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Perioperative Management of Patient with Systemic Mastocytosis Going for Total Thyroidectomy

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Abstract: Systemic mastocytosis (SM) is a rare disorder affecting 1 in 10,000 individuals, characterised by abnormal mast cell proliferation in extra-cutaneous organs, resulting in a variety of presentations ranging from mild asymptomatic disease to life-threatening tissue-destructive disease. Severe anaphylaxis caused by the release of histamine from mast cells can lead to poor postoperative outcomes, especially during general anaesthesia, as the severe manifestations of anaphylaxis, such as cardiovascular collapse and bronchospasm, may potentially be the first symptoms to present. In this case report, we describe the perioperative and intraoperative management of a 70-year-old female with SM, undergoing total thyroidectomy.

Keywords: Anaphylaxis; Perioperative Management; Systemic Mastocytosis.

1. INTRODUCTION

Systemic mastocytosis (SM) is a rare disorder characterised by abnormal mast cell proliferation in extra-cutaneous organs [1]. The clinical manifestation of SM varies, depending on the different tissues infiltrated by abnormal mast cells [2]. Mastocytosis is associated with immediate hypersensitivity reactions (IHR), which may arise from non-allergic mechanisms or allergic pathways (IgE-mediated immune responses) [3].

Tryptases are neutral serine proteases stored in mast cells, and serve as indicators of mastocytosis. There are two main forms of tryptase: The first, pro-α tryptase, typically shows increased levels in cases of mastocytosis. The second form, mature β-tryptase, is generally released during IgE-mediated anaphylactic reactions [4]. Total tryptase, measured in serum via fluoroimmunoassay, includes both forms and typically peaks between 30–60 minutes, with a half-life of 2 hours. Median serum tryptase levels are reported at 5.1 μg/L (range: 1–30.7 μg/L). Tryptase can remain elevated for 24–48 hours following a mastocytosis event, with levels depending on the extent and severity of mast cell degranulation [4]. Guidelines for safe surgery in patients with systemic mastocytosis can help ensure safer surgery for affected patients [5]. For example, *"A practical approach to systemic mastocytosis complications in cardiac surgery"* can be used to improve anaesthetic care for such patients [6]. In this case report, we describe a 70-year-old female with a known case of systemic mastocytosis (SM) who underwent an elective total thyroidectomy. We highlight the perioperative challenges and strategies to minimise potential complications. The aim of this report is to discuss the management options for such cases, emphasising the importance of careful perioperative assessment, multidisciplinary collaboration, and vigilant intraoperative monitoring.

II. CASE PRESENTATION

A 70-year-old female patient with a known history of papillary thyroid carcinoma and atrial fibrillation on apixaban 5 mg OD presented to our preoperative clinic for a scheduled total thyroidectomy. She disclosed a diagnosis of systemic mastocytosis (SM), established two years ago on the basis of gastrointestinal biopsy findings, and currently managed with

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midostaurin 50 mg BID and a protein kinase inhibitor. The patient reported no episodes of anaphylaxis since the diagnosis. Furthermore, she denied any cardiac or respiratory symptoms, and had no past history of allergies to medications or food. Her medical history was significant for papillary thyroid carcinoma, atrial fibrillation, and unexplained syncope, for which an implantable loop recorder was placed. Additionally, she had undergone a hysterectomy, appendectomy and bilateral knee replacement over 15 years ago, prior to her mastocytosis diagnosis, with no perioperative complications.

On physical examination the patient appeared well, showing normal vital signs with a heart rate (HR) of 66 bpm. There were no significant cutaneous manifestations or signs of heart failure. Airway examination revealed a Mallampati score of 2, with normal thyromental distance and neck mobility, and no signs of airway obstruction or obvious anatomical abnormalities. Laboratory results were mostly within normal limits. However, electrocardiography showed atrial fibrillation rhythm, while the echocardiogram showed normal bi-ventricular size and systolic function, left ventricle ejection fraction above 55%, mild to moderate tricuspid regurgitation and bi-atrial enlargement.

