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CASE REPORTS

Anaesthetic management of emergency caesarean section in a parturient with systemic mastocytosis



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ABSTRACT

Mastocytosis is a rare disorder caused by the proliferation and accumulation of mast cells in various organs. It has a broad variety of clinical manifestations, including cardiovascular collapse. Diverse stimuli trigger the release of vasoactive substances and parturients with systemic mastocytosis are at high risk for precipitating mast cell degranulation. As a result, women with systemic mastocytosis should have an anaesthetic plan for labour and delivery. Anxiety, stress, sleep deprivation, pain and numerous pharmacological agents are all triggers for mast cell degranulation. For pain relief in labour, epidural analgesia is recommended. Pharmacological agents with a high potential for triggering mast cell degranulation should be avoided. This is particularly important in the case of an emergency caesarean section. Resuscitation equipment must be available should life-threatening haemodynamic instability occur during surgery. We report the case of a pregnant woman with systemic mastocytosis who required emergency caesarean section.

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Introduction

Mastocytosis is a rare disorder of haematopoietic stem cells. The exact incidence is unknown although it is estimated that annually 5–10 new cases per million are diagnosed.¹ Mastocytosis is caused by the proliferation and accumulation of mast cells in one or more organs. Frequently affected are the skin and bone marrow; rarely the gastrointestinal tract, liver, spleen and lymph nodes.^{1,2} Cutaneous mastocytosis, frequently known as urticaria pigmentosa, is the most common form and usually occurs in childhood.^{1,3} Approximately 10% of patients with urticaria pigmentosa also have systemic manifestations;⁴ this condition is more common in adults and clinical presentation varies immensely. A number of stimuli such as trauma, mechanical irritation, emotional stress, pain, fluctuating body temperature, nutrients, venom and pharmacological agents trigger the release of histamine, prostaglandin and other vasoactive substances. Minor symptoms include weakness, fatigue, urticaria, pruritus, abdominal cramps, vomiting, diarrhoea, mental confusion and fever.⁵ Patients

are at risk of fulminant mast cell degranulation, which can lead to bronchospasm, profound anaphylactoid reactions and even cardiovascular collapse.^{1,4–6} Parturients with systemic mastocytosis are at particularly high risk for developing mast cell degranulation because of the physiological stress of labour.⁵ An anaesthetic plan for delivery should be established at the earliest opportunity. We describe the case of a pregnant woman with systemic mastocytosis requiring emergency caesarean section.

Case report

A 33-year-old woman with systemic mastocytosis presented to the preoperative assessment clinic at our anaesthetic department at 29 weeks of gestation. A repeat caesarean section was planned at term. The woman reported having multiple exacerbations of her condition during pregnancy with abdominal pain, nausea and vomiting, bronchospasm, tachycardia and low blood pressure. She reported no anaphylactoid reactions. At the time of assessment she was on multiple medications (Table 1) and her condition was stable. Two years previously, the patient delivered a healthy infant by caesarean section under spinal anaesthesia. For this she had received intrathecal hyperbaric bupivacaine with fentanyl together with intravenous midazolam 2 mg for sedation.

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Table 1 Current medications

<i>Regular</i>	Budesonide 3 mg oral 8-hourly Prednisolone 7.5 mg oral daily Ranitidine 300 mg oral 12-hourly
<i>As required</i>	Budesonide inhalation Prednisolone 200 mg oral Diphenhydramine 50 mg oral Ondansetron 4 mg oral Flunitrazepam 0.5 mg oral Vitamin C 750 mg oral

Postoperatively she had mild pruritus that did not require treatment.

After discussion, an anaesthetic plan was formulated for her caesarean delivery. The preferred option was for spinal anaesthesia with the possibility of general anaesthesia, if required. A list of anaesthetic agents considered safe in systemic mastocytosis was placed in her medical notes (Table 2).

At 39 weeks of gestation the woman presented as an emergency to the hospital with severe continuous abdominal pain. Agitation made communication difficult and cardiotocography could not be performed. At first she refused medical investigation but was convinced

of the necessity by her midwife. Auscultation of the fetal heart rate revealed decelerations and on vaginal examination her cervix was 3 cm dilated. Ultrasonography showed no fetal heart rate. The duty anaesthetist was notified of the need for emergency caesarean section 19 min after the patient arrived in the hospital.

In the operating room the patient was uncooperative and complained of severe and continuous abdominal pain, possibly due to labour and an acute exacerbation of her systemic mastocytosis. Intravenous midazolam 3 mg was administered for sedation and to facilitate pre-oxygenation. Baseline blood pressure was 142/72 mmHg and heart rate 128 beats/min. Her weight was estimated to be 76 kg.

With thiopental and suxamethonium contraindicated in systemic mastocytosis, general anaesthesia was induced with a fractionated, large dose of propofol (200 + 100 + 100 mg) and tracheal intubation was carried out without the use of a muscle relaxant. Laryngoscopy and intubation were uneventful (Cormack and Lehane grade 1). Cricoid pressure was not applied. After verification of correct tube position by capnography, the obstetricians started the operation, six min after raising the emergency alarm. Anaesthesia was maintained with 1.0% end-tidal concentration of isoflurane in oxygen.

Table 2 Safety of medications in systemic mastocytosis

	Safe	Contraindicated	Uncertain
Alpha-2 adrenoreceptor agonists	Clonidine		
Analgesics	Alfentanil	Codeine	
	Fentanyl	Morphine	
	Sufentanil	Pethidine	
	Remifentanyl	Tramadol	
	Piritamid	NSAIDs	
	Paracetamol	Metamizole	
	Erythromycin	Penicillin	
Antibiotics		Atropine	
Anticholinergics		Beta blockers	
Antihypertensives	Urapidil		
	Dihydralacine		
	Glyceryl trinitrate		
Benzodiazepines	Midazolam		
	Flunitrazepam		
Catecholamines	Phenylephrine		
	Ephedrine		
Hypnotics	Propofol	Etomidine	
	Ketamine	Thiopental	
Local anaesthetics	Amide local anaesthetics	Ester local anaesthetics	
Minerals	Magnesium		
Muscle relaxants	Vecuronium	Mivacurium	Rocuronium
	Cisatracurium	Suxamethonium	
Tocolytics	Fenoterol		
Uterotonics			Oxytocin
			Methylergonovine
			Prostaglandins
Volatile agents	Isoflurane		
	Sevoflurane		
	Desflurane		

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