Case Report

# Indolent Systemic Mastocytosis Manifesting as Protracted Anaphylactic Shock

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## Abstract

Systemic mastocytosis is a rare disease due to abnormal proliferation of mast cells (MCs). A case of indolent systemic mastocytosis is presented here. After anesthetic induction for elective thyroid swelling with propofol and atracurium followed by endotracheal intubation, a 57-year-old female patient developed acute hypotension, sinus tachycardia, red rashes, increased airway pressure along with difficult ventilation, and desaturation. She developed multiorgan failure subsequently. MC tryptase level was persistently high. Bone marrow study revealed mastocytosis. She required antihistaminic, steroid, and organ support. With treatment, organ functions recovered gradually. Atracurium precipitated anaphylactic shock causing severe morbidity in this patient.

**Keywords:** Atracurium, mast cell tryptase, mastocytosis

## Introduction

Mastocytosis, a rare clonal disorder of abnormal mast cell (MC) proliferation, occurs at reported incidence of approximately 5–10 cases/$10^6$ people/year.[1] We report a case of an indolent systemic mastocytosis (ISM) who experienced anaphylactic shock during anesthetic induction. Written consent was obtained from the patient's relatives.

## Case Report

A 57-year-old female patient (height 5'2" and body weight 55 kg) without any known comorbidity and any known drug allergy was posted for elective thyroidectomy for a colloid goiter. Computed tomography of the neck region showed the huge thyroid swelling compressing the airways [Figure 1]. She was induced with injection propofol 120 mg intravenously followed by direct laryngoscopy and endotracheal intubation with a bougie. After successful intubation confirmed by capnography and bilateral air entry, she was connected to anesthesia machine. She was hemodynamically stable at this point and adequately ventilated. For adequate skeletal muscle relaxation for surgery, injection atracurium 30 mg intravenously was administered slowly, approximately 2 min after she developed profound hypotension (50/32 mmHg) and tachycardia (153/min regular). It was associated with high airway pressure, acute desaturation, and hematuria. She was started on adrenaline infusion and ventilated with 100% oxygen. With the suspicion of an acute anaphylaxis, blood for MC tryptase (MCT) was sent 45 min after the above episode. She was treated with bronchodilator nebulization and hydrocortisone. Operative procedure was postponed, and she was transferred to Intensive Care Unit (ICU) for further management. In ICU, the patient was brought in completely unresponsive state. She was ventilated in assist-control mode. She was reviewed by a team of experts including intensivists, nephrologist, hematologist-oncologist, immunologist, and neurologist. Magnetic resonance imaging of brain revealed bilateral watershed infarct [Figure 2]. Initial MCT was >200 ng/ml. Bone marrow study revealed huge number of MCs scattered throughout the field [Figure 3]. A working diagnosis of systemic mastocytosis was made. Serial MCT monitoring was started. The patient was started on high-dose

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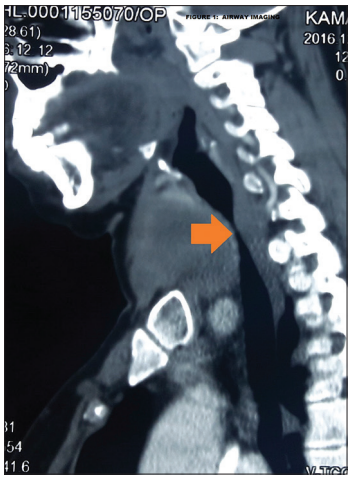


**Figure 1:** Airway imaging

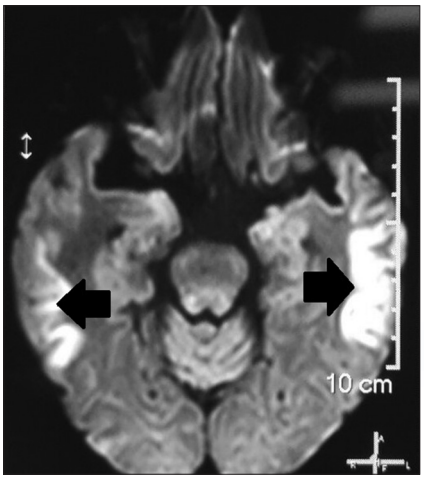


**Figure 2:** Magnetic resonance imaging brain

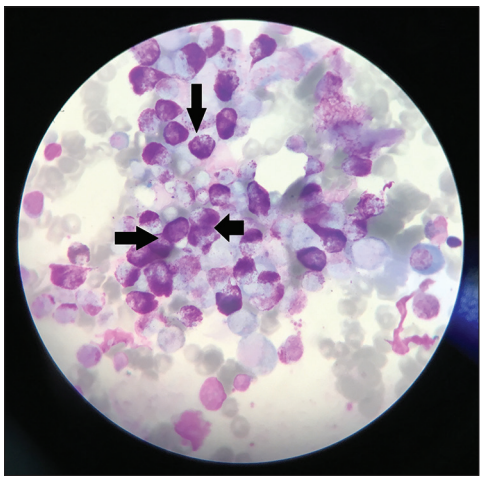

**Figure 3:** Mast cells

steroids and antihistaminics. Repeat MCT at 6 h was >200 ng/ml as well. She needed hemodialysis for acute renal failure. Gradually, adrenaline infusion was tapered off. Ventilator demands started getting reduced. MCT value at 24 h was 161 ng/ml. Because of anticipated requirement of prolonged airway protection, elective tracheostomy was planned. Provocative incremental intradermal test of lignocaine done before tracheostomy revealed hypersensitivity to lignocaine as well. Therefore, tracheostomy was carried out under inhalational anesthetic sevoflurane and injection ketamine. MCT was repeated after 7 days which showed >200 ng/ml value. With intensive management, patient's neurological condition improved remarkably albeit with a residual weakness in proximal muscles of extremities. Renal parameters and lung functions were normalized. Steroid doses and antihistaminics were tapered. During ICU stay, she had nosocomial infections which were treated by appropriate antibiotic as per culture sensitivity and after allergic testing.

## Discussion

ISM is the most frequent variant of mastocytosis in adults. Mostly, symptomatic management is advocated for its management.[2] In 1869, mastocytosis was first described.[3] Typical MCs are round or oval cells with small central nucleus and metachromatic granules dispersed in large cytoplasm. These granules contain inflammatory mediators including tryptase. Elevated levels of serum tryptase (>20 ng/ml) are consistent with the diagnosis according to WHO system.[4,5] Very high levels (>200 ng/ml) correlate with more severe course and poor prognosis.[6]

## Conclusion

ISM can be life threatening in several situations including anesthesia and surgery. In our case, atracurium precipitated this protracted anaphylactic shock causing severe morbidity in this patient. We recommend testing of serial measurement of MCT in the setting of perioperative anaphylaxis.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's relatives have given consent for images and other clinical information to be reported in the journal. The patient's relatives understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

### Acknowledgment

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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