Cardiac risk stratification was done with enhancement of PRO-BNP, with a pro B-type natriuretic peptide (BNP) level of 1000 ng/L, and revised cardiac risk index score of 0. Her CHADS-65 score was 1 (for age), per the Canadian Society of Cardiology Guidelines. The patient was considered high risk for major perioperative adverse cardiovascular events including anaphylactic shock, tachyarrhythmias induced by treatment of hypersensitivity manifestations, in addition to CHF, stroke, and myocardial injury.

Regarding perioperative medication management, it was planned that the patient would continue using midostaurin during the preoperative period.

Apixaban was discontinued for 48 hours preoperatively without bridging. For thromboprophylaxis, it was planned that the patient would be on a pneumatic device intra- and postoperatively on the day of the surgery, and on the first and second day post-surgery she would receive prophylaxis with low molecular weight heparin (LMWH). Apixaban would be resumed on the third day post-surgery, after achieving haemostasis. One day before the opera tion, the patient was started on loratadine 20 mg p.o. BID and systemic steroids. Prior to her arrival in the operating room, an anaphylaxis tray was prepared, which included epinephrine, diphenhydramine, esmolol, and hydrocortisone. Latex-free instruments were used throughout the procedure. Once in the operating room, she received 1.5 mg midazolam, along with 1.5 mg cefazolin, prior to an A-line insertion at the right dorsalis pedis. General anaesthesia was induced using a remifentanil infusion, ketamine 30 mg, fentanyl 50 mcg, propofol 150 mg, and succinylcholine 50 mg. Following a successful induction, an 18G midline was inserted in the right leg. Intraoperative anaesthesia was maintained with remifentanil 0.05-0.1 mcg/min, and sevoflurane (MAC 0.8-1). Pain management during the procedure was achieved using paracetamol and fentanyl boluses.

The patient's lower extremities were exposed for continuous observation for any cutaneous signs, and careful observation of vitals and peak pressure was maintained throughout the procedure. The procedure was uneventful. After extubation, a brief period of hypertension was managed using a total of 30 mg esmolol. The patient was admitted to the ICU for postoperative observation, and during her first 24 hours she was fully conscious, vitally stable off vasopressors, maintaining oxygen saturation on room air, and had no active complaints. She was therefore discharged to the ward and resumed her home medication.

III. DISCUSSION

In this case, a 70-year-old female with systemic mastocytosis presented for an elective total thyroidectomy. The perioperative management of patients with SM presents significant challenges due to the potential for mast cell degranulation, which can be triggered by factors such as anaesthesia, surgical stress, temperature changes, and physical manipulation during surgery. This case highlights the im-

portance of perioperative optimisation and a multidisciplinary approach to minimise the risk of adverse events.

Systemic mastocytosis (SM) is a rare disease affecting 1 in 10,000 individuals; however, underdiagnosis is common [7]. It is not directly an inherited disease; rather, it is related to a somatic gain-of-function mutation in the KIT gene, a tyrosine kinase receptor expressed by mast cells, which plays a role in mast cell proliferation, maturation, adhesion, chemotaxis, and survival [1]. Mast cells are found throughout the body, predominantly around the blood vessels and tissues exposed to the external environment, including cutaneous tissue, and respiratory and gastrointestinal mucosa [8]. The severity of the disease ranges from mild, with either no symptoms or mild cutaneous manifestations, to fatal, tissue-destructive disease [2]. The clinical manifestations of SM depend directly on the tissue infiltrated by defective mast cells. For example, gastrointestinal tract involvement can lead to symptoms such as abdominal cramping, nausea, vomiting, diarrhoea, and heartburn, as seen in our patient. Other common manifestations include flushing and pruritus of the skin, as well as cardiovascular symptoms such as recurrent hypotension and cardiovascular collapse [2].

