Anaesthesia for children with Paediatric Mastocytosis

Dr. Nargis Ahmad Consultant Anaesthetist

Great Ormond Street Hospital for Children NHS Trust

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This document provides guidance for the anaesthetic management of children with mastocytosis.

There is very little published information about anaesthesia in this patient group - the evidence base is limited. At best it is Grade D i.e. extrapolated from non-analytic studies, e.g. case reports, case series and expert opinion. Reported experience is largely limited to children with cutaneous mastocytosis (CM) and this experience suggests that the risks of anaesthesia in this group are low. Significant morbidity and mortality associated with anaesthesia in adults with systemic mastocytosis has been reported. Systemic mastocytosis (SM) is extremely rare in children. Given the limited evidence available it not possible to determine the risks of anaesthesia in children with systemic mastocytosis.

1. Multidisciplinary team care with involment with paediatricians and paediatric dermatologists should be in place.

2. Assess whether the patient has CM or SM?
    a. SM is extremely rare in children. If SM is suspected then the serum tryptase should be measured. If possible it should be established whether the patient fulfils the WHO criteria for SM, however invasive tests cannot be justified simply for the purposes of anaesthesia. If SM is present then the risk of widespread mast cell degranulation may be much higher than in CM.

3. Evaluate baseline disease and factors known to precipitate mast cell mediator release e.g. temperature, physical exertion, friction. How is the disease managed. How is the condition manifest e.g. urticaria, pruritus, flushing, gastrointestinal disturbance? Is there a history of syncopal episodes or shock.

4. Details of previous anaesthesia should be explored. It may be helpful to ascertain whether drugs such as NSAIDs have been taken and whether any sensitivity has been encountered as there is a range of tolerance to this group of drugs.

5. Preoperative skin testing has not been found to be predictive. Pre-operative tryptase levels should be measured where possible.

6. Continue scheduled medication but it is not felt that prophylactic medication with anti-histamines or steroids are of benefit.

7. Many drugs (listed below) have been used during anaesthesia with no serious complications, some of these drugs are known to precipitate mast cell degranulation in experimental models.

    - Midazolam, chloral hydrate
    - Propofol, sodium thiopentone, ketamine
    - Volatile anaesthetic gases and nitrous oxide
    - Bupivicaine, lignocaine
    - Rocuronium, vecuronium, cis-atracurium, succinylcholine
    - Remifentanil, fentanyl, morphine, codeine
    - Neostigmine, glycoppyrolate, atropine
    - Paracetamol, ibuprofen, diclofenac
    - Ondansetron, dexamethasone

8. Avoid any known triggers including environmental factors e.g. temperature changes or friction. Anxiety can promote mast cell degranulation, consider sedative premedication.

9. Any known pharmacological triggers should be avoided. Although in vitro and in vivo studies of drug induced mast cell degranulation do not correlate with clinical experience, agents known to promote mast cell degranulation should be avoided where a feasible alternative option exists e.g. fentanyl not morphine and not codeine
   e.g. cis-atracurium not atracurium.

10. Great care must be taken with positioning and handling.

11. The importance of vigilance and preparedness for widespread mast cell degranulation with cardiovascular collapse cannot be overemphasised. Treatment should be in line with national guidelines for the management of anaphylaxis.

12. If mast cell degranulation occurs it may manifest as urticaria, pruritus or gastrointestinal disturbance. Serum tryptase may be helpful if there is uncertainty particularly if a baseline tryptase has been measured. Exacerbation of skin manifestations although regarded as minor complications may be extremely upsetting and uncomfortable for the patient and symptom control is an important aspect of peri-operative care.

References

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