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A case report of Knee Replacement Surgery in Indolent Systemic Mastocytosis: the Anaesthesiologic Management as an obstacle race

References:

¹Pascale Dewachter, Mariana C. Castells, David L. Hepner, Claudie Mouton-Faivre; Perioperative Management of Patients with Mastocytosis. *Anesthesiology* 2014; 120:753-759. ²Bocca-Tjeertes IFA, van de Ven AAJM, Koppelman GH, Sprikkelman AB, Oude Elberink HJNG. Medical algorithm: Peri-operative management of mastocytosis patients. *Allergy*. 2021 Oct;76(10):3233-3235.

Author Block: Emanuele Pisello¹, Matteo Ciuffreda², Luca Brugiaferri³, Jessica Silvestri³, Sergio Sorrenti³, Cristiano Piangatelli², Dario Galante⁴.

¹Anaesthesia, Resuscitation, Intensive Care and Pain Management, AST Ancona - UNIVPM, Fabriano, Italy, ²Anaesthesia, Resuscitation, Intensive Care and Pain Management, AST Ancona, Fabriano, Italy, ³Anaesthesia, Resuscitation, Intensive Care and Pain Management, UNIVPM, Ancona, Italy, ⁴Anaesthesia, Resuscitation, Intensive Care and Pain Management, ASL Foggia, Cerignola, Italy.

Abstract:

Background: Systemic Mastocytosis (SM) results from clonal proliferation of abnormal, aberrant mast cells in various organs and tissues, with or without skin involvement. An acute release of mast cell mediators causing anaphylaxis is a sudden and life-threatening manifestation of the disease, especially during anaesthesia. The prevalence is estimated between 13:100000 and 1:364000 whereas its worldwide incidence is estimated to be 1:150000¹.

Case Description: We describe a case of anaesthesiologic management for knee replacement surgery in a 76-years-old woman diagnosed with Indolent Systemic Mastocytosis due to past anaphylaxis and elevated serum tryptase levels. Anaesthesiology management started from a detailed preoperative evaluation of patient's medical history. Allergy to wasp venom (past anaphylactic shock triggered by wasp sting), NSAIDs intolerance, ischemic cardiopathy (non-critical single vessel coronary artery disease), hypertension, dyslipidemia, hypothyroidism, fibromyalgia and anxious-depressive syndrome were found. Echocardiographic evaluation showed a moderate aortic stenosis, bicuspid aortic valve and mild diastolic dysfunction. Before surgery, patient underwent infective endocarditis prophylaxis and premedication recommended by the allergist consultant (Prednisone 50mg, Chlorphenamine 10mg and Pantoprazole 40mg). We opted for loco-regional anesthesia with execution of a single-shot lumbar plexus ultrasound-assisted by administering 300 mg Mepivacaine with Epinephrine (formulation 10 mg/5 mcg/ml) and Dexamethasone 4 mg and subsequent placement of a perineural catheter with elastomeric pump infusing 300 mg Mepivacaine in 200 ml NaCl plus 4 mg Dexamethasone with a rate of 4 ml/h for 48 hours. In addition, spinal anesthesia was performed with 12 mg Bupivacaine 1% at the level of L3-L4. During the operation NIBP, oxygen saturation, heart rate, body temperature were monitored and crystalloids were administered for a total of 600 ml. Tranexamic acid 500 mg was given to reduce intraoperative bleeding.

Discussion: In Systemic Mastocytosis, an acute release of mast cell mediators causing anaphylaxis is a sudden and life-threatening manifestation of the disease, especially during anaesthesia. Body temperature control, antimediation therapy, avoiding histamin-releasing drugs (e.g. atracurium, succinylcholine, mivacurium, morphine, thiopental and neostigmine) is required to lower both pharmacological and non-pharmacological stressors and subsequent risk of intraoperative anaphylaxis². Regional anesthesia techniques allow fewer drugs usage comparing to general anesthesia. Using Mepivacaine with Epinephrine in lumbar plexus ultrasound-assisted block, both in single shot and continuous perineural infusion, allowed us to reduce NSAID and opioids post-operative needing.

Learning Point: In agreement with most recent update in management around invasive procedure in Mastocytosis, we investigated for any known triggers to anticipate and avoid mast cell activation; we monitored body temperature and administered a prophylactic antimediation therapy to lower non-pharmacological stressors and subsequent risk of intraoperative anaphylaxis; we preferred regional anesthesia techniques to use fewer drugs comparing to general anesthesia, thus also reducing the risk for acute degranulation; we avoided use of histamin-releasing drugs (e.g. atracurium, succinylcholine, mivacurium, morphine, thiopental and neostigmine) to prevent histamine-induced reactions; we guaranteed perioperative availability of epinephrine, antihistamines, bronchodilators, iv fluid, videolaryngoscope with disposable blades of various sizes and additional devices for difficult intubation to face any anaphylactic events ensuring adequate airway management.

Author Disclosure Information:

E. Pisello: None.

Track (Complete): Regional Anaesthesia

Declaration (Complete):

*Was this abstract presented before March 2023?: No

*Patient Consent: Yes

Status: Finalized

