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Challenging Anesthesia for a Lapidus Bunionectomy in a patient with Systemic Mastocytosis

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Abstract: Systemic Mastocytosis, a rare lymphoproliferative disorder that results in spontaneous mast cell degranulation, presents a unique challenge for the Anesthesiologist. We describe a patient with this disease who presented for routine outpatient foot surgery. In the past, the patient had experienced anaphylaxis as well as episodic mast cell mediator release, resulting in significant nausea, vomiting, pruritis, urticaria pigmentosa, and ostealgia. While precise guidelines for premedicating these patients are lacking, we were able to achieve a successful outcome by premedicating the patient with both H1 and H2 blockers, a mast cell stabilizer, and steroids.
Keywords: Anesthesia, Systemic Mastocytosis

INTRODUCTION

Mastocytosis is a rare myeloproliferative disorder characterized by accumulation of mast cells in the skin and/or internal organs. Symptoms result from excessive mast cell mediator release, especially histamine, and range from mild pruritus to full blown anaphylaxis [1].

Anesthetic management of these patients can be challenging since mast cell activation and histamine release can be triggered by trauma, stress, extremes of temperature, and drugs [2]. Potentially problematic medications include opiates, anesthetics, and antibiotics. We report a patient with systemic mastocytosis and multiple drug allergies presenting for outpatient foot surgery, and describe the measures taken to minimize the potential for triggering mast cell degranulation.

CASE REPORT

A 52 year-old female with hallux valgus deformity presented for left foot bunionectomy. She had systemic mastocytosis, diagnosed by bone marrow biopsy after years of urticaria pigmentosa and GI complaints; including diarrhea and gastritis; and asthma.

The patient had multiple drug allergies including penicillin, cephalosporins, percocet, oxycontin, lidocaine, cortisone, macrodontin, and iodine contrast media. Her drug allergy symptoms included urticaria pigmentosa, pruritis, and ostealagia. She developed anaphylaxis in the past following ant bites and tessalon use. The patient underwent a bunionectomy on her right foot 5 months earlier which was complicated by severe post-operative nausea and vomiting, pruritis, and ostealgia.

The patient was 168 cm tall, weighed 65 kg, and had a Mallampati class 2 airway. After consultation with her hematologist, she was premedicated with 200mg PO cromolyn the night before surgery; 150mg PO Zantac and 10mg PO Zyrtec the morning of surgery; and 50mg IV Benadryl and 125mg IV Solumedrol before anesthesia induction. Clindamycin 600 mg IV, which was safely used during her previous procedure, was given for prophylaxis. Epinephrine was readily available.

Anesthesia was induced with 2.0 mg/kg propofol and was maintained with sevoflurane after placement of a laryngeal mask airway. Intraoperatively, the patient received Fentanyl, Ondansetron, and intravenous Tylenol.
Her blood pressure, heart rate, and oxygen saturation remained stable throughout the 2-hour procedure and the immediate postoperative period. She experienced mild pruritus in her left upper extremity on postoperative day 1, which was successfully treated with two doses of PO Benadryl.

DISCUSSION

Systemic Mastocytosis (SM) is a rare myeloproliferative disorder characterized by excessive

mast cell accumulation in the skin and/or one or more extracutaneous tissues. Mast cell degranulation can result in both chronic and episodic mediator release. These mediators, including histamine and heparin, can result in a variety of allergic and anaphylactic reactions [1].

In children, 80% of mastocytosis cases appear during the first year of life, and the majority is limited to the skin usually in the form of urticaria pigmentosa. Most eventually improve or resolve completely by adolescence[3]. Adults who develop mastocytosis more often have persistent systemic disease. Frequently, patients present with a history of unexplained allergic or anaphylactic reactions or with symptoms caused by episodic mediator release

Release of mast cell mediators can induce vasodilation, hypotension, flushing, pruritus, syncope, abdominal pain, nausea, vomiting, diarrhea, fatigue, and headache [3]. Over time, chronic mediator release is associated with cachexia, chronic gastrointestinal symptoms, diffuse musculoskeletal pains, and/or tissue remodeling and fibrosis of some organs. For example, mast cell infiltration of the liver can lead to hepatosplenomegaly, portal hypertension and ascites [4]. Systemic mastocytosis can present in an aggressive form, as a mast-cell leukemia or mast-cell sarcoma, which carries a poor prognosis [5]. A bone marrow biopsy revealing evidence of mast cell aggregation is diagnostic.

Triggers of mediators release include foods (23%), insect venom (21%), alcohol ingestion (14%) and medications (14%), specifically: narcotics, opioids, nonsteroidal antiinflammatory drugs (NSAIDs), iodinated contrast agents, vancomycin, and muscle relaxants used in anesthesia. Additionally, physical factors, such as exercise, extremes of temperature, sudden temperature changes, very spicy foods; surgical procedures, infections, and emotional stress can induce mediator release. Systemic mastocytosis, therefore, has serious anesthetic implications and could result in serious or fatal complications [6].

If a patient with systemic mastocytosis is scheduled for surgery, it is prudent to retrieve any previous anesthesia records to identify which drugs he/she has tolerated during the previous anesthetic. While precise guidelines for premedicating these patients are lacking, data support the benefit of premedicating them with steroids and H1 and H2 blockers [7]. In addition, our patient received a mast cell stabilizer the night before surgery.

CONCLUSION

We presented a patient with systemic mastocytosis who underwent outpatient foot surgery under general anesthesia. Identifying and using medications she tolerated during a previous anesthetic as well as medicating her with steroids, H1 and H2 blockers, and cromolyn helped produce a successful outcome.

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