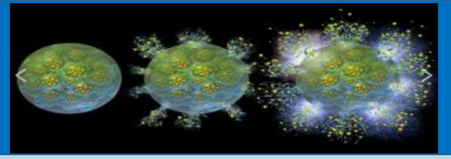
George Eliot Hospital NHS Trust

Anaesthesia for caesarean section in a parturient with systemic mastocytosis

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BACKGROUND

Mastocytosis is a rare disorder (1 in 20,000) of mast cell proliferation that occurs in cutaneous or systemic forms. The systemic variant occurs in about 10% of cases and particularly affects the reticuloendothelial system. The substances that cause the symptoms of allergy are stored in granules within the mast cells and are released into the body during an allergy attack and this is called 'mast cell degranulation'. In people with mastocytosis, mast cells are triggered much more easily which causes severe allergy-type symptoms.

Our parturient had diagnosis of systemic mastocytosis. A written consent was taken from her for publication of this case report.

CASE REPORT

A 37-year-old, G2P1 diagnosed to have systemic mastocytosis after immunology and bone marrow examination was admitted to the obstetric unit for an elective caesarean section. She was previously evaluated in the Obstetric Anaesthetic clinic and deemed to be suitable for central neuraxial anaesthesia and peripheral nerve blocks. In the operating theatre after commencing standard AAGBI monitoring, IV cannulation and betadine skin preparation, a combined spinal epidural was performed with 2.2 mL hyperbaric 0.5% bupivacaine and 20 mcg of fentanyl in the intrathecal space. Intra-operative cell salvage was used. Post-operative pain relief was achieved by bilateral transversus abdominis block with ropivacaine, regular oral paracetamol and ibuprofen. The lady tolerated the anaesthesia and surgery well. She was monitored in the labour suite for next 24 hours for any delayed reactions and was discharged on post-operative day two after an uneventful recovery.

DISCUSSION

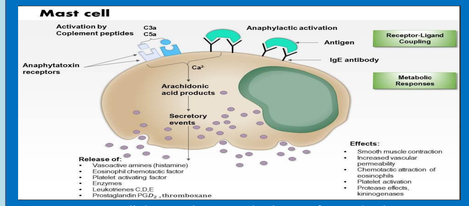


Figure 1: Mast cell degranulation and release of stored substances

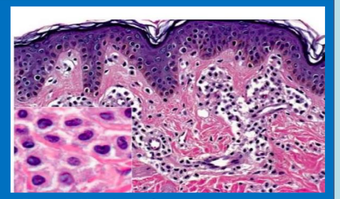


Figure 2: Histology of cutaneous mastocytosis

PATHOPHYSIOLOGY

Systemic mastocytosis (SM) is a distinctive myeloproliferative disorder that is associated with point mutation in the c-KIT tyrosine kinase resulting in c-KIT kinase activation. It causes increased growth and decreased apoptosis of mast cells. SM is one of eight subcategories of myeloproliferative neoplasms resulting in clonal, neoplastic proliferation of morphologically and immunophenotypically abnormal mast cells that accumulate in one or more organ systems.

World Health Organization classification of mastocytosis (1)

1. Cutaneous mastocytosis (CM)
2. Indolent systemic mastocytosis (ISM)
3. Systemic mastocytosis with an associated clonal haematological non-mast cell lineage disease (SM-AHNMD).
4. Aggressive systemic mastocytosis (ASM)
5. Mast cell leukaemia (MCL)
6. Mast cell sarcoma (MCS)

CLINICAL FEATURES

It has got heterogeneous clinical picture, ranging from skin-limited disease (cutaneous mastocytosis), particularly in paediatric cases where the majority have disease-onset within the first 2 years of life and commonly experience spontaneous regression of skin lesions, to a more aggressive variant with extra cutaneous involvement (systemic mastocytosis) that may be associated with multiorgan dysfunction/failure and shortened survival, that is generally seen in adult patients (2).

Table 1: common symptoms and signs of mastocytosis (2)

Dermatologic	Flushing, pruritus, hives
Cardiovascular	Near syncope or syncope, palpitations, chest pain, dysrhythmias, hypotension, hypertension
Pulmonary	Cough, wheezing
Eyes, ENT	Postnasal drip, inflammation (conjunctivitis, rhinitis, sinusitis, pharyngitis, laryngitis), throat itching and swelling
Neurologic	Headache, seizures, tremors
Psychiatric	Cognitive dysfunction, memory difficulties, anxiety, depression, psychoses
Gastrointestinal	Nausea, vomiting, reflux, constipation, diarrhea, abdominal pain, malabsorption
Musculoskeletal	Bone or muscle pain, arthritis, myositis
Immunologic	Types I, II, III, and IV hypersensitivity reaction





Figure 3 and 4 : cutaneous mastocytosis

DIAGNOSTIC CRITERIA

Table 2: Proposed diagnostic criteria for mastocytosis (3)

(1)Episodic symptoms of mast cell mediator release involving ≥2 organ systems.

(2)Appropriate response to antimediator therapy.

(3)Documented increase in validated systemic markers of mast cell activation during episode (e.g., serum tryptase or urinary markers such as histamine metabolites, prostaglandin D_2 or its metabolite, and leukotriene E_4).

(4)Primary and secondary causes ruled out.

PRECIPITATING FACTORS

Non-pharmacological: stress, exercise, excessive heat or cold, latex , certain foods.

Pharmacological: alcohol, morphine, codeine, pethidine, buprenorphine. atracurium, mivacurium, suxamethonium, amethocaine, chlorhexidine skin antiseptic, NSAIDS, radio contrast media.

TREATMENT

Includes H-1 and H-2 receptor antagonists and corticosteroids.
Adrenaline is typically used by patients for breakthrough degranulation and life threatening allergic reactions.

ANAESTHETIC CONSIDERATION (4)

Preoperative assessment

Detailed history may suggest organ involvement.
Any known triggers
FBC and peripheral smear to exclude associated haematological disorder
Elevated Serum alpha tryptase: to assess total body mast cell burden. (>20mcg/L is cut off)
Bone marrow biopsy.

Preop preparation

Pre-procedural skin testing is not recommended.
Avoidance of triggers and emergency preparedness.
Adrenaline and other resuscitative drugs and equipment made available.
Premedication with H1 and H2 antagonists
Sodium cromoglycate- effective for some of the effects of mastocytosis on the gut
No evidence for prophylactic steroids.

Choice of anaesthetic and post-operative care.

Central and peripheral neuraxial techniques reduce risk of multiple drug administration (2)
Medications for general anesthesia should include non–histamine-releasing drugs.
Active patient warming.
HDU bed is booked for the first night after surgery to look out for delayed reactions.

Safer medications:

Fentanyl, remifentanil, paracetamol, NSAIDs, rocuronium, vecuronium, cisatracurium, propofol, inhalational agents, benzodiazepine, amide Local anaesthetics, skin antiseptic- povidone iodine.

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