Anaesthesia in a patient with mastocytosis

Dear Editor,

MASTOCYTOSIS, a rare disorder characterized by abnormal proliferation of mast cells in cutaneous and extracutaneous sites with incidence 1:1000-8000.[1] With knowledge beforehand, anaphylactic reactions can be prevented. We present our experience with 27-year-old woman, undergoing laparoscopic cholecystectomy under general anesthesia.

One year back she developed painful palpable maculopapular rash involving neck, trunk and both extremities without fever. Lesion's punch biopsy showed mild perivascular inflammation including mast cells in dermis with negative serum tryptase. Pathological diagnosis of cutaneous mastocytosis was made after bone marrow cytology and negative KIT, FLIP1 and PDGFR. She had no obvious systemic manifestations and history of anaphylactic-like reactions. Patient and family consent was obtained after thorough preoperative evaluation and discussion regarding possibility of immediate or delayed hypersensitivity reactions. Tablet ketotifen 1mg bd and fexofenadine 180mg od started and negative serum tryptase prior to surgery.

Premedication done with intravenous (i.v) ranitidine 50mg 40 minutes prior, hydrocortisone 100mg, pheniramine

maleate 22.5mg before induction and 1mg midazolam. For any anaphylactoid reaction, epinephrine 100μg/ml was prepared. Coamoxiclav 1.2g given i.v after intradermal testing. Inhalational induction was done using sevoflurane and fentanyl 75μg and vecuronium 6mg with sevoflurane, oxygen and air as maintenance (MAC=0.7). Normothermia ensured with warm Ringer lactate, surgical draping and ambient temperature (24°C). Paracetamol 1g given for analgesia and ondansetron 4mg for antiemesis. Reversal done with neostigmine 3.0 mg and glycopyrrolate 0.4 mg. Duration of surgery was 45 minutes with uneventful perioperative course.

World Health Organization (WHO) defines seven types of mastocytosis.10% of those with cutaneous mastocytosis will have systemic involvement.[2] Histamine release can be triggered by variety of stimuli, including mechanical irritation, temperature changes, stress, alcohol, vomiting, and certain drugs. Clinical features include most commonly pruritis, flushing, and urticarial rashes. Less frequent but more serious complications are hypotension, tachycardia, syncope and shock. In response to mast cell release and the local mast cell burden, histamine and other inflammatory mediators are released leading to profound vasodilatation and blood pooling in splanchnic and peripheral vascular beds, resulting in life-threatening condition. Bronchospasm does not usually occur. For diagnosis molecular, immunohistochemistry, and histological tests should be done.[2] Chronic elevations of serum tryptase levels (>20μg/ml) strongly suggest anaphylaxis and mast cell degranulation.[3] Antihistaminics, anti inflammatories glucocorticoids and mast cell stabilizers, are recommended. Antibiotic and anaesthetic drugs, like local anesthetics, should be tested intradermally since positive results are conclusive, but negative results do not reliably exclude sensitization. Initial wheal of >3mm is considered positive.[4] Volatile anesthetic agents, fentanyl, sufentanil, remifentanil, and naloxone do not release histamine and propofol, ketamine, barbiturates and midazolam can also be used.[5] Morphine, atracurium, mivacurium, and succinylcholine known to cause anaphylactic reactions, vecuronium, local anesthetics does not. Temperature changes and extremes are also triggering factors. When anaphylaxis occurs, adrenaline is administered early, followed by fluid challenge of 20 ml/kg and repeated if necessary. In refractory hypotension, vasopressin 2U, repeated as necessary. Corticosteroids also as part of resuscitation.[6] Activating cell surface receptors, epinephrine regulates hemodynamics, relieves bronchospasm, and stops mast cell degranulation.[7]

Thus, mastocytosis management involves knowledge, thorough preanaesthesia assessment, vigilant monitoring, safe anesthesia techniques, and avoidance of physical and pharmacological triggers.

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Conflicts of interest
There are no conflicts of interest.

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References

1. Carter MC, Uzzaman A, Scott LM, Metcalfe D, Quezado Z. Pediatric mastocytosis: Routine anesthetic management for a complex disease. Anesth Analg 2008;107:422-7.
2. Longley J, Duffy TP, Kohn S. The mast cell and mast cell disease. J Amer Acad Derm 1995;32:545-61.
3. Horny HP, Sotlar K, Valent P, Hartmann K. Mastocytosis: A disease of the hematopoietic stem cell. Dtsch Arztebl Int 2008;105:686-92.
4. Laguna JJ, Archilla J, Doña I, Corominas M, Gastaminza G, Mayorga C, *et al*. Practical guidelines for perioperative hypersensitivity reactions. J Investig Allergol Clin Immunol 2018;28:216-32.
5. Stellato C, Casolaro V, Ciccarelli A, Mastronardi P, Mazzarella B, Marone G, *et al*. General anaesthetics induce only histamine release selectively from human mast cells. Br J Anaesth 1991;67:751-8.
6. Harper NJN, Cook TM, Garcez T, Farmer L, Floss K, Marinho S, *et al*. Anaesthesia, surgery, and life-threatening allergic reactions: epidemiology and clinical features of perioperative anaphylaxis in the 6th National Audit Project (NAP6). Br J Anaesth. 2018 Jul; 121:159-171.
7. Turk JW, Oates JA, Roberts JL. Intervention with epinephrine in hypotension associated with mastocytosis. J Allerg Clinl Immun1983;71:189-92.

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