

ANAESTHETIC ASPECTS OF CUTANEOUS MASTOCYTOSIS IN A CHILD PRESENTING FOR DAY SURGERY: CASE PRESENTATION

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INTRODUCTION

Cutaneous mastocytosis or urticaria pigmentosa is one of the three major categories of the disease mastocytosis.¹ This is a rare and usually benign disease, presenting with a reddish to brown macular rash, mainly over the trunk, that sometimes spreads to the limbs. The lesions may become itchy and red when rubbed (Darier's sign).² The rash usually presents within the first 6 months of life and can be quite prominent, but often fades after a few years. The disease regresses spontaneously in over 50% of affected children. Biopsy shows an increased accumulation of mast cells in the skin lesions. Certain triggers induce release of mediators from the mast cells, causing a range of symptoms including flushing, pruritus, urticarial swellings and even blistering.³ In rare cases there may be wheezing, syncope and anaphylaxis. Treatment is mainly symptomatic, with control of the rash with H₁ and H₂ blockers and avoidance of physical trigger factors (rubbing, extremes of temperatures) and chemical triggers (histamine-releasing agents such as opiates, codeine, muscle relaxants and insect venom).¹

The possibility of anaphylaxis during anaesthesia from exposure to certain triggers makes a management plan for the peri-operative period essential. The risk of anaphylaxis is greatly reduced when H₁ and H₂ blockers^{4,5} and corticosteroids⁵ are administered pre-operatively. Disodium cromoglycate is a mast-cell-stabilising drug and may be of value when used prophylactically.⁴ Extremes of temperatures in theatre, as well as contact with cold, rough surfaces should be avoided. There are numerous anaesthetic-related drugs which trigger mast-cell degranulation. These include muscle relaxants, opiates, codeine, atropine, ephedrine and phenylephrine.⁶ If any of these agents, especially the muscle relaxants, has to be used, it should be diluted and administered slowly. The agent with the lowest histamine-releasing qualities should be chosen. Intravenous access must be established as soon as possible. Adrenaline should be at hand in case of anaphylaxis, and can be given intramuscularly if there is no vascular access or as an intravenous infusion. Analgesics to be avoided are codeine-containing mixtures, salicylates and opiates. Non-steroidal anti-inflammatory drugs (NSAIDs) may cause a severe reaction, but have been used in the treatment of prostaglandin-induced flushing in cutaneous mastocytosis. If necessary they should be started under close supervision to ascertain if they are tolerated or whether they cause a severe reaction. In the event of an anaphylactic reaction additional resuscitative measures need to be instituted. These include adrenaline, additional intravenous antihistamines, steroids, vasopressors, intravenous fluids, oxygen and respiratory control.

CASE REPORT

A 4-year-old boy, who had been diagnosed at 3 months of age with cutaneous mastocytosis, presented for elective tonsillectomy and adenoidectomy. His symptoms were occasional extreme flushing and pruritus in response to triggers of heat and pressure. There had been no episodes of anaphylaxis. The rash, which had been quite remarkable when he was a baby, was beginning to fade. His parents were careful, and had never used medications containing codeine, salicylates or NSAIDs. Their internet search (www.mastokids.com) had made them aware of the possibility of anaphylaxis with certain medications and particularly under anaesthesia. Several bouts of tonsillitis had been treated with antibiotics and he had a recurrent wheezy chest, which was treated with montelukast (Singulair). At the time of surgery he was on a short course of Celestamine (betamethasone and dextchlorpheniramine, i.e. a steroid and H₁ blocker) for his chest.

On examination the child was a normal healthy 4-year-old with a faint brownish, macular rash on the trunk. His lungs were clear. Two hours pre-operatively he was given an H₂ blocker (ranitidine 75 mg in 5 ml), an additional H₁ blocker (desloratidine 1.25 mg in 2.5 ml) orally and was nebulised with ipratropium and fenoterol. Anaesthesia was induced with the inhalational agent sevoflurane and intravenous access was secured. A small dose (20 mg) of propofol was given intravenously and the trachea intubated. Anaesthesia was maintained with sevoflurane. Anti-emetics (ondansetron 0.1 mg/kg and dexamethasone 0.15 mg/kg) were given intravenously. Muscle relaxants were not necessary and the child breathed spontaneously throughout the operation. Adrenaline was at hand but not needed. Haemodynamic and respiratory parameters were stable throughout and there was no flushing of the skin. An NSAID in the form of a 25 mg diclofenac suppository was given rectally as medical supervision was deemed optimal. He remained stable throughout the procedure as well as postoperatively, when he was observed in the ward for 6 hours. Paracetamol syrup and diclofenac suppositories were prescribed as analgesia for the postoperative period, which remained uneventful. He was discharged home into the care of his parents.

The parents were concerned that he might still develop an anaphylactic reaction at home. Reassurance was given that this was most unlikely, that the child had a very mild form of the condition and was already outgrowing it. However, they were counselled, shown how to use adrenaline, and supplied with adrenaline (1 ml of 1:1000), a graded 1 ml insulin syringe and a needle.

DISCUSSION

This child presented as a very mild case of cutaneous mastocytosis. Measures were taken to minimise his response on exposure to trigger agents during surgery. He was already covered by steroids and an H₁ blocker because of the Celestamine. An hour before surgery he was given an H₂ blocker (ranitidine) as well as an H₁

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blocker (desloratidine), and he was nebulised to lessen airway hyperreactivity in response to tracheal intubation or anaesthetic trigger agents. Although the dexamethasone was used for its anti-emetic qualities, it may possibly be of value for the prevention of non-immune mediated reactions.⁵ Adrenaline was at hand. Pethidine is often used as an analgesic after tonsillectomy, but was avoided in this case. After tonsillectomy, the worst pain occurs on day 4 or 5 postoperatively; by this time the child would obviously be at home in the care of his parents. Because an agent more effective than paracetamol might then be needed, an NSAID was given in hospital so that the child's response could be observed; there was no untoward reaction. A severe reaction at a later stage was unlikely, but the parents were instructed how to administer adrenaline correctly should an emergency arise.

Declaration of conflict of interest

The author declares no conflict of interest.

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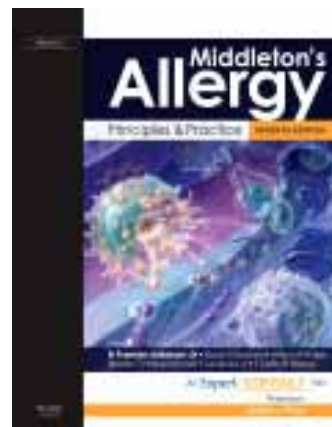
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