Perioperative management in our patient began with a comprehensive assessment of the severity of her disease and the optimisation of any comorbid conditions. The patient's current treatment with midostaurin, which had previously controlled her condition, was continued. One day prior to surgery, the patient was given an H1-receptor antagonist (loratadine 20 mg orally twice daily) and systemic steroids to stabilise mast cell activity and reduce the likelihood of anaphylaxis. In a similar case study, an allergist recommended a regimen that included, one day before the operation, prophylaxis with an H1-receptor antagonist (clemastine 1 mg orally at night), and on the day of the operation, a leukotriene receptor antagonist (montelukast 10 mg orally) [3]. Additionally, they used IV prophylaxis, including an H1-receptor antagonist (clemastine 2 mg), an H2-receptor antagonist (ranitidine 50 mg), and corticosteroids (dexamethasone 4 mg) 20 minutes before the induction of anaesthesia. Afterwards, H1, H2, and leukotriene receptor antagonists were continued for five days [3]. In another case of undiagnosed mastocytosis, a patient undergoing total knee replacement experienced severe hypotension and required vasopressor support after receiving clonidine, suggesting anaphylaxis [9]. Blood samples confirmed mast cell degranulation, and the patient was later diagnosed with systemic mastocytosis based on elevated tryptase levels and a bone marrow biopsy showing abnormal mast cell clusters [9]. This case highlights the importance of recognising potential mast cell activation in the perioperative setting, even when the diagnosis of SM is not known prior to surgery.

In our case, preoperative preparation included not only pharmacological interventions but also the preparation of an anaphylaxis tray in the operating room, containing epinephrine, diphenhydramine, esmolol, and hydrocortisone, to be used in case of an emergency. We also used latex-free instruments throughout the procedure to avoid potential allergic reactions.

Due to a patient's unconsciousness during general anaesthesia, manifestations of anaphylaxis such as cardiovascular collapse and bronchospasm could be extremely dangerous, potentially presenting as the first symptoms, making vigilant monitoring even more critical. This is in contrast to the more subtle cutaneous manifestations, which could be masked by surgical draping [10, 11].

Preventing anaphylactic shock was of huge importance in our patient's care, given her high risk for perioperative major adverse cardiovascular events, including the potential for tachyarrhythmias induced by treatment of hypersensitivity manifestations. Additionally, she was at risk for congestive heart failure, stroke, and myocardial injury. Our primary goal was to minimise the risk of perioperative anaphylaxis by blocking the cascade of mast cell activation and avoiding triggering factors. This we achieved through prophylactic administration of antihistamines and steroids preoperatively, and by avoiding the most common drugs involved in perioperative anaphylaxis, such as morphine and neuromuscular

blocking drugs (NMBDs) [12].

Finally, close postoperative monitoring for signs of anaphylaxis is necessary. Although the patient in this case had an uneventful postoperative course, the use of a multidisciplinary approach and careful perioperative management was key to minimising risk and ensuring a favourable outcome.

IV. LIMITATIONS

This case report has two potential limitations that should be considered. First, it concerns a single patient, and second, there is a lack of long-term follow-up data to assess the outcomes of the management approach in this case. Our patient's postoperative course may not be representative of all patients with systemic mastocytosis.

V. CONCLUSION

In conclusion, the perioperative and intraoperative management of mastocytosis presents unique challenges due to the rarity of the condition and the limited literature available. This case highlights the importance of tailored perioperative strategies for patients with systemic mastocytosis. Careful planning, a multidisciplinary approach, and vigilant monitoring are essential in such cases. Furthermore, preoperative optimisation with antihistamines and corticosteroids, along with careful anaesthesia management, can significantly reduce the risk of complications.

Given the scarcity of comprehensive studies, there is a pressing need for further retrospective and prospective research to enhance evidence-based management strategies for surgical procedures involving mastocytosis patients. Establishing a robust body of evidence will not only improve clinical outcomes, but also contribute to the development of standardised protocols for use in similar cases, ultimately advancing the field of perioperative medicine for this rare condition.

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