].
- **Pathophysiology**: Mutation of the mast cell receptor which induces constitutive activation (found in 50% of CM cases and >90% of SM cases)[2]. These mast cells have the potential to degranulate and release inflammatory mediators such as histamine, tryptase, and cytokines throughout the body[1] (Figure 1).
- **Anesthetic management**: Requires knowledge of triggers/drugs that can cause mast cell degranulation and preparedness to treat cardiovascular collapse.

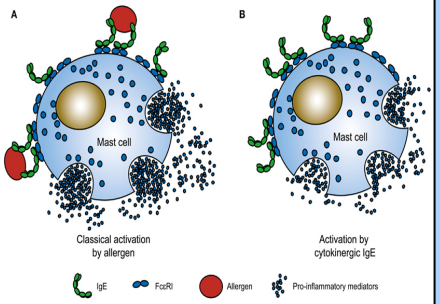

Figure 1: Mast Cell Degranulation. Reprinted from "Cytokinergic IgE action in mast cell activation" by H.J. Bax, 2012, *Frontiers in Immunology,* Copyright 2012. https://doi.org/10.3389/fimmu.2012.00229

Case Presentation

17-year-old female with CM presents with worsening urticarial rash and new systemic symptoms over the last year for a diagnostic bone marrow biopsy to evaluate for SM.

- **Past Medical History**: Diagnosed with CM in childhood with symptoms of urticarial rash flares 2-3 times a year.
- **History of Present Illness**: For the last 12 months, flares occur 2-3 times weekly with gastrointestinal discomfort, flushing, headaches and arthralgias.
- **Home medications**: Ranitidine, Montelukast and an Epi Pen.
- **Pre-anesthetic Plan**: Maintain normothermia, pain control, minimize stimulation, and encourage anxiolysis to prevent mast cell degranulation and histamine release.
- **Pre-medications**: Midazolam and dexmedetomidine for anxiolysis, fentanyl for analgesia, and diphenhydramine and dexamethasone to prevent histamine release.
- **Anesthetic**: Monitored anesthesia care with a native airway. Propofol was used for induction and maintenance of anesthesia.
- **Intraoperative Course**:
  - Hemodynamic stability constant throughout.
  - New urticarial rash observed at the site of betadine prep for the biopsy.
  - Pre-existing urticarial rash on her neck increased in size (Figure 2).
- **Postoperative Course**: The patient was monitored under the care of an anesthesiologist in the postoperative area for 1 hour with no further issues. Rashes resolved in a few hours.

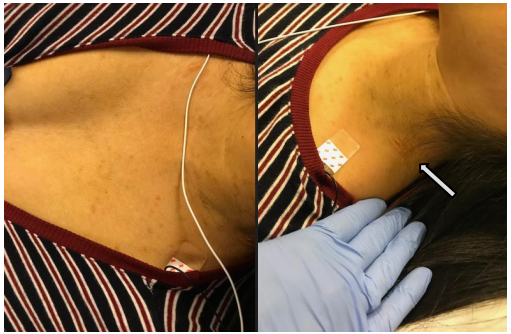

Figure 2: Evolution of rash before (left) and after (right) anesthetic

Discussion

- Mastocytosis patients are sensitive to triggers of mast cell degranulation.
- Avoid stress, pain, changes in temperature, light anesthesia, or friction to the skin.
- Knowledge of patient specific triggers is important and drugs that can cause histamine release should be avoided or administered with caution[3].
- While many anesthetic drugs can instigate mast cell degranulation (ex opioids, muscle relaxants, analgesics, and volatile anesthetics), studies have shown that routine anesthetic techniques can be used in these patients as long as providers:
  1) Pre-medicate patients thoughtfully
  2) Anticipate potential reactions
  3) Prepare for possible cardiovascular collapse[1]
- Patients should be monitored after the procedure because symptoms may take 1-2 hours to appear.

References